Vertebral and epidural hemangioma with paraplegia in Klippel-Trenaunay-Weber syndrome

Case report

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A patient with a rare case of Klippel-Trenaunay-Weber syndrome presented with paraplegia due to compression by a vertebral and epidural cavernous hemangioma. The metameric distribution of the large cutaneous vascular nevus provided the clinical clue to the nature of the spinal lesion. The association of the two lesions is explained on the basis of developmental anomaly.

KEY WORDS • Klippel-Trenaunay-Weber syndrome • vertebral hemangioma • spinal cord compression • hemangiectatic hypertrophy • vascular nevus

Cutaneous nevus associated with hypertrophy of the affected limb was first described by Klippel and Trenaunay in 1900, and was subsequently further elaborated by Weber who coined the term “hemangiectatic hypertrophy.” The essential features are vascular nevus extending over an entire limb in a segmental distribution, and varicosities and hypertrophy of all the tissues of the involved limb. Various associated features including scoliosis, hyperhidrosis, paresthesiae of the affected limb, pulmonary hypertension, and polydactyly have been observed. Involvement of the nervous system has also been reported. Mental deficiency has been observed to occur frequently in these patients, and is attributed to a developmental defect. Psychosis and peripheral neuropathy have been reported in isolated cases. A rare combination of cutis laxa, aneurysmal dilatation of the terminal part of the basilar artery, and ectasia at the junction of the right vertebral artery producing ischemic infarction of the brain stem has recently been documented.

This report is of a case of Klippel-Trenaunay-Weber syndrome with vertebral and epidural hemangioma producing cord compression and paraplegia.

Case Report

This 25-year-old man was admitted on June 21, 1972. He was an athlete, but for the previous 6 months he had experienced increasing difficulty while running, and subsequently in walking for the past 2 months. The disability had considerably worsened in the 2 weeks before admission, when he noticed that he was dragging his feet even while walking at
a slow pace. He also noticed a sensation of heaviness in the abdomen spreading down to both lower limbs. He had not observed any wasting of the legs, numbness, tingling, alterations of sensation, or sweating of the lower limbs, nor was there any back pain. There was no difficulty in passing urine or stool. At birth he had had a deep purple nevus involving the whole of the right upper extremity and neck in the distribution from C-4 to T-2, and it increased in size proportionately with the growth of the limb (Fig. 1). No other members of his family have been known to have nevi.

Examination. The girth of the involved limb was 5 cm greater than the left side but there were no symptoms pertaining to the limb. A careful inspection failed to reveal any more nevi on the body surface, or congenital anomalies like polydactyly or syndactyly. There were no varicose veins, and no evidence of congestive cardiac failure or pulmonary hypertension.

The neurological examination did not reveal any involvement of the mental function, cranial nerves, or upper limbs. The tone was increased in the lower limbs and there was no wasting of any of the muscle groups. The power was slightly reduced in flexors and extensors of hip, knee, ankle, and toes (Grade 4). The knee and ankle jerks were increased in both lower limbs, more on the left (++++) than the right (+++). The sensation of touch, pain, temperature, joint, and position were normal, and Romberg's test was negative. The patient's gait was spastic, and he dragged his legs, especially the left one, while walking. There was no tenderness or deformity of the spine.

Blood examination showed hemoglobin 10.6 gm, total leukocyte count 7600/cu mm, polymorphonuclear cells 65, lymphocytes 30, eosinophils 2, and monocytes 3; erythrocyte sedimentation rate was 22 mm in the first hour. X-ray films of the chest and cervical and dorsal spine did not reveal any abnormality. Lumbar cerebrospinal fluid showed 3 lymphocytes/cu mm, protein 80 mg%, sugar 42 mg%, and chloride 700 mg%, venereal disease research laboratory (VDRL) test was negative. Lumbar myelogram showed a complete block at the lower border of T-2 (Fig. 2). The entire picture was suggestive of an extradural lesion.

Operation. Upper dorsal laminectomy was performed. The cutaneous hemangioma ex-
FIG. 2. Anteroposterior view of lumbar myelogram showing complete block to flow of dye at the upper border of the T-2 vertebral body. No appreciable change of the architecture of the T-1 and T-2 vertebrae is noticeable.

tended to the deeper tissues, involving the subcutaneous tissues and paravertebral muscles. The laminae and spinous processes were abnormally vascular. The mass arose from the T-2 vertebra, and encircled the spinal cord from C-7 to T-2. It was exclusively extradural, soft, friable, and extremely vascular. Most of the mass on the posterior and lateral aspect of cord was removed.

Postoperative Course. In the postoperative period the patient developed weakness of the small muscles of the hand, but this gradually improved after a course of irradiation. Weakness of the legs and the gait disturbances gradually improved, and by the end of 6 months there was no residual neurological deficit. The patient has now been followed for 4½ years and is completely free of symptoms.

Pathological Examination. Large cavernous vascular spaces partly or completely filled with blood were seen to be lined by normal-appearing endothelial cells. The endothelial cells were separated from each other by scanty connective tissue stroma. The microscopic appearance was suggestive of cavernous hemangioma.

Discussion

Association of skin angioma or vascular nevus in the metameric distribution corresponding to the spinal angioma is a rare occurrence and has been noted in only 17 reported cases, described as Cobb syndrome (cutaneomeningeal angiomatosis). In 19 other cases, although the skin lesions were present they did not conform to the segmental distribution. The commonest pathological lesion in these cases is intradural arteriovenous angioma, and next in frequency is intradural venous angioma. Epidural and vertebral angioma are rarely encountered. In 1906, Devic and Tolot reported a patient who had an epidural angioma and a large vascular nevus on the arm, justifying the diagnosis of Klippel-Trenaunay-Weber syndrome.

Of the triad of vascular nevus, hypertrophy, and varicosities, only the first two were present in our case. However, when varicosities are not obvious on clinical examination they can be visualized by phlebography in almost all cases. In 1918, Weber named the condition “hemangiectatic hypertrophy” where few large arteriovenous fistulae contributed to the hypertrophy of the affