A Case Report Of Cobb Syndrome

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INTRODUCTION:
Cobb syndrome or Cutaneomeningospinal Angiomatosis or Spinal Arteriovenous Metameric Syndrome is a rare non-inherited metameric disorder, of which only 50 cases have been documented worldwide. It is characterised by coupling of a vascular abnormality of the spinal cord, with associated vascular skin lesion of the same metamere.

MATERIALS & METHODS:
A 12-year-old boy was referred to us to rule out liposarcoma. He had a lipoma excised from his left upper back at 4-year-old and presented with increasing swelling over his right back and gradual onset spastic paraplegia over 1 month. Upon examination, a 18x15cm hyperpigmented vascular lesion was noted in the right posterolateral lower thoracic chest wall associated with a large subcutaneous soft tissue mass extending from the lower thoracic spine medially in continuity with the hyperpigmented lesion laterally.

RESULTS:
CT TAP and angiography demonstrated an AVM at the right lateral and posterior upper chest and upper abdominal wall, measuring 17.6 x 19.3 x 21.1cm (APxWxCC).
Spinal angiography demonstrated a capillary malformation arising from the right T8-T11 posterior intercostal and right L1-L2 lumbar arteries.

Figure 1: CT-angiogram showing the spinal cord vascular abnormality

The patient was diagnosed with Cobb syndrome. A multidisciplinary approach to patient care was undertaken, with the involvement of neurosurgical, interventional radiologist and orthopaedic oncology. Treatment is ongoing at the time of preparation of this abstract.

DISCUSSIONS:
Cobb syndrome was first described by Cobb in 1915. Unlike spinal trauma, infection or tumour, the neurological deficit in Cobb Syndrome can evolve over a long period of time, sometimes as long as months. Neurology is believed to result from venous hypertension, cord compression and blood steal syndrome.

CONCLUSION:
Although it is very rare, any encounter of a cutaneous vascular lesion with gradual neurological deficit must raise a suspicion of Cobb Syndrome.

REFERENCES: