Cobb Syndrome Associated with Spinal Cavernoma – Case Report

Síndrome de Cobb associada a cavernoma medular – Relato de caso

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Abstract

Cobb syndrome, or cutaneomeningospinal angiomatosis, is a rare condition that affects young adults, and its etiology has not been completely elucidated. It is characterized by a cutaneous sign, or stigma, associated with spinal or intracranial malformations. The symptoms are quite diverse, but, in most cases, the disease presents motor deficit and pain. The present study reports the case of a 48-year-old female patient, who initially sought dermatological medical care for a single skin lesion in the posterior cervical region. During the excision, it was noticed that the lesion had contiguous behavior to the deep anatomical planes, thus requiring the evaluation of the neurosurgical team. The purpose of this report is to describe this rare disease, covering more details about diagnosis and therapy.

Keywords
- arteriovenous malformation
- nevus
- angiomatosis
- Cobb syndrome

Resumo

Síndrome de Cobb, ou angiomatose cutâneo-meningo-espinhal, é uma condição rara que afeta adultos jovens, e sua etiologia não foi completamente elucidada. Caracteriza-se por um sinal cutâneo, ou estigma, associado a malformações espinhais ou intracranianas. Os sintomas são bastante diversos, mas, na maioria dos casos, a doença apresenta déficit motor e dor. O presente estudo relata o caso de uma paciente do sexo feminino de 48 anos, que inicialmente procurou atendimento médico dermatológico para uma única lesão cutânea na região cervical posterior. Durante a excisão, notou-se que a lesão apresentava comportamento contíguo aos planos anatômicos profundos, exigindo assim a avaliação da equipe neurocirúrgica. O objetivo deste relatório é descrever essa doença rara, abrangendo mais detalhes sobre diagnóstico e terapia.

Palavras-chave
- malformação arteriovenosa
- nevo
- angiomatose
- Síndrome de Cobb

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Introduction

Cutaneomeningospinal angiomatosis, a rare pathology that occurs in the spine, is characterized by the association of a vascular malformation affecting the skin, bone, and spinal cord, and it can also reach nerve roots in the same metamer.[1] This syndrome was first described in 1895 and was characterized by Stanley Cobb in 1915,[1] which is why it is also called Cobb syndrome (CS).

To date, only a few cases have been reported, with less than 50 cases in the English language, a series of 61 cases in China, and only one report in South America, all of which report one skin lesion associated with an arteriovenous malformation (AVM) involving the spinal cord.[2] The present study concerns a report of CS associated with a spinal cavernoma. This is the first case reported in Brazil, and no similar cases have been found until the date of this study.

Case Report

A 48-year-old female patient sought dermatological medical attention for the excision of a lesion in the posterior cervical region, which caused sporadic mild pain. During the procedure, the professional noticed that the lesion had visceral contiguity in depth, and requested additional tests for investigation, referring the patient for neurosurgical evaluation.

On physical examination, the lesion accompanied the C1 metamer, causing a bulging in the posterior cervical region (►**Fig 1**). The patient also had monoparesis (grade 4 +/5) in the left upper limb and diffuse hyperreflexia in the same limb. There were no structural changes, flaccidity, or tone abnormalities and no other health issues.

The magnetic resonance imaging (MRI) evidenced an association of the superficial lesion with adjacent tissues, up to the vertebra at the same level (C1), in addition to a vascular lesion, compatible with cavernoma (►**Fig 2**). This association confirmed the criteria for CS. A spinal digital angiography was also performed, and revealed the absence of contrast uptake, corroborating the diagnosis of cavernoma.

The patient was then submitted to neurosurgical treatment, with intraoperative neuromonitoring, and microsurgical technique (►**Fig 3**). Surgical access was performed through a 2-level laminectomy (C1 and C2) to expose the lesion, with subsequent opening of the dura mater and direct visualization of the lesion. The lesion was completely resected, and the patient evolved with improvement in the previous neurological deficit.

