Self-assessment questions

The uncommon denominator

D O'Driscoll, M Duddy

A 70-year-old man presented with anorexia, altered bowel habit and weight loss. He had a history of heart failure and rheumatoid arthritis. Clinical examination showed rheumatoid deformity and pedal oedema. Laboratory investigations demonstrated microcytic anaemia, eosinophilia and hypoalbuminaemia. Barium enema and contrast-enhanced abdominal computed tomography (CT) were performed (figures 1 and 2, respectively).

Figure 1 (A and B) Barium enema

Figure 2 CT scan

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Questions

- 1 What are the radiological signs on figures 1 and 2 ?
- 2 What is the differential diagnosis for each ?
- 3 Are there any common denominators ?

Answers

QUESTION 1

The barium enema (figure 1) shows loss of the normal haustral pattern and the mucosa appears granular. There is widening of the presacral space (A). The bladder, opacified by prior CT enhancement, is narrowed and elongated (B). On the CT scan (figure 2), the intra-abdominal fat is of similar attenuation to the water density of a simple right renal cyst. The mesenteric and pararenal fat has a misty appearance with thickening of Gerota's fascia.

QUESTION 2

The differential diagnosis for the CT appearances is summarised in box 1.¹ The differential diagnosis for the changes on the barium enema is summarised in box 2.² The correct diagnosis is suggested radiographically by radiating strands of soft-tissue density that surround the central vessels without displacing them. Generalised adenopathy is not seen. Typically a halo of fat surrounds the vessels in the mesentery

QUESTION 3

The common denominators are ulcerative colitis and amyloidosis. Amyloidosis is supported by the clinical history of rheumatoid arthritis.

Outcome

Rectal biopsy showed no evidence of ulcerative colitis or amyloidosis. Laparoscopic biopsy of the mesenteric fat confirmed amyloid deposited in the vessel walls and as rings in the adipose tissue. Further stains confirmed amyloidosis associated with inflammatory disease (AA amyloidosis). Amyloid was also seen in biopsies of the pleura, bladder wall and prostate. Over the next 20 months he became

Differential diagnosis of a 'misty mesentery' adapted from¹

- mesenteric oedema: cirrhosis, heart failure, nephrosis, tricuspid disease, constrictive pericarditis, portal hypertension, portal vein thrombosis, mesenteric artery and vein thrombosis, vasculitis, Budd-Chiari syndrome, inferior vena cava obstruction and trauma
- lymphoedema: congenital anomalies, inflammation, neoplasm, surgery, radiation therapy
- inflammation: pancreatitis, appendicitis, inflammatory bowel disease, diverticulitis, abdominal tuberculosis
- haemorrhage and trauma
- neoplasms: non-Hodgkin's lymphoma, carcinoma of the pancreas, colon, breast or ovary, gastrointestinal stromal tumour, carcinoid, melanoma, leukaemia, mesothelioma, mesenteric dermoid (one third of these lesions will diffusely involve the mesentery)
- idiopathic disorders: mesenteric panniculitis (pathologically lipid-laden macrophages infiltrate throughout the mesentery); amyloidosis

Box 1

Differential diagnosis of complete loss of colonic haustrations with a granular mucosa as in figure 1²

- inflammatory: ulcerative colitis, infection, Crohn's disease, non-specific forms of colitis
- cathartic colon
- scleroderma
- amyloid
- dermatomyositis
- myotonic dystrophy
- Fabry's disease

Box 2

increasingly symptomatic with cardiac failure, became refractive to treatment, and died.

Discussion

The clinical and pathological manifestations of amyloid disease were first reported in the mid-1800s. Amyloidosis is a rare systemic disease caused by extracellular deposition of a protein polysaccharide in various organs leading to hypoxia, mucosal oedema, haemorrhage, ulceration, mucosal atrophy and muscle atrophy. Although it is usually seen in a systemic form, 10–20% of cases can be localised. The localised form of amyloidosis is site-limited and does not progress to systemic involvement. Systemic amyloidosis is sub-classified into an idiopathic primary form and a secondary or reactive form. Patients with primary amyloidosis have no underlying disease. Men are affected more often than women, the mean age at presentation is 55–60 years. Some causes of secondary amyloidosis are multiple myeloma (10–15%), rheumatoid arthritis (20–25%), tuberculosis (50%), or familial Mediterranean fever (26–40%).

Infiltration of the gastrointestinal tract (GIT) with amyloid, as demonstrated by microscopic examination, is not unusual. Symmers reported GIT findings in 70% of autopsy cases of primary amyloidosis,³ while 75% of rectal biopsies were positive for amyloid in a study of 200 patients reported by Blum and Sohar.⁴ Aspiration biopsy of abdominal fat has a sensitivity of 82% and 100% specificity.⁵ Gilat *et al* demonstrated involvement of the digestive tract at autopsy in 68 of 70 cases of systemic amyloidosis.⁶ Amyloid is deposited around small blood vessels in the submucosa and can result in occlusion, ischaemia, and ultimately infarction of bowel wall. Damage to musculature and nervous elements may cause alterations in gut motility.

Patients with GIT involvement are often asymptomatic, but widespread dysfunction may occur. The clinical manifestations are equally non-specific and diverse, including dysphagia, bowel obstruction, diarrhoea, haemorrhage, or simply diffuse abdominal discomfort with weight loss.

The radiographic abnormalities most commonly reported include motor dysfunction, nodular and thickened mucosal folds, ulcerations, mucosal atrophy, malabsorption pattern,

Table X-Ray abnormalities in 20 patients with primary amyloidosis (from⁷)

Site	No of patients	Radiographic findings
Oesophagus	4	Decreased or absent peristalsis
	2	Tertiary contractions with decreased peristalsis
	1	Reflux of barium into the nasopharynx
Stomach	2	Masses or prominent folds
	1	Multiple small ulcers
Small bowel	1	Atrophic folds
	5	Thickened folds
	2	Malabsorption pattern
Colon	1	Dilated loops
	3	Changes suggestive of long-standing ulcerative colitis

hepatosplenomegaly, distinct intraluminal and intramural masses, and obstruction at multiple sites in the GIT. Amyloid fibrils have an affinity for calcium, and radiographically detectable calcified amyloid deposits may occur in both primary and secondary amyloidosis. Carlson *et*

al reviewed 230 cases of primary systemic amyloidosis seen at the Mayo Clinic over a 10-year period. In 47 patients who had amyloidosis diagnosed from a GIT biopsy site or had GIT involvement at autopsy, 20 had abnormal radiological reports (summarised in the table).⁷

CT and magnetic resonance imaging findings are non-specific, often mimicking both inflammatory and neoplastic processes. GIT involvement with amyloidosis is common. As radiological findings, when positive, are non-specific, definitive diagnosis depends on histology.

Final diagnosis

Mesenteric amyloidosis secondary to rheumatoid arthritis.

Keywords: amyloidosis; rheumatoid arthritis

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Difficulties in the diagnosis of an intra-abdominal mass

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The discovery of an intra-abdominal mass often poses significant diagnostic difficulties. The following case demonstrates this, and highlights some of the pitfalls.

Case report

A 73-year-old man presented with a 2-year history of lower limb paraesthesiae, macrocytosis and a recent onset of left upper quadrant pain. Examination revealed a large, slightly tender, smooth mass arising from the left upper quadrant of the abdomen with the lower edge in the left iliac fossa. Laboratory investigation, including the assessment of catecholamine levels, was non-contributory.

Questions

- 1 What investigations might help to determine the nature of the mass?

2 What is the probable diagnosis and what other imaging techniques might be useful?

3 What is the nature and presumed aetiology of these lesions?

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Answers

QUESTION 1

Grey-scale ultrasound examination is a useful primary investigation in determining the nature of discrete intra-abdominal masses, yielding information such as the size of the lesion and determining the mass to be solid or cystic. The structure from which the mass originates may be identified. In many cases ultrasound characteristics may be diagnostic.

In this case ultrasound was misleading, suggesting an enlarged spleen containing a partly sub-capsular/partly sub-splenic collection, with displacement of the kidney into the pelvis. In most cases involving an abdominal mass computed tomography (CT) and magnetic resonance imaging (MRI) are adequate to enable a confident diagnosis.¹ However, CT also suggested massive splenic enlargement as the cause of this lesion.

A gadolinium-enhanced MRI scan (see figure) demonstrated a 20-cm mainly cystic retroperitoneal mass displacing the spleen superiorly, the left kidney inferiorly, the tail of the pancreas anteriorly, but arising from none of these. The left adrenal gland could not be identified and was thus considered to be the probable site of origin of the mass.

QUESTION 2

The probable diagnosis, confirmed subsequently after excision, is of an adrenal pseudocyst. Pre-operative diagnosis of adrenal pseudocysts can be problematic, though a number of imaging modalities are helpful. Plain radiographs may show curvilinear calcification above the renal shadow²; speckled calcification suggests malignancy. Excretion urography may show downward displacement and rotation of the kidney, suggesting an often incidental suprarenal mass.³ Usually, particularly in the case of non-functioning adrenal lesions, CT and MRI are adequate. Both will demonstrate a mass with a thick wall and/or prominent sep-

tations, which may contain calcification. MRI has the advantage of being able to image in any anatomic plane, hence permitting evaluation of the relationship of the mass with adjacent retroperitoneal structures. This was vital in this case in making the diagnosis before operation. Pitfalls have been reported with both, however. Repeated haemorrhage with resultant multiple central punctate calcifications has caused the CT appearances of a pseudocyst to mimic those of a hydatid cyst.⁴ Unusual appearances at ultrasound, CT, and MRI have also caused benign lesions to be diagnosed as malignant.⁵⁻⁶ In such cases angiography and adrenal venography, showing the characteristic arching of venules over and around an avascular adrenal mass, may be helpful.²

QUESTION 3

Adrenal pseudocysts account for 39% of non-neoplastic adrenal cysts. They are cystic lesions with neither an epithelial nor an endothelial lining, thus precluding a more precise classification. Initial reports were of small cysts found incidentally at post mortem, though improved imaging techniques are resulting in a higher reported incidence.⁷ Most (75%) occur in women, with some suggesting them to be more common in the young and middle-aged. The cyst found here is one of the largest reported, the largest being 33 cm, removed from a West Indian woman.⁸

The aetiology of adrenal pseudocysts has been the subject of considerable debate. Studies suggest that the majority are of vascular origin,⁹ due to haemorrhage from a pre-existing vascular anomaly. Organisation of the haematoma results in the formation of a thick-walled pseudocyst. This is borne out by the histological analysis of the cyst reported here. The specimen was 21 cm in diameter, sections confirming a fibrous capsule without endo- or epithelium. The adrenal gland within the specimen was divided into nodules by bands of acellular collagen. A large blood vessel with an abnormal muscle wall was identified, one section of which showed haemorrhage. Haemorrhage in the absence of a vascular anomaly may occur due to crushing injury, birth trauma, or in systemic illnesses such as severe shock, endotoxaemia or haemorrhagic diatheses. Pseudocysts may also develop after bleeding into adrenal tumours. Thus the diagnosis of malignancy must be considered in all cases of pseudocyst, and exclusion of malignancy may, in the absence of symptoms, be the sole indication for surgical excision.

Final diagnosis

An adrenal pseudocyst.

Keywords: adrenal lesion; pseudocyst

Figure MRI scan demonstrating the large intra-abdominal cystic mass

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Lung mass in a short woman

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A 26-year-old woman presented with a 6-week history of a dry cough and several recent episodes of scant haemoptysis. She admitted to mild exertional dyspnoea, night sweats and right-sided anterior pleuritic chest pain. A 20 lb weight loss was noted. She smoked 10 cigarettes daily (4 pack-years in total). She had been diagnosed as having Swyer's syndrome (a mosaic variant of Turner's syndrome with a Y chromosome present) at the age of 20 years following investigation for oligomenorrhoea. She had had a gonadectomy performed 4 years earlier and a skin biopsy confirmed a 45XO/46XY karyotype. She subsequently attended a Turner's syndrome clinic and received vitamin D preparations for a low bone mineral density. On admission vital signs were normal with a temperature of 37.3°C. She measured 152 cm and had a Turner's syndrome habitus with partial neck webbing, poor breast development and several naevi on the chest wall. She had poor dentition and halitosis. Two small lymph nodes were present in the right supraclavicular fossa. Air entry was reduced in the right hemithorax anteriorly, but no rales were audible. Laboratory data included a white blood cell count of $23.7 \times 10^9/l$ with a neutrophilia, slightly reduced serum albumin of 31 g/l, elevated globulins of 50 g/l and an erythrocyte sedimentation rate of 95 mm in the first hour. CA-125 was slightly elevated at 37 IU (normal 0–25). Her chest radiograph and thoracic computed tomography (CT) scan are shown in figures 1 and 2, respectively.

Figure 1 Chest X-ray

Figure 2 Thoracic CT scan

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Questions

- 1 What is the most probable diagnosis?
- 2 What diagnostic intervention would you undertake?

Answers

QUESTION 1

The most probable diagnosis is right middle lobe consolidation/lung abscess secondary to dental caries. The X-ray (figure 1) demonstrates a large mass involving the right middle lobe. The thoracic CT scan (figure 2) shows consolidation and cavitation of the right middle lobe with reactive lymphadenopathy of the pre-carinal region.

QUESTION 2

Fibre-optic bronchoscopy is indicated. This procedure revealed an inflamed right middle lobe orifice with impacted purulent secretions. Aspiration and lavage removed the mucus plug and released a large amount of foul-smelling secretions which grew *Streptococcus milleri* when cultured. The patient was treated with physiotherapy and appropriate antibiotics and made a full recovery. She was referred for dental treatment.

