A large pelvic arteriovenous malformation in an adult patient with cystic fibrosis

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Summary
We present a prepubertal male cystic fibrosis patient with high circulating oestrogen levels (as a consequence of severe cystic-fibrosis-related hepatobiliary disease) who subsequently developed a large pelvic arteriovenous malformation. This has not previously been described in patients with cystic fibrosis, despite the association between high oestrogen levels and arteriovenous malformations. The aetiology and treatment options of arteriovenous malformations are discussed.

Keywords: arteriovenous malformation; cystic fibrosis

Although life expectancy in cystic fibrosis (CF) continues to improve, most patients still have considerable morbidity from multiorgan disease. Vascular abnormalities are well known in CF, and usually involve the bronchopulmonary or portal circulation. It is also recognised that arteriovenous malformations (AVMs) occur more frequently where there are high levels of circulating oestrogens, a state which can occur in CF patients with advanced liver disease. Despite this, AVMs have not previously been described in CF patients. We present a case of a pelvic AVM developing in an 18-year-old CF patient with delayed puberty and severe respiratory and hepatobiliary disease. The pathogenesis, diagnosis, and treatment of these lesions are discussed.

Case report

An 18-year-old male CF patient had secondary biliary cirrhosis noted aged 8 years, and subsequently developed oesophageal varices. Following this he had several upper gut bleeds and multiple courses of sclerotherapy. By the age of 14 years his main problem was symptomatic massive splenomegaly which was corrected by splenectomy. Male hormone treatment (Sustenon 250) was commenced for delayed puberty (no pubic hair and Tanner stage II genitalia) at 16 years, given by intramuscular deltoid injection monthly for six doses. Despite this, he still has no signs of secondary sexual development and recent assay of his sex hormones reveal him to have very low luteinising hormone, follicle-stimulating hormone, and testosterone levels, in keeping with delayed puberty. Furthermore, he had a normal serum zinc level but raised oestradiol level secondary to his severe liver disease.

At 18 years a painful lump appeared on his left buttock and examination revealed a large firm swelling affecting the whole of the left buttock and to a lesser extent the right buttock and lumbar region (figure 1). The swelling was non-pulsatile, warm, mildly tender, and not fixed to the overlying skin. Rectal examination revealed no evidence of haemorrhoidal varices. Computed tomography (CT) showed a large vascular lesion involving the gluteal muscles, the skin and subcutaneous tissues (figure 2). Angiography demonstrated extensive arteriovenous communications within the pelvic and gluteal areas fed by both internal iliac systems and a large median sacral artery.

Discussion
Arteriovenous malformations can be either congenital or acquired. In the primitive mesen-
chyme interlacing blood spaces are partly absorbed and coalesce, and separate venous and arterial conduits appear around the capillary network. Developmental arrest or misdirection causes formation of communicating channels between mature arteries and veins, associated with the appearance of supernumerary branches due to overgrowth of vascular elements. Increase in size of congenital AVMs is due to engorgement and ageing of the component elements. Acquired AVMs are rarer and may be due to trauma: Natali et al. described a woman who developed a pelvic AVM a year after trauma to her buttock. However, some authors believe that trauma simply calls to attention congenital lesions.

The majority of AVMs involve the extremities, head and neck, and lungs. Pelvic AVMs are very rare (< 2% of the total); there are less than 60 cases described in the world literature. Most are diagnosed in the second and third decades of life. Symptoms depend on the size and site of the lesion: pelvic lesions may grow to a large size before they occur. Paradoxically, systemic haemodynamic effects rarely occur because communications are multiple, tortuous, and narrow, maintaining peripheral vascular resistance. High output cardiac failure has been observed only in very bulky lesions and in pelvic lesions during pregnancy.

Angiographic investigation demonstrates the flow characteristics of the lesion, feeding arteries, draining veins, and their relationship to the normal circulation. Magnetic resonance imaging or CT will reveal the extent of the lesion. Treatment requires a team approach and a combination of pre-operative embolisation and surgery is advocated, with complete excision of isolated lesions resulting in the best cure rates. Recurrence is common if there is simply ligation of feeding vessels. However, asymptomatic static lesions can be monitored and may not require active treatment.

Two-thirds of AVMs occur in women and there is a relationship to parity, suggesting an effect of fluctuating oestrogen and progesterone levels on the vascular wall, and oophorectomy may lead to regression of an AVM. Despite the association between AVMs and high oestrogen levels, oestrogen blocking agents have not been used to treat these lesions.

