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Neuro-vascular Abnormalities with a Metameric Nevus: A Case Report and Review of Cobb Syndrome

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Introduction

Cobb syndrome, also known as cutaneomeningospinal angiomas, first described by Berenbruch in 1890 and reported by Cobb in 1915, is a rare disorder characterized by spinal vascular malformations and a corresponding dermatomal vascular nevus.^{1,2} It usually presents in children and adults as lower extremity weakness, back pain, sensory loss below the affected level, or paralysis. Onset may be acute, gradual, or follow a stuttering course. Symptoms are thought to result from rupture or expansion of the angioma leading to direct compression of the spinal cord or ischemia due to diverted blood flow.^{3,4} We report a case of a 29-year-old female with a constellation of congenital neuro-vascular abnormalities who presented with lower extremity paresthesias and urinary retention and was found to have a portwine stain in the S1 and S2 dermatomes without a corresponding spinal angioma on imaging.

Case Report

A 29-year-old female presented in 2006

with a chief complaint of neck pain, headache, and numbness and tingling of both hands without prior trauma. Magnetic resonance imaging (MRI) of the cervical spine revealed a Chiari type I malformation with herniation of the cerebellar tonsils 15 mm below the foramen magnum as well as a syrinx from C2 to C4 (Figure 1). She underwent a suboccipital craniectomy with C1 laminectomy and duraplasty. Her headaches, described as bioccipital, pulsatile, and exacerbated by exertion, persisted after the surgery despite treatment with multiple prophylactic agents, trigger point injections, and occipital nerve blocks. Magnetic resonance angiography (MRA) revealed two incidental aneurysms in the right internal carotid artery (Figure 2). Over the next five years, she underwent four additional decompressions and revisions of the posterior fossa with subsequent periods of relief followed by inevitable return of her headache. The patient is adopted so she does not have her family history.

In 2009, the patient began to experience urinary retention and difficulty initiating micturition. She also developed paresthesias in the

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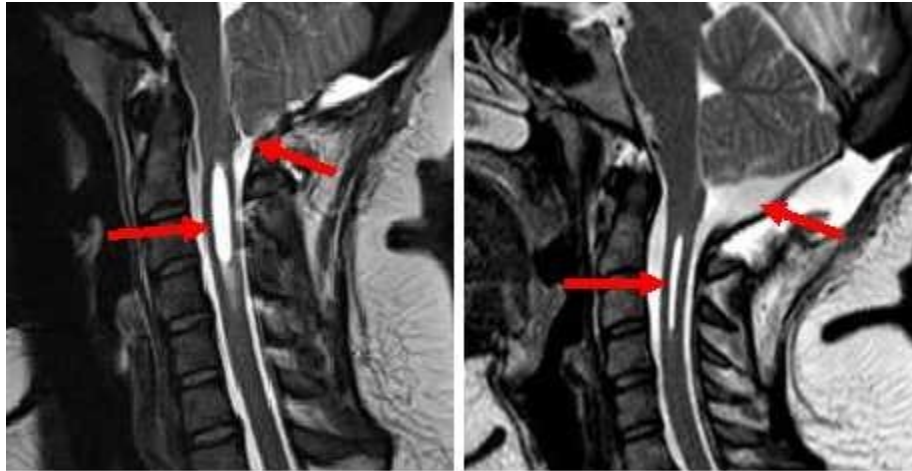


Figure 1: **Chiari type I and syrinx pre-op (left) and post-op (right).** T2-weighted images of cervical spine pre- and post-op demonstrate reduction in size of syrinx and decompression of posterior fossa and foramen magnum.

groin radiating down to the left leg as well as her bilateral soles. After a negative workup including multiple lumbar punctures, nerve conduction studies, electromyography, autoimmune panel, infectious serologies, and body imaging, she was referred to our clinic for management. A skin exam at the time revealed a flat and patchy blanchable rash (port-wine stain) in the S1 and S2 dermatomes in her left posterior thigh and calf, reminiscent of the cutaneous lesion in Cobb syndrome (Figure 3). Aside from her subjective sensory complaints, a neurological exam was unremarkable. An MRI and MRA with and without contrast of her lumbar spine demonstrated no evidence of a vascular abnormality in the spinal canal. Pre and post contrast MRA/MRV sequences utilizing 4d TRAK (time resolved angiography) and delayed MRV images were obtained. This does not rule out subradiographic vascular malformations in the spinal canal or cauda equina as a potential explanation for her symptoms since it would be unusual for a treated Chiari malformation and cervical syrinx to involve the bladder and lower extremities. The patient currently returns to clinic every six months for follow-up. Her symp-

toms have remained stable over the past three years.

