Cobb syndrome: A rare cause of paraplegia

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ABSTRACT

Cobb syndrome is an exceedingly rare clinical condition defined by the presence of a vascular skin nevus and an angioma in the spinal canal at the same metamere. We report the case of a 14-year-old boy who presented with sudden onset paraplegia. Physical examination showed port-wine stains over buttock and thigh. Magnetic resonance (MR) angiogram of the dorso-lumbar spine revealed a large arteriovenous malformation (AVM) at D_{11}-D_{12} to L_2-L_3 levels. These concurrent findings led to the diagnosis of Cobb's syndrome. The patient received orally administered prednisolone therapy and underwent endovascular embolization of spinal angioma. Cobb's syndrome is a rare disease entity and literature search revealed only a few case reports and series mentioning this condition to date. The importance lies in the recognition that cutaneous vascular lesions may clue to an associated spinal cord angioma or AVM that may lead to weakness or paralysis.

KEY WORDS: Cobb syndrome, magnetic resonance angiogram, spine, arteriovenous malformation, surgery, embolization

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Introduction

Cutaneomeningospinal angiomatosis, also known as Cobb syndrome, describes the presence of spinal arteriovenous malformation (AVM) and a vascular skin lesion affecting the corresponding dermatome. Although first described by Berenbruch in 1890, the recognition is largely attributed to Cobb's description of a case of "hemangiomata" of the spine associated with a skin nevi in 1915. [1] Cobb syndrome probably has an underreported prevalence, and the number of documented cases of Cobb syndrome in the literature is less than 50. Owing to the rarity of this syndrome, definitive management recommendations remain unclear.

The present case describes a 14-year-old boy who presented with sudden-onset paraplegia and presence of multiple patchy angiomas which raised the suspicion of Cobb syndrome that was subsequently confirmed by imaging studies. Recognition of the coincidence of the port-wine stain and spinal vascular lesions is important for accurate diagnosis of this syndrome because most of the common presenting symptoms, such as pain and motor deficits, are non-specific. We have reviewed the literature on Cobb syndrome and discussed the currently available treatment modalities.

Case Report

A 14-year-old boy presented with sudden-onset weakness of the lower limbs associated with loss of sensation of all modalities up to the pelvis and with marked bladder/fecal dysfunction. There was a history of frequent cramps in both legs since 5 years of age. On examination, there were conspicuous port-wine stains in the buttock, measuring 60 × 45 cm and over the right thigh, measuring 40 × 25 cm (Figure 1). There was no palpable thrill or audible bruit over this area. These spots were initially blue after birth which gradually faded with time to evolve into the current appearance. Neurologically, the patient had marked flaccid weakness of both upper and lower limbs (grade 1/5). Deep tendon reflexes were absent, with plantar and cremasteric reflexes being non responsive. Definite sensory level was present corresponding to L_1 segment of spinal cord. Fundoscopic examination was normal.

A lumbar puncture was performed in a guarded fashion in L_4-L_5 interspace as per our hospital protocol for investigation of paraplegia to exclude chronic infections which revealed xanthochromic cerebrospinal fluid (CSF) with cells 8/mm^3 (all lymphocytes). Protein and glucose levels were 440 mg/dl and 40 mg/dl respectively. The lumbar puncture could have been avoided if we had the magnetic resonance imaging (MRI) of dorso-lumbar spine findings available with us beforehand. It revealed a large arteriovenous malformation (AVM). The AVM was extending from D_{11}-D_{12} to L_2-L_3 levels (Figure 2) with associated hematomyelia in the lower dorsal cord and hematoma in the conus with low lying conus terminating at L_3. A dilated and tortuous vessel was seen arising from one of the right sided intercostal artery and going into the dorsal spinal canal through the right sided neural foramina (Figure 3A). Intrathecal enhancing vessels were seen on post contrast T1W images (Figure 3B).

The patient received orally administered prednisolone therapy (initial dose, 30 mg/d). Paraparesis partially improved and the skin nevus showed some degree of regression thereafter. A multidisciplinary approach to the patient's care was undertaken, involving the involvement of neurology, neurosurgical, and interventional radiology teams. He underwent endovascular embolization of the feeding vessels with the use of *n*-butyl-2-cyanoacrylate (NBCA). Postoperatively, the patient did well; no neurological deterioration was observed. One month after embolization, he started to regain strength with physiotherapy. He was discharged from the hospital and continued to receive orally administered prednisolone; the dose was tapered to 5 mg/d. At the time of discharge, he was able to lift his legs against gravity (Grade 3/5). A written

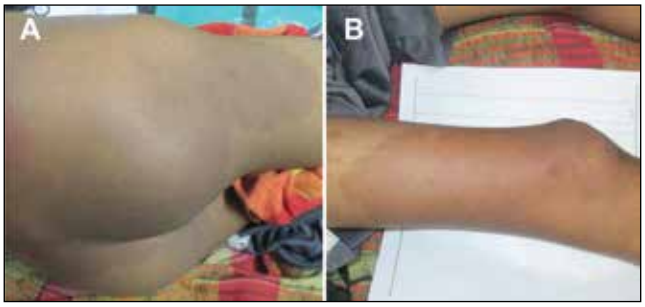

Fig. 1: Large port-wine stains on his buttock **(A)** and over the anterior aspect of right thigh **(B)**.

