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Case Report

Cervicothoracic cutaneomeningospinal angiomatosis in adults (Cobb's syndrome): A case report of acute quadriparesis

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Abstract

Background: Cutaneomeningospinal angiomatosis or Cobb syndrome is a rare, not well understood phacomatosis that features metameric cutaneous and spinal arteriovenous malformations (AVMs). The first case was described in Boston in 1915, and since then, few more cases have been reported in the English literature. No case was found to be from Argentina.

Case Description: The authors present a 16-year-old boy with acute quadriparesis and respiratory failure who was diagnosed as Cobb syndrome and treated with microsurgery alone with very good results.

Conclusion: Authors believe that microsurgery, alone or combined with embolization, should be the mainstay of treatment. They also acknowledge Harvey Cushing's contribution to the description of the syndrome and propose the syndrome to be renamed as Cobb–Cushing syndrome.

Key Words: Acute quadriparesis, Cobb syndrome, cutaneomeningospinal angiomatosis, Harvey Cushing, phacomatosis, spinal arteriovenous malformation

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INTRODUCTION

The first case of a cutaneomeningospinal angiomatosis was described by Stanley Cobb in 1915,[2] and was treated by Harvey Cushing at the Peter Bent Brigham Hospital in Boston. The patient was an 8-year-old boy who developed acute back pain followed by flaccid paraplegia and was admitted on May 23, after 8 days of onset of symptoms with a presumptive diagnosis of anterior poliomyelitis.[2] However, in the original clinical notes prepared by Cushing himself, he remarked that "the definite upper level of anesthesia, the complete lower limb paralysis with exaggerated reflexes, visceral paralysis and priapism, make it fairly definite that there must be pressure against the cord" and "the presumptive diagnosis is of a congenital lesion pressing upon the cord, either a dermoid or an angioma," previously describing a 5-inch diameter nevus under the right scapula.[7]

Very little information is present in the medical literature since then, and the syndrome remains in obscurity regarding its physiopathology, genetics, natural history, and even diagnosis criteria and optimal treatment modality. We present a 16-year-old boy with undiagnosed Cobb syndrome who developed acute chest and right arm pain followed by severe quadriparesis and respiratory failure; the patient

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recovered substantially after microsurgical treatment without embolization. This is the first case presented from an institution of Argentina. We agree with Maramatton *et al*. that the contribution of Harvey Cushing to the description of this rare syndrome should be acknowledged,[7] and propose it to be renamed as Cobb–Cushing syndrome.

CASE DESCRIPTION

History and presentation

A 16-year-old boy was admitted to our center from another hospital to start plasmapheresis treatment, with the presumptive diagnosis of acute transverse myelitis. The symptoms started 8 days prior to his admittance in our institution, with sudden onset of severe right arm and chest pain, interpreted as a precordialgia secondary to ventricular arrhythmia, followed by progressive severe quadriparesis and respiratory failure. He was admitted few hours after the onset of symptoms to the regional local hospital and was sent to the Neurology Department at our hospital. Previous magnetic resonance imaging (MRI) findings were interpreted as a cevicodorsal acute transverse myelitis. The boy was previously healthy and had no medical history.

Examination

On examination, the patient presented flaccid paraplegia, with areflexia of the lower limbs, monoplegia of the right arm, and monoparesis of the left arm 2/5 proximal, 1/5 distal. He exhibited a complete sensory level at D4 and sphincter paralysis. He displayed a port-wine angioma in the right cervicodorsal and chest region [Figure 1]. The patient was already intubated on arrival. The spinal MRI (repeated in our institution for lack of quality of the first one) showed intraspinal tortuous blood vessels in the saggital plane from C5-D2, with no predominant variceal component and many flow voids images in the axial plane, occupying the whole thickness of the cord at the aforementioned levels [Figure 2]. It is noteworthy that the intraspinal signal hyperintensity was well above and below the lesion, making it probably responsible for the upper cervical symptoms, as well as the misdiagnosis of acute transverse myelitis from general practitioners who may lack experience in visualizing such images, in the setting of a poorly performed cervical-only MRI (not displayed in this paper).

The patient had no improvement from the symptoms after a 4-day treatment with plasmapheresis, and was presented to our department, where the final diagnosis was made. We programmed surgery 9 days after arrival prior to obtaining a cervical DSA showing right-sided predominance of arterial feeding pedicles and several tortuous varicosal drain veins [Figure 3].