Discussion

Aspiration from the oropharynx is the most common cause of lung abscess, and is often associated with dental caries.^{1,2} *Streptococcus milleri* is probably the most common microbe found in lung abscesses. The foul smell of the secretions suggests anaerobic infection although none was cultured (a mixed growth is particularly common). The wide differential diagnosis of a cavitating lesion on a chest film is well described, but the age of the subject makes an infective process the most likely scenario.

The history of weight loss associated with the evidence of an inflammatory process should always raise the possibility of an abscess, even if there is no evidence of cavitation on the plain chest film (as in this case). Occasionally, a lesion appears solid on the plain film but cavitation is only visualised on CT scanning. The obstruction of the right middle lobe orifice makes the diagnosis more difficult because of the lack of sputum production or signs of consolidation.

The history of Swyer's syndrome is a 'red herring' in that there is no known risk factor for the development of a lung abscess in this condition. Swyer first described the condition in 1955 as a condition of pure gonadal dysgenesis with the karyotype 46XY.³ Turner's syndrome represents gonadal dysgenesis in its classical form but many chromosomal variants and mosaics are known, of which Swyer's syndrome is just one. These phenotypic females are at significant risk of developing gonadal tumours if there is a Y chromosome or a fragment of one present. Gonadoblastoma, dysgerminoma, and embryonal carcinoma have an incidence of at least 25% in XY karyotypes with the risk increasing with age.⁴⁻⁶

Malignancy is a possible diagnosis in this female given the history of palpable lymphadenopathy and CT visualised pre-carinal nodes, but a primary lung carcinoma would be exceptionally rare in this age group. A gonadal or mesenchymal tumour would be a distinct possibility, although the gonadal tissue had previously been surgically removed as a precaution.

The role of the bronchoscopy in this situation is to obtain bacteriological cultures whilst excluding inhaled foreign bodies and tumour obstruction. Bronchoscopy has also been used as a means to enter or place a catheter into an abscess cavity and using suction,⁷ although CT-guided procedures are now the treatment of choice.

In summary, this case demonstrates the risk of lung abscess formation secondary to poor dentition and highlights the use of the CT scan to show cavitation when the plain chest film does not. The trap in this history is looking for a more exotic condition in a rare syndrome.

Final diagnosis

Right middle lobe consolidation/lung abscess secondary to dental caries.

Keywords: lung abscess; dental caries; Swyer's syndrome.

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A child with multiple bony swellings

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A 5-year-old boy presented with multiple bony swellings around his knees, wrists, shoulders, and rib cage. He was in good general health and there was no history of trauma, but his father was concerned about increasing deformity of the child's knees and the gradual increase in size of the swellings around his wrists, chest and knees. On examination, there were bilateral multiple fixed bony outgrowths from the distal radius, medial side of lower femur, upper end of humerus and costochondral junctions. The child had mild genu valgum on the left side. There was no deformity or swelling of the fingers. Movements at the shoulder, hip and knee were within normal limits and there was no neurovascular problem. The father had similar bony swellings around his knees and shoulder but was not bothered about these because they were not growing and he had no functional problems. Plain X-rays of the knees, wrists and chest of the patient and a photograph of him and his father are shown in figures 1-4.

Figure 1 X-Ray of both knees

Figure 2 X-Ray of both wrists

Figure 3 X-Ray of the chest

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Figure 4 Photograph of the child and his father

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Questions

- 1 What are the radiological and clinical findings?
- 2 What is the diagnosis?
- 3 What are the prognosis and treatment?

Answers

QUESTION 1

The X-ray of the knees (figure 1) shows bony outgrowths from the lower femoral metaphyses mainly on the medial aspect. The wrist X-ray shows similar outgrowths involving the distal metaphyses of radius and ulna in both hands. There is also some shortening of the right third metacarpal with broadening of the distal metaphysis. Chest X-ray reveals bilateral outgrowths from the proximal humeral metaphyses with slight broadening of the anterior ends of the lower ribs on the left side.

The clinical photograph of the child shows left-sided genu valgum with diffuse swelling over the lower thighs. There are multiple bony outgrowths at costochondral junctions. The photograph of the father shows bilateral multiple bony growths around the knees and right shoulder.

QUESTION 2

The clinical presentation and the radiological findings are typical of osteochondromatosis, also known as hereditary multiple exostoses, diaphyseal aclasis, and metaphyseal aclasis. The disease should be differentiated from solitary osteochondroma, myositis ossificans and multiple enchondromatosis, the latter involving mainly the hand bones.

QUESTION 3

Most of the time the lesions are asymptomatic and do not affect the general health of the patient. Disability may be produced if the growth is near the joint, interfering with joint movements or irritating the tendon or muscle belly.

Neurological symptoms may be produced by mechanical pressure, especially in vertebral lesions. Rarely, the osteochondroma may become painful because of fracture of the stalk. Malignant transformation to chondrosarcoma is possible, although rare. Surgical treatment is indicated if the tumour is symptomatic or if there is suspicion of malignancy.

Discussion

Osteochondroma is a benign developmental growth defect involving the metaphyseal area of long bones, producing a bony outgrowth with a cartilage cap. It is the commonest benign bone tumour.¹ Osteochondromatosis has a marked hereditary tendency with autosomal dominant inheritance.² The penetrance is almost 100%

Learning points

- osteochondroma is the commonest benign bone tumour
- multiple osteochondroma (osteochondromatosis) differs from solitary lesions in mode of inheritance and chance of malignant transformation
- surgical excision is required for symptomatic and possibly malignant growths

by the age of 12 years.³ In this case, both the boy and his father had multiple osteochondroma. This disorder is usually detected in early childhood around the age of 8-10 years. Long bones are most commonly involved. The majority of patients are asymptomatic and present with fixed bony lumps around the joints. The tumour grows with the growth of the patient and any increase in size of the lesion after skeletal maturity is suggestive of possible malignant transformation.⁴

Osteochondroma has typical radiographic features. The growth may be sessile or pedunculated. Characteristically, the cortex of the lesion is continuous with the cortex of the host bone. The actual size of the growth is greater than the radiological size because of the cartilage cap over the bony outgrowth. The bone harbouring the tumour may be shortened, causing deformity or limb length discrepancy.^{1,2}

If the tumour is symptomatic it should be excised. Excision is done through the base of the tumour along with the cartilage and the perichondrium. The chances of recurrence are greater if the perichondrium is removed incompletely. The exact figure for malignant transformation of the growth is not known but figures usually quoted are 1% for solitary lesions and 6% for multiple lesions.⁵

The relative frequency of chondrosarcoma is greater in central lesions involving the pelvis, shoulder girdle, or spine.³ Malignant transformation should also be suspected if the lesion suddenly becomes symptomatic or begins to grow rapidly. If there is any suspicion of malignancy, the lesion should be excised.

Final diagnosis

Hereditary multiple exostoses.

Keywords: exostoses; bony swellings; chondrosarcoma; osteochondromatosis

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Diarrhoea, fever, shock and bullous skin lesions after ingestion of raw oysters

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A 38-year-old man presented to the emergency department with a 2-day history of cramping abdominal pain, severe vomiting, diarrhoea, and fever with chills. He reported eating a large quantity of raw oysters at a local restaurant, one day prior to the onset of symptoms. The patient had a significant history of daily alcohol consumption (12 cans of beer and one bottle of wine) for many years. On examination, the patient appeared toxic, with a systolic blood pressure of 90 mmHg, pulse of 116 beats/min, respiration rate of 22 breaths/min and an oral temperature of 39°C. Right basal crackles were present on auscultation of the chest. Cardiovascular examination showed tachycardia with normal heart sounds. Abdominal examination revealed a tense abdomen without evidence of ascites, with diffuse tenderness and hyperactive bowel sounds. The patient's skeletal muscles were tender to palpation and movement. Joint examination revealed no evidence of synovitis. A few hours after admission, multiple 1–2 cm skin lesions were noted predominantly on the truncal area (figure). Over the next 24 hours, these evolved into haemorrhagic bullae with purpuric centres.

Laboratory findings were significant for severe leucopenia (1000 cells/mm³), hypoalbuminaemia (1.7 g/dl), abnormal liver function tests (total bilirubin 5.3 g/dl; lactate dehydrogenase 1544 IU/l; aspartate transaminase 615 IU/l; alanine transaminase 465 IU/l), elevated creatine phosphokinase 14 600 IU/l, prolonged prothrombin time 15.9 s, INR 1.6, and lactacidaemia (4.6 mEq/l). Empiric antibiotics using a third generation cephalosporin and doxycycline were started immediately after blood cultures were drawn. However, within 8 hours of admission the patient became diaphoretic, tachypnoeic, hypoxic, and his mental status deteriorated.

Figure Skin showing early target lesions and bullae over the truncal area

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Questions

- 1 What is the diagnosis and what are the predisposing factors?
- 2 What is the treatment and prognosis?

Answers

QUESTION 1

This patient had *Vibrio vulnificus* septicaemia after ingestion of contaminated raw shell-fish. Individuals with pre-existing liver disease are at 80 times greater risk for illness and at over 200 times greater risk of death from *V. vulnificus* oyster-associated infection.¹ Patients with cirrhosis of the liver, haemochromatosis and immunocompromised states are especially susceptible.

QUESTION 2

Current recommendations include intravenous administration of doxycycline (100 mg q 12 h) and ceftazidime (2.0 g q 8 h). Early and aggressive treatment is recommended, as the case fatality rate for patients with septicaemia has been shown to increase with greater delays between illness onset and initiation of antibiotic treatment.² Fatality rates exceed 50% and are greater than 90% in patients who develop shock, even with appropriate treatment.^{1,3}

Outcome

One day after admission, blood cultures grew *V. vulnificus*, although stool cultures were negative. The organism was sensitive to all antibiotics tested, including third generation cephalosporins, tetracycline and gentamicin. Despite immediate resuscitation with intravenous fluids and appropriate antibiotics, the patient rapidly developed fulminant septicaemia, with refractory hypotension requiring vasopressors. He subsequently developed adult respiratory distress syndrome, requiring mechanical ventilation. The patient died on day 16 after a complicated hospital course.

Discussion

V. vulnificus, a halophilic, lactose-fermenting, marine organism, is known to cause two distinct clinical syndromes. The first is primary bacteraemia with secondary seeding of the soft tissues. This usually occurs in patients with chronic liver disease and a history of recent ingestion of raw oysters. The disease is rapid in onset with high fever, chills and shock as well as haemorrhagic bullous skin lesions. The second syndrome is characterised by primary wound infection after exposure to sea-water.³ The organism is aptly named 'vulnificus' (Latin for 'wounding'), since it may cause extensive soft-tissue destruction.³

V. vulnificus is known to inhabit coastal waters and estuaries throughout the world. This bacteria is found in sea-water as well as contaminated sea-food, particularly oysters, fish, shell-fish and crustaceans. Like other vibrios, *V. vulnificus* is concentrated in filter feeders, such as oysters.³ Studies have found that more than 50% of the oyster lots sampled in the US contain *V. vulnificus*. Infections are seasonal, with the peak onset of the illness from April to October in the Gulf Coast areas of the North American continent.⁴ *V. vulnificus* infec-

Learning points

- *Vibrio vulnificus* infection is usually seen in coastal areas, but can occur anywhere with ingestion of raw sea-food, particularly oysters
- it can cause serious and fatal infection in people with chronic liver disease
- fever, shock and bullous skin lesions should raise suspicion for the diagnosis
- fatality rates exceed 50% and are more than 90% in patients who develop shock
- early institution of appropriate antibiotics and surgical debridement can decrease mortality
- prevention relies upon educating patients and thorough cooking of sea-food

tions occur most commonly in persons exposed to sea-water along the Gulf of Mexico and the Southern Atlantic and Pacific coasts. Infections have also originated from other American coastal waters and from Europe, Asia, Australia and South America.⁵ A water temperature above 20°C and a saline content of 0.7–1.6‰ is required for colonisation.⁶

The organism is the most virulent of the vibrios, which may account for the high mortality in infected patients. The presence of a polysaccharide capsule may increase the organism's resistance to phagocytosis and to the bactericidal activity of human serum. *V. vulnificus* also produces a cytotoxin–haemolysin, collagenase, phospholipases and a protease that lyses elastin, thus increasing tissue penetration.³ High frequency of infection is seen in elderly men with an underlying liver disease. This is especially true of patients with cirrhosis and haemochromatosis who have an elevated serum iron concentration.⁷ Iron is essential for bacterial growth, and the ability to obtain iron from the host is essential for pathogenicity.⁸

This patient had primary bacteraemia with secondary seeding of the soft tissues as a consequence of ingesting contaminated raw oysters. The clinical course was characteristic with rapid onset of high fevers, chills and shock as well as the development of haemorrhagic bullae. He also developed myositis which is often seen with this infection. This case displayed many of the classic features seen in primary *V. vulnificus* septicaemia and had a fatal outcome.

Early treatment with antibiotics, debridement and amputation when necessary may improve survival. The duration of the antibiotic therapy depends on the clinical response of the patient. Surgical debridement and good wound care facilitate the healing of the necrotic lesions.¹

Final diagnosis

Fatal *Vibrio vulnificus* septicaemia after ingestion of raw oysters.

