Self-assessment questions

A 40-year-old woman with lung cavitation

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A 40-year-old woman presented to the emergency room with intermittent fever, cough, haemoptysis and dyspnoea for one week. She also had a history of episodic breathlessness with wheezing for the last 5 years. She did not drink alcohol or smoke tobacco, denied intravenous drug abuse or use of steroids. On physical examination, the patient was febrile (temperature, 38.5°C), tachypnoeic and tachycardic. Blood pressure was 150/90 mmHg. Examination of lungs revealed bilateral coarse crackles and rhonchi. The rest of the systemic examination including cardiovascular system was normal. Serial chest X-rays are shown in figures 1 to 3.

Figure 1 Chest X-ray (PA view) at presentation

Figure 2 Chest X-ray (PA view), one week later

Figure 3 Chest X-ray (lateral view), one week later

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Questions

- 1 What are the findings on serial chest X-ray?
- 2 What is the differential diagnosis?

Answers

QUESTION 1

The first chest X-ray (figure 1), which was taken at presentation, shows bilateral non-homogenous opacities and a thick walled cavity in the left mid-zone with surrounding consolidation. The second X-ray (figure 2), taken a week later, shows bilateral, multiple lung cavitation with air–fluid levels. Pulmonary cavities in both upper and lower lobes are also seen on lateral view (figure 3). The chest X-ray in figure 4, taken approximately 4 weeks after starting treatment, shows significant resolution of pulmonary lesions with antibiotic therapy.

QUESTION 2

The differential diagnosis of cavitating lesion in lung is given in the box. Inflammatory lesions are the most common cause of lung cavities. The number of cavities may vary from one to many. If the lesion is single, abscess from necrotising Gram-negative or staphylococcal pneumonia should be the first consideration, especially if patient is acutely ill with a severe pneumonia that cavitates.

If multiple cavities are present, the infection is likely due to haematogenous dissemination (septic emboli), and a source for this dissemination should be sought. The source could be right-sided endocarditis or infected venous thrombi.

Tubercular cavities are usually located in the upper zone, either the posterior segment of the upper lobe or apical segment of lower lobe. Because of the high concentration of tuberculous bacilli in the cavity, patients usually have a smear strongly positive for acid-fast bacilli. Lung abscesses secondary to aspiration are frequently right-sided and most often involve the lower zone. Most patients with anaerobic lung abscess would have risk factors for aspiration

Differential diagnosis of cavitation on chest X-ray

- cavitating pneumonia: *S aureus*, Gram-negative bacilli (*Klebsiella*, *Pseudomonas*, *Legionella*), anaerobes, mycobacteria, fungi, pneumocystis
- septic emboli, bacterial or fungal
- Wegener's granulomatosis or pulmonary vasculitis
- pulmonary infarction
- infected bullae or cysts
- neoplasia: primary or secondary

such as poor dental hygiene, seizure disorder, alcoholic blackouts, etc. The manifestation is often indolent, with complaints of cough, fever, and malaise lasting for weeks to months. A variety of fungi may cause cavitory lesions. *Aspergillus*, *Mucor*, and *Candida* spp rarely cause severe disease in patients without neutropenia. The endemic fungi (histoplasma, blastomycetes, and coccidioides), however, may cause cavitory lesions in immunocompetent hosts. These organisms should be considered in the differential diagnosis of a cavitory lesion if travel to an endemic area has occurred.

Pulmonary lesions due to infected bullae/cysts are thin-walled cavities with smooth outline. Cavitation may rarely complicate pulmonary embolism with infarct, which are usually seen in the lower zone. Thick-walled cavities may result from pulmonary vasculitis, of which Wegener's granulomatosis is the prototype. Wegener's granulomatosis often results in bilateral multiple cavitory lesions, single lesion being less common. Neoplasia, either primary (bronchogenic carcinoma, lymphoma) or metastatic (from squamous cell) involving the lung, also may cavitate. These are usually thick walled and fluid level is seen more commonly with primary tumours than with metastasis.

Follow-up

Initial laboratory work-up revealed normocytic anaemia, leucocytosis with shift to left and toxic granulation. Arterial blood gas analysis showed mild hypoxaemia which improved with oxygen therapy. Blood biochemical laboratory values were normal. Sputum Gram stain showed Gram-positive cocci in clusters and culture grew *Staphylococcus aureus* (methicillin sensitive) on four occasions. Sputum smears and cultures were negative for acid-fast bacilli. Repeated blood cultures did not grow any organisms. Echocardiogram did not show any abnormalities.

The patient was treated with antibiotics, initially with cefotaxime/amikacin, later changed to cloxacillin. She became afebrile after 2 weeks of therapy, but antibiotics were continued for 6 weeks to promote cure. In addition, chest physiotherapy was helpful in drainage of the lung abscess.

Figure 4 Chest X-ray (PA view), 4 weeks after therapy

Discussion

Staphylococcal pneumonia is an uncommon, but serious infection. Despite appropriate antibiotic treatment, mortality in staphylococcal pneumonia remains high.¹⁻³ Staphylococcal pulmonary infections are typically nosocomial and usually occur in older adults (sixth decade or older) with concomitant medical illnesses.¹ Watanakunakorn³ reported the cases of 44 patients with bacteraemic *S aureus* pneumonia (positive sputum and blood culture). All patients had at least one underlying condition (intravenous drug abusers were excluded from the study). Virtually all patients in a study by Kaye *et al*¹ had some underlying illness, particularly chronic pulmonary disease. Our patient was relatively younger, however she did have a history of chronic pulmonary airway disease.

No single radiological presentation is diagnostic of *S aureus* pneumonia. However, multiple cavitation or pneumatoceles are characteristically seen, particularly in children and intravenous drug abusers. These cavities are usually thin walled (2–4 mm) and are slightly indistinct on their outer border. Abscess formation has been reported in 23–70% of adult patients presenting with *S aureus* pulmo-

nary infections.⁴⁻⁶ However, in recent studies,^{1,2} abscess formation was seen in 16–20% of patients and multiple abscesses occurred infrequently. In the study by Kaye *et al*¹ all patients had single abscesses. The chest X-rays in these patients typically show multilobar infiltrates, predominantly in the lower lobes, and often bilateral.^{1,5,6} The frequency of pleural involvement is variable, with estimates ranging from 5% to 48%.^{1,5,6} Our patient had bilateral involvement on presentation and went on to develop multiple abscesses, however, she did not have pleural involvement.

Despite the availability of effective antibiotics, mortality continues to be 30–32%.^{1,2} This may reflect the fact that patients with *S aureus* pneumonia tend to be elderly and have other significant illnesses. Treatment is with a penicillinase-resistant penicillin such as nafcillin, cefazolin or vancomycin (penicillin allergic or methicillin-resistant *S aureus*), and 4–6 weeks of therapy is required to promote cure.

Final diagnosis

Multiple staphylococcal lung abscesses.

Keywords: pneumonia; *Staphylococcus aureus*

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The chilling tale of a patient with thrombocytopenia

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An 86-year-old woman was admitted having been found collapsed at home. On examination, she was pale, dehydrated, confused and disorientated with a rectal temperature of 28°C. There was no lymphadenopathy or bruising. Her pulse was regular at 50 beats/min and blood pressure was 150/100 mmHg. Cardiorespiratory, abdominal and neurological examinations were all unremarkable. Review of this patient's previous hospital notes revealed no significant illnesses in the past and she was on no regular medication. There was no history of excess alcohol intake. Social assessment revealed that she lived alone with inadequate heating provisions. Investigations showed thrombocytopenia, with a platelet count of $88 \times 10^9/l$ (reference range $150\text{--}450 \times 10^9/l$), confirmed by analysis of two venous blood samples. Her haemoglobin was 14 g/dl, white blood count $8.2 \times 10^9/l$, with a normal differential count, and an erythrocyte sedimentation rate of 6 mm in the first hour. Serum urea was 11.1 mmol/l ($3.5\text{--}8\text{ mmol/l}$) with a serum creatinine of 127 $\mu\text{mol/l}$ ($50\text{--}105\text{ }\mu\text{mol/l}$). Serum electrolytes, liver function tests, coagulation studies, electrocardiogram and chest X-ray were all normal, as were vitamin B12 and folate levels. Urine microscopy and culture revealed no evidence of urinary tract infection. Serum thyroid-stimulating hormone was 18.93 mU/l ($0.5\text{--}5.0\text{ mU/l}$) and free thyroxine 14 pmol/l ($9\text{--}24\text{ pmol/l}$).

Questions

- 1 What is the cause of thrombocytopenia?
- 2 What complications may occur in this patient?

Answers

QUESTION 1

The potential causes of thrombocytopenia are numerous (box 1). In this patient, the cause of thrombocytopenia was hypothermia. Following admission, she was rewarmed and rehydrated and the following day, her resting oral temperature had risen to 34°C and although she remained lethargic, she was no longer confused or disorientated. On day three her resting oral temperature was normal. On day nine, her platelet count had risen to normal ($295 \times 10^9/l$).

QUESTION 2

Apart from the complications of thrombocytopenia, which include skin purpura, mucosal haemorrhage and prolonged bleeding after trauma, hypothermia may lead to other clinical features, shown in box 2.

Discussion

Thrombocytopenia occurring with low body temperature was first described 58 years ago.¹ Since then, animal studies have attempted to identify a cause for this phenomenon.^{2,3} In 1958, Villalobos used radioactively labeled platelets to demonstrate hepatic and splenic sequestration during hypothermia in dogs, with

80% of the platelets returning to the circulation on rewarming.² A later study showed the liver to be the major site of sequestration.³ Disseminated intravascular coagulation and bone marrow failure have also been postulated as causes of hypothermia induced thrombocytopenia.

Platelet sequestration as a result of hypothermia is rare and only a few recorded cases exist.^{4,5} This was thought to be the cause of our patient's thrombocytopenia. She never exhibited any clinical features suggestive of disseminated intravascular coagulation and clotting studies were normal. Her initial full blood count did not show any evidence of bone marrow suppression and the rapid return of the platelet count to normal, mirroring the rise in body temperature, would suggest a causal relationship.

Although our patient experienced no problems as a result of her low platelet count, this case illustrates that thrombocytopenia is a potentially serious complication of hypothermia.

Final diagnosis

Hypothermia-induced thrombocytopenia.

Keywords: hypothermia; thrombocytopenia

Causes of thrombocytopenia

Reduced platelet production

- viral infections: measles, varicella, infectious mononucleosis, congenital rubella, mumps
- drugs: thiazide diuretics
- aplastic anaemia
- leukaemia
- myelodysplasia
- myelosclerosis
- marrow infiltration: carcinoma, lymphoma
- multiple myeloma
- megaloblastic anaemia
- alcohol

Decreased platelet survival

Increased destruction

- auto-immune idiopathic thrombocytopenic purpura
- secondary immune thrombocytopenia: SLE, CLL, lymphoma
- post-transfusion purpura
- drug-induced immune thrombocytopenia: quinine, quinidine, sulphonamides, rifampicin
- heparin

Consumption

- disseminated intravascular coagulation
- thrombotic thrombocytopenic purpura
- haemangioma (Kasabach-Merritt syndrome)

Platelet sequestration

- splenomegaly
- hypothermia

Box 1

Complications of hypothermia

Haematological

- polycythaemia
- isolated thrombocytopenia or leucopenia
- bone marrow suppression
- disseminated intravascular coagulation

Nervous system

- listlessness
- confusion
- amnesia
- loss of consciousness
- motor paralysis
- sensory impairment

Cardiovascular

- initial rise in cardiac output and blood pressure and tachycardia
- later fall in cardiac output and hypotension and bradycardia
- arrhythmias: atrial fibrillation, ventricular fibrillation
- ischaemic necrosis

Respiratory

- depressed respiration
- tissue anoxia
- respiratory acidosis
- pneumonia

Metabolic

- hyperglycaemia
- hyperkalaemia
- renal failure

Gastrointestinal

- gastric erosions
- haemorrhage
- pancreatitis

Box 2

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Hypotension and intractable vomiting in the first trimester of pregnancy

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A 24-year-old woman, gravida 1, para 1, presented at 12 weeks gestation with abdominal pain, weakness, fatigue, nausea and vomiting of several days duration. Ten days earlier she had been admitted to another hospital with the same complaints; she had been diagnosed as having hyperemesis gravidarum and hospitalised for 3 days, after which she was stabilized. Her medical history was unremarkable and she had no family history of relevant illness. On initial evaluation, she was in rather poor clinical condition and was pigmented to an abnormal degree, particularly around the eyes and in the skin creases of the hands; buccal pigmentation was also pronounced. Blood pressure was 100/60 mmHg, pulse 108 beats/min and body temperature was 38.7°C. Physical examination was unremarkable except for dehydration and hypotension. The laboratory examinations are summarised in the table. The results of urinalysis were: specific gravity 1.016, pH 5, 2+ protein, and urine sediment examination revealed a large number of erythrocytes and leukocytes. The urine contained Gram-negative bacteria.

Within two hours, the patient was confused, sweating and tachypnoeic. A repeat blood sugar level was 52 mg/dl. She was given a rapid infusion of intravenous 5% dextrose in 0.9% sodium chloride, but did not improve. Arterial blood gases showed a profound metabolic acidosis. Her acid-base status was as follows: pH 7.12, HCO₃ 6 mmol/l, pO₂ 154 mmHg, pCO₂ 20 mmHg, base excess –22.