Intervention and postintervention course

The patient was treated with a standard microsurgical procedure for spinal AVMs performed by the senior author (FRP) with no prior embolization. He was positioned supine, and C6-D1 bilateral laminectomy was performed. The dura was exposed and incised in the regular cefalocaudal fashion, showing underneath the varicosal draining shunted veins in the dorsal archnopial compartment [Figure 4]. With the use of bipolar cautery and the aid of small-sized transitory aneurismal clips, all the arterial feeders, both right and less predominant left-sided, were ligated and coagulated. Then, dissection proceeded in the usual manner as for any AVM, leaving the ligation of the main venous drainages as the last step before piecemeal removal of the lesion. The lesion was removed completely and surgical bed hemostasis was performed with oxidized cellulose polymer, taking extreme caution not to use cautery in the remaining cord. The patient did well in the immediate postoperative period, showing motor amelioration in the right arm at 1/5. His neurological function continued to improve in the following weeks after the surgery, with the residual

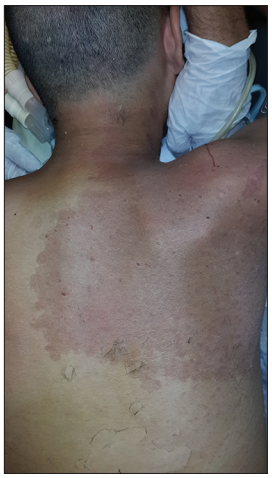

Figure 1: Port-wine angioma in the right cervicodorsal region of the patient compromising scapular region and shoulder. The angioma extends to the homolateral pectoral region (not seen in this picture)

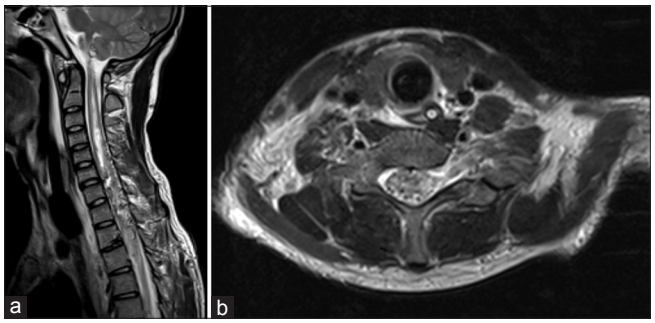

Figure 2: (a) Sagittal cervicodorsal T2-weighted MRI revealing the AVM from C5-D2, with multiple flow voids compromising the whole thickness of the spine and severe intensity changes up to well above in the first cervical spinal segment. (b) Axial plane T2-weighted MRI at the level of C7

capability of moving both upper limbs: 4/5 proximal, 3/5 distal left arm; 3/5 proximal, 2/5 distal right arm. He also showed improved respiratory function, being capable to breathe for his own at first and swallowing soft food afterwards. He was discharged to his local regional hospital 7 weeks after the surgery to continue physiotherapy and medical treatment for intercurrent nosocomial urinary tract infection.

In the subsequent control MRI and angiography, there was absence of lesion and significant lowering of the signal intensity above and below the lesion in the remaining spinal cord [Figure 5].

At the 6-month follow-up after surgery, the patient continued rehabilitating in his home town in an outpatient basis, with both upper extremities improving functional capacity to the extent of being capable of self-feeding and performing daily activities. In the last follow-up 1 year after the surgery, the patient functions in a wheelchair, moves both arms with minor deficits, and can attend school.

DISCUSSION

After the first published description of this rare syndrome made by Stanley Cobb more than 100 years ago, there have been several adult and pediatric case reports and cases series in the English literature, with literature reviews reporting 18 adult cases for 2009 and 45 cases in 2012, along with one article in Chinese reporting a series of 61 cases.[3,5,6] Most of the cases reported "typical" presentation of angiomatous-cutaneous stains and spinal AVM corresponding to the same metamere, whereas other cases reported Cobb syndrome cases with cutaneous cavernous angiomas (CAs) associated with spinal Cas.[9] A few cases have been presented as acute paraplegia, but none as acute quadriparesis.[1,6,10] Although the most widely accepted diagnosis criteria is that proposed by Kissel and Dureux,[4] which includes a skin nevus in the same segment as the spinal angioma (with or without visceral angiomatosis), the type and pathology of spinal angioma is still ill-defined. The genetic framework of this syndrome also remains unclear, although one study suggested an inherited disposition based in a family case of Cobb syndrome.[8,7] In the spinal arteriovenous lesions classification proposed by Spetzler *et al.*,[11] they correspond to the true spinal cord AVMs, extradural-intradural type (formerly known as juvenile or metameric type).