Keywords: *Vibrio vulnificus*; septicaemia; oysters; poisoning

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A rare but important cause of focal hepatic lesions

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A 57-year-old man presented with a 3-month history of right hypochondrial pain which was a constant dull ache often radiating to the back. There was no history of nausea, vomiting, gastro-intestinal bleeding, jaundice or weight loss. Clinical examination was unremarkable apart from hepatomegaly; the liver was felt 2 cm below the right costal margin with a vertical span of 10 cm and was smooth, firm and non-tender. There were no signs of chronic liver disease. A basic haematological and biochemical screen was normal, with the exception of an isolated rise in γ -glutamyl transpeptidase (γ -GT) at 104 IU/l (normal 10–50 IU/l). He had not taken any hepatotoxic or enzyme-inducing drugs. His admitted alcohol intake was less than 15 units per week. An abdominal ultrasound (figure 1) and computed tomography (CT) (figure 2) were done. A liver biopsy was done under ultrasound guidance (figure 3). The patient was well after 6 months and had improved with symptomatic treatment.

Figure 1 Abdominal ultrasound

Figure 2 Abdominal CT scan

Figure 3 Liver histology

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Questions

- 1 What are the findings on the abdominal ultrasound?
- 2 What are the findings on the abdominal CT scan?
- 3 What does the liver biopsy show?

Answers

QUESTION 1

The abdominal ultrasound (figure 1) shows abnormal liver parenchyma with multiple hyperechoic areas.

QUESTION 2

The abdominal CT scan (figure 2) shows multiple low attenuated areas in the liver. The appearances on the ultrasound and CT can easily be mistaken for multiple abscesses or metastases.

QUESTION 3

Liver biopsy shows dilated bile ducts lined by attenuated bland epithelium. Inspissated bile can be seen in one bile duct. These findings are consistent with bile duct microhamartomas or von Meyenburg complexes (VMC).

Discussion

Bile duct microhamartomas were described by von Meyenburg in 1918.¹ They are small, usually multiple, greyish hepatic nodules and result from the maldevelopment of ductal plates, thus explaining the similarities or association with congenital hepatic fibrosis, hepatic and choledochal cysts, polycystic kidneys, medullary sponge kidney, Caroli's disease, etc.² The persistence or lack of remodeling of embryonic duct plates is an essential precursor of this lesion. Histologically, they are characterised by cystically dilated bile ducts lying in a collagenous stroma. The bile duct structures are lined by regular cuboidal epithelium and may contain bile plugs.³

VMC do not usually produce symptoms or abnormalities in liver function tests. The isolated rise in γ -GT in our patient, in the absence of an attributable cause such as alcohol, drugs, etc, can be regarded as non-specific. It is well known that sometimes even very high values of γ -GT can be seen without an obvious cause. The diagnosis of VMC is usually made at autopsy as an incidental finding. The lesions are not uncommon with a prevalence ranging from 0.7% to 2.8% in different autopsy series.³

The increasing use of ultrasound and CT has led to a rise in the diagnosis of VMC. The ultrasound appearance of VMC has been described as multiple hyper- or hypo-echoic areas.³ The CT appearance consists of multiple, irregular, small, low attenuated areas that do not normally enhance on contrast injection,⁴ although exceptions have been reported.⁵ These appearances are likely to be mistaken particularly for hepatic abscesses or metastases. On magnetic resonance imaging (MRI), bile duct hamartomas are hypointense

von Meyenburg complexes

- von Meyenburg complexes are hepatic microhamartomas and are a part of a family of hepatic and renal fibropolycystic disorders
- they are a relatively uncommon incidental finding and do not normally cause symptoms or disturbances in liver function
- the histological appearances are specific, consisting of dilated bile ducts lined by attenuated epithelium with inspissated bile
- abdominal ultrasound, CT, and MRI are used increasingly for diagnosis

on T1-weighted spin echo sequences and hyperintense on T2-weighted ones. Dynamic MRI (Gd-DTPA) displays a slow perfusion pattern in the nodules.⁵

It is probable that VMC will be more frequently recognised as ultrasound and CT are increasingly used in the diagnosis of abdominal diseases. Therefore, VMC should be included in the differential diagnosis of multiple hyper- or hypo-echoic lesions on radiological investigations, particularly if these lesions are seen in patients with otherwise good health and normal liver function tests. Interestingly, Lef-Toaff *et al*⁶ reported 18 patients with primary extra-hepatic malignancy who had liver biopsy for evaluation of possible metastases which showed biliary hamartomas without any evidence of metastatic disease. The spectrum of findings varied from one or two circumscribed lesions to innumerable lesions of varying sizes from 2–15 mm.⁶ The final diagnosis, however, must be confirmed by liver histology. VMC can very rarely produce portal hypertension but these cases cannot be differentiated from congenital hepatic fibrosis.³ Some cases of cholangiocarcinoma have been reported in patients with VMC, suggesting that malignant change is possible, but this is very unusual.^{3,7}

In summary, VMC are normally asymptomatic, and are a chance finding at autopsy or during investigation for another disorder. They are a part of a family of fibropolycystic diseases and the diagnosis rests on histological confirmation.