Table Initial laboratory findings

Variables	Patient	Normal range
Haemoglobin (g/dl)	14.6	12–18
Mean corpuscular volume (fl)	84	80–100
White cell count ($\times 10^9/l$)	16.6	3.6–10
Differential count (%)		
Neutrophils	78	40–70
Band forms	10	0–5
Lymphocytes	12	20–40
Platelet count ($\times 10^9/l$)	452	150–450
Sedimentation rate (mm/h)	90	0–15
Urea nitrogen (mg/dl)	28	8–23
Creatinine (mg/dl)	1.0	0.6–1.2
Total protein (g/dl)	6.8	6–7.8
Albumin (g/dl)	3.8	3.2–4.8
Uric acid (mg/dl)	6.2	2.7–7.8
Glucose (mg/dl)	62	70–110
Sodium (mmol/l)	124	136–147
Potassium (mmol/l)	5.6	3.5–5.5
Alanine transaminase (IU/l)	28	5–40
Aspartate transaminase (IU/l)	32	8–33

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Questions

- 1 What is the differential diagnosis compatible with the history, clinical and laboratory findings?
- 2 Which clues suggest an underlying endocrinologic illness?
- 3 What could be a precipitating factor leading to the aggravation of the underlying illness?

Answers

QUESTION 1

The differential diagnosis is septicaemia, severe hyperemesis gravidarum, adrenal crisis, acute thyrotoxicosis, or hyponatraemic encephalopathy.

QUESTION 2

The classical features of adrenal crisis (box 1) are present in this patient and the severe degree of metabolic acidosis suggests an underlying precipitating factor.

Classical clinical features of adrenal crisis

- symptoms: rapid worsening of preceding symptoms; severe weakness, fatigue, confusion, abdominal pain, nausea, vomiting, diarrhoea
- signs: fever, low blood pressure, dehydration, skin pigmentation
- laboratory findings: hyperkalaemia, hyponatraemia, high blood urea nitrogen, hypoglycaemia, metabolic acidosis (sometimes), low serum cortisol with high ACTH level (primary adrenal insufficiency), or low serum cortisol with low ACTH level (secondary adrenal insufficiency)

Box 1

QUESTION 3

The precipitating factor could be infection (sepsis, urinary tract infection, etc), trauma, surgery, withdrawal of therapy, or drugs (box 2).

Precipitating causes of adrenal crisis

- in diagnosed chronic adrenal insufficiency: withdrawal of therapy, infection, trauma, surgery, dehydration, drugs
- in a normal person: blockage of hormone synthesis (eg, by drugs such as aminoglutethimide, ketoconazole, etc, or as a result of surgery, eg, adrenalectomy), or increased degradation of hormones due to drugs such as rifampicin, dilantin, phenobarbital, etc
- following massive bilateral adrenal haemorrhage as may occur in severe infection (septicaemia, meningococcaemia, etc), HIV, coagulation disorders, intra-abdominal surgery, adrenal metastasis
- hypophyseal causes: surgery, radiotherapy, apoplexy, thyroid hormone replacement without steroids in panhypopituitarism

Box 2

Outcome

The patient was treated with intravenous bicarbonate infusion, and intravenous ceftriaxone for suspected sepsis due to a urinary tract

infection. Because her clinical and laboratory status were suggestive of acute adrenal insufficiency, we performed a rapid adrenocorticotropin (ACTH) stimulation test with a 0.25 mg intravenous bolus of tetracosactrin after taking blood samples for basal serum cortisol and ACTH. The test showed the basal plasma cortisol to be 5 µg/dl, with no evidence of a response to the tetracosactrin after 30 minutes. While the serum cortisol concentration was inappropriately low, the ACTH level was elevated. These results suggested acute adrenal insufficiency. A bolus intravenous infusion of 100 mg hydrocortisone followed by a continuous infusion of hydrocortisone at a rate of 10 mg/h was administered. Within 3 hours, there was restoration of blood pressure and body temperature and a general improvement was seen. Therapy was maintained with a continuous infusion of dextrose in 0.9% saline, antibiotics, and hydrocortisone. On day 2 of the admission the patient continued to improve. Her pH was normal, serum lactic acid levels had dropped significantly, and blood sugar levels were normal. Her electrolyte status also improved. Maternal and foetal monitoring during the pregnancy did not show any profound effects, and she delivered vaginally a healthy infant at 38 weeks gestation.

Discussion

A wide variety of metabolic and endocrine disorders may complicate pregnancy. Acute adrenal insufficiency is an emergency and is caused by a sudden, marked decline in levels of adrenocortical hormones.¹ In some cases, the condition first appears during the pregnancy, and in others acute adrenal insufficiency can occur in the course of a chronic insufficiency. Rarely, acute adrenal insufficiency may be the initial manifestation of new adrenal disease in pregnancy.² Acute adrenal crisis during pregnancy may mimic hyperemesis gravidarum.³

Previous case reports have suggested that acute adrenal insufficiency does not become manifest until the postpartum period. This may be partly due to a delay in diagnosis due to the similarity of symptoms common to pregnancy with those of Addison's disease, and partly because placental foetal steroid production protects the mother from crisis, although this latter point is controversial.¹⁻⁷ Alternatively, patients may develop acute adrenal crisis during pregnancy in the presence of precipitating factors such as severe infection, as in our patient.

Clinical symptoms, signs and laboratory findings in adrenal crisis are presented in box 1. In cases of adrenal crisis, the serum cortisol concentration is inappropriately low, and the ACTH levels will be elevated if the disease is primary and low in secondary adrenal insufficiency.² However, in the pregnant patient with hypoadrenalism, cortisol levels can be within the normal range for non-pregnant patients and the diagnosis will be based on the lack of a rise of plasma cortisol after ACTH stimulation.⁵⁻⁶ In a patient with no known history of adrenal insufficiency, it can be difficult

to distinguish between Addison's disease and acute adrenal insufficiency, especially during the first trimester of pregnancy. It is important to remember that mild cases can go undetected during pregnancy and become manifest as crises at parturition or in the presence of other illness such as urinary tract infection, dehydration, pre-eclampsia, etc.^{6,7}

In conclusion, acute adrenal insufficiency is a rare disorder whose diagnosis can be difficult during pregnancy. On the other hand, it is associated with high maternal and/or foetal

morbidity and mortality if allowed to progress. For this reason, early recognition and intervention are critical.

Final diagnosis

Acute adrenal insufficiency precipitated by urinary tract infection.

Keywords: adrenal insufficiency; pregnancy; urinary tract infection; vomiting; hypotension

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Fatal complication of coincidental operative finding

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A 65-year-old man with an established diagnosis of Crohn's colitis presented as an emergency with peritonitis and free subdiaphragmatic air on erect chest X-ray. At laparotomy, a grossly distended colon was noted, with two sites of perforation at the rectosigmoid and splenic flexure areas. Multiple non-dilated jejunal diverticula were noted coincidentally. Subtotal colectomy with cross-stapling of the rectum and formation of an end ileostomy was performed. He made an uneventful early postoperative recovery but during the fourth postoperative week he developed intermittent, colicky abdominal pain, vomiting and reduced ileostomy output. Subacute small bowel obstruction secondary to adhesions was diagnosed after clinical and radiological examination. Conservative treatment with intravenous fluids and nasogastric decompression produced an initial improvement but on the 32nd postoperative day the patient suddenly collapsed and died from a cardiorespiratory arrest secondary to acute renal failure and septic shock. At post-mortem examination, evidence of proximal jejunal diverticular disease with signs of acute diverticulitis and perforation were noted along with widespread peritonitis. No histological features of Crohn's disease were identified in the segment of perforated jejunum, or elsewhere in the bowel, and the inflammatory process was centred around the diverticulum (figure).