Our patient did not have zinc deficiency which has been associated with delayed puberty in CF, but did have severe liver disease which has been associated with delayed puberty and high oestrogen levels, which may have contributed to the condition. To our knowledge there have been no previous case reports of AVMs in patients with CF. As survival prospects for patients with CF continue to improve, there will be more individuals with severe CF-related liver disease and therefore it is possible that more of these unusual complications will occur within this patient group. In our patient, a combination of embolisation with surgical resection and plastic reconstruction of the buttocks would be required. Indications for this would be haemorrhage, heart failure, skin breakdown, or pain intolerable to the patient. Since he has many concomitant problems which limit his survival prospects, surgery would be a hazardous undertaking and therefore a conservative approach has been adopted.

Management of pelvic AVMs

**Investigations**
- angiography
- CT/MRI

**Treatment**
- asymptomatic non-enlarging lesions: monitor only
- indications for intervention: relative (pain, functional impairment) or absolute (haemorrhage, high output cardiac failure)

**Intervention options**
- percutaneous arterial embolisation
- surgical devascularisation ± excision of mass
- percutaneous arterial embolisation to diminish size of AVM then surgical excision

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Intractable oesophageal variceal bleeding caused by splenic arteriovenous fistula: treatment by transcatheter arterial embolization

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Summary

We describe a rare case of splenic arteriovenous fistula and venous aneurysm which developed after splenectomy in a 40-year-old woman who presented with epigastralgia, watery diarrhoea, repeated haematemesis and melena caused by hyperkinetic status of the portal system and bleeding of oesophageal varices. It was diagnosed by computed tomography and angiography, and obliterated with giant Gianturco steel coils.

Keywords: splenic arteriovenous fistula; gastrointestinal bleeding; transcatheter arterial embolization

Splenic arteriovenous fistula (SAVF) and splenic venous aneurysm after splenectomy are extremely rare. To our knowledge, only five cases have been reported in the English literature.1–5 SAVF is unusual but curable. It may cause portal hypertension. The hyperkinetic state of flow through SAVF may cause gastrointestinal bleeding, diarrhoea, ascites, or life-threatening heart failure. Elimination of SAVF by transcatheter arterial embolization has been advocated as an alternative to surgical resection. We present a patient with SAVF who presented unusually with epigastralgia associated with variceal bleeding.

Case report

A 40-year-old woman complained of intermittent epigastric pain associated with watery diarrhoea, haematemesis and melaena. She had undergone splenectomy 17 years earlier due to splenic rupture after a motor vehicle accident. Endoscopy performed at a local hospital revealed variceal bleeding in the lower third of the oesophagus. She was transferred to our hospital. Endoscopic variceal ligation was performed several times within a month. A grade II systolic murmur was heard over the left lower chest. The abdomen was soft and tender. There was no ascites. Liver was not palpable. Laboratory examination showed haemoglobin 10.5 g/dl, haematocrit 29.1%, leucocytes 11.6 × 10^9/l and platelets 76 × 10^9/l. Liver function tests were normal. Computed tomography (CT) scan revealed an engorged splenic artery, 1 cm in diameter, and a fusiform dilated splenic vein, 5 × 6 × 9 cm in size (figure 1). A fistula between the splenic artery and vein was tentatively diagnosed.

Because of continuing melaena, embolization of the splenic artery was performed using eight giant Gianturco steel coils. Initially, a coil with a diameter of 10 mm and a length of 10 cm was used because the splenic artery measured around 10 mm in diameter on CT images. Subsequently, a smaller coil (5 mm × 5 cm) was used to occlude the lumen of the 10 mm × 10 cm coil. This was not successful because of hyperkinetic flow, and the smaller coil floated into the aneurysm. Finally, for further thorough embolization, six larger coils (10 cm × 15 mm) were deployed into the splenic artery to establish a complete meshwork. After the embolization by coils, Gelfoam pledgets were added to the coil tangle for further closure of the fistula (figure 3). The bruit in the left lower chest was absent after embolization. The

Figure 1 Serial CT images (from left to right) show an engorged splenic artery (white arrows) which is contiguous to the strongly enhanced mass or aneurysm (A) and dilated splenic vein (v). Note the accessory spleen or splenosis (white arrowheads)
patient was finally discharged without any symptoms.