We believe that this syndrome, rare and often misdiagnosed, should be highly suspected in any patient presenting port-wine stains on the skin in a metameric disposition, and screening diagnostic spinal MRI should be warranted for both the patient and close family members.

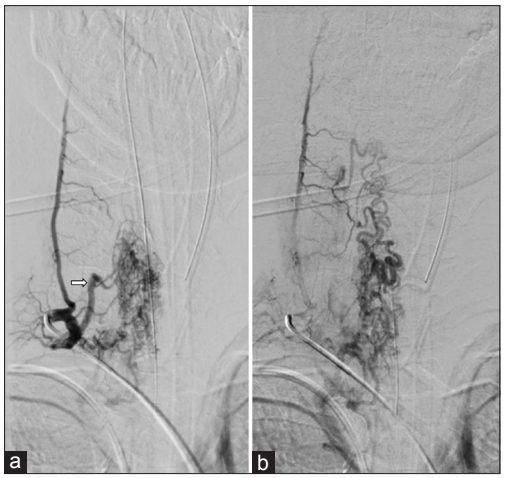

Figure 3: (a) Early arterial phase DSA in the right cervical region, showing a high-flow compact spinal AVM nidus with a main feeding trunk (white arrow) and multiple smaller ones. (b) Venous phase of the same angiogram, displaying multiple abnormal varicoceal draining veins extending well above the compact nidus in the cervical region

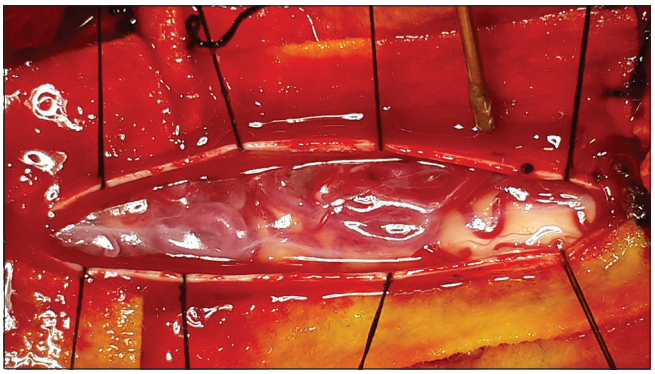

Figure 4: Intraoperative photography before the microsurgical resection, showing prominent tortuous vessels in the subarachnoid space

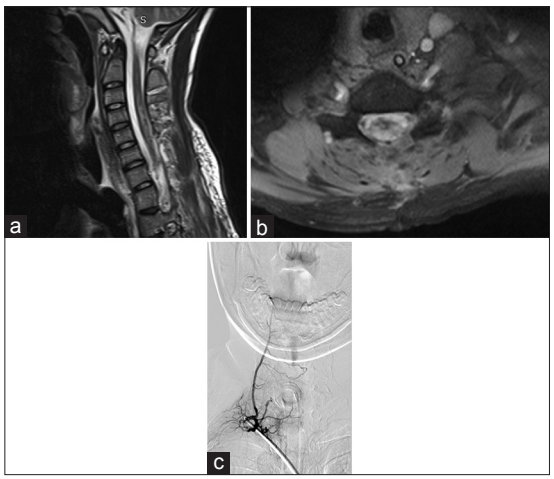

Figure 5: (a) Ten days postoperative sagittal MRI showing complete removal of the AVM and significant improvement of the hyperintensity in the cervical spinal cord [Figure 2]. (b) Same postoperative MRI in the axial plane at the level of C7 demonstrating no flow voids inside the spinal cord. (c) Seven weeks postoperative angiogram showing no residual AVM nidus

We also think that the optimal treatment modality is, as with most spinal AVMs, microsurgery (alone or combined with previous embolization) to both ligate and coagulate the feeding vessels and remove the mass effect upon the spinal cord, ideally before the bleeding occurs, with its devastating clinical outcome once the damage is established. We emphasize the role of microsurgery because it is the only treatment modality that offers (in selected cases of compact AVMs) complete removal of the malformation and can achieve very good results if diagnosis is followed by treatment in a timely manner. Further studies are needed to determine the best modality of treatment and better demarcate diagnostic criteria.

Finally, we agree with other authors opinion, that the contribution of Harvey Cushing should be acknowledged,[7] and propose the syndrome to be renamed as "Cobb–Cushing" syndrome, in view of the increasing number of cases presented so far and the importance of the lifetime work of the "Father of Neurosurgery."

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Conflicts of interest

There are no conflicts of interest.

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