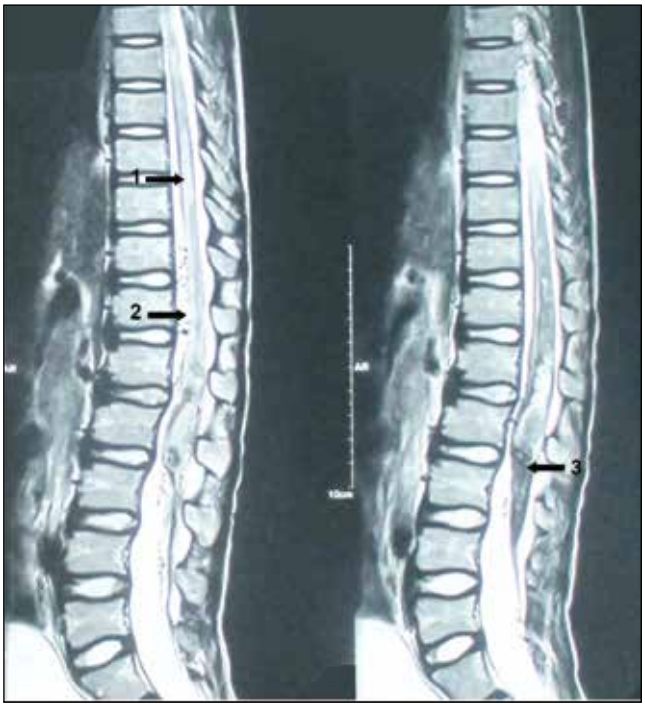

Fig. 2: Magnetic resonance imaging scan of dorso-lumbar spine. T2 weighted sagittal imaging: (1 = Lower dorsal and upper lumbar cords showing heterogenous T2 hyperintensities suggestive of developing syrinx; 2 = Vascular flow voids on ventral aspect of thecal sac). Conus appears to be low down and expanded with internal heterogenous signal intensity (3 = Resolving cord hematoma.).

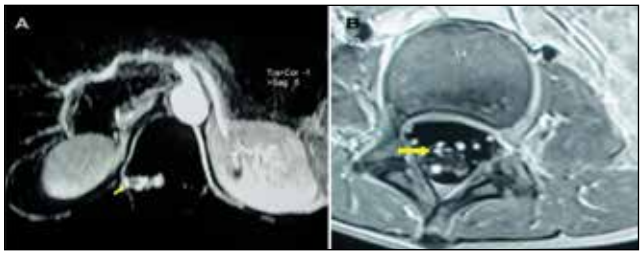

Fig. 3: (A). MR angiogram (MIP image) showing a dilated and tortuous feeding artery of the AVM arising from lower intercostal arteries (yellow arrow); **(B)**. Axial fat-suppressed post-contrast T1 weighted image showing multiple intra thecal dilated vessels (yellow arrow) situated both dorsal and ventral to the cauda and conus.

consent was obtained from a boy's parents before carrying out the study.

Discussion

Cobb's syndrome is a rare, non-inherited disorder which represents the concurrent findings of a spinal AVM in the same metamere as the cutaneous lesion.[2] To date, although only 45 cases have been reported in world literature, but the actual incidence may be higher as only symptomatic cases are diagnosed.[3–6] More recently, Cobb syndrome was recognized to be a part of a larger group of multiple arteriovenous shunts with metameric links, and the syndrome has been renamed to "spinal arteriovenous metameric syndrome".[7]

The cutaneous manifestations range from macular port-wine stains to various types of papular or nodular vascular lesions including angiomas, angiokeratomas, angiolipomas, and lymphangioma circumscriptum.[2] Vascular lesions associated with Cobb syndrome do not tend to resolve or involute spontaneously, like the usual capillary and cavernous malformations of childhood.[1] Cutaneous lesions are either noted incidentally or brought to medical attention because of cosmetic concerns or susceptibility to bleeding with trauma. It is essential to point out that the embryological origin of the blood supply to the vertebrae and spinal cord comes from the segmental dorsal arteries. This finding may explain the common metameric origin of the AVM vessels that create the cutaneomeningospinal angiomas.[7] The vascular skin nevus found with Cobb syndrome is accompanied by a large variety of vascular pathologies. The intraspinal lesions are usually AVMs (high flow lesion) and rarely angiomas (low- flow lesion).[3,7,8]

