lesson of the month

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A simple case of upper limb venous thrombosis requiring anticoagulation?

In patients with arm swelling Paget-von Schroetter syndrome (PvSS) should be suspected and multidisciplinary treatment involving radiologists and surgeons considered.

Lesson

A 27-year-old right-handed man, who undertook regular weightlifting, presented with a seven week history of intermittent left arm pain and swelling exacerbated by exercise. Two weeks prior to presentation this became sustained and progressive. There were no risk factors for venous thrombosis.

The left upper limb appeared dusky and was swollen with obvious collateral blood vessels. The arm was neurovascularly intact. Examination was otherwise unremarkable.

Full blood count, chest radiography, D-dimer and thrombophilia screening were normal. Left arm unsubtracted venogram images demonstrated hold-up of contrast in the left axillary vein with multiple collaterals at this site and no demonstration of the central veins (Fig 1).

Repeat examination using digital subtraction confirmed short occlusion of the axillary vein passing the first rib with collaterals. The patient received anticoagulation with warfarin as presentation was considered too late for intravenous thrombolysis.

Comment

Primary venous thrombosis presents as swelling, pain, discolouration and distension of superficial veins of the limb involved. Many patients have a history of transient episodes of swelling ('herald episodes'). Differential diagnoses include fracture, crush injury, muscular sprain and lymphatic obstruction.

Paget-von Schroetter syndrome (PvSS), synonymous with spontaneous or effort-induced thrombosis of the subclavian-axillary vein,[1] is associated with numerous sports activities including weightlifting.[2] Developmental anomalies including fibromuscular bands result in dynamic obstruction of the subclavian vein at the thoracic outlet.[3] Strain on the subclavian and axillary veins, greatest when the arm is hyperabducted and externally rotated (the 'I surrender' or 'Roos' position[3]) causes them to be compressed between the clavicle and the first rib. This induces microtrauma of the venous intima causing coagulation and thrombogenesis.[2] Compression induced fibrosis and stenosis precede thrombus formation.

Plain radiographs of the thoracic outlet can exclude a bony cervical rib. Axillary thrombosis can be diagnosed with duplex ultrasound and standard venography. Digital subtraction venography is required to image the underlying venous stenosis. Magnetic resonance venography is increasingly used.[4]

Patients with a short history of herald symptoms (the pathological correlate being compression and stenosis without thrombosis) need early digital subtraction venography, anticoagulation and consideration for rib resection.

When thrombosis has occurred catheter-directed thrombolysis is associated with marked relief of symptoms and a prolonged symptom-free period compared with anticoagulation alone.[5] It carries a small risk of haemorrhage and stroke and requires overnight monitoring in a high dependency unit. Attempts to deploy stents into the axillary/subclavian vein prior to decompressive surgery have been disappointing as external compression causes stent fracture or re-occlusion with a high risk of recurrent thrombosis.[6]

This patient presented too late for thrombolysis to re-canalise his axillary vein. Anticoagulation was used to prevent pulmonary embolus.

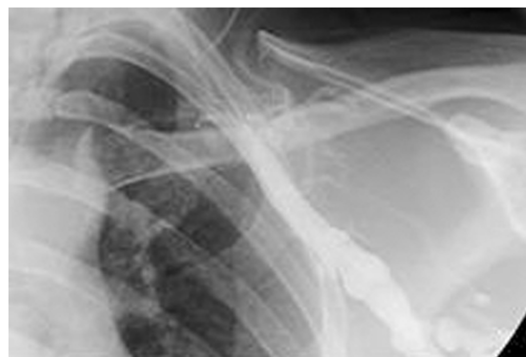

Fig 1. Left arm venogram demonstrating axillary vein thrombosis.

Patients with PvSS are often considered for rib resection, especially when symptoms persist despite thrombolysis and anticoagulation when balloon angioplasty may improve flow pending surgical decompression.[7] However, not all patients require surgery.[8] Following decompression, if the vein remains narrowed, balloon angioplasty and stent placement have been shown to have good long-term results.[9]

Paget-von Schroetter syndrome usually occurs in young, healthy individuals. Prompt recognition and referral for urgent imaging enables treatment to re-canalise the vein and may prevent venous thrombosis if identified at the 'herald symptom' stage. Surgical assessment enables consideration for rib resection if necessary.

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Literature and medicine

Medical anthropology in Jane Austen's *Emma*

Neil Vickers

Jane Austen's interest in medicine is well known. Her comic masterpiece *Emma*, first published in 1815, wears its medical interests on its sleeve with references to health and sickness found in every chapter.[1] And yet the novel seems oddly and knowingly reticent about its medical content.

Perhaps the most striking feature of the little group Austen describes is the way it responds to sickness. Emma's father, Mr Woodhouse, is a hypochondriac who devotes every waking moment to health worries. He urges those around him to adopt his regimen of thin gruel, warm rooms and comically moderate exercise. Much of his time is spent in consultation with the local apothecary, Mr Perry, who – if Mr Woodhouse is to be believed – acquiesces in all his patient's opinions. (But Woodhouse is a bad listener, 'he could never believe other people to be different from himself' (16), and we never hear Perry say anything in his own voice. His deeds and sayings are only ever reported.)

Mr Woodhouse attributes his survival into late middle age to a life of extreme caution and to Perry's ministrations. Austen archly describes Mr Woodhouse as 'a valetudinarian all his life, without activity of mind or body' (5); if he has any physical complaint, the book does not disclose it. Mentally he seems blocked. He hates to leave his own house and he dislikes making new acquaintances. When he takes a walk he makes it a rule never to go beyond his shrubbery. He commiserates with his friend Mr Knightley on having walked half a mile in the moonlight. And he lives in constant fear of catching colds.

What appears to irk Mr Woodhouse more than anything else is marriage. Early on in the novel we find him depressed that Miss Taylor, who had acted as governess since his wife's death 16 years earlier, has left him to marry one of his neighbours, Mr Weston. The marriage is a source of joy to everyone but him. 'Ah! poor Miss Taylor!' he exclaims, ' 'tis a sad business' (16). The couple make him a present of a large portion of their wedding cake. When his daughter invites their friends to share it, he earnestly tries 'to dissuade them from having any wedding-cake at all, and when that proved vain, as earnestly tried to prevent any body's eating it'. The