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CASE REPORT

Unusual cause of lower extremity wounds: Cobb syndrome

Bahareh Abtahi-naeini[1,2], Ali Saffaei[2] & Mohsen Pourazizi[3]

1 Skin diseases and Leishmaniasis Research Center, Isfahan University of Medical Sciences, Isfahan, Iran
2 Students' Research Committee, Isfahan University of Medical Sciences, Isfahan, Iran
3 Cancer Research Center, Semnan University of Medical Sciences, Semnan, Iran

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Correspondence to
Dr M Pourazizi
Cancer Research Center
Semnan University of Medical Sciences
Semnan
Iran
E-mail: m.pourazizi@yahoo.com

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Abstract

Cobb syndrome (Cutaneomeningospinal Angiomatosis) is a rare segmental neurocutaneous syndrome associated with metameric cutaneous and spinal cord arteriovenous malformations (AVMs). In this syndrome, capillary malformation or angiokeratoma-like lesions are formed in a dermatomal distribution, with an AVM in the corresponding segment of the spinal cord. The spinal cord lesions can cause neurological disorder and paraplegia, which typically develop during young adulthood. We report a 32-year-old male with the Cobb syndrome associated with lower extremity painful wounds and acute-onset paraplegia due to metameric vascular malformations.

Cobb syndrome (cutaneomeningospinal angiomatosis) is a rare metameric disorder characterised by an association with cutaneous angiomas in the same metamere (1,2). The syndrome was not recognised until Cobb's description of it in 1915 (1). The common neurological manifestations of this syndrome are pain and motor deficits caused by spinal angioma. Nevus flammeus and angiokeratomas are the known skin manifestations of Cobb syndrome (1–4). We report a 32-year-old male, who presented to the dermatology clinic of Alzahra hospital, Isfahan University of Medical Sciences, with a malodor cauliflower-like bulky lesion at the intergluteal cleft and lower back from 6 months ago. The lesion was classified as spinal arteriovenous lesion and required surgical management. In addition, he had a painful ulcer on the right lateral malleolus (Figure 1).

He had a history of acute paraplegia since 4 months ago that presented with sudden development of weakness and decreased muscle strength in both lower extremities, which was diagnosed as arteriovenous malformation (AVM) distributed from T9 to L1–L4. After embolisation of the AVM, the L2 and L3 intercostal arteries were cannulated and the feeders were embolised by Gluebran mixed with Lipiodol. The patient's motor nerve activity gradually returned to normal, but the painful leg ulcers did not respond to conventional treatments. Cardiovascular, respiratory and abdominal examinations produced normal results. On skin examination, a recently developed malodor cauliflower-like plaque, measuring 13 cm × 6 cm, was found in the right lumbar region. There was no palpable thrill or audible bruit over this area. He recalled that the slightly raised plaque on his lower back had developed recently with current appearance since 6 months ago. Histopathological examination of the skin lesions showed keratinocyte proliferation and rete ridge elongation with dilated vascular spaces of different sizes suggesting verrucous vascular malformation (angiokeratoma).

Another notable lesion was a painful, round, leg ulcer with sharply demarcated border and punched-out appearance on the right lateral malleolus, from 18 months ago. Color Doppler ultrasonography of the right lower limb showed that superficial femoral vein (SFV), common femoral vein (CFV), popliteal vein and posterior and anterior tibialis were normal in size and compressibility. Dilation and torsion of tibialis posterior vein were remarkable. Based on the clinical and diagnostic findings the patient was given a diagnosis of Cobb syndrome. Treatment

Key Messages

- this report emphasise the importance of recognising cutaneous lesions of the spinal axis, which should alert the clinician to the possibility of spinal dysraphism or spinal arteriovenous malformation (AVM)
- patients were presented with unexplained radicular pain and neurologic symptoms combined with lower extremities ulceration, which can alert the clinician to the possibility of Cobb syndrome and lead to earlier detection and treatment of an associated spinal AVM and prevention of neurologic sequels

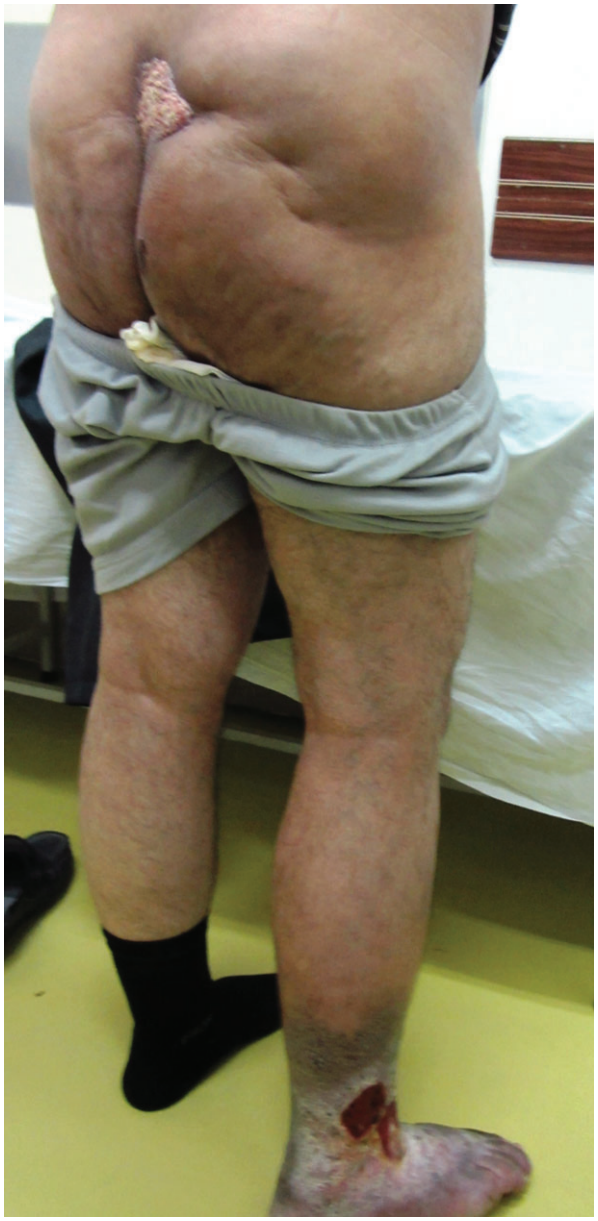

Figure 1 Cobb syndrome. Lumbosacral angiokeratoma-like lesion and leg ulcer are seen.

included embolisation of AVM of the lumbosacral spinal cord for improving the underlying vascular problem. The distal leg ulcer was managed with conservative treatment which included cleaning the wound with normal saline, applying Burow solution and bandaging twice daily. Because the lumbosacral lesion was malodorous, systemic antibiotic including clindamycin and ciprofloxacin were prescribed. Kissel and Dureux reviewed 18 cases of Cobb syndrome. They stated that the neurological manifestations of this syndrome are caused by spinal angioma and that the most common initial symptoms are pain and motor deficit (5,6). The range of presentations in symptomatic Cobb syndrome is diverse, and the course is unpredictable, thus making firm prognoses difficult (2,7). In our patient, initial symptom was radicular pain in the lumbar region, and followed by urinary symptoms due to neurogenic bladder and ultimately paraplegia. The findings were associated with a clinically significant spinal AVM including motor and sensory deficits, with the onset of symptomatic deterioration in neurologic performance which developed insidiously in 6 months. Treatment options include neurosurgery, embolisation, radiation and corticosteroid therapy. Because spinal AVMs are often extensive, curative therapy may not be possible (3). It is therefore important to manage the patient in the context of a multidisciplinary team, with involvement from neurology, neurosurgery, interventional neuroradiology and also dermatology specialists (3,8).

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