Figure Perforated jejunal diverticulum lined by inflamed granulation tissue. Some residual diverticular mucosa is visible on the right (arrow). The lumen of the perforation is indicated by *

Questions

- 1 What is the incidence of small bowel diverticula?
- 2 What is the aetiology of small bowel diverticulosis?
- 3 List the common pre-operative clinical manifestations of small bowel diverticula.

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Answers

QUESTION 1

Small bowel diverticula are uncommon, occurring in 0.5–2.3% of small bowel contrast studies¹ and 0.06–1.3% of post-mortem examinations.² They manifest most frequently in the sixth and seventh decades of life and were thought to occur more commonly in males,³ although one study has shown a preponderance for females.¹

QUESTION 2

Non-Meckelian small bowel diverticula are thought to be acquired pulsion defects caused by possible underlying abnormalities in peristalsis which in turn produce segmentally high intraluminal pressures.⁴ Jejunal diverticulosis has been described in association with systemic sclerosis and visceral neuropathy or myopathy.⁵ The diverticula of mucosa and submucosa emerge at natural weak sites where mesenteric vessels penetrate the intestinal wall explaining the observation that they are most commonly located at the mesenteric side of the bowel.⁶ Small bowel diverticula occur more frequently in the proximal jejunum and distal ileum where the vasa recti of greatest diameter are found.⁶

QUESTION 3

Common pre-operative complications associated with jejuno-ileal diverticula include haemorrhage, obstruction, diverticulitis, perforation, malabsorption, anaemia and chronic vague abdominal pain (box 1).⁶ The most common clinical signs include hyperactive bowel sounds, hyper-resonance with epigastric percussion and vague epigastric discomfort on palpation.¹ Complications of small bowel diverticula may manifest in different ways with varying frequencies according to their anatomical location (box 2).

Discussion

There are no reliable diagnostic tests to confirm the presence of small bowel diverticula. Erect abdominal X-rays may show air–fluid levels throughout the small bowel. The classic triad of features associated with jejuno-ileal diverticular disease consists of vague abdominal pain, anaemia and dilated loops of small bowel on abdominal X-ray.⁷ If the presence of small bowel diverticula is suspected then a small bowel barium follow-through study or enteroclysis can be performed. While the former easily demonstrates large diverticula, smaller ones may not be identified because of inadequate filling with contrast or due to extrinsic abdominal compression.⁸ Thus, a negative barium follow-through study does not exclude small bowel diverticular disease. Workers have shown that enteroclysis has a greater sensitivity at detecting diverticulae.⁸ This investigation should be borne in mind for the patient with persisting abdominal symptoms who has undergone negative endoscopic and standard contrast studies of both upper and lower gastrointestinal tracts.¹ Small bowel diverticulitis should also be included in the differential diagnosis of a small

Learning points

- the majority of small bowel diverticula are asymptomatic
- jejuno-ileal diverticula are more likely to have complications than duodenal diverticula
- duodenal diverticula are more likely to bleed
- jejuno-ileal diverticula are more likely to perforate/form abscesses
- asymptomatic small bowel diverticula can cause postoperative complications

Box 2

bowel inflammatory mass demonstrated by computed axial tomography.⁹ Upper gastrointestinal endoscopy and laparotomy are the two other main methods for diagnosing the presence of small bowel diverticula.⁵

In jejunal diverticular disease, obvious perforation, bleeding, or mechanical complications require resection with primary anastomosis. Failure of medical therapy in cases of blind loop syndrome or nonmechanical obstruction from severe jejunal dysmotility in the presence of diverticula may also benefit from resection of the affected segment. A dilated, hypertrophied segment of jejunum with large diverticula, suggestive of a progressive form of diverticular disease, which is found coincidentally at laparotomy should also be resected.⁶ The majority of ileal diverticulae on the other hand do not require operative intervention, except in cases of perforation, bleeding, or obstruction which occur less frequently than in jejunal disease. Excision of a coincidental, solitary, non-Meckelian ileal diverticulum in an adult is not warranted.^{1, 6}

Jejunal diverticula are probably more common than reported.⁷ They may be easily overlooked at operation as they are frequently present between the leaves of mesentery.⁴ In asymptomatic jejuno-ileal diverticular disease, noted coincidentally at operation or during radiological investigations, the risk of complications ensuing was 18% at a mean follow-up of 4.8 years in one series.¹⁰ The above case illustrates that serious complications of jejunal diverticular disease can arise in the early postoperative period. To date, there have been no reports in the literature of jejunal diverticular disease causing early postoperative complications. The coincidental finding of previously asymptomatic jejunal diverticula at laparotomy, whilst uncommon, should always be considered in the differential diagnosis of subsequent early postoperative symptoms suggestive of subacute small bowel obstruction. Failure to do so, with attention and management focussed on more likely causes for such symptoms may mask the true diagnosis with fatal consequences.

Final diagnosis

Fatal perforated acute jejunal diverticulitis complicating postoperative recovery following emergency subtotal colectomy for perforated fulminant Crohn's colitis.

Clinical manifestations of small bowel diverticulosis

- asymptomatic
- bleed
- obstruction
- perforation +/- abscess formation
- chronic abdominal pain
- volvulus
- enterolith formation
- anaemia
- malabsorption

Box 1

Keywords: jejunoileal diverticulitis; diverticulitis; small bowel perforation; Crohn's disease; enterocolitis

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Multinodular goitre, dysphagia and nodular shadows in the lung

A Bhansali, S Bhadada, R Kochhar, R Muralidharan, R J Dash

A 41-year-old woman presented in December 1997 with dry cough with streaky haemoptysis of 15 days duration. She had had a goitre for the last 25 years and had been diagnosed as having thyrotoxicosis on clinical and biochemical grounds (triiodothyronine (T_3) 2.8 ng/ml and thyroxine (T_4) 220 ng/ml) in September 1995. She had received carbimazole for 6 months and remained asymptomatic thereafter.

