A Case of Cobb Syndrome

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Cobb syndrome is a rare neurocutaneous angiomatosis characterized by a vascular skin nevus associated with a spinal cord angioma of the same metamere.

A 14-year-old girl had an asymptomatic large cutaneous hemangioma distributed from the T1 dermatome downward to the L3 dermatome since birth and complained of a gait disturbance and urination difficulty for 1 year.

A biopsy specimen in the skin lesion revealed the findings of capillary hemangioma. From C7 downward to L4 posterior epidural hemangioma composed of arteriovenous and cavernous components was diagnosed by radiological examination and surgical exploration.

Because of very extensive cord hemangioma, only partial removal of the tumor at T11, T12 and L1 level was performed and postoperatively she was transferred to a special facility for rehabilitative therapy. (Ann Dermatol 9:(1)64~68, 1997).

Key Word : Cobb syndrome, Neurocutanous angiomatosis

Cobb syndrome is a rare neurocutaneous disorder characterized by a vascular skin nevus associated with a spinal cord angioma of the same metamere[1]. This disorder was first coined by Cobb[2] in 1915 but the first case was described by Berenbruch[3] in 1890. So far it has been rarely reported in the world literature, especially in English.

We report a case of Cobb syndrome which consists of cutaneous hemangioma and epidural hemangioma in the spinal cord accompanied by vertebral hemangioma of the corresponding metamere and is the first report in the Korean literature to our knowledge.

CASE REPORT

A 14-year-old girl has had an asymptomatic diffuse large bright or dark red-colored plaque with a rough surface and nearby small similar lesions on the trunk since birth(Fig. 1-A). In the course of her life multiple nodular outgrowths or warty excrescences developed within the lesion and the cutaneous lesions had no evidence of involution. Two protruding nodules in the lesion were excised from the back for cosmetic purposes three years ago. All the skin lesions were distributed from the T1 dermatome downward to the L3 dermatome and located more in the right side of her trunk than in the left(Fig. 1-B).

She had also been suffering from a gait disturbance and urination difficulty for 1 year. On neurological examination, paraparesis, hypoesthesia below the T6 dermatome and upper motor neuron signs were present and the right lower extremity showed more severe involvement than the left corresponding one with the distribution of the cutaneous lesion. Other family members with similar lesions were not noted.

Routine laboratory examinations including a complete blood count, urinalysis, renal and liver function tests were within normal limits. Chest, cervical, thoracic and lumbosacral spine X-ray films were insignificant. MRI of the brain showed no evidence of definite abnormal signal intensity.

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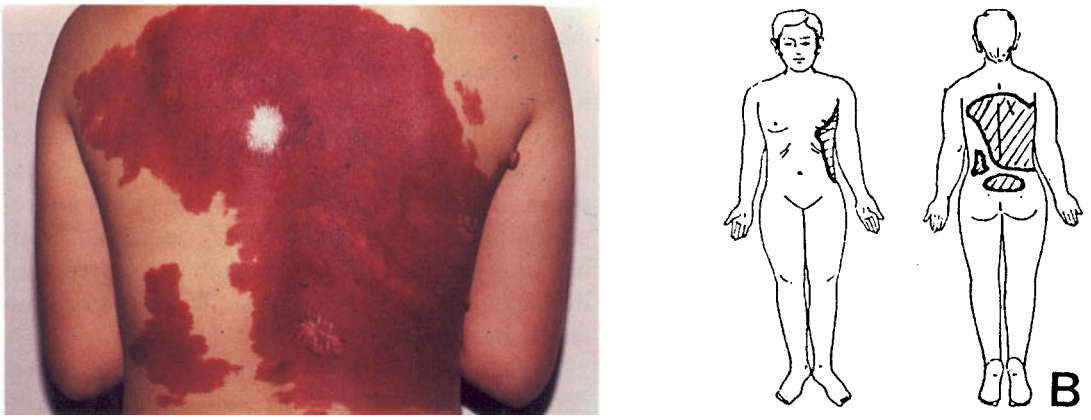

Fig. 1-A. Diffuse, large, bright or dark red-colored plaque with a rough surface and nearby similar same lesions from the T1 dermatome downward to the L3 dermatome of the trunk. Multiple nodular outgrowths and two previous excision scars in the lesion are shown.
Fig. 1-B. Schematic diagram of the distribution of the cutaneous hemangioma.