Vascular anomalies can be divided into vascular neoplasms (e.g. hemangioma) and vascular malformations. Whereas vascular malformations are generally subdivided into: (1) slow- or low-flow and (2) fast- or high-flow malformations. High-flow malformations contain arterial components, (e.g. AVMs and arterio-venous fistula) may present with pain, ulceration, ischemic changes, bleeding, congestive heart failure, warm pink patches on the skin with an underlying vascular murmur or thrill and treated with surgical resection or embolization. Low-flow malformations include combinations of capillary, venous, and lymphatic components which may present at birth or later with slow growth, may increase in size with crying or Valsalva. They are treated expectantly with compression garments, sclerotherapy or surgical excision for pain, swelling or if life threatening (e.g. airway compromise).[9]

Cobb syndrome is typically diagnosed following the onset of neurological symptoms. Although this order is most commonly seen during late childhood, it may occur at any age. Onset of signs usually manifest over weeks to years, but a sudden onset of weakness with rapid progression has also been reported.[10] Neurological presentations can vary from monoparesis to sudden-onset paraplegia or quadriplegia. Bladder and bowel involvement is common but occurs late as the disease progress. Less common signs include meningismus, headache, fever, and gluteal and limb hypertrophy. With regard to neurological symptoms, cord compression due to spinal angioma *per se* may not be the sole mechanism underlying the spinal cord symptoms. Other factors may include compression, venous hypertension and cord ischemia due to steal syndrome are the speculated mechanisms that would explain the myelopathy.[11]

Computed tomography (CT) and MRI are useful modalities to assess the extension of the lesions. MRI is better than CT in displaying deformed vessels, angiomas and the feeding artery. The final diagnosis of the syndrome depends on angiography. MRI can show intramedullary signal changes and most of the vessels and is safer than invasive angiography with intravascular contrast. However, selective spinal angiography facilitates understanding of the complex angioarchitecture and pathophysiology as well as the embolization procedure.[11,12] Lumbar puncture is contraindicated in suspected spinal mass lesion especially spinal vascular malformation.

The optimal management of the disease entity largely remains enigmatic. With the limited cases described in the literature, it is difficult to formulate a best possible treatment algorithm. The role of definitive surgical excision is limited because surgical morbidity remains a concern. Treatment options reported in the literature have ranged from palliation without intervention to surgical resection and more recently endovascular embolization.[4,11,13,14] The therapeutic efficacy of endovascular treatment for spinal AVMs in Cobb syndrome has been reviewed by Linfante *et al*, who reported restoration of ambulation in a patient with long-standing paraplegia related to Cobb syndrome after successful embolization of a large spinal AVM.[15] As the spinal AVMs are often extensive and at multiple spinal levels, curative therapy may not be achieved, and combined modality intervention is intended to halt the symptom progression and minimize the neurological sequelae by reducing mass effect, venous hypertension and vascular steal along the spinal cord.[14,16] Surgical resection has been successfully combined with endovascular embolization for large spinal AVMs.[6,7] Endovascular embolization had been combined with pre-embolization steroid therapy by Soeda A et al.[13] The varying modalities have been combined effectively with steroids to halt progression of symptoms, allowing quick recovery of some degree of neurological function and reduce the number of embolization procedures.[13,17] Systemic corticosteroid therapy was found to be effective in other cutaneous vascular malformations like Kasabach-Merritt syndrome.[18] Given the rarity of Cobb syndrome and report of only two cases previous to our case showing benefit of steroids, the role of corticosteroids in Cobb syndrome at present is mostly empirical and needs further study.[13,19] Earlier only 2 cases of Cobb syndrome has been reported from India in 1996 by Bassapa et al.[20] and in 2006 by Sardana *et al*.[19] This is only the third case report of Cobb syndrome from India and the first one to be treated with combined endovascular embolization and steroid therapy. In a recent study from India on spinal arteriovenous malformation, 22 patient underwent surgical occlusion of spinal dural arteriovenous fistula (SDAVF) and there was either improvement or stabilization of motor weakness in all of them.[21]

Conclusion

A diverse range of presentation is reported in symptomatic Cobb syndrome, and the course is often unpredictable. The cutaneous lesion may provide a clue when a patient comes with sudden onset of paraplegia or subarachnoid hemorrhage. A multidisciplinary approach balancing the patient's current neurological status against the potential risks and probable gains from any interventional and surgical procedure, is recommended. Endovascular embolization of spinal angioma in Cobb syndrome along with oral corticosteroid therapy seems useful in reducing morbidity and hastens recovery.

Authorship Contribution

Partha Pal, Sayantan Ray: Conception and design, **Sayantan Ray:** Drafting of the article, **Subhasish Dey:** Literature search, **Sumit Chakraborty**, **Sayantan Ray:** Analysis and interpretation of data, **Sayantan Ray, Sumit Chakraborty, Arunansu Talukdar:** Critical revision of the article for important intellectual content, **Partha pal**, **Sayantan Ray**, **Sumit Chakraborty**, **Subhasish Dey**, and **Arunansu Talukdar:** Final approval of the article

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