Discussion

Fistula between the splenic artery and vein are rare. The first case was reported by Wiegert in 1886.\(^6\) Only 42 patients with SAVF have been reported in the English literature. The majority of these occurred after rupture of a splenic artery aneurysm (29 cases). Other less common causes included splenectomy (\(n = 5\)), gun-shot (\(n = 3\)), congenital origin (\(n = 3\)), mycotic infection (\(n = 1\)) and splenopancreatectomy (\(n = 1\)).\(^6\) The communication of artery and vein usually remains asymptomatic for a long time.

The main clinical manifestations are upper abdominal pain, diarrhoea, oesophageal variceal bleeding and abdominal bruit. The bruit, as noted in this patient, can be either machinery or systolic murmur. It is a diagnostic hallmark and is present in only one-third of patients.\(^6\) It may be heard over the epigastrium, left upper abdomen, left lower chest or left flank. These clinical manifestations may disappear after the development of collateral venous channels. Gastrointestinal bleeding may be caused by an increased portal pressure and hepatopetal sclerosis.\(^10\) High blood flow through the central splenic shunt may lead to development of nontransmural small bowel ischaemia and mesenteric steal syndrome.

Arteriovenous fistula after surgery most probably occurs when vessels are ligated with transfixion sutures or en masse ligation of arteries and veins. Occasionally, penetrating injury of vessels by needles, wires, or pins, may also be the cause of fistulae. SAVF may be avoided by individual ligation of the splenic artery and vein in cases of splenectomy.\(^7\)

SAVF must be confirmed by selective celiac or splenic arteriography. An elongated, tortuous splenic artery, early filling of the splenic vein during the arterial phase, dense opacification of splenoportal venous system and aneurysm-like splenic vein are the characteristic findings of SAVF. If the splenic vein is grossly dilated, the contrast medium may fill in with a ‘turbo-whirl’ pattern. Angiography should be carried out in all patients with portal hypertension, normal liver function, audible abdominal bruit, and traumatic or surgical history.

In most instances, elimination of SAVF is recommended, even in asymptomatic patients, to prevent complications. Surgical excision of SAVF is technically difficult and is sometimes unsuccessful because of the remote location of the lesion, presence of numerous portal collaterals and adhesion. Interventional radiologic techniques, such as transcatheter occlusion of vessels, offer alternatives to surgery. Oblitera-

### Learning points

- SAVF should be suspected in cases of portal hypertension with normal liver function, audible abdominal bruit, and a traumatic or surgical history.
- Characteristic angiographic findings of SAVF include a tortuous splenic artery, early filling of the splenic vein during the arterial phase, dense opacification of splenoportal venous system and aneurysm-like splenic vein.
- Transcatheter arterial embolization is a safe and effective alternative to surgery in the management of SAVF.

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**Figure 2** Celiac angiogram shows the venous aneurysm (black arrowheads) and early opacification of the dilated portal venous system (white arrows). Hepatofugal opacification of superior mesenteric vein (small black arrows) and inferior mesenteric vein (large black arrows) was noted.

**Figure 3** (A) Radiograph shows the giant Gianturco steel coils (arrowheads) deployed into the splenic artery to create a meshwork. (B) Follow-up angiogram reveals the stasis of contrast medium in the splenic artery with occlusion of the arteriovenous fistula. The aneurysm was no longer visualized. Note the smaller coil floated into the aneurysm.
Postpericardiotomy syndrome following temporary and permanent transvenous pacing

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Summary
The postpericardiotomy syndrome may occur as a complication of temporary and permanent pacing. Physicians involved in procedures which may be complicated by this condition therefore need to be aware of its diagnosis and management.

Keywords: postpericardiotomy syndrome; cardiac pacing

Postpericardiotomy syndrome is recognised as one of the commonest complications following cardiac surgery, with an incidence of between 10–50% being reported.1 Its description following the insertion of a transvenous pacemaker, however, is relatively rare. We present a case of temporary and subsequent permanent transvenous pacing resulting in the postpericardiotomy syndrome.

Case report
A 68-year-old man was admitted with a history of presyncopal and syncopal symptoms. There was a previous history of atrial fibrillation, transient ischaemic attacks (for which he was anticoagulated with warfarin) and ischaemic heart disease. On examination, he had a heart rate of 35 beats/min, the electrocardiograph (ECG) showing slow atrial fibrillation. A temperature of 37.8°C, associated with intermittent neck and chest pains and a temperature of 37.8°C.