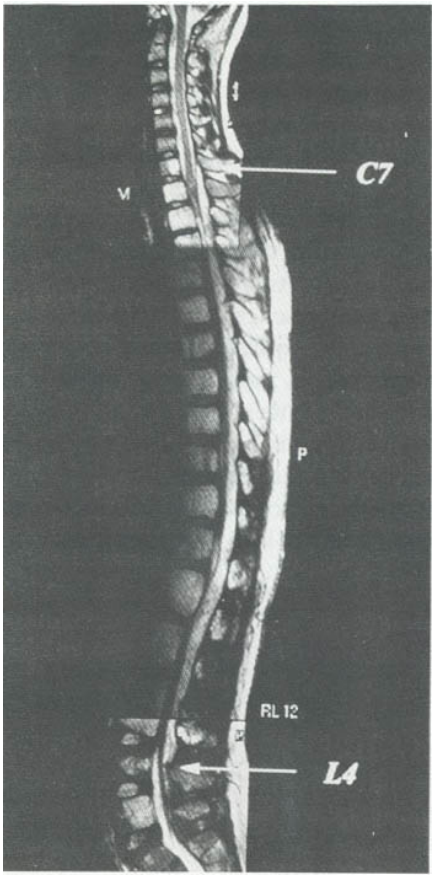

Fig. 2. MRI of the cervical, thoracic and lumbosacral spines shows a posterior epidural fusiform mass from C7 downward to L4(arrows).

However, MRI of the cervical, thoracic and lumbosacral spine revealed a posterior epidural fusiform mass from C7 downward to L4(Fig. 2). Selective spinal angiography by the right transfemoral approach showed diffuse trabeculated or spiculated dense enhancement along the vertebral bodies of T3-L1 levels suggestive of the hemangioma involving the vertebral bodies and no enhancement in the epidural mass(Fig. 3).

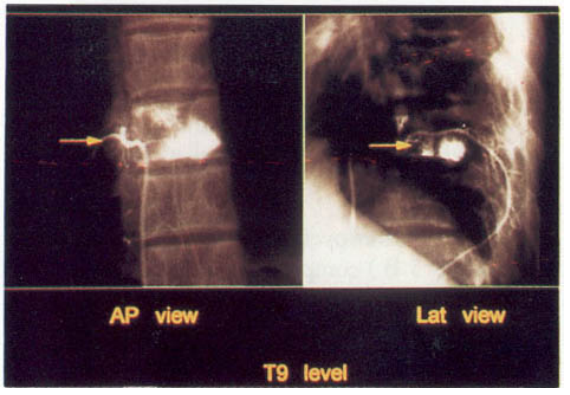

Fig. 3. Selective spinal angiography by the right transfemoral approach shows diffuse trabeculated or spiculated dense enhancement of the T9 vertebral body(arrows).

A biopsy specimen of the skin lesion revealed multiple lobulated vascular masses in the mid to deep dermis(Fig. 4). The mass was made up of considerable proliferation of endothelial cells forming a few capillary lumina.

In the department of neurosurgery, total laminectomy and partial removal of the tumor at T11, T12 and L1 level was performed. On the microscopic examination of the tumor, hemangioma

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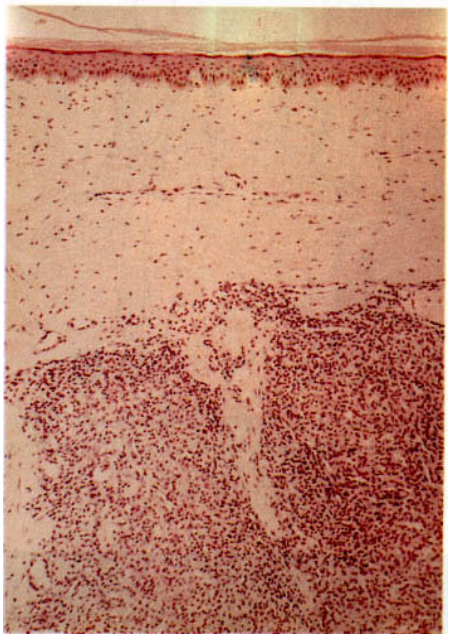

Fig. 4. A biopsy specimen of the skin lesion reveals vascular mass made up of considerable proliferation of endothelial cells forming a few capillary lumina in the dermis(H & E stain, × 100).

composed of arteriovenous(Fig. 5-A) and cavernous(Fig. 5-B) components was diagnosed.

Postoperatively her neurological symptoms were slightly improved and she was transferred to the department of rehabilitative medicine for further treatment.

DISCUSSION

Cobb syndrome is a rare neurocutaneous angiomatosis made up of cutaneous hemangioma accompanied by angioma in the spinal cord segments corresponding to the dermatomes involved[1,2]. It may occur mainly in childhood or adolescence and slightly more in males than in females[1]. This disorder is not familial in most cases reported[4]. However, two cases associated with hereditary cutaneous hemangioma were reported by Kaplan et al[5] and Mercer et al[6]. In our case no family history was noted.

The cutaneous lesions in most of the cases reported were mainly the port wine stains or angiokeratomas which occured within the same dermatome at the site of spinal cord angioma[1]. However, other vascular tumors such as angiolipoma, carvernous hemangioma, and arteriovenous malformation may occur[1,3]. In the case with arteriovenous malformation, bony and soft tissue hypertrophy of the extremities may develop[7,8]. Cutaneous angiomas are usually asymptomatic and do not tend to resolve spontaneously.