Discussion

Currently, the diagnostic criteria accepted for CS, proposed by Kissel and Dureux, are based on the presence of a skin nevus, in the same segment (dermatome/metamer) as the spinal vascular malformation, including or not visceral angioma. The diagnosis can be confirmed by magnetic resonance imaging.[1]

The incidence of this disease is unknown.[1,3] It is known that CS is a disease that affects mostly children and young adults,[3] and it is believed that there is a higher incidence in men.[2] The pathophysiology and vascular subtype of spinal angiomas are still poorly defined. The relationship of familiar genetic factors is not clear, although one study suggested an inherited disposition, when presenting an embryonic connection with ectodermal structures.[4] Although the mechanisms that lead to symptomatic manifestation are unknown, it is believed that the neurological deficit present in CS is a consequence of ischemia, secondary to diversion of the blood flow to the vascular injury, or to mechanical compression of the spinal cord.[2]

The clinical presentation is diverse, ranging from progressive evolution to sudden symptoms. Motor deficit (such as paraplegia), pain, fever, and signs of meningeal irritation may be present.[1] Motor disorders correspond to the earliest and most frequent symptoms.[1,3] The neurological physical examination does not present a characteristic finding, but bilateral patellar and Babinski sign have been described bilaterally.[5] However, there is no specific and determining sign or symptom for the characterization of CS.[5]

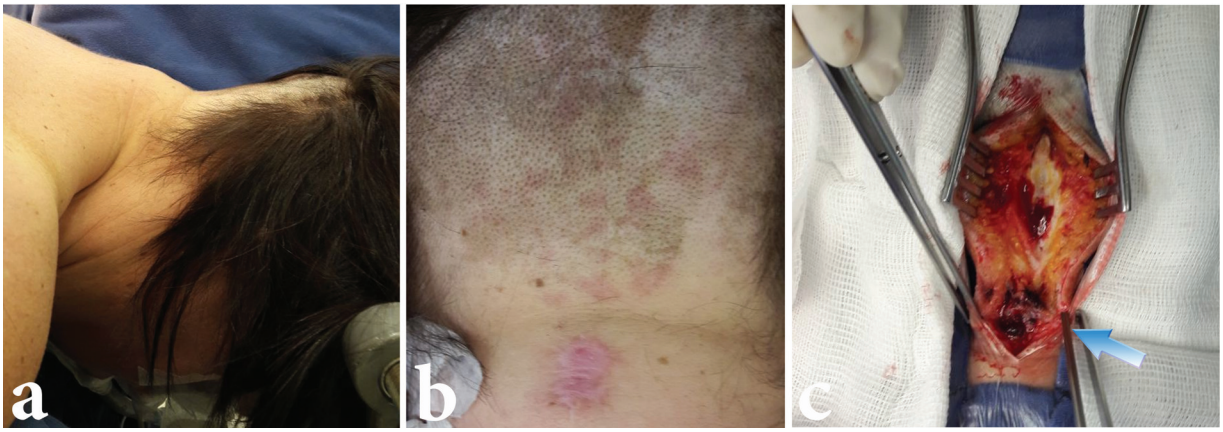

Fig. 1 (**a**) Bulking in the posterior cervical region evidenced after prone positioning of the patient. (**b**) Cutaneous nevus at the skin covering the suboccipital region and upper cervical region, in the C2 dermatome. Below, a scar from previous biopsy. (**c**) Subcutaneous exposure after surgical incision, evidencing contiguity of the lesion, extending from the superficial to deep planes (blue arrow).

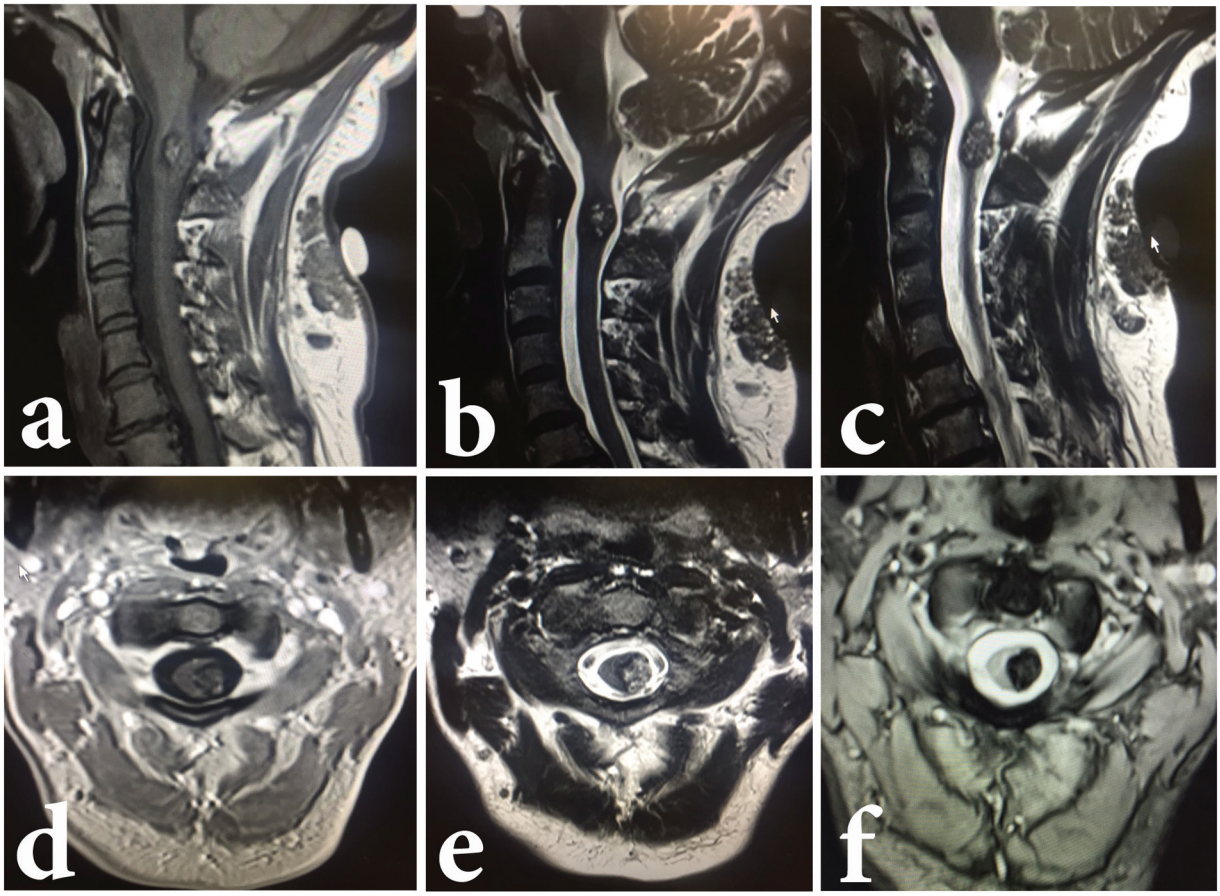

Fig. 2 (**a**) Sagittal T1-weighted magnetic resonance imaging (MRI) of the cervical spine, evidencing a hyperintense lesion, with a hypointense halo at the C1-C2 level. (**b, c**) Sagittal T2-weighted MRI of the cervical spine, evidencing the typical blueberry aspect of cavernomas, with a mixture of hyperintense and hypointense signals at the C1-C2 level. (**d**) Axial T1-weighted MRI of the cervical spine, showing the right lateral aspect of the lesion in relation to the cervical spine. (**e**) Axial T2-weighted MRI of the cervical spine, highlighting the cleavage plane between the lesion and the cervical spine, and the irregular signal. (**f**) Axial T2*-weighted MRI of the cervical spine, evidencing the vascular pattern (hyposignal) of the lesion.

The differential diagnosis should include diseases that present with cutaneous sign and spinal or intracranial malformations, such as Klippel-Trenaunay-Weber syndrome, Sturge-Weber syndrome, Wyburn-Mason syndrome, and Osler-Rendu-Weber syndrome.[4]

There is no consensus regarding a current treatment modality, since the syndrome remains obscure from the genetic point of view, natural history, and even specific diagnostic criteria. The application of the classification of AVMs is made in order to guide the treatment, since the majority of the reports described have been made with spinal AVMs, and not with cavernomas, as in the present case.[3]