A VVI permanent pacemaker was inserted (via the left subclavian route) and temporary wire removed 5 days following admission. This was complicated by dull central chest pain following the procedure. A pneumothorax was excluded and a small posterior pericardial effusion was identified with mild mitral regurgitation. The white cell count (WCC) was 109/l (neutrophilia) and the erythrocyte sedimentation rate (ESR) was elevated at 115 mm/h. He was discharged 11 days later following resolution of his symptoms and pyrexia. A swelling at the right side of his neck (site of temporary pacing) was shown by Doppler ultrasonography to be due to a thrombosis involving the internal jugular vein. The erythrocyte sedimentation rate (ESR) was elevated at 115 mm/h. He was discharged 11 days later following resolution of his symptoms and pyrexia, although the ESR remained elevated at 100 mm/h.

At re-admission, 3 weeks later, he gave a history of acute pleuritic chest discomfort and dyspnoea. On examination, he had a temperature of 38°C, a pericardial friction rub and reduced air entry at the left pulmonary base. Chest radiography revealed cardiomegaly and a small left pleural effusion. The white cell count (WCC) was 13 × 10^9/l (neutrophilia) and cardiac enzymes normal. Serial blood cultures remained sterile. The ECG revealed ventricular pacing on a background of atrial fibrillation. At echocardiography, a 3.5 cm pericardial effusion was identified with mild mitral regurgitation and good left ventricular function. The ESR was 107 mm/h, C-reactive protein 251 mg/l (normal <10 mg/l), complement C3/C4 levels normal and autoantibodies (including antineutrophil cytoplasmic antibody and anti-cardiolipin IgG and IgM) negative. Right heart catheterization revealed moderate pulmonary hypertension (pulmonary artery pressure

References
52/10 mmHg) with normal pulmonary and right ventricular angiography. Although no objective evidence for an infection was identified (negative cultures of sputum, urine and blood) an empirical course of amoxicillin/amoxicillin-clavulanic acid combination was administered for 9 days. Although intermittently pyrexial, this gradually subsided with symptomatic improvement. Serial echocardiography confirmed resolution of the pericardial effusion over a 4-week period associated with a decline in the levels of the inflammatory indices and WCC. A diagnosis of postpericardiotomy syndrome secondary to transvenous pacing was felt to be consistent with this presentation.

Discussion

The postpericardiotomy syndrome was originally reported following closed mitral valvotomy. In the classical description, it occurs as fever and pleuropericarditis over a week following cardiac surgery which has involved pericardiectomy and cardiac manipulation. An identical syndrome has been reported following transvenous pacing,1 cardiac catheterization, blunt chest trauma and insertion of epicardial pacing wires.4 There is close similarity to Dressler’s syndrome, which presents in the weeks/months following myocardial infarction.7 Both syndromes include endothelial injury and haemopericardium followed by a delayed response incorporating fever and pericarditis. Antimyocardial antibodies have been reported in both syndromes.

The aetiology for these syndromes is not entirely resolved although an autoimmune reaction has been postulated, based on the associated findings of antimyocardial antibodies. The possible involvement of a viral aetiology with the autoimmune process has also been proposed. An alternative hypothesis has suggested the importance of the leakage of blood into the pericardial space. A higher incidence of Dressler’s syndrome when oral anticoagulation was in common use following myocardial infarction is suggested as corroborative evidence for this theory.7

It is presumed that a prerequisite for the development of this syndrome following transvenous pacing requires perforation of or damage to the right ventricular myocardium. The exposure to myocardial antigens would give rise to an autoimmune reaction with anti-heart antibodies and pleurepericarditis. Pericardial tamponade complicating postpericardiotomy syndrome following permanent transvenous pacing, has been reported.6 Of the cases of this syndrome reported following transvenous pacing, approximately 50% reported the use of temporary wires.