Final diagnosis

von Meyenburg complexes or bile duct microhamartomas.

Keywords: bile duct microhamartomas; von Meyenburg complexes; hepatic abscesses; metastatic liver disease

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Acute abdominal pain following anticoagulation

I S Cook, K Amar

An 87-year-old woman was admitted with a 2-month history of worsening shortness of breath. She had a non-productive cough and denied any history of chest pain. She had a history of pulmonary tuberculosis, diverticular disease, and a ventral suspension. Respiratory function testing identified severe airways obstruction and she was commenced on inhaled bronchodilators to good effect. Arterial blood gas examination on air revealed pH 7.44, pCO₂ 6.7 and pO₂ 9.0. Full blood count, urea and electrolytes, and liver function tests were all normal. Two days after admission she complained of left pleuritic chest pain although with no increase in breathlessness. Repeat arterial blood gas examination showed pH 7.38, pCO₂ 5.9 and pO₂ 7.7. A diagnosis of possible pulmonary embolism was made and she was anticoagulated with intravenous heparin. The following morning she complained of severe left lower abdominal pain and nausea. Photographs of her abdomen are shown in figure 1. On examination she was tachycardic and had tenderness over the left lower quadrant where a 15 × 20 cm mass was felt. An abdominal X-ray showed no evidence of intestinal obstruction and there was no sign of air under the diaphragm on chest X-ray. Blood investigations revealed haemoglobin 7.9 g/dl and an activated partial thromboplastin time ratio of 4.63 (ideal therapeutic range 1–2.5). Platelets were normal. She had no history of trauma. Ultrasound examination was carried out and a computed tomography (CT) scan made (figures 2 and 3).

Figure 1 Photographs of the patient's abdomen (reproduced with her permission)

Figure 2 Ultrasound findings

Figure 3 CT appearance

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Questions

- 1 Describe the appearances of the patient's abdomen in figure 1.
- 2 Describe the radiological appearances in figures 2 and 3.
- 3 What is the cause of the patient's abdominal pain?

Answers

QUESTION 1

There is blue/red discolouration of the skin around the umbilicus, anterior abdominal wall scar from previous surgery and on the flank. These are Cullen's and Grey Turner's signs. The commonest causes of these signs are shown in box 1.

Causes of Cullen's / Grey Turner's sign

- haemorrhagic pancreatitis
- retroperitoneal haematoma
- abdominal wall haematoma
- leaking abdominal aortic aneurysm
- ruptured ectopic pregnancy
- ruptured kidney

Box 1

QUESTION 2

The ultrasound examination shows a large mixed echogenic collection within the anterior abdominal wall. CT scan shows an extensive anterior abdominal wall collection principally involving the left rectus abdominis. There is no abdominal, pelvic or retroperitoneal pathology.

QUESTION 3

The patient has developed a spontaneous haematoma affecting the anterior abdominal wall.

Discussion

Intravenous heparin is commonly used within a hospital setting for patients requiring systemic anticoagulation. However, it has many potential complications (box 2), some of which can be catastrophic.¹ Excessive bleeding following anticoagulation is one of the commonest and most serious of these.

Complications of anticoagulation with heparin

- excessive bleeding
- skin necrosis
- heparin-induced thrombocytopenia and thrombosis
- hypersensitivity reactions

After long-term use:

- osteoporosis
- alopecia

Box 2

Abdominal wall haematomas are uncommon although when they do occur usually follow trauma or anticoagulation.^{2,3} They are visible on ultrasonography but CT is often required to clarify or confirm the findings as they may be confused for other conditions such as tumours or abscesses. One case has been reported where acute abdominal pain in a patient anticoagulated with dicoumarol was thought on ultrasonography and CT to be secondary to a large pelvic cystic tumour, possibly of ovarian origin, invading the abdominal wall. On laparotomy, however, a large abdominal wall and retroperitoneal haematoma penetrating into the free abdomen was found.⁴

In the case we have presented above, the patient had a history of diverticular disease and CT was required to confirm that this was a simple abdominal wall collection and not, for example, an abscess associated with bowel pathology. The majority of abdominal wall haematomas are managed conservatively³ and so accurate diagnosis is necessary.

Final diagnosis

Abdominal wall haematoma following anticoagulation with heparin.

Keywords: haematoma; abdominal wall; anticoagulation; heparin

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