COBB SYNDROME
SPECTRUM OF THORACIC IMAGING FINDINGS

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INTRODUCTION

• Cobb syndrome, or cutaneomeningospinal angiomatosis, is a nonfamilial neurocutaneous disorder characterized by spinal vascular abnormalities in association with a vascular lesion of the skin at the same metamere.\(^1,2\)

• More recently, was recognized to be a part of a larger group of multiple arteriovenous shunts with metameric link, and the syndrome has been renamed to “spinal arteriovenous metameric syndrome” (SAMS)\(^3\)

• Patients are diagnosed at any age but most commonly in late childhood and typically after the onset of neurologic symptoms such as paraplegia, quadriplegia, back pain, and, less frequently, fatigue due to heart failure.\(^4,5\)

1. Niimi Y et al, 2013
2. Komiyama M, 2014
5. Schirmer CM et al, 2012
OBJECTIVES

• This report aims to review a series of cases focusing on computed tomography (CT) and magnetic resonance (MRI) imaging findings, correlating to the literature to support the diagnosis of Cobb syndrome, in the context of thoracic/intramedullary arteriovenous malformations (AVM) investigation.
METHODS

• Review of current literature on "PubMed" database.
  
  o Using the descriptors defined by MeSH (Medical Subject Headings), the research was performed as follows: ((magnetic resonance OR computed tomography)) AND (("Cobb syndrome") OR cutaneomeningospinal angiomatosis OR spinal arteriovenous metameric syndrome).

• Retrospective analysis of two cases from our hospital, identifying the main structural MRI and CT imaging findings to support the Cobb Syndrome diagnosis.
CASE REPORT – CASE 1

• An 23-year-old male patient with progressive paraparesis and paresthesia of the lower extremities for 9 years, intensified in the past 13 days.

• None other symptoms related or familial history were observed.

• Physical examination:
  o Multiple vascular skin lesions, some of them “port-wine stain” over his left upper midback (T3–T10).
  o Muscle strength rating scale: 0(zero) (paraplegia).
  o Reduced tactile sensitivity and exalted deep reflexes.
Hemangiomatous skin lesion, telangiectasias and “port-wine stain” in left hemithorax.

Axial CT C+, lung window, demonstrating mosaic attenuation pattern (perfusion disturbance) and ectasia of intrapulmonary arterial branches.

Axial MR T1WI C+ showing an intramedullary vascular malformation with spinal cord compression and extension to the left chest wall. Note also enlargement of foraminal basis and spinal canal.

MRI MIP C+ demonstrating a vascular malformation with brachiocephalic vein and intercostal vessels communication in the axillary region and left chest wall of the same metamere.
A 16-year-old male patient with vascular malformations of the upper limb and right shoulder girdle since childhood. Complaining of "overweight", pain and paresthesia without loss of muscle strength.

None other symptoms related or familial history were observed.

Physical examination:
- Hemangiomatous and telangiectatic skin lesion in posterior region of the right hemithorax.
- Muscle strength, tactile sensitivity and deep reflexes preserved.
Figure 5. Hemangiomaticous and telangiectasic skin lesion in the posterior region of right hemithorax.

Figure 6. Axial CTA showing the previously described findings. Note also cervical bone remodeling with enlargament of foraminal basis.

Figure 7. Sagital T2WI MRI demonstrating an intramedullary vascular malformation determining spinal cord compression and enlargement of spinal canal (yellow arrow).

Figure 8. MRI Coronal view MIP C+ demonstrating a vascular malformation communicating to the brachiocephalic trunk, cervical and intercostal vessels. Note the supraclavicular soft tissue bulging, as well as the infiltrative aspect in the chest wall and right shoulder girdle of the same metamere.
DISCUSSION

- Cobb syndrome is a rare disease which cutaneous, muscular and/or bony vascular lesions, as well as spinal or paraespinal vascular lesions are found in the same metamere.

- It is classified, according to the International Society for the Study of Vascular Anomalies (ISSVA), into the subgroup of vascular malformations associated with other anomalies. 1-3

- Treatment is a great challenge and in many cases requires the combination of embolisations and subsequent surgeries to reduce the morbidity and mortality of this pathology 4,5

1. Niimi Y et al, 2013
2. Komiyama M, 2014
5. Schirmer CM et al, 2012
DISCUSSION – Clinical Findings

- The cutaneous manifestations range from macular “port-wine stains” to various types of vascular lesions/flammeus nevus. This findings have a common metameric origin of the AVM that create the cutaneomeningospinal angiomas. 3,6

- Cobb syndrome is typically diagnosed following the onset of neurological symptoms. Onset of signs usually manifest over weeks to years, but a sudden onset of weakness with rapid progression has also been reported. 3,7

- Neurological presentations can vary from monoparesis to sudden-onset paraplegia or quadriplegia. Less common signs include meningismus, headache, fever, and gluteal and limb hypertrophy. 3

7. Rodesch G et al, 2013
Figure 1 (image of case 1). Hemangiomatous skin lesion, telangiectasias and “port-wine stain” in left hemithorax.

