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Cobb Syndrome Associated With a Verrucous (Angiokeratomalike) Vascular Malformation

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Cobb syndrome is defined by a cutaneous vascular lesion and a corresponding spinal cord vascular malformation within a segment or two of the involved dermatome. Even though Cobb syndrome has been reported in association with various cutaneous vascular lesions, to our knowledge, only one other previous report describes verrucous changes overlying the cutaneous vascular lesion. We describe a patient with verrucous vascular malformation extending from his right chest to his right upper back in a dermatomal distribution. The patient's medical history included excision of a cavernous vascular malformation from the corresponding segment of his thoracic spinal cord 4 years prior, reversing 7 years of progressive leg weakness and neural deficits. Cobb syndrome was diagnosed based on the dermatomally distributed vascular malformation and the corresponding spinal cord vascular malformation. His skin lesions initially improved with erbium: YAG laser treatment but later recurred. This is the second reported case of Cobb syndrome associated with verrucous angiokeratomalike changes overlying the cutaneous vascular malformation. Because of the potentially severe neurologic sequelae caused by spinal cord lesions, clinicians should evaluate patients with dermatomally distributed cutaneous lesions on the trunk or extremities, including vascular malformations with verrucous features.

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Cobb syndrome, or cutaneomeningospinal angiomatosis, consists of a cutaneous vascular lesion and an associated spinal cord vascular malformation corresponding within a segment or two of the involved dermatome.[1] The cutaneous lesion may be macular, such as port-wine stains, or raised papular or nodular malformations. Although a number of various types of cutaneous vascular lesions have been described in association with Cobb syndrome, a vascular malformation with verrucous angiokeratomalike changes is a rare finding, to our knowledge reported only once previously, in 1977.[1] We report the second case of Cobb syndrome with this association to illustrate the need for prompt diagnosis of dermatomal vascular proliferations with associated spinal cord lesions to prevent neurologic sequelae.

Case Report

A 17-year-old Hispanic boy presented for the treatment of erythematous-to-violaceous papules and plaques extending from his right chest to his right upper back in a dermatomal distribution. The vascular malformation was present at birth, where it was initially macular, but over time had developed progressively to a verrucous angiokeratomalike surface (Figure 1). The patient denied any pain or pruritus associated with the lesion.

A biopsy of the lesion down to the dermis was performed. Histopathology results showed vascular proliferation with superficial and deep components. The superficial component contained dilated capillaries with overlying hyperkeratosis, papillomatosis, and irregular acanthosis (Figure 2). The deep component contained telangiectatic venous-sized and capillary-sized vessels involving the deep dermis

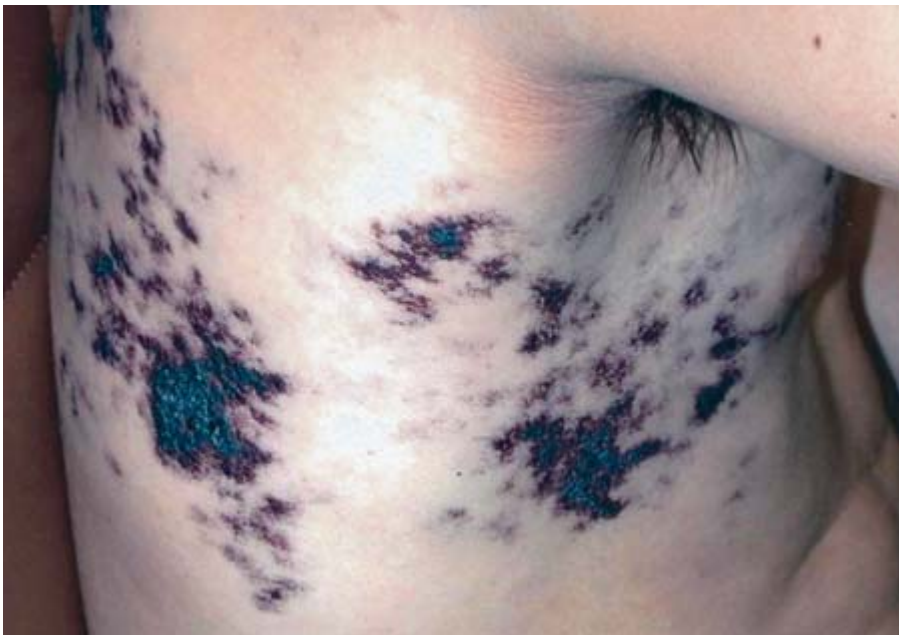

Figure 1. Verrucous angiokeratomalike surface and dermatomal distribution of the lesions.

