

Paraplegia Revealing Cobb's Syndrome

Capatas S*, Hali F, Baline K, Chiheb S

Department of Dermatology and Venereology, UHC Ibn Rochd of Casablanca

DOI: [10.36347/SJMCR.2019.v07i10.004](https://doi.org/10.36347/SJMCR.2019.v07i10.004)

| Received: 20.09.2019 | Accepted: 27.09.2019 | Published: 29.10.2019

*Corresponding author: Sophia Capatas

Abstract

Case Report

Cobb syndrome is an arteriovenous malformation defined by association of vascular cutaneous, muscular, osseous and medullary lesions at the same metamere or spinal segment. We report a case of 42-year-old woman who presented fifteen years ago an angiomatous cutaneous patch in abdominal and dorsal region associated with paraplegia and sphincter disorders. Clinical examination revealed a vegetal papulo-nodular angiomatous patch extending from the right hypochondrium to the dorsal region D10-D11. An abdominal angiography scan showed a thickening of the subcutaneous adipose tissue in band affecting the anterior, lateral and posterior abdominal wall. The medullary MRI has objectified a dorsal intramedullary process compatible with a lesion of vascular origin. The patient was referred for neurosurgery for management. Cobb syndrome is a rare arteriovenous malformation that can remain asymptomatic for a long time. Clinical follow-up and radiological exploration are necessary in any patient with metameric vascular lesions to prevent neurological deficit.

Keywords: Cobb syndrome, abdominal angiography, medullary magnetic resonance imaging, spine, arteriovenous malformation, surgery.

Copyright © 2019: This is an open-access article distributed under the terms of the Creative Commons Attribution license which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use (NonCommercial, or CC-BY-NC) provided the original author and source are credited.

INTRODUCTION

Cobb syndrome is a complex arteriovenous malformation (AVM) with a metameric distribution involving the skin and the spinal cord. The common neurological manifestations are pain and motor deficits due to medullary compression. This entity was first described by Stanley Cobb in 1915, presenting the case of an 8-year-old boy who developed flaccid paralysis [1]. Mean age at diagnosis is 36.4 years, with a peak between the third and fifth decades [2]. Cobb syndrome is rare; it has an underreported prevalence, and the number of documented cases of Cobb syndrome in the literature is less than 50. We describe the case of a 42-year-old woman who presented a dark red patch affecting dermatome D10-D11 and neurological signs as paraplegia and sphincter disorders. Early diagnosis in our patient would have prevented permanent sequelae, this is why it is important to conduct imaging tests when cutaneous vascular display a segmental distribution.

CASE SYNOPSIS

A 42-year-old woman living in the countryside, with no significant past or family history, presented five years ago a progressive neurological deficit in the legs leading to paraplegia. She developed

urinary and fecal incontinence. There was no history of previous trauma or infectious disease. It started first by sudden development of weakness and decreased muscle strength in both lower extremities. Despite the motor disability, she had never seen a doctor before. She also had a history of lesions like a "bunch of grapes" that were painful, extending from abdominal to dorsal region. Clinical examination revealed a papulo-nodular dark red patch with a metameric layout involving the right abdomen to the dorsal vertebrae D10-D11 (Fig 1, 2, 3). It was ulcerated giving exit to pus and blood in the right flank. Palpation noted an infiltration in some places and a presence of a thrill. Neurological examination found paraplegia with a sensory deficit in her legs and an umbilical sensitive level. An abdominal angiography scan showed a thickening of the subcutaneous adipose tissue in band affecting the anterior, lateral and posterior abdominal wall. The medullary MRI has objectified a dorsal intramedullary process compatible with a vascular lesion. Surgery wasn't recommended in her case. The patient was referred to interventional radiology department for embolization.



Fig-1: Papulo-nodular abdominal patch as a “bunch of grapes”



Fig-2: Metameric disposition of the abdominal patch



Fig-3: Ulceration of the patch next to D10-D11 vertebrae



Fig-4: A dorsal intramedullary process compatible with a vascular lesion in medullary MRI

CASE DISCUSSION

Cobb syndrome or “Spinal Arteriovenous Metameric Syndrome” was first described in 1895. Stanley Cobb reported in 1915 the case of an 8-year-old boy who presented with acute paraplegia to Harvey Cushing’s surgical service at the Peter Bent Brigham Hospital. Cushing’s resident, Stanley Cobb, noticed “areas of dark reddish skin” over the ninth to 12th ribs on the right side of the patient’s back, which he retrospectively identified as nevi. Cobb reported the case and subsequently was credited with the eponym “Cobb syndrome [3].”

