Segmental cutaneous hemangioma and spinal arteriovenous malformation (Cobb syndrome)

Case report and historical perspective

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The discovery of either a dermatomal cutaneous nevus or a spinal arteriovenous malformation (AVM) should raise the suspicion of Cobb syndrome. The Cobb syndrome is a neurocutaneous syndrome in which there are metameric cutaneous and spinal AVMs. The authors present the case of a patient with acute cervical myelopathy and subtle cutaneous hemangiomas in whom a cervical perimedullary fistula was discovered. They also acknowledge Harvey Cushing’s contribution to the recognition of this syndrome.

KEY WORDS • hemangioma • arteriovenous malformation • Cobb syndrome • Harvey Cushing

Cobb syndrome exhibits a resemblance to other neurocutaneous syndromes such as Klippel-Trenaunay-Weber syndrome, Wyburn-Mason syndrome, Sturge-Weber syndrome, and Osler-Rendu-Weber syndrome. In each of these syndromes, cutaneous stigmata accompany underlying spinal or intracranial vascular malformations. In particular, as Cushing noted, this syndrome parallels Sturge-Weber syndrome as leptomeningeal angiomatosis coexists with facial hemangiomas. We present the case of a patient with Cobb syndrome in whom we observed a subtle dermatomal cutaneous hemangioma and a cervical cord AVM. We will acknowledge, for the first time, Cushing’s contribution to the recognition of this syndrome.

Case Report

History and Presentation. This 17-year-old boy was admitted for acute quadriplegia accompanied by neck pain. He had an 8-year history of intermittent tingling and numbness in his right arm and neck pain on exertion. Occasionally, he would awaken from sleep with right-arm numbness of 5 to 10 minutes’ duration. His history was remarkable for nocturnal enuresis until the age of 16
years. On the day prior to admission, his neck pain worsened, becoming continuous. Overnight, flaccid weakness developed in all four limbs.

**Examination.** On examination, he exhibited a flaccid quadriplegia, areflexia, and a sensory level at C-4. Spinal MR imaging revealed tortuous blood vessels in the cervical subarachnoid space with a large variceal component dorsal to the C5–7 spinal cord levels (Fig. 2 left). The varix compressed and flattened the cervical cord. Spinal angiography demonstrated a C5–7 perimedullary fistula with feeding structures arising from a right superior intercostal vessel (Fig. 2 center). The venous plexus drained cranially through multiple dilated channels in the posterior fossa into the superior petrosal sinus. Subsequent examination revealed small flat irregular cutaneous hemangiomas on his right fourth finger along the C-8 dermatome and another one on his right shoulder in the T-1 distribution (Fig. 2 right).

**Intervention and Postintervention Course.** The AVM was embolized by placing platinum Micro and hydrocoils to obliterate the variceal component of the AVM. Subsequent spinal MR imaging demonstrated progressive resolution of the tortuous vessels and the compressing varix in the cervical subarachnoid space. Three-month follow-up angiography revealed no residual flow into the fistula (Fig. 3). The patient’s two siblings were noted to have facial port-wine stains in the ophthalmic and maxillary divisions of the trigeminal nerve, raising concerns about coexisting intracranial AVMs. Cranial computerized tomography scans obtained in these individuals were normal.

During the six-month follow-up examination we ob-
served ongoing improvement in spastic quadriplegia, but
the patient remained unable to ambulate.

**Discussion**

In 1915, an 8-year-old boy presented with acute para-
plegia 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 retro-
spectively identified as nevi. Cobb reported the case and
subsequently was credited with the eponym “Cobb syn-
drome.” We found this patient’s original hospital record
in the Cushing tumor registry at the Yale Department of
Neurosurgery. A review of this original case record yields
an insight into the thought process and intricate document-
tation that characterized Cushing’s work. It contains dia-
grammatic representations of the nevus and operative
findings. In his operative note Cushing wrote, “It seemed
futile to attempt to ligate any of the vessels and the dura
therefore was left widely open and the wound closed as
usual in layers without a drain.”

Cushing used the term “cavernous pial arachnoid angi-
oma” to describe this entity. It is unfortunate that Cush-
ing’s contribution was omitted. We can only speculate that
the eponym would have been more widely recognized if
the contribution of the “Father of Neurosurgery” was in-
cluded.

The literature contains numerous case reports of Cobb
syndrome, but only one collated series, falling far short
of earlier estimates. Nevertheless, identification of this
syndrome is imperative as it reiterates the embryological
linkage of neuroectodermal structures and the indolent
nature of these malformations. More importantly, the pre-
ence of dermatomal cutaneous nevi should lower the
threshold for imaging of the neuraxis and encourage exami-
nation of close family members for similar stigmata.

Our patient harbored cutaneous hemangiomas in a
C8–T1 dermatomal pattern in conjunction with a lower
cervical AVM. The hemangiomas were subtle but linear,
and they corresponded to the spinal metamere containing
the AVM. A similar situation arises in the Klippel-Tre-
naunay syndrome, where venous varicosities and limb
hypertrophy accompany cutaneous vascular nevi or port-
wine stains. Cobb’s syndrome is distinguished from Klip-
pegel-Trenaunay syndrome by the absence of these addi-
tional findings; indeed cutaneous hemangiomas are the
sole clues to metameric AVMs. Cutaneous angiomas can
be acral, small, and overlooked unless their segmental dis-
tribution is recognized.

The genetic framework of Cobb syndrome is unclear,
although the authors of one study found a clustering of
Cobb syndrome with flame angiomas in a family, hinting
at an inherited predisposition. The patient in our case also
had two siblings with unusual facial angiomas, which rais-
es the suspicion of a familial phacomatosis. Although the
results of the neuroimaging studies obtained in the sib-
lings were normal, we believe that the discovery of unusu-
al cutaneous angiomas in isolated or multiple family
members warrants imaging of the neuraxis to exclude the
presence of concomitant vascular malformations.

The optimal treatment modality for the spinal vascular
malformations found in Cobb syndrome remains un-
known because of the syndrome’s rarity and poorly under-
stood pathophysiology. For the same reasons, the natural
history of this condition remains elusive. Cord compres-
sion secondary to venous angioma, venous hypertension,
and vascular steal syndrome may account for neurological
deficits. Because total resection of the racemose angiomas
may be associated with unacceptable risks, endovascular
embolization remains a reasonable therapeutic approach.
Long-term evaluation of embolization therapy may clari-
fy its efficacy.
Conclusions

Cobb syndrome is a segmental neurocutaneous syndrome associated with metameric spinal cord vascular malformations. Endovascular embolization may provide a reasonable option to treat the spinal AVM. Although Cobb alone has been credited with the description of this syndrome, we believe that Cushing’s description and contribution to the syndrome warrant acknowledgment.

References


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