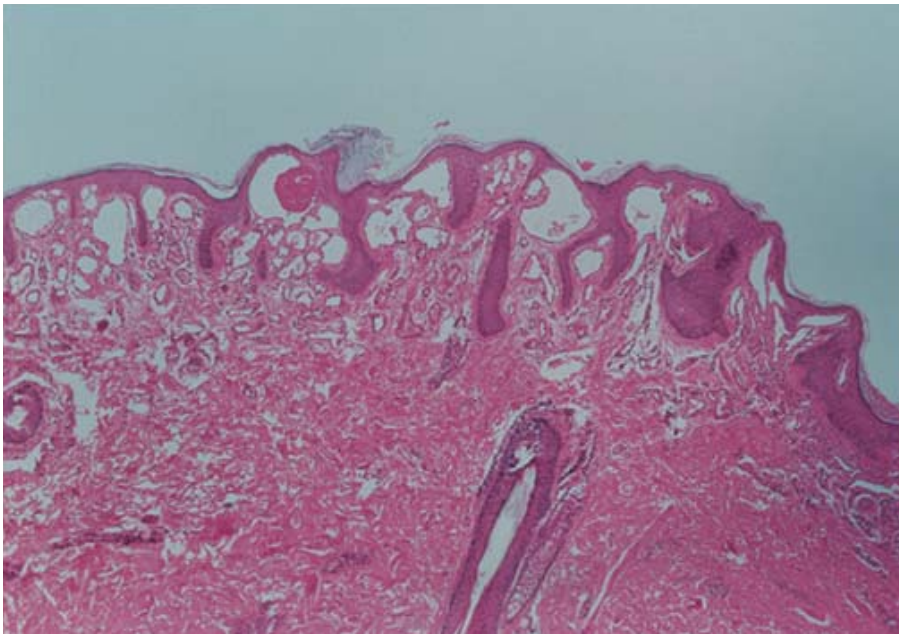

Figure 2. Skin biopsy specimen shows a dermal vascular proliferation with overlying hyperkeratosis, papillomatosis, and irregular acanthosis (H&E, original magnification ×40).

and subcutaneous fat extending to the base of the biopsy specimen (Figure 3). These findings were consistent with a combined capillary-venous malformation with verrucous angiokeratomalike features.

A review of the patient's medical and surgical history showed that 4 years earlier he had presented complaining that his limp had become worse and that there had been swelling in his right leg for approximately 7 years (from 6 through 13 years of age). Results of the physical examination at that time revealed atrophy of his right quadriceps and gastrocnemius muscle groups, along with decreased strength and sensation of his right lower extremity. Findings of magnetic resonance imaging (MRI) showed a complex intramedullary spinal cord vascular lesion at the level of the T2 through T5 vertebrae (Figure 4). Resection of the spinal cord vascular malformation was performed. Results of the excisional biopsy showed a benign vascular proliferation with prominent irregular telangiectasias; furthermore, the larger vascular spaces—lined with a single layer of thin endothelial cells—contained red blood cells and fibrinous material. The pathology diagnosis was that of an intramedullary cavernous venous

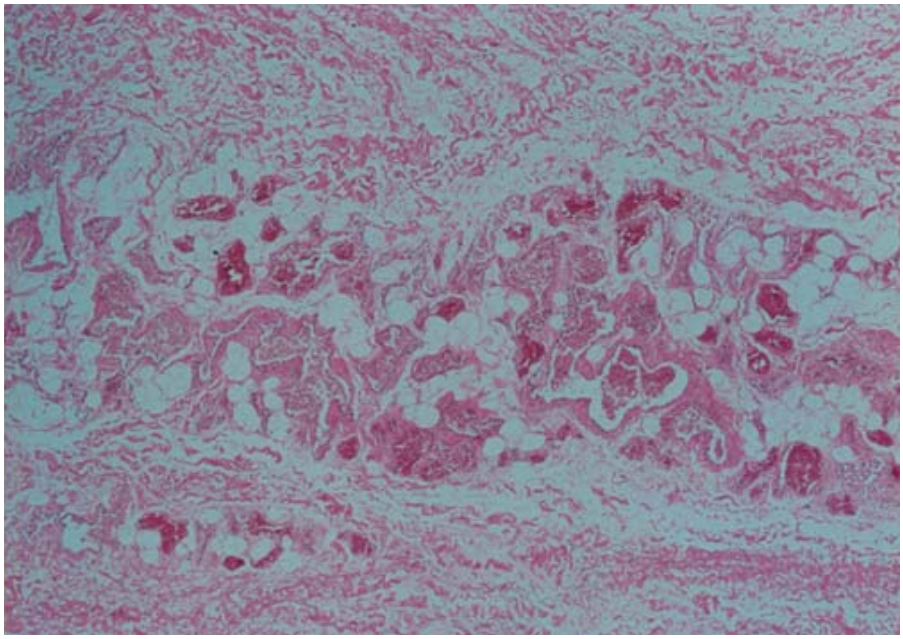

Figure 3. Telangiectatic venous-sized and capillary-sized vessels found in the deep dermis (H&E, original magnification ×100).

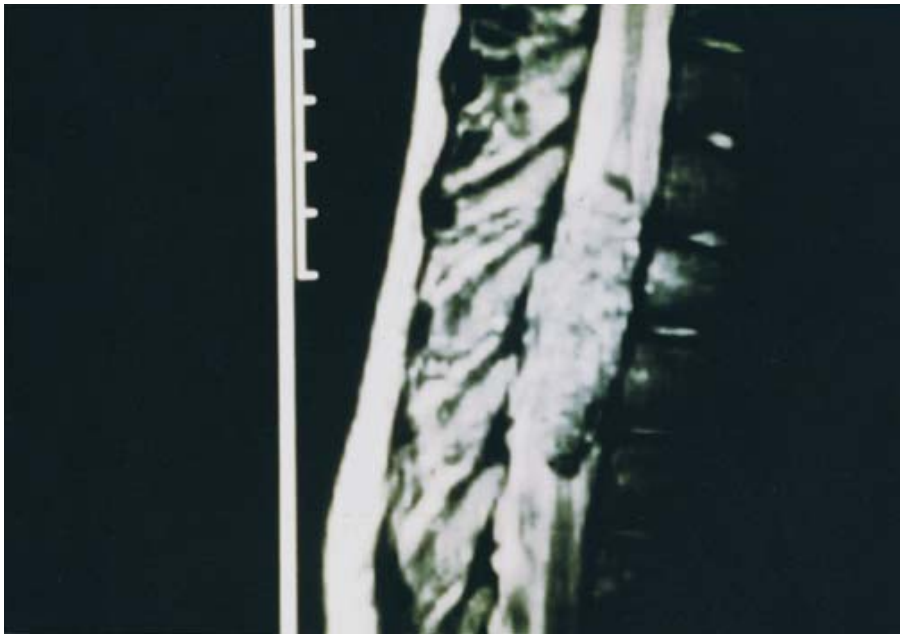

Figure 4. An intraspinal cavernous vascular malformation at the level of the T2 through T5 vertebrae (magnetic resonance imaging, T2-weighted sagittal image).

malformation. The patient gradually regained most of his neurologic functioning, with the exception of a mild limp in his right leg.

Based on the history of a T2 through T5 spinal cord cavernous vascular malformation with a cutaneous verrucous angiokeratomalike vascular malformation in the corresponding dermatomes, the diagnosis of Cobb syndrome was made. Because the spinal cord lesion had already been excised successfully, the patient requested treatment to improve the cosmetic appearance of the cutaneous vascular lesion. Treatment was attempted with the erbium:YAG laser, which was chosen over the vascular lesion laser because of its deeper tissue penetration. The patient was pleased with the initial improvement, but recurrence was noted at the 3-month follow-up visit. The patient did not return for further treatment.

Comment

Although the first case of cutaneomeningospinal angiomatosis was reported by Berenbruch in 1890, the syndrome did not receive recognition until Cobb reported his case in 1915.[1,2] Cobb syndrome

is rare, with only 33 cases reported in the English literature.[1,3-8] However, Doppman et al[9] studied 28 patients with spinal cord arteriovenous malformations and found 6 (21%) with cutaneous vascular lesions in corresponding dermatomes, suggesting that the prevalence may be higher than recognized. Although there have been 2 reports of Cobb syndrome where each case apparently had a strong family history of cutaneous malformations,[10,11] familial cases of the full syndrome have not been reported.[1-8]

The cutaneous manifestations can range from macular lesions, such as port-wine stains, to various types of papular or nodular vascular lesions, including capillary malformations, cavernous malformations, and lymphangioma circumscriptum.[1,2,4-8,10,11] To our knowledge, only one prior case of Cobb syndrome associated with a verrucous vascular malformation as the cutaneous manifestation has been reported, in addition to the current case.[1] Since 1998, under the new International Society for the Study of Vascular Anomalies classification system, vascular lesions previously diagnosed as Cobb syndrome would now be better characterized as vascular malformations rather than true hemangiomas.[12,13] Vascular lesions associated with Cobb syndrome do not tend to resolve or involute spontaneously, as is seen typically with the usual capillary and cavernous malformations of childhood.[1] Cutaneous lesions are either noted incidentally or brought to medical attention because of cosmetic concerns or susceptibility to bleeding with trauma.

Verrucous vascular malformations are typically congenital lesions that start as bluish discrete to confluent papules or nodules, usually unilaterally, on a lower limb. Over time, they become verrucous and hyperkeratotic with an angiokeratomalike appearance, probably as a result of trauma or secondary infections.[14,15] Verrucous malformations do not regress spontaneously and tend to enlarge with body growth; therefore, early excision is recommended. Larger lesions may require wide deep excision with grafting. Overall, excision is associated with a 30% recurrence rate because of frequent extension into subcutaneous fat planes.[14,16] Various lasers, such as carbon dioxide and vascular, are associated with a high recurrence rate because of their inability to ablate this subcutaneous extension completely.[16] Our attempt to ablate the lesions with the deeper penetrating erbium:YAG laser also resulted in eventual recurrence.

Symptoms associated with the spinal cord vascular malformation tend to begin during childhood or adolescence; the initial events usually occur spontaneously, though they sometimes occur with strong physical effort or during the last months of pregnancy.[5] Initial symptoms can include pain in the area supplied by the involved spinal cord segment, paraplegia or monoplegia, or decreased sensation in the lower extremities. Involvement of the urethral sphincter, rectal sphincter, or both, is not common early in the course but often develops as the symptoms progress. The neurologic symptom course can follow 1 of 3 patterns: (1) sudden onset of symptoms progressing to severe motor and sensory deficits within hours, (2) series of mild to moderate episodes separated by weeks or years, or (3) gradual onset and progression of neurologic deficits.[1] Our patient's symptoms followed the third pattern.

Findings of the neurologic examination of spinal cord vascular malformations in Cobb syndrome are typical of those detected with intraspinal vascular malformations. Patients usually have a sharp sensory level, decreased sensation to all modalities, spastic paresis or paralysis, and hyperreflexia; the findings may be in one or both lower extremities.[1] The pathogenesis of the deficits is believed to be neural tissue ischemia related to the shunting of blood into the malformation. In cases where the spinal cord lesion is an arteriovenous malformation, limb hypertrophy and length discrepancy may be present.[1]

Spinal cord lesions may be visualized with either MRI or selective spinal angiography. MRI can show intramedullar signal changes and most of the vessels and is safer than invasive angiography with intravascular contrast. However, angiography can demonstrate the extent of the lesion fully, as well as allow occlusion of the lesion with embolization.[5,8] Spinal cord lesions also can be excised if not too extensive. Lesions fed by the posterior spinal artery are often juxtamedullary and can be excised without appreciable damage to the spinal cord, whereas lesions fed by the anterior spinal artery are usually intramedullary, and critical motor pathways may be damaged during the excision procedure.[1] Most spinal cord lesions in Cobb syndrome are described as high-flow malformations on angiography. Because our patient did not undergo angiography before excision, we cannot comment on the flow status of his spinal vascular malformation. However, the pathologic diagnosis of cavernous venous malformation on the biopsy specimen suggests that our patient's lesion was a slow-flow malformation. Although arteriovenous malformations comprise the majority of spinal cord lesions described in association with Cobb syndrome, spinal cord slow-flow venous malformations with cutaneous vascular lesions in the corresponding dermatomes are also within the definition of Cobb syndrome.[17,18]

Despite the variety of skin lesions associated with Cobb syndrome, the association with a verrucous angiokeratomalike vascular malformation is a rare event. One may speculate that our patient's neurologic deficits may have been averted if an investigation for possible spinal cord lesions had been conducted after noting the dermatomal skin lesion during his childhood. Given that the prevalence of Cobb syndrome may be higher than recognized, clinicians should investigate for possible spinal cord lesions when noting dermatomally distributed vascular lesions on the trunk or extremities of their patients, including vascular malformations with verrucous angiokeratomalike cutaneous surfaces. The importance of further investigation is emphasized when considering the potentially severe neural deficits caused by spinal cord lesions.

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