Haemorrhagic myositis associated with prophylactic heparin use in dermatomyositis

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An 80 year old man was admitted with a short history of fever, dyspnoea, and productive cough. A chest radiograph disclosed right lower lobe infiltrates, and a diagnosis of community acquired pneumonia was made. However, it was noted that he had profound weakness of the proximal, respiratory, and bulbar muscles together with typical features of dermatomyositis, including periorbital oedema and heliotrope discolouration, nailfold infarcts, and Gottron's papules. The serum creatine kinase (CK) level was raised at 1850 IU (reference range<300 IU). An electromyogram and muscle biopsy of the left vastus lateralis confirmed inflammatory myositis. There was no excess bleeding at the site. The diagnosis was revised to aspiration pneumonia secondary to bulbar weakness associated with dermatomyositis. Respiratory muscle weakness progressed after admission, with a fall in the forced expiratory volume (1 second) to 0.95 litres (predicted = 2.5 litres).

Intravenous methylprednisolone (500 mg/day for 3 days) was given, followed by oral prednisone at 80 mg/day. Treatment with prophylactic unfractionated heparin (UFH) (5000 IU twice a day subcutaneously) was started (the patient's weight was 80 kg).

After 9 days of corticosteroid treatment, the patient's strength and serum CK levels were improving. However, on day 10 he complained of a painful right hip of gradual onset. Plain radiographs of the right hip were normal. Avascular necrosis was suspected and magnetic resonance imaging (MRI) of the region was arranged for the next day. Examination before MRI showed a palpable small mass in the left rectus sheath, a tense swollen right thigh, and extensive bruising affecting the left flank. MRI subsequently showed extensive haemorrhagic change in the muscles of the right thigh (fig 1), and computed tomography of the abdomen showed haemorrhage in the rectus sheath and oblique muscles (fig 2). His haemoglobin had fallen to 50 g/l from 130 g/l overnight, platelets were within the reference range, and the activated partial thromboplastin time (APTT) was slightly raised at 42 seconds (reference range<38 seconds). The APTT, prothrombin, and bleeding time were normal before the muscle biopsy.

The subcutaneous heparin was stopped and 3 units of packed cells and 4 units of fresh frozen plasma were transfused. The patient subsequently developed pulmonary oedema requiring assisted ventilation, but made a successful recovery and returned home after a short period of intensive rehabilitation. There was no history suggestive of a coagulation disorder and after the transfusion his coagulation profile returned to normal.

DISCUSSION

Our patient had a major complication associated with standard dose UFH use for deep vein thrombosis prophylaxis. Significant muscle haemorrhage has not been previously reported in patients with myositis, though recently has been found in a patient receiving therapeutic low molecular weight heparin (LMWH) and warfarin.[1] Heparin treatment is known to be associated with an increased risk of major bleeding, about 0.3%,[2] with both UFH and LMWH, mostly occurring in the gastrointestinal tract, though haemorrhage at the site of injection has been reported.[3]

Given the serious nature of this event, we would advise caution in using prophylactic heparin in patients with acute myositis.

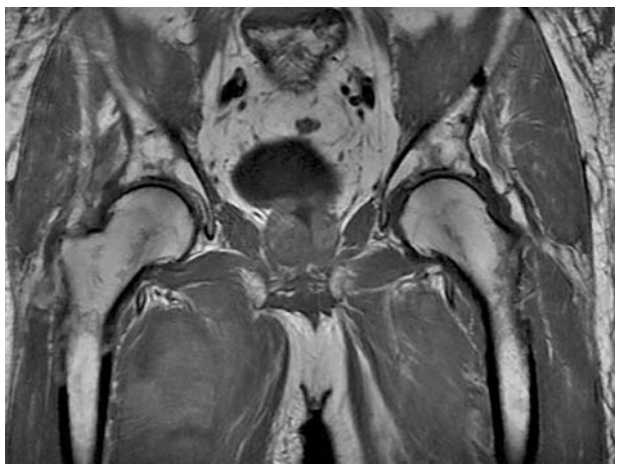

Figure 1 MRI of the pelvis disclosing a large right thigh haematoma.

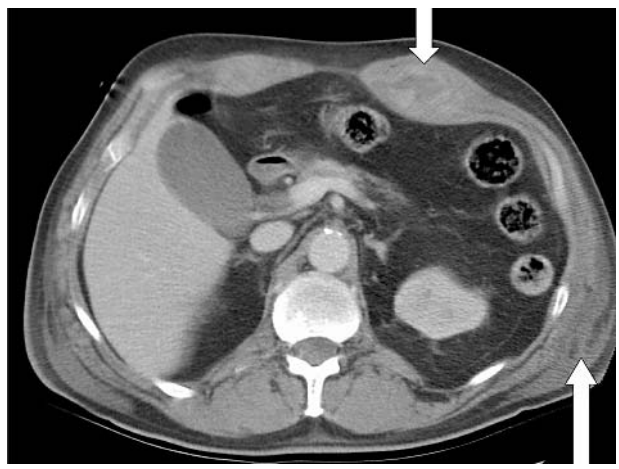

Figure 2 A computed tomographic scan of the abdomen showing left rectus abdominus and oblique musculature haemorrhage.

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Isolated cranial nerve syndromes without proximal carotid involvement in aortoarteritis

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Takayasu's arteritis or aortoarteritis affects the aorta and its proximal branches.[1] Cranial nerve involvement in the form of ischaemic optic neuropathy secondary to proximal carotid involvement is common.[2] Involvement of other cranial nerves has been reported only once before.[3] We report two patients with aortoarteritis who developed isolated cranial nerve palsies. Notably, in both cases it occurred without proximal carotid involvement.

CASE REPORTS

Patient 1, a 30 year old businessman, presented with malignant hypertension because of right renal artery stenosis due to aortoarteritis affecting the abdominal aorta. After successful balloon angioplasty, blood pressure was well controlled with antihypertensive drugs. Two years later he developed sudden headache and diplopia on looking towards the left. Examination showed a convergent squint in the left eye due to abducens nerve palsy. Contrast enhanced brain computed tomography (CT) scan and cerebrospinal fluid (CSF) analysis were normal. Angiography showed that the aortic arch and its branches were normal; the suprarenal abdominal aorta showed irregular narrowing with stenosis of the left renal artery and the previously balloon-dilated right renal artery. On treatment with 1 mg/kg/day of prednisolone he recovered completely in 10 days.

One year later he developed diplopia again. This time he was found to have right abducens nerve palsy. Angiography showed normal carotid vessels but complete occlusion of the left subclavian artery 1 cm distal to its origin. Again, he made a complete recovery with 1 mg/kg of prednisolone over 20 days.

Patient 2, an 18 year old female student, was admitted for accelerated hypertension. A renal Doppler examination showed left renal artery stenosis. On aortography she had an irregular contour with narrowing of the abdominal aorta and occlusion from the origin of the left renal artery, suggestive of aortoarteritis. She refused renal angioplasty and was treated with antihypertensive drugs. Four years later she developed sudden diplopia and right eyelid ptosis. Examination showed complete ptosis on the right, divergent squint, and ophthalmoplegia, suggestive of isolated right oculomotor palsy. The pupil, however, was normal in size and constricted briskly in light. Contrast enhanced brain CT scan, CSF analysis, and four-vessel angiogram were normal. She received 1 mg/kg of prednisolone for 30 days with complete recovery.

DISCUSSION

Both patients satisfied Ishikawa's diagnostic criteria for aortoarteritis,[4] and presented with sudden onset of isolated cranial nerve palsies. The normal CT scans and CSF examination excluded structural and infective aetiologies of cranial nerve palsies. The sudden onset of deficits in both patients suggested a vascular cause. Moreover, recurrence of the cranial nerve deficit on the contralateral side in patient 1 and the presence of pupillary sparing in patient 2 further supported a vascular aetiology. The pupil is characteristically unaffected in ischaemic oculomotor nerve palsy because the pupillary fibres, which are outermost, receive additional blood supply from the pial (meningeal) vessels and are therefore spared in oculomotor nerve infarction.[5]

The oculomotor and abducens nerves derive their arterial supply from the inferolateral trunk, which originates from the intracavernous siphon of the internal carotid artery.[6] In our patients the extracranial carotid circulation and the siphon of the internal carotid artery were angiographically normal, suggesting involvement of small sized vessels supplying the cranial nerves. Nuclear brainstem infarcts could also cause cranial nerve deficits[7] and a magnetic resonance imaging study (not done in our patients) would be most sensitive to diagnose them. However, this is very unlikely owing to the absence of signs of involvement of the brainstem ascending or descending tracts and rapid complete recovery of deficits with corticosteroids.

Similar isolated palsies of the nerves innervating the extraocular muscles may occur in other vasculitides, including Behçet's disease and giant cell arteritis.[8 9]

Small vessel involvement has been described in some patients with aortoarteritis. Manifestations of these include glomerulonephritis,[10] interstitial lung disease,[11] and necrotising vasculitic lesions of the skin.[12] It is likely that cranial nerve palsies in our patients too might be due to similar small vessel involvement in the central nervous system.

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