Figure 5 (image of case 2). Hemangiomatous and telangiectasic skin lesion in posterior region of the right hemithorax.

Figure 9. Photograph of the patients “port-wine-colored” cutaneous angioma. (Soeda et al, 2003)
• Vascular skin nevus found with Cobb syndrome is accompanied by a large variety of vascular pathologies, however arteriovenous malformations are a main spinal vascular pathology. 3,7,9

• Vascular anomalies can be divided into vascular neoplasms (e.g. hemangioma) and vascular malformations. Vascular malformations are subdivided into: 3,9,10
  
  (a) low-flow and
  
  (b) high-flow malformations (contain arterial components).

• The intraspinal lesions are usually AVMs (high flow lesion) and rarely angiomas (low-flow lesion). 3,10

7. Rodesch G et al, 2013
**Figure 3 (image case 1).** Axial MRI T1WI C+ showing an intramedullary vascular malformation with spinal cord compression and extension to the left chest wall. Note also enlargement of foraminal basis and spinal canal.

**Figure 8 (image case 2).** MRI Coronal view MIP C+ demonstrating a vascular malformation communicating to the brachiocephalic trunk, cervical and intercostal vessels. Note the supraclavicular soft tissue bulging, as well as the infiltrative aspect in the chest wall and right shoulder girdle of the same metamere.

**Figure 10.** Sagittal T2WI MRI demonstrating an intramedullary AVM between T6-T9 (black arrow). *(Linfante I et al, 2012)*
DISCUSSION – Imaging Findings: thoracic

- Although vascular anomalies are the most frequent imaging findings, some other findings can also be observed at thoracic investigation.

- Theses are usually due to high-flow malformations – perfusion disturbance, cardiomegaly, vascular ectasia, congestive heart failure, others.

**DISCUSSION – Imaging Findings: thoracic**

**Figure 2 (image case 1).** Axial post-contrast chest CT, lung window, demonstrating mosaic attenuation pattern (perfusion disturbance) and intrapulmonary artery branches ectasia.

**Figure 11 (image of case 1).** Coronal post-contrast chest CT, mediastinum window, demonstrating azigos venous system and superior vena cava ectasia, associated with cardiomegaly (right chambers enlargement).

**Figure 12 (image of case 2).** Coronal Computed Tomography Angiography (CTA) demonstrating also suevena cava ectasia, associated with cardiomegaly.
Figure 13 (image of case 2). Axial CT, bone window, demonstrating intra and extra-bone marrow vascular communication determining bone remodeling areas and lytic lesions in the right superolateral scapula portion and right humeral head.
DISCUSSION – Diagnostic

- Computed tomography (CT) and MRI are useful modalities to assess the extension of the lesions. ³,¹⁰
  - MRI is better in displaying soft-tissue, AVM, angiomas.
  - CT is better in showing a pulmonary parenchyma, mediastinum and bone.

- The final diagnosis of the syndrome depends on angiography. ³
DISCUSSION – Differenciel Diagnosis

- In general, the differencial diagnosis should be made on the basis of clinical history, physical and imaging finding. Nevertheless, most cases may cause diagnostic doubt. ⁹,¹⁰

- In vascular malformations associated with other anomalies group, the differential diagnosis may be made on basis of vascular malformations: ³,⁷,⁹,¹⁰

(a). low-flow malformations:
- Klippel-Trénaunay Syndrome
- Proteus Syndrome

(b). high-flow malformations:
- Parkes Weber Syndrome
- Rendu-Osler-Weber
- Vascular Metameric Sd.

7. Rodesch G et al, 2013
LIMITATIONS OF THE STUDY

• Due to rarity of disease, the limitations were:
  o Sample size.
  o Lack of available and/or reliable data.
  o Lack of a standard pattern to confirm or exclude the diagnosis.
Neurological symptoms
- Paresia, parenthesias...

Cutaneous manifestations
- “Port-wine stain” to vascular lesions...

Imaging findings
- Vascular malformation; AVM...

Cobb syndrome
• Cobb syndrome is an unusual entity and should be considered when there is an association of cutaneous manifestation and underlying neurological deficit.

• Presence of asymmetric vascular skin nevus associated to Cobb syndrome should incite the investigation of corresponding intramedullary AVM to approach the diagnosis of this metameric pathology.
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