Learning points

● although postpericardiotomy syndrome most frequently occurs following cardiac surgery it may also, on rare occasions, complicate transvenous pacing
● of the cases of this syndrome reported following transvenous pacing approximately 50% report the use of temporary wires
● post pericardiotomy syndrome occurs as fever and pleuropericarditis over one week following the initiation procedure.
Efficacy of statin therapy: possible effect of phenytoin

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Summary
Statins are currently the most widely prescribed lipid-lowering drugs. Individual statins are known to be metabolised by the CYP3A4 isoform of the cytochrome P450 system. The effect of CYP3A4 inducers such as phenytoin on the metabolism and efficacy of these agents is unknown. We report a patient with familial hypercholesterolaemia and epilepsy in whom the introduction and subsequent discontinuation of phenytoin were associated with marked changes in the lipid response to treatment with simvastatin and atorvastatin. The serum activity of γ-glutamyl transpeptidase may have acted as a marker of microsomal induction by phenytoin, since it rose markedly when phenytoin was introduced and returned to normal after it was discontinued.

Keywords: statins; phenytoin; drug interaction

Case report
A 50-year-old woman was referred to the Lipid Clinic in 1995. She was taking simvastatin 10 mg; her total cholesterol was 9.4 mmol/l and triglycerides 1.87 mmol/l. She had never smoked. Her father had died at 45 years of age from a myocardial infarct; one brother had coronary artery bypass grafting at 35 years of age. The patient had gross thickening of both Achilles tendons and a diagnosis of familial hypercholesterolaemia was made. Epilepsy had been diagnosed 6 years previously for which the treatment was sodium valproate 200 mg tid.

On review 3 months later, the patient’s anticonvulsant medication had been changed to phenytoin 325 mg. Her total cholesterol was now 15.99 mmol/l; she claimed to be complying fully with simvastatin 10 mg. Serum activity of γ-glutamyl transpeptidase (γGT), which had previously been normal, was 175 U/l (reference range 5–50 U/l). Significant alcohol intake was denied, and there was no other evidence of this; mean red cell volume was 85 fl (80–100 fl).

Successive subsequent changes to her lipid-lowering therapy included increasing the dose of simvastatin step-wise to 40 mg, switching to...
fluvastatin 40 mg, and finally to atorvastatin, the dose of which was increased step-wise to 80 mg. During this period, the patient developed angina; coronary angiography revealed the presence of significant coronary heart disease. Her total cholesterol remained in excess of 10 mmol/l throughout. Finally, phenytoin was discontinued, in two steps: initially the dose was reduced to 225 mg (simultaneously, the dose of atorvastatin was increased from 40 to 80 mg); phenytoin was then withdrawn altogether. A marked reduction in her total cholesterol (to 6.24 mmol/l) coincided with a return to normal of the serum activity of γGT. Total cholesterol has remained less than 7 mmol/l since the discontinuation of phenytoin; the patient remains on atorvastatin 80 mg. Selected results are shown in the figure.

**Discussion**

The evidence presented here that phenytoin may affect the efficacy of some statins is important for several reasons. Firstly, the deterioration in total cholesterol that accompanied the introduction of phenytoin was clinically significant. The risk of coronary heart disease in familial hypercholesterolaemia and the role of statins in its treatment have both long been recognised. Secondly, such an interaction has not previously been described, despite the fact that phenytoin is known to be an inducer of the isoenzyme CYP3A4 which is involved in statin metabolism. Thirdly, phenytoin appeared in this case to alter the efficacy of both atorvastatin and simvastatin, two of the statins with the greatest cholesterol-lowering potency. Fluvastatin 40 mg was no better in this regard, raising the possibility of a class effect.

Alternative explanations for the initial unexpected deterioration in our patient’s total cholesterol (and marked rise in serum γGT) were sought. There was no other evidence of liver dysfunction, although it is possible that the rise in serum γGT was the sole manifestation of this. Stated compliance with simvastatin was 100%, and alcohol intake was denied. These findings did not, of themselves, provide strong evidence that phenytoin was responsible for the observed deterioration. As we could not test our hypothesis by discontinuing phenytoin, alternative approaches to lipid-lowering were tried. Successively, we increased the dose of simvastatin step-wise to 40 mg daily, switched to fluvastatin 40 mg, and finally introduced atorvastatin, starting at the lowest dose of 10 mg and increasing step-wise to the maximum dose of 80 mg. Despite these changes, the total cholesterol remained unacceptably high. When finally phenytoin was discontinued, in two steps, there was a marked improvement in total cholesterol; simultaneously, the serum activity of γGT returned to normal. These findings provide stronger circumstantial evidence that phenytoin alters the efficacy of atorvastatin at least.

Demonstration of pharmacokinetic interactions between phenytoin and the individual statins has not been possible: in the case of simvastatin, the possibility of an interaction only came to light long after the pre-phenytoin sample had been discarded, while the cost of assaying atorvastatin in the presence and absence of phenytoin is prohibitive. Phenytoin levels remained within the therapeutic range while our patient remained on this agent, and were undetectable following discontinuation.

It is not clear what extra information would be provided by pharmacokinetic studies involving statins. The relationships between serum concentrations of individual statins and concentrations within the hepatocyte (the site of action of statins) are not fully understood. Although the relative activities of some metabolites of individual statins are known, our understanding of statin metabolism, and of the ability of enzyme-inducing drugs to influence it, is likewise far from complete.

In conclusion, we are currently not in a position to test our hypothesis that phenytoin induced CYP3A4 to metabolise atorvastatin and simvastatin (and possibly also fluvastatin) to less active metabolites, thereby reducing their cholesterol-lowering efficacy in our patient. However, we believe that the circumstantial evidence presented here makes such an interaction the most plausible explanation for the sequence of events described.

The authors would like to thank Ms Kathy McFall of the Department of Medical Illustration, West Glasgow Hospitals University NHS Trust, for her help in the preparation of the figure.

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Pentazocine-induced fibromyositis and contracture

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Summary
We report a case of myopathy, accompanied by widespread contractures predominantly involving the elbow and knee joints, following long-standing pentazocine abuse.

Keywords: pentazocine; myopathy; contractures

Complications of parenteral narcotic abuse such as focal tissue damage (localised sclerosis of skin and subcutaneous tissue, indurations and nonseptic ulcerations) are well documented.1 2 There are a few reports of myopathy following chronic pentazocine administration,3 4 but the association with contractures around the shoulder and hip joints is still rare.5 6 We report a patient with myopathy, accompanied by widespread contractures predominantly involving the elbow and knee joints, following long-standing pentazocine abuse. A detailed electrophysiological study of different muscle groups and biopsies from involved and healthy sites was also carried out.

Case report
A 38-year-old doctor (figure 1) presented with a painless and progressive restriction of flexion around both the knee joints for 4 years. This had been followed by a similar problem involving both the elbow joints a year later. His main complaints were inability to squat, difficulty in dressing/undressing and generalised stiffness. Over the last 6 months he had mild limitation at the extremes of abduction of the right shoulder, and bilateral hip extension for which he had developed a compensatory lumbar lordosis. He had no complaints of any muscle weakness within the limited range of possible movements. He had been addicted to injection of Fortwin (pentazocine) 1–2 ml intramuscular bid for the past 18–20 years. The site of injection was the right deltoid for the first 8 years, followed by sequential use of both glutei, quadriceps and the left deltoid. He had a history of frozen shoulder on the right side which subsequently improved. He was non-diabetic and non-hypertensive. There was no family history of any neuromuscular disorder.

Physical examination revealed a well-built man with marked contractures involving both the knee and elbow joints and mild limitations of abduction of right shoulder and extension of both hip joints. He walked with a lordotic gait and had marked woody indurations of the deltoids, biceps, glutei and quadriceps. Both elbows were semi-flexed with a 20° range of movement on either side. The knees could not be flexed beyond 80° and the right hand could not be abducted beyond 90°. Distal joints were normal. Muscle power was normal within the limited range of movements, and there was no sensory deficit. Laboratory studies disclosed the following values: haemoglobin 15.0 g/dl; white cell count 6.0 × 10⁹/l (neutrophils 67%, lymphocytes 20%, monocytes 3%, eosinophils 10%); platelets 275 × 10⁹/l; erythrocyte sedimentation rate 2 mm 1st hour; serum urea 8.9 mmol/l; serum creatinine 0.09 mmol/l; aspartate transaminase 27 U/l; alanine transaminase 9 U/l; alkaline phosphatase 56.8 U/l; creatine kinase 324 U/l. Radiography revealed normal joint architecture. Electromyography (EMG) of the deltoids, triceps and quadriceps showed a reduced number of potentials, low in amplitude and of short duration. Polyphasias were increased (>40%) with a full recruitment pattern. EMG of the supraspinati was normal. Muscle biopsy showed a normal right soleus, whereas there was extensive fibrosis involving the left quadriceps (figure 2).

It was explained to the patient that his drug addiction was responsible for the condition, and he received drug rehabilitation treatment as well as vigorous physiotherapy with passive and active stretching exercises. At 6 months follow-up he had successfully overcome his addiction, but there was no change in his deformity apart from some increase in range of mobility around both the elbow joints to 40–45°, from the previous range of 20°.