On examination, her body mass index (BMI) was 24.1 kg/m². She had no pallor, lymphadenopathy, clubbing, oedema or hyperkeratosis. Her pulse was 96 beats/min regular, blood pressure 110/70 mmHg. She denied a history of fever, weight loss, loss of appetite, or recent increase in goitre size, but had been dysphagic to solids for 2–3 days. She was a non-smoker and never consumed alcohol. She had a firm, grade III (WHO) non-tender multinodular goitre with retrosternal extension. Systemic examinations were normal except for diffuse bilateral rhonchi. Her haemogram and serum biochemistry, including thyroid hormone profile (T_3 0.86 ng/ml, T_4 78 ng/ml, thyroid-stimulating hormone 0.3 μ U/ml) were normal. Ultrasonography of thyroid confirmed the multinodular goitre with retrosternal extension with some areas of necrosis and specks of calcification. Ultrasound-guided fine needle aspiration cytology (FNAC) revealed follicular cells with nuclear grooving and psammoma bodies. Her radiological profile is shown in figure 1. Bronchoscopy could not be completed because of laryngeal spasm. Computed tomography (CT)-guided FNAC from pulmonary lesions was inconclusive.

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Figure 1 Clinical and radiological profile of the patient

Questions

- 1 What is shown in figure 1?
- 2 What is your diagnosis?
- 3 What would be the approach to confirm the diagnosis?

Answers

QUESTION 1

Figure 1 shows bilateral multiple nodular opacities in lung fields on chest X-ray, further substantiated on high-resolution CT. Cervical X-ray shows a large goitre with retrosternal extension and calcification.

QUESTION 2

With the available clinical and radiological profile, a diagnosis of euthyroid multinodular goitre with retrosternal extension and metastatic lesions in the lung can be made. Further work-up to determine the primary source, whether from thyroid or elsewhere, is required.

In the presence of a long-standing euthyroid goitre, the possibility of thyroid carcinoma with pulmonary metastases is high. However, multinodularity and absence of cervical lymphadenopathy in the face of pulmonary metastases decreases this probability.¹ The presence of nuclear grooving and psammoma bodies can also be seen in long-standing multinodular goitre.^{2,3}

QUESTION 3

The clue to the primary site of malignancy came from the widening of the prevertebral space in the cricopharyngeal region and the presence of malignant keratinizing squamous cells in the sputum. Barium swallow showed a narrowed segment with irregular mucosa in the upper third of the oesophagus (figure 2). Fibre-optic endoscopy revealed a growth in the upper cervical oesophagus. Brush cytology smears from the growth confirmed squamous cell carcinoma. Her disease was classified as stage IV (any T, any N & M1) according to the TNM staging system. She received cisplatin and etoposide cyclically (3 days treatment at 4-week intervals) and palliative radiation therapy.

Figure 2 Barium study showing a narrowed segment with irregular mucosa in the upper third of the oesophagus

Learning points

- when metastatic pulmonary lesions are seen in a patient with long-standing euthyroid nodular goitre without enlarged lymph nodes in the neck, a non-thyroidal primary source needs to be considered
- nuclear grooving and psammoma bodies, which are considered a cytological hallmark of papillary carcinoma, can also be present in long-standing multinodular goitre
- upper oesophageal carcinoma with lung metastases may present with subtle symptomatology

Discussion

Patients with oesophageal carcinoma with metastatic disease usually present with dysphagia, odynophagia, weight loss and marked emaciation.⁴ Our patient had maintained reasonably good health (BMI 24.1 kg/m², haemoglobin 10.6 g/dl) at presentation. She experienced transient dysphagia to solids, attributable to compression from the multinodular goitre. Dysphagia, however, is not a presenting manifestation of upper cervical oesophageal carcinoma as this segment of the oesophagus is capacious.

Squamous cell carcinoma of the oesophagus, oropharyngeal leucoplakia and palmo-plantar tylosis are genetically linked. However, except for a positive family history of dysphagia in two of her first degree relatives, there were no indications for this syndrome in our patient.⁵ The relative frequencies of squamous cell carcinoma in the upper, middle and lower portions of oesophagus are 17%, 55% and 33%, respectively. Based on gross appearance, the fungating variety accounts for 60%, ulcerative type 25%, and infiltrative type 15%.⁵

Chronic cough in squamous cell carcinoma of the oesophagus occurs due to aspiration, tracheo-oesophageal fistula, or pulmonary metastases. In view of ready access to blood vessels, haematogenous metastases to lungs, liver, kidneys and bone are common.⁵

Chemotherapy combined with palliative radiation yields better results than radiation therapy alone in patients with localised carcinoma of the oesophagus.⁵ Only about 20% of lesions are resectable but surgical intervention does not lead to increased survival except when the lymph nodes are not involved. The overall prognosis is poor, with 5-year survival rate less than 5%.⁵

Final diagnosis

Euthyroid multinodular goitre and squamous cell carcinoma of oesophagus with pulmonary metastases.

Keywords: multinodular goitre; squamous cell carcinoma; oesophageal carcinoma; pulmonary metastases

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An unusual cause of dysphagia

Israel Gotsman, Paul Mogle, Michael Y Shapira

A 74-year-old woman was referred for evaluation of dysphagia and weight loss. She had a history of dysphagia for solid foods, which had become worse over the past year. She had lost 5 kg in weight but her appetite was good. She suffered from mild heartburn without pain and her bowel movements were normal. Gastroscopy showed mild gastritis with a positive culture for *Helicobacter pylori*. A short course of triple antibiotic therapy and omeprazole was prescribed. Follow-up endoscopy appeared normal. Her heartburn improved but the dysphagia persisted.

She had had a myocardial infarction 4 years earlier, complicated by acute mitral regurgitation due to papillary muscle rupture. This was repaired by urgent surgery. A post-operative echocardiogram showed slight left ventricular enlargement with decreased global function and moderate mitral regurgitation. She was treated with frusemide and digoxin and had minimal complaints of heart failure. She also suffered from type II diabetes and hypertension controlled by atenolol.

Physical examination revealed a thin woman with normal vital signs. The thyroid and lymph nodes were not enlarged. Heart sounds were normal with a 3/6 blowing systolic murmur at the apex radiating to the axilla. Breath sounds were reduced at the base of the right lung. The abdomen was mildly distended but not tender. The liver and spleen were not enlarged, peristalsis was normal. There was mild oedema of both legs. Peripheral pulses were present. Laboratory studies showed mild elevation of alkaline phosphatase and γ -glutamyl transpeptidase. Albumin and cholesterol levels were normal. Haemoglobin was 14.5 g/dl. Electrocardiogram showed sinus rhythm with a non-specific intraventricular block. Chest X-ray (figure 1) demonstrated an enlarged cardiac silhouette with a small right pleural effusion.

Questions

- 1 What is evident on the lateral chest X-ray that may explain the dysphagia?
- 2 What further examinations should be made in order to make a diagnosis?