Discussion

In 1915, Stanley Cobb described an eight-year-old boy who had a sharp pain in his lower back while playing ball. He had recurrence of the pain three days later followed by flaccid paralysis, anesthesia, and areflexia of both legs. The patient also experienced urinary retention, bowel incontinence, and priapism. He was admitted to the hospital with a presumptive diagnosis of poliomyelitis. Physical exam revealed a port-wine birthmark on his left back. Over ten and a half weeks, his legs became spastic and atrophied with hyperreflexia and upgoing toes. Five months after hospitalization, the patient underwent an exploratory laminectomy by Harvey Cushing who noted that the “dura was tense, bulging, and transmitted an unusually dark subdural coloration,” which when exposed, revealed “an extraordinary tangle of huge pulsating vessels filling the canal.” The dura was left open and the wound was closed. The only symptom relieved by the surgery was priapism. The patient was dis-

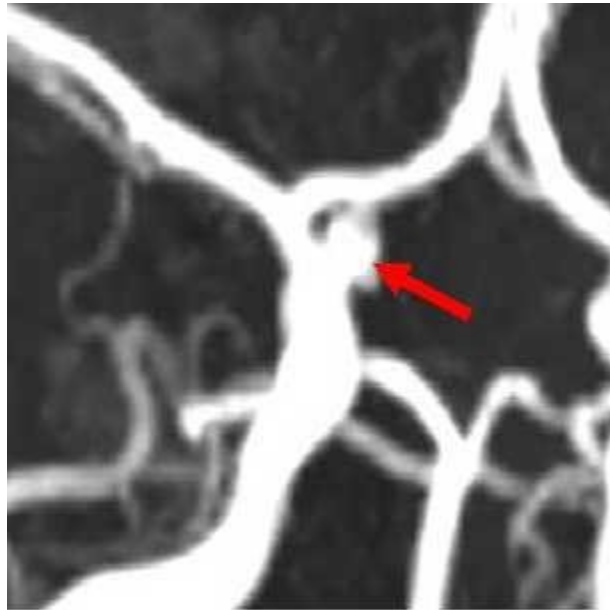


Figure 2a: **Intracranial aneurysms.** MRA head shows aneurysms in the right ICA.

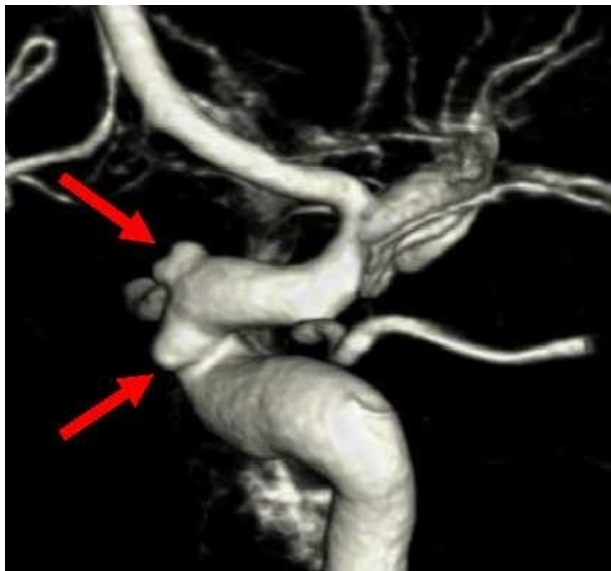


Figure 2b: **Intracranial aneurysms.** Reconstructed images from cerebral angiography reveal a right proximal medially-pointing superior hypophyseal broad-based aneurysm 2.5 mm in width and 2 mm in depth, and an ophthalmic artery aneurysmal origin dilatation 1.5 mm in depth.



Figure 3: **Vascular nevus.** Flat patchy blanchable nevus (port-wine stain) distributed in S1 and S2 dermatomes.

charged home with unchanged spastic paralysis, incontinence, and anesthesia one year after admission. Cobb cited seven other cases, including the original dissertation by Berenbruch, where a vascular nevus occurred in the same metamere as an angioma of the spinal cord. The various presenting symptoms included weakness, paralysis, anesthesia, and pain, which were thought to be triggered by expansion or rupture of the angioma causing either direct compression of the cord or ischemia to the cord secondary to diverted blood flow. Cobb concluded that vascular nevi on the skin may be of diagnostic value when neurologic deficits corresponding to a

spinal level are present.⁵

There are less than fifty cases of Cobb syndrome reported in the literature.¹ The disorder appears to have a slight male predominance, usually presenting in childhood but can be seen in adults, and most frequently involves the thoracic segment of the spinal cord.^{6,7} Along with the classic port-wine stain, other skin lesions that have been described include angiokeratomas, nevus flammeus, and lymphangioma circumscriptum.⁸⁻¹⁰ The most common vascular malformation in the spine is an arteriovenous angioma but other anomalies such as dural arterio-

venous fistula, telangiectasia, cavernous hemangioma, epidural venous angioma, and intraspinal hemangioma have all been reported.^{10,11} Current treatment options include embolization, resection, radiation, or steroids; the exact management depends on the patient's clinical signs and characteristics of the angioma.¹⁴

There have been no reported cases of Chiari malformation associated with a dermatomal vascular nevus. Other cutaneous disorders that have been described in conjunction with Chiari malformations include neurofibromatosis, which has been cited to be as high as 8%.¹⁵ There are also reported associations with macrocephaly-cutis marmorata telangiectatica congenita, which are blue to purplish reticulated telangiectasias that superficially ulcerate due to loss of underlying dermal substance.¹⁶ Other cutaneous findings include acanthosis nigricans, giant congenital melanocytic nevi, and phakomatosis pigmentovascularis.¹⁶ Spinal defects have also been reported including atlantoaxial assimilation, caudal regression syndrome, odontoid retroflexion, and spondyloepiphyseal dysplasia.¹⁷ Due to the rarity of these cases, it is unclear if there is a single pathophysiogenic mechanism that is responsible for the Chiari malformation and these other abnormalities or whether they are merely coincidental. Our patient does not exhibit any of these findings on her exam.

There has been one case report of Cobb syndrome in association with neurofibromatosis type I¹³ and another with tethered cord.¹² Tethered cord syndrome was excluded from our patient by her prior neurologist. She did not demonstrate any cutaneous abnormalities in the midline lumbar region. She did not have other associated signs of tethered cord such as lower extremity weakness or deformities, and she did not demonstrate any upper or lower motor neuron signs on physical ex-

am.¹⁸ The level of her conus on the lumbar spine MRI was normal and no thickened or fatty filum was identified. There was not enough clinical suspicion for us to reconsider this diagnosis.

In the reported cases of Cobb syndrome we reviewed, the dermatome of the rash always corresponded with the level of the AVM in the spinal cord. It is possible that a developmental abnormality early on disrupts the neuro-vascular axis, leading to malformation of blood vessels and nervous tissue at a specific segment. Despite our patient's symptoms of urinary retention, the absence of any cutaneous findings in the thoracic dermatomes and the presence of her vascular nevus in the S1 and S2 dermatomes placed the level of the lesion in the lumbar, and not the thoracic, spinal cord. Further imaging of her thoracic spinal cord could be considered in the future if her sensory symptoms were to ascend or if she were to develop signs of myelopathy.

Our patient was also evaluated by a neurointerventionist for the possibility of a spinal angiogram after her spinal MRA revealed no evidence of any vascular malformation during both early and late phase enhancement. Spinal MRA has been reported to be accurate in terms of diagnosing and excluding both dural arteriovenous fistulas and spinal arteriovenous malformations.²¹ Significant limitations of spinal MRA include difficulty detecting small arterial feeders, inability to classify the various types of AVMs, and difficulty differentiating dural AVF and spinal AVM.²⁰ The neurointerventionist did not proceed with spinal angiography because of its risk of ischemia to the spinal cord, high radiation exposure in a young female, as well as the potentially nephrotoxic contrast load.¹⁹ It is a future diagnostic option if our patient's symptoms continue to worsen.

In summary, our patient was found to have a

port-wine stain in her left leg that appeared classic for Cobb syndrome, coupled with symptoms of numbness and tingling and urinary retention, implying possible pathology of the cauda equina or exiting sacral nerve roots. Absence of an angioma on lumbar MRA does not rule out microscopic or subradiographic vascular anomalies in the spinal canal or within the nerves themselves as an etiology of her symptoms. It is interesting to note that her constellation of abnormalities – Chiari malformation, syrinx, intracranial aneurysms, and port-wine stain – seems to originate from defects in early development along a neuro-vascular axis.

Conclusion

We conclude that the presence of a dermatomal vascular nevus should prompt the clinician to obtain spine imaging particularly in the setting of neurologic symptoms. Most patients with spinal vascular malformations usually remain asymptomatic until there is rupture or rapid expansion of the angioma. Timely diagnosis allows for closer monitoring of changes in size, prompt treatment, and earlier considerations for surgery.

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