This entity is a rare, non-familial condition coupling cutaneous vascular lesions with arteriovenous malformations involving the spinal cord in the same metamere as the cutaneous lesion [4]. The incidence of Cobb syndrome is unknown, but may be more common than the 39 cases in the literature suggest, insofar as only symptomatic patients are likely to be diagnosed [5]. The genetic framework is unclear, although the authors of one study found a clustering of Cobb syndrome in a family, hinting at an inherited predisposition [6]. The embryologic origin of the blood supply to the vertebral and spinal cord derives from the segmental dorsal arteries. This finding corresponds to the metameric origin of the arteriovenous malformation vessels that create the Cobb syndrome lesions. Many

theories exist regarding the etiology of the neurologic manifestations of Cobb syndrome, including blood steal syndrome -that produces cord ischemia-, cord compression and venous hypertension [6, 7]. There is a slight male predominance associated with this syndrome [8]. The most frequent vascular lesion location is thoracic spine. Pathophysiology of this syndrome is poorly understood so the correct management remains unknown. Cord compression secondary to venous malformation, venous hypertension, and vascular steal syndrome may account for neurological deficits. Most symptoms in adults appear as a gradual progressive neurological deficit, as in our case. The most common dermatologic manifestations comprise nevus flammeus and angiokeratomas that can ulcerate and become infected. Concerning radiological explorations, angiography increases understanding of the angioarchitecture because spinal vascular malformation in Cobb's syndrome has a distinct blood supply from the normal spinal cord [6]. The optimal treatment modality for this entity remains unknown because of the syndrome's rarity and poorly understood pathophysiology [9].

Because surgery may be associated with unacceptable risks, endovascular embolization remains a reasonable therapeutic approach. Corticosteroids therapy before intramedullary embolization may reduce extension and number of spinal arteriovenous embolization procedures [10, 11]. For our patient, the surgery was contraindicated and she was referred to the interventional radiology department for embolization.

CONCLUSION

Cobb syndrome is rare vascular malformation that is still misdiagnosed. It accounts for less than 15% of arteriovenous malformations in the spinal cord, and can remain asymptomatic for a long time. It's characterized by a spinal vascular abnormality in association with a vascular skin lesion of the same metamere. Current treatment options are embolization and neurosurgical intervention in some cases. Clinical follow-up and radiological exploration are necessary in any patient with metameric vascular lesions in order to prevent neurological deficit.

REFERENCES

1. Cobb S. Haemangioma of the spinal cord: associated with skin naevi of the same metamere. *Annals of surgery*. 1915 Dec;62(6):641.
2. Zevgaridis D, Medele RJ, Hamburger C, Steiger HJ, Reulen HJ. Cavernous haemangiomas of the spinal cord. A review of 117 cases. *Acta neurochirurgica*. 1999 Mar 1;141(3):237-45.
3. Maramattom BV, Cohen-Gadol AA, Wijdicks EF, Kallmes D. Segmental cutaneous hemangioma and spinal arteriovenous malformation (Cobb syndrome): Case report and historical perspective. *J Neurosurg* 2005;3:249-52.
4. Shim JH, Lee DW, Cho BK. A case of Cobb syndrome associated with lymphangioma circumscriptum. *Dermatology*. 1996;193(1):45-7.
5. Veznedaroglu E, Nelson PK, Jabbour PM, Rossenwasser RH. Endovascular treatment of spinal cord arteriovenous malformations. *Neurosurgery*. 2006;59 (Suppl.):S3202-9
6. Mercer R, Rothner AD, Cook SA, Alfidi RJ. The Cobb syndrome. *Cleveland Clinic Quarterly*. 1978;45(1).
7. Bao YH, Ling F. Classification and therapeutic modalities of spinal vascular malformations in 80 patients. *Neurosurgery*. 1997 Jan 1;40(1):75-81.
8. Halbach VV, Higashida RT, Dowd CF, Fraser KW, Edwards MS, Barnwell SL. Treatment of giant intradural (perimedullary) arteriovenous fistulas. *Neurosurgery*. 1993 Dec 1;33(6):972-80.
9. Johnson WD, Petrie MM. Variety of spinal vascular pathology seen in adult Cobb syndrome: report of 2 cases. *Journal of Neurosurgery: Spine*. 2009 May 1;10(5):430-5.
10. Soeda A, Sakai N, Iihara K, Nagata I. Cobb syndrome in an infant: treatment with endovascular embolization and corticosteroid therapy: case report. *Neurosurgery*. 2003 Mar 1;52(3):711-5.
11. Akyüz C, Yarış N, Kutluk MT, Büyükpamukçu M. Management of cutaneous hemangiomas: a retrospective analysis of 1109 cases and comparison of conventional dose prednisolone with high-dose methylprednisolone therapy. *Pediatric hematology and oncology*. 2001 Jan 1;18(1):47-55.
12. Weber TR, Connors RH, Tracy TF, Bailey PV. Complex hemangiomas of infants and children: individualized management in 22 cases. *Archives of Surgery*. 1990 Aug 1;125(8):1017-21.