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Figure 1 Lateral chest X-ray

Answers

QUESTION 1

The lateral X-ray (figure 1) shows an enlarged left atrium, consistent with mitral valve disease. An enlarged left atrium can cause dysphagia to solids due to external compression of the oesophagus. This rare cause of dysphagia is known as cardiovascular dysphagia.¹

QUESTION 2

The patient's predominant complaint was dysphagia to solids. This usually implies mechanical obstruction of the oesophagus. Intrinsic lesions that obstruct the oesophagus include peptic stricture, lower oesophageal (Schatzki's) ring, or oesophageal carcinoma. Extrinsic lesions include vascular abnormalities, mediastinal abnormalities or cervical osteoarthritis. The first step in the diagnosis is to demonstrate the anatomy of the oesophagus by barium swallow. Video barium swallow showed normal movement of the oropharyngeal muscles and normal peristalsis of the oesophagus. A pulsatile bulge was noted in the distal posterior oesophagus consistent with external pressure from an enlarged left atrium (figure 2). No reflux was apparent. A computed tomography (CT) scan of the chest revealed an enlarged left atrium compressing the oesophagus. A space-occupying lesion was not evident. A manometric study of the oesophagus, performed to complete the diagnosis and exclude oesophageal dysmotility due to diabetes, revealed normal peristaltic pressures in the oesophagus. A transthoracic echocardiogram showed an enlarged left ventricle with moderate global dysfunction and severe mitral regurgitation. The left atrium was enlarged.

Figure 2 Barium swallow of the oesophagus showing external compression of the distal posterior oesophagus from an enlarged, left atrium (arrow). The repaired annulus of the mitral valve can be seen due to a ring placed there

The patient was treated for mitral regurgitation with after-load reduction and diuretics. She improved, the dysphagia gradually subsided, and she gained weight. The final diagnosis was cardiovascular dysphagia.

Discussion

Cardiovascular dysphagia is an uncommon clinical entity that is often unrecognised (box 1).¹⁻⁴ The left atrium is a mid-line posterior chamber of the heart, in front of the oesophagus. Left atrial enlargement causes dysphagia by external compression of the oesophagus. A manometric study of left atrial enlargement⁵ showed that mechanical compression causes a localised high pressure zone in the oesophagus at the level of atrium with pressure oscillations corresponding to the electrocardiogram.

Other suggested mechanisms for cardiovascular dysphagia include deranged peristalsis due to local ischaemia of the oesophageal mucosa and nerve plexus caused by the elevated external pressure (which was not evident in our case). Prolonged exposure of the distal oesophagus to a high external pressure may cause proximal oesophageal muscle fatigue and dysphagia.

This patient presented with dysphagia due to mechanical compression of the oesophagus by an enlarged left atrium from mitral incompetence. Reduction of mitral incompetence by afterload reduction ameliorated the dysphagia. Cardiovascular dysphagia is uncommon, but should be suspected in patients with an enlarged left atrium.

Causes of cardiovascular dysphagia

- left atrial enlargement
- aberrant left or right subclavian artery
- tortuous atherosclerotic aorta
- thoracic aortic aneurysm

Box 1

Cardiovascular dysphagia (left atrial enlargement)

- occurs in rheumatic heart patients with mitral valve disease and enlarged left atrium
- is caused by luminal obstruction due to oesophageal compression by an enlarged left atrium

Diagnosis:

- barium swallow: posterior displaced oesophagus, a smoothly compressed oesophageal lumen, delayed passage of barium
- endoscopy: pulsatile bulging mass in the mid-oesophagus, normal mucosa
- oesophageal manometry: elevated mid-oesophageal base line pressure with superimposed cyclic pressure waves simultaneous with the ECG

Box 2

Final diagnosis

Cardiovascular dysphagia.

Keywords: cardiovascular dysphagia; mitral valve disease; weight loss

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Lessons from a case of tetanus in an elderly woman

J Kwan, S Lim, S C Allen

A healthy 84-year-old Caucasian woman fell whilst gardening at home and lacerated her right leg. The wound was deep and heavily contaminated with soil and gravel. She attended an Accident and Emergency (A&E) department for treatment and the wound was cleaned and dressed. She had never received tetanus vaccination in the past and at the A&E department she was given one dose of tetanus toxoid. She was sent home without any follow up. Over the next week she became increasingly immobile and complained of dysphagia, stiff neck and breathlessness. Her general practitioner admitted her into a nursing home with a presumed diagnosis of stroke. Her condition deteriorated over the next week and she was admitted into hospital with a provisional diagnosis of septicaemia from her original leg wound.

On admission she was dehydrated, hypoxic, hypertensive (230/125 mmHg) and tachycardic. She had generalised muscular spasms including trismus and neck stiffness as well as hyperexcitability to touch and noise. Spirometry showed a restrictive lung defect. Initial laboratory results were as follows: leucocytes $15.5 \times 10^9/l$, platelets $653 \times 10^9/l$, urea 15.4 mmol/l, creatinine 227 $\mu\text{mol/l}$ and corrected calcium 2.48 mmol/l. A diagnosis of tetanus was made and she was transferred to the Intensive Care Unit where she was managed according to recommended guidelines for the treatment of tetanus. Her pulse and blood pressure observations were monitored closely (table).

Table Blood pressure and pulse observations on day 7

Time	Blood pressure (mmHg)	Pulse (beats/min)
01:00	122 / 50	80
04:00	90 / 40	70
07:00	125 / 60	80
12:00	150 / 50	100
20:00	160 / 70	120
22:00	130 / 70	88

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Questions

- 1 Comment on the original A&E department management of this patient.
- 2 What caused the abnormally unstable blood pressure and pulse?
- 3 What are the most important aspects of the management of tetanus?
- 4 What is the differential diagnosis and how may the initial diagnosis be confirmed?

Answers

QUESTION 1

The patient's wound was heavily contaminated with soil and gravel and, by definition,¹ the wound was tetanus prone. The patient had never received vaccination against tetanus. According to the Department of Health guidelines,¹ patients who have never been vaccinated and have a tetanus-prone wound should receive an immediate dose of human tetanus immunoglobulin followed by a complete primary course (three doses) of tetanus toxoid. The patient in our case, however, had not been given a dose of human tetanus immunoglobulin and was discharged without follow-up.

QUESTION 2

The labile blood pressure and tachycardia were most likely a result of a combination of pain (from both the wound and muscular spasms), anxiety, sepsis and autonomic dysfunction. Autonomic dysfunction is common in patients with tetanus and it may also present as pyrexia, dry mouth, profuse sweating, urinary retention and cardiac arrhythmia, which is associated with a mortality of over 50%.³ The unstable blood pressure of autonomic dysfunction is difficult to control, but some benefit has been demonstrated with the use of propranolol and clonidine.^{2,4}

QUESTION 3

The immediate management for severe tetanus should include surgical debridement of the wound, intravenous metronidazole and human tetanus immunoglobulin, antispasmodic therapy, early mechanical ventilation and tracheostomy. Nursing the patient in a quiet room prevents muscular spasms from hyperexcitability. Metronidazole has been shown to be more effective than penicillin in preventing death when used in the treatment of tetanus. It has been postulated that penicillin is a centrally acting GABA antagonist, and may therefore act synergistically with tetanospasmin in producing muscular spasms.² Early enteral or parenteral feeding is also recommended because of the high caloric requirement secondary to hypermetabolism in patients with tetanus. Subsequently, since the amount of tetanus toxin released is insufficient to induce an adequate immune response, the patient will also need immunisation with tetanus toxoid.