Discussion
Cutaneous complications of pentazocine injections were first described by Schlicher et al.1 Swanson et al., in their study of psychiatric aspects of pentazocine abuse,7 noted a 33%
incidence of brawny induration of skin and underlying tissue. The list of complications was later extended by Steiner et al and Joong et al who described a fibrous myopathy with intramuscular pentazocine injections. Their patients had woody induration of muscles with secondary contractures. Deltoid contracture causes a fixed abducted posture, aptly termed ‘arm levitation sign’, a signal of chronic injection myopathy. The main histopathologic change is extensive fibrosis in skin and muscle. Endarteritis, vascular thrombosis, granulomatous inflammation and fat necrosis may also be seen but are non-specific. The history of progressive decline in mobility with generalised stiffness and a lordotic gait in our patient might suggest joint disease. However, the extensive induration of soft tissue with resultant contractures and normal radiographic studies point to the nonarticular nature of his deformity. Although he did not have the arm levitation sign, he did have a history of frozen shoulder. Differential diagnosis of this clinical picture would include infiltrative myopathies such as amyloidosis, eosinophilic fasciitis and the stiff man syndrome. Rarely, an adult form with a forme-fruste manifestation of Emery Dreifuss muscular dystrophy may bear a close resemblance. The history of chronic pentazocine injection in conjunction with extensive fibrosis on biopsy is sufficient to exclude these conditions. The normal EMG of the supraspinatius and normal biopsy of the soleus point against a generalised muscle involvement, despite the raised creatine kinase.

The mechanism of this condition remains unclear. Repeated injections of other medications are not commonly reported to result in diffuse muscle fibrosis. Pentazocine is most soluble under acidic conditions, and precipitation in the alkaline pH of extracellular tissue with secondary inflammation has been postulated. Birefringent crystals have been noted in the areas of induration. The role of repeated muscle trauma, microhaemorrhages, and infections is unknown. Elevations of muscle enzymes, as seen in our patient, have been recorded and suggest ongoing muscle destruction. Animal studies utilising repeated injections of pentazocine show induction of localised sclerosis in guinea pigs but not in rats. Pentazocine-induced fibrous myopathy should be included in diagnostic considerations when a parenteral narcotic agent is required for management of chronic pain.

Learning points

- pentazocine-induced myopathy is a rare condition and should be looked for in all patients using intramuscular pentazocine
- it should be considered in the differential diagnosis of any atypical form of myopathy with contractures
- multiple muscle groups should be evaluated by EMG/biopsy as the disease may be restricted to specific muscle groups
- differentiating this iatrogenic myopathy from polymyositis/dystrophy is of immense prognostic value

Spontaneous rupture of a saphenous vein graft

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Summary
We present a case of spontaneous rupture of a right coronary bypass vein graft in a 57-year-old woman 10 years after coronary by-pass surgery. Although rare, this diagnosis should be considered in such patients presenting with appropriate symptoms.

Keywords: coronary artery bypass graft; false aneurysm; saphenous vein graft

Coronary artery rupture as a consequence of interventional procedures is well described both for native coronary vessels and for bypass grafts. Spontaneous rupture in the absence of intervention is rare, though described for native vessels. Spontaneous rupture of a bypass graft is an even rarer occurrence.

Case report
A 57-year-old woman presented with sudden onset of chest pain, described by her as ‘explosive’, and so severe as to ‘knock her to the ground’. The pain was in the anterior chest, radiated through to the back, and was exacerbated by deep respiration and by movement. She felt nauseated and was breathless. Moderately intense pain continued until admission to hospital 4 hours later, by which time she had experienced some minor haemoptysis. She had undergone coronary surgery 15 years earlier, with vein grafts to the left anterior descending artery and diagonal branch; 10 years ago the vein grafts were occluded, and she underwent further coronary surgery, with grafts to the left anterior descending artery (left internal mammary artery) and the right coronary artery (saphenous vein graft). Two years prior to the current admission, angiography demonstrated impaired left ventricular function, with an ejection fraction of 45%, occluded left anterior descending artery, with a good distal internal mammary artery graft, severe native circumflex disease, and an occluded native right coronary with the distal vessel supplied by an ectatic bypass vein graft, measuring up to 2 cm in diameter (figure 1). Medical management was pursued.