In the case presently described, microsurgical treatment was proposed for excision of the spinal cord injury, with intraoperative neuromonitoring. The aim was to remove the mass effect causing spinal compression, and, also, prophylaxis of bleeding, minimizing neurological damage. The treatment modalities described by the literature review include the combination of embolization, neurosurgical intervention, use of corticosteroids, and radiotherapy.[1] It is not possible to infer which treatment leads to the highest clinical improvement, minimizing neurological sequelae, a fact that highlights the importance and the need for further studies about this entity.[1] The report published by Linfante et al.[6] suggest that endovascular therapy, through embolization, can be performed as a preoperative treatment, and possibly as a definitive treatment. In the present case, because the vascular lesion was configured as a cavernoma, endovascular treatment could not be applied.

The present report aims to highlight that CS is remotely suspected; however, it should be kept in mind when patients present with violet skin lesions in the spinal topography. Screening should be encouraged to avoid major surgical complications or neurological sequelae.[3]

Conclusion

Although it is rare, in patients with signs and symptoms of spinal compression, associated with vascular lesions in the skin in the same segment of the motor deficit, CS should be considered as a diagnostic hypothesis for early diagnosis and appropriate treatment.

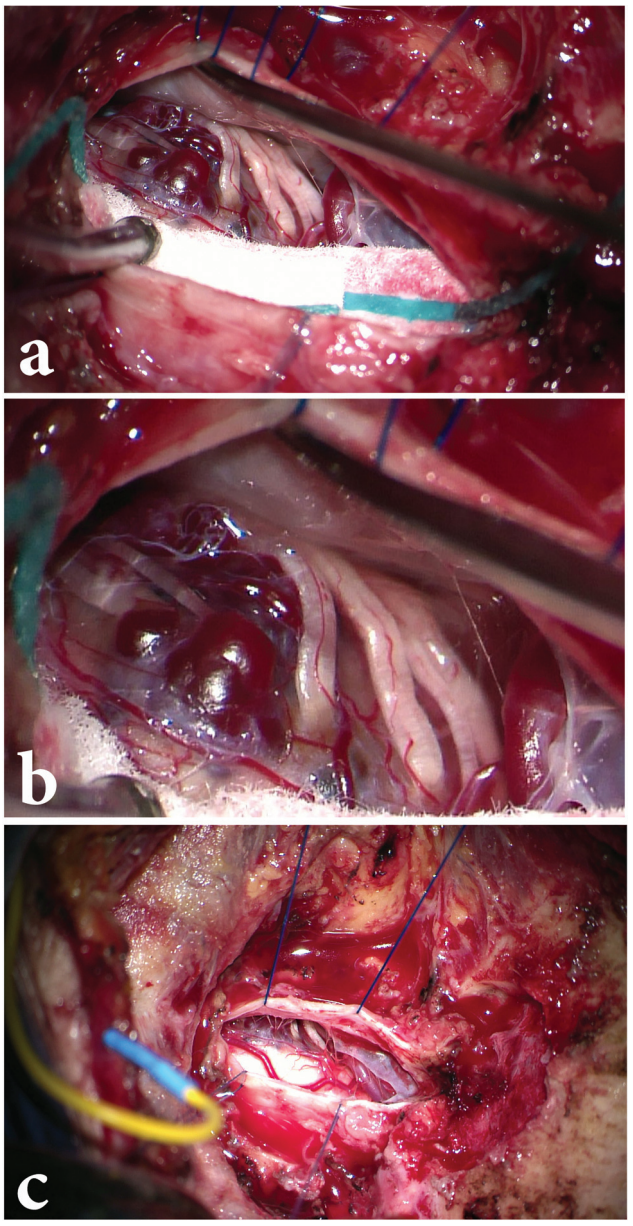

Fig. 3 (**a**) Intraoperative image. Initial exposure, with the cavernoma appearing in the lateral aspect of the cervical spine, in close contact with the C2 roots, on the right side. (**b**) Intraoperative image. After initial dissection, the lesion became more apparent, and the comprised nerve roots, superficial and deep, are highlighted. (**c**) Intraoperative final image. Complete excision of the lesion with preservation of the nerve roots and arterial supply of the cervical spine.

Conflict of Interests

The authors have no conflict of interests to declare.

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