QUESTION 4

Tetanus is a clinical diagnosis. The combination of a history of injury with a contaminated wound, muscular spasms (with or without risus sardonicus, trismus or opisthotonus) and signs of sepsis should raise the possibility of the diagnosis. Tetanus immunisation history is a poor predictor of immune status in elderly

Learning points

- tetanus is a disease of the elderly population
- tetanus is preventable with appropriate wound care and prophylaxis
- admission into a nursing home without a proper diagnosis should be avoided
- autonomic dysfunction can cause labile blood pressure and pulse in tetanus

people with a positive predictive value of 50% and a negative value of 76%.⁵ It is important to exclude other differential diagnoses such as hypocalcaemic tetany, orofacial infection, status epilepticus and drug-induced dystonia (eg, phenothiazines, metoclopramide). Strychnine poisoning can mimic tetanus but it does not cause trismus or abdominal rigidity. Culture of *Clostridium tetani* from the affected wound is positive in less than 30% of cases. Moreover, a positive culture is not diagnostic since not all patients with wound colonisation develop tetanus. Serological tests may be used to detect the level of anti-tetanus antibody but their use in diagnosis is limited.

Discussion

In the UK, tetanus is a disease of the elderly population and elderly women are most at risk probably due to a combination of inadequate immunity and the increasing risk associated with a physically active old age. Tetanus immunisation began in 1938 for those who served in the Armed Forces. In 1961, primary tetanus immunisation of infants was introduced nationally. In 1970, tetanus immunisation was recommended as part of the routine management of all wounds. Between 1984 and 1995, there were 145 notified cases of tetanus in England and Wales; 53% of the cases were in individuals over 65 years and two-thirds of them were women.

Although tetanus is now rare, it is preventable by undertaking appropriate wound care and tetanus prophylaxis as recommended by the Department of Health.¹ Our case illustrates that admission of any elderly person into a private sector nursing home without a proper diagnosis may delay investigations and treatment of the underlying illness. The most important step in diagnosing tetanus is the maintenance of a high suspicion for the disease.

Final diagnosis

Autonomic dysfunction in an elderly patient with severe tetanus.

Keywords: tetanus; elderly; autonomic dysfunction

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Unusual presentations to a lipid clinic

M A Crook, A Mukherjee, K Marshall.

We describe the case-histories of two young men who both presented to a lipid clinic with mixed hyperlipidaemias.

Case 1

A 33-year-old man had been referred to the lipid clinic by the dermatology unit because of planar xanthoma that been found on a skin biopsy. Apart from previously seeing the Ear, Nose and Throat department for an 18-month history of snoring, there was no other relevant medical history nor other symptoms. He was a non-smoker, rarely took alcohol and at the time was not taking any medication. On examination he was overweight with a body mass index (BMI) of 29 kg/m². His blood pressure at presentation was 140/90 mmHg and pulse 72 beats/min. He was clinically euthyroid and had planar xanthoma on his hands and tuberous xanthoma on his elbows. He had been commenced on a lipid-lowering diet and a number of investigations were performed. His fasting serum cholesterol was 12.6 mmol/l, triglyceride 5.3 mmol/l and high-density lipoprotein (HDL) cholesterol 1.31 mmol/l. Plasma urea, creatinine and fasting glucose were normal, as were his liver function tests. However, thyroid function tests showed a thyroid-stimulating hormone of 257.7 mU/l (normal range 0.3–5.0) and free thyroxine of 1.1 pmol/l (10.3–19.4). Anti-thyroglobulin and antithyroid peroxidase antibodies were 396 and 240 IU/ml, respectively (0–180 and 0–50). Apolipoprotein (apo) E phenotype performed by Western blotting showed him to be a E2/E2 homozygote. On a lipid-lowering diet instigated by a dietician and thyroxine 150 µg per day his fasting serum cholesterol improved to 5.11 mmol/l and triglyceride to 2.15 mmol/l and his thyroid function tests were rendered normal.

Case 2

A 33-year-old man was referred to the lipid clinic by his general practitioner because of hyperlipidaemia and a family history of premature coronary heart disease. His mother had hypothyroidism and pernicious anaemia. On examination he was euthyroid and had no lipid stigmata. His BMI was 31 kg/m², blood pressure was 150/92 mmHg, and pulse 72 beats/min. Initial fasting serum cholesterol was 16.49 mmol/l, triglyceride 20.83 mmol/l and HDL-cholesterol 0.59 mmol/l. His renal and liver function were normal, as was a fasting plasma glucose. However, his TSH was 37.6 mU/l and free T4 8.5 pmol/l. Anti-thyroglobulin and anti-thyroid peroxidase antibodies were 2153 and 711 IU/ml, respectively. He was commenced on thyroxine which resulted in normalisation of his thyroid function, although his fasting serum cholesterol remained elevated at 11.8 mmol/l with triglyceride of 16.7 mmol/l. His apoE phenotype also by Western blotting was E2/E2. He was subsequently started on bezafibrate 400 mg (MONO) nocte with considerable improvement in his lipids.

Questions

- 1 What lipid disorder do these two patients display?
- 2 How can the diagnosis be confirmed?
- 3 What other conditions is this lipid disorder associated with?

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Answers

QUESTION 1

They were both diagnosed as having a type III hyperlipoproteinaemia. Type III hyperlipoproteinaemia, also called broad beta or remnant dyslipidaemia, is a rare familial hyperlipidaemia whose recognition is important as it is usually responsive to therapy. The treatment is usually dietary, in conjunction with a lipid-lowering drug using a fibrate or possibly a statin.¹ The palmar striae (palmar xanthomata) are considered pathognomonic for the disorder and occur in less than 50% of patients but tubero-eruptive xanthomata, typically on the elbows and knees, as well as xanthelasma and corneal arcus have been described in this condition. Peripheral vascular disease is a typical feature of this hyperlipidaemic disorder as is premature coronary artery disease. Serum lipid determination will frequently reveal hypercholesterolaemia and hypertriglyceridaemia, often in similar molar proportions. Serum HDL cholesterol is usually low. Serum low-density lipoprotein (LDL) cholesterol may also be low due to the fact that there is reduced conversion from intermediate-density lipoprotein particles, although LDL cholesterol may also be normal or elevated.²⁻⁴

The underlying biochemical defect is one of a reduced clearance of chylomicron and VLDL remnants. This is also known as broad beta hyperlipidaemia because of the characteristic serum lipoprotein electrophoretic pattern often observed (the broad beta band that is seen being predominately remnant particles).

An association with type III/broad beta hyperlipidaemia and homozygosity for apoE2 or apoE2 variants has been described. ApoE shows three common alleles, E2, E3, and E4, coded for on chromosome 19 and which are important for the binding of remnant particles to the remnant receptor.

The mechanism for the disorder seems to be that apoE2-bearing particles have poor binding to the apoB/E (remnant) receptor and thus are not effectively cleared from the circulation.

QUESTION 2

Serum lipoprotein electrophoresis can show the classic type III picture with a broad beta band composed of remnant particles, although this is not always present. An association of type III broad beta hyperlipidaemia with homozygosity for apolipoprotein E2 has been described and thus apoE phenotyping or genotyping by a specialised laboratory can be useful, although some patients with broad beta hyperlipidaemia can show other apoE pheno-

types or variants. Another investigation that can be useful in establishing the diagnosis is ultracentrifugation to separate the lipoprotein particles. The cholesterol of the VLDL particles is then quantitated and expressed as a total of the serum triglyceride concentration. In molar terms, normal individuals show a ratio of below 0.30 while ratios over 0.30 are more likely in broad beta hyperlipidaemia, particularly if this is nearer 0.60.

QUESTION 3

It is becoming apparent that it is not just inheriting the apoE2 genotype that is important in developing broad beta hyperlipidaemia. The prevalence of the apoE2/E2 genotype is about 1 in 100 in the general population, yet only about 1 in 5–10 000 individuals manifest type III hyperlipidaemia. A concurrent increase in serum VLDL also seems necessary for the condition to be expressed, such as might occur in diabetes mellitus, hypothyroidism or obesity. Some patients may show either an autosomal recessive or dominant mode of inheritance of the condition.

The presentation of primary hypothyroidism in two young euthyroid males with type III hyperlipoproteinaemia is remarkable. It is known that the prevalence of newly diagnosed overt hypothyroidism in patients referred to a lipid clinic is approximately twice that seen in the general population.⁵ In view of our report we would suggest that secondary causes of hyperlipidaemia should be sought in a lipid clinic. In cases of type III hyperlipoproteinaemia, secondary causes such as obesity, diabetes mellitus and hypothyroidism should always be considered.

Interestingly, the lipid profile considerably improved in the first patient due to treatment of his hypothyroidism with thyroxine in conjunction with dietary measures. However, the second patient required the addition of a fibrate lipid-lowering drug. Finally, the occurrence of hyperlipidaemia in these young patients provides an unusual presentation of primary hypothyroidism which could have resulted in catastrophic sequelae in the long term if not identified.

Final diagnosis

Primary hypothyroidism in two young clinically euthyroid males with type III hyperlipoproteinaemia.

Keywords: hyperlipidaemia; hypothyroidism; hyperlipoproteinaemia

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Extracranial cerebrovascular disease – a management dilemma

A G Speers, S K Das

A 71-year-old man presented to the vascular clinic with a 5-year history of amaurosis fugax in his right eye. He described this as a blanket descending over his visual field, which would come on slowly, lasting approximately 5 minutes before rapidly resolving, and was associated with a feeling of dizziness. Initially, attacks had occurred once every 2 months but recently the frequency had increased to twice a week. The patient was known to suffer from hypertension and hypercholesterolaemia, both of which were controlled medically. Ten years earlier, the patient had undergone an aorto-iliac graft for peripheral vascular disease.

Examination revealed a regular pulse of 76 beats/min and a blood pressure of 136/84 mmHg; a late systolic murmur was detected in the mitral region. The right carotid artery pulse was absent, with a low pitched bruit. A left carotid thrill was detected with a high-pitched murmur. An ophthalmic examination confirmed embolic events in the right eye.

Duplex scan showed a diseased innominate artery with total occlusion of the right common carotid, 75% stenosis of the left internal carotid and occlusion of the left subclavian artery with reversal of flow through the vertebral artery. An aortic arch angiogram (figures 1 and 2) was performed.

Figure 1 Aortic arch angiogram

Figure 2 Aortic arch angiogram

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Questions

- 1 What is the cause of the visual symptoms?
- 2 What does the arteriogram show?
- 3 What is subclavian steal and how would you treat it?
- 4 How can the patient's symptoms be explained?

Answers

QUESTION 1

The patient is suffering from amaurosis fugax or temporary blindness secondary to embolisation within the retinal artery. Of significance in this patient was the fact that he was exhibiting evidence of hypoperfusion (dizziness) which is only seen in 5% of cases of carotid disease.

QUESTION 2

Currently, duplex scan remains the first choice of investigation for anatomical and functional assessment of the arterial tree, as it is non-invasive, reliable and can be repeated, thus it has almost replaced arteriogram in most situations.^{1,2} However, arteriogram is often obtained to confirm the duplex scan findings in complex situations, such as in this case. The arteriogram (figures 1 and 2) in this case confirmed the findings of the duplex scan accurately and demonstrated that the right common carotid is occluded and there is no filling of the right internal or external carotid artery. There is 75% stenosis of the left internal carotid artery and occlusion of the left subclavian artery at its origin. There is distal filling of the left subclavian by reverse flow from the left vertebral artery.

QUESTION 3

Subclavian steal (figure 2) is a phenomenon where the distal blood supply in the subclavian (usually on the left) is derived from a reversal of blood flow from the vertebral to the subclavian artery distal to the occlusion. This occurs because the proximal segment of the subclavian artery has been occluded and therefore the arterial pressure gradient favours blood flow in the reverse direction. It is usually only treated if the patient is symptomatic (clumsiness, dizziness or drop attacks), and is usually brought on by exertion. The main form of treatment is arterial reconstruction of the occluded segment of the subclavian artery or angioplasty or stenting of the occluded segment.

QUESTION 4

The patient's symptoms are the result of embolisation from one of his carotid arteries. The most likely source of the emboli was felt to

be the left internal carotid with emboli traversing the circle of Willis to impact in the right retinal artery. The other possibility was that emboli were being released by the occluded right internal carotid artery, a phenomena known as carotid stump syndrome. Thus, the surgical options available were to either tie off the occluded stump on the right, perform a carotid endarterectomy on the left, or do both.^{3,4}

A carotid endarterectomy was performed on the left, with no complications in the postoperative period. Out-patient follow-up has revealed that the patient's symptoms of embolisation and hypoperfusion have resolved since discharge.

Discussion

This case illustrates the extensive and complex nature of atheromatous disease and that management of these patients often can be difficult. In this category of patient the risk of not operating was that these transient episodes of blindness could lead to an event causing permanent neurological damage. It has been estimated that 20% of patients with amaurosis fugax will suffer from a stroke if untreated.⁵ This has to be weighed against the risk of mortality and stroke from performing a carotid endarterectomy. Mortality from this procedure has been estimated at 1.3% after 30 days for those with asymptomatic disease and 1.8% for those with symptomatic disease with a risk of fatal stroke of 0.47% and 0.91%, respectively.⁶

Attempts have been made to evaluate the effectiveness of anticoagulating this cohort of patients. The general consensus appears that, although anticoagulation reduces the risk of a cerebral event, it does not do this to a significant enough degree.

Final diagnosis

Amaurosis fugax secondary to embolisation within the retinal artery.

Keywords: carotid artery; subclavian steal; amaurosis fugax

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