Examination showed her to be in mild distress, warm, heart rate 90 beats/min, with no signs of heart failure and normal cardiac auscultation. Saturation on finger plethysmography was 76%. Chest X-ray showed cardiomegaly with clear lung fields. Electrocardiography (ECG) showed no new changes.

The most likely diagnosis was felt to be a pulmonary embolus, though aortic dissection was also considered. Computed tomography (CT) scan revealed a unremarkable aorta, with an abnormal structure lying behind the heart. Transthoracic echocardiography (TTE) revealed a small 1 × 2 cm fluid-filled structure lying behind the left atrium. Twelve hours later repeat TTE showed that this structure had enlarged dramatically, resulting in severe compression of the left atrium. Transoesophageal echocardiography (TOE, figure 2) showed a large mass behind and to the right side of the heart. This structure had a thick rim, with spontaneous contractecho. Laterally there was a thick rimmed structure divided by a thick septum. Repeat CT scan of the chest (figure 2) showed that contrast leaked into the posterior structure. At urgent thoracotomy, a large false aneurysm of the right coronary saphenous by-pass graft was found, with extensive extravasation of blood into the surrounding tissues. Haemostasis was achieved, but it was not possible to wean her from bypass, and she died.

Discussion
Late rupture of a coronary bypass graft seems very rare. In this case, rupture led to extrinsic compression of the left atrium, and subsequent left heart failure, hypoxaemia, haemoptysis and death. Of particular interest was the difficulty in establishing the correct diagnosis, which was in part due to its rarity. At presentation, the sudden onset of pleuritic chest pain, with hypoxaemia and haemoptysis led to an initial diagnosis of pulmonary embolism, though common forms of vascular catastrophe, including aortic dissection, were considered.
The orientation of the probe is such that structures on the right side of the heart are shown on the left side of this photo, and those behind the heart at the top of the photo. A large thick walled mass is seen immediately adjacent to the probe, extending anteriorly for up to 5 cm, and virtually obliterating the left atrium. The ectatic vein graft is seen, together with its false aneurysm, to the right of the heart. Below: Thoracic CT scan, with contrast. This CT scan demonstrates the contrast-filled right coronary vein graft and, posterior and medial to the graft, the crescent-shaped false aneurysm. There is a 5 x 5 cm mass of extravasated blood behind the heart, which is lightly stained with contrast.

investigations were inconclusive, and it was not until 12 hours later, after further bleeding into the mediastinum had occurred, that investigations revealed the true nature of the problem. Diagnosis was established by demonstrating blood in the mediastinum on CT scanning as well as a false aneurysm of the coronary graft. In other case reports diagnosis has usually been suggested by CT scanning, though TOE has been used, and diagnosis has been confirmed either by coronary angiography or at surgery. The usual treatment is surgery to tie off both ends of the ruptured graft, with or without the application of further grafts. In patients unfit for surgery trans-catheter embolisation of the false aneurysms may be a viable alternative.

Rupture of vascular structures depends on several factors, including wall stress, which in turn will depend on blood pressure, and the thickness of the vessel wall, as well as the size of the vessel, according to the law of Laplace. An important factor in the rupture of this bypass graft may have been the antecedent aneurysmal dilatation. Large numbers of coronary operations use veins as conduits and, as veins are thin-walled structures, and as aneurysmal dilatation of vein grafts is not infrequent, one might expect to see spontaneous rupture more commonly. Current data suggest, however, that spontaneous rupture is unusual. It is possible that the true incidence is underestimated, as many patients with previous coronary surgery who present with further chest pain may be thought to have further myocardial ischaemia, particularly if there are new ECG changes. Death in these circumstances is rarely followed by a post mortem, and thus the true diagnosis may not be established. However in subjects presenting with symptoms suggestive of a vascular catastrophe, this diagnosis is worth consideration. Furthermore, presentation can be atypical, and therefore the diagnosis should be considered in all patients who have had coronary surgery with saphenous vein bypass grafts who present with atypical chest pain, superior vena caval obstruction, or mediastinal masses.

Learning points

- spontaneous rupture of a saphenous bypass vein graft is rare, but does occur
- the most important aspect of diagnosis is awareness of this late complication of coronary artery bypass surgery
- diagnosis can be made by CT, MRI, TOE, or coronary angiography
- haemostasis can be achieved either by surgery or by trans-catheter embolisation