The spinal cord lesion is predominantly arteriovenous or venous hemangioma and is mostly situated in the intradural space[1,9,10]. It can cause neurologic deficits similar to that of the intraspinal tumor. Neurological deficits are frequent initial symptoms indicating the presence of the cord lesion. A majority of patients may develop neurological symptoms such as paraplegia, monoplegia, rectal or urethral sphincter disturbances, hypoesthesia, and neuralgia suddenly or gradually. The pathogenesis of neurological deficits are thought to be cord compression by the tumor mass or cord ischemia due to diversion of blood into the hemangioma[1,11]. Our

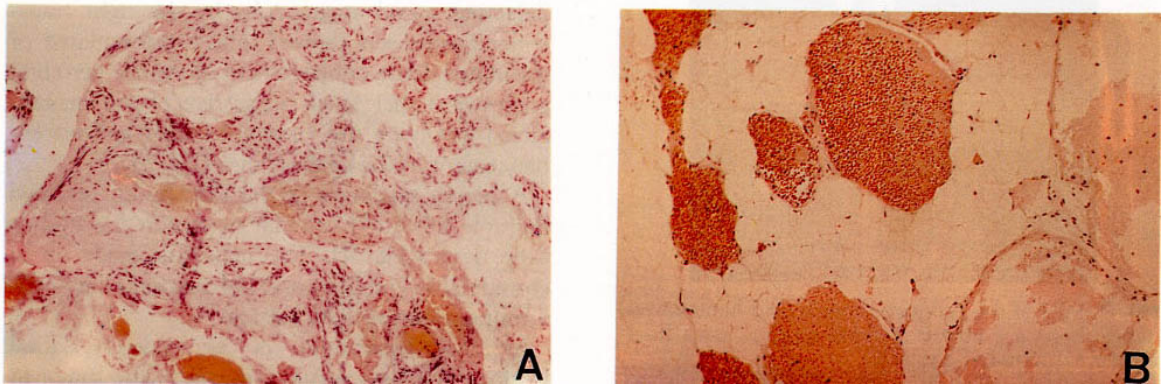

Fig. 5-A & B. On microscopical examination of the tumor in the spinal cord, densely aggregated thick-walled and thin-walled vessels containing red blood cells or thrombi(A) and large irregular spaces filled with blood and lined by a single layer of thin endothelial cells(B) are seen(H & E stain, × 100).

patient showed cutaneous capillary hemangioma associated with epidural hemangioma in the spinal cord and also complained of neurological symptoms such as paraparesis, hypoesthesia below the T6 dermatome, and urination difficulty caused by cord compression of the hemangioma.

Because Cobb syndrome consists of two basic conditions previously described, the clincally visible vascular skin nevus is of significance in that it may indicate the presence of spinal cord hemangioma of a corresponding metamere. In the review of 28 cases of spinal cord arteriovenous malformations, Doppman et al[12] reported that about 40% of patients also had a cutaneous hamangioma, which was located in the corresponding dermatome in about half of these cases. Our patient also had vertebral hemangioma at the T3-L1 level diagnosed by spinal angiography. Associated anomalies with Cobb syndrome may also include renal angioma, and kyphoscoliosis[13].

The pathogenesis of Cobb syndrome is believed to be due to developmental derangements of vascular stuructures in the skin and nervous system which is the failure of vascular regression in particular regions during embryonic life[14]. Doppman et al[12] found that the cutaneous and the spinal angiomas were supplied by posterior branches of the same intercostal artery on selective arteriography and both angiomas had common segmental blood supply origins.

The diagnosis can be achieved by clinical presentation of vascular skin nevi with a dermatomal distribution and identification of spinal cord angioma of the same metamere by radiological examinations such as a lateral spine X-ray, selective spinal angiography, myelography and MRI of the spine. Recently developed MRI examinations can not only facilitate the diagnosis of this disorder but also indicate the exact extent of the involved spinal cord. Surgical exploration also helps to confirm the diagnosis of the spinal cord hemangioma grossly and histologically. In our patient cord hemangioma was easily diagnosed by spine MRI and surgical exploration and not by spinal angiography.

Cobb syndrome should be differentiated from other neurocutaneous disorders including Sturge-Weber syndrome, Fabry-Anderson disease, Osler-Weber-Rendu disease, von Hippel-Lindau syndrome and ataxia telangiectasia. However, these diseases have abnormalities or lesions in the brain different from Cobb syndrome.

The treatment of Cobb syndrome is performed mainly when the progressive neurological deficits are developed. Surgical intervention to remove the tumor in the spinal cord can be used and preoperative embolization may be helpful[15]. But when the angioma involves the spinal cord extensively it is impossible to remove the tumor as a whole. Postoperatively rehabilitative therapy can be performed for better life quality of the patients. In our case only partial removal of the tumor at T11, T12 and L1 level was done because of very extensive cord hemangioma and postoperatively the patient transferred to the department of rehabilitative medicine for further therapy.

In conclusion, we report a case of Cobb syndrome which showed cutaneous capillary hemangioma accompanied by very extensive epidural hemangioma with vertebral hemangioma in the spinal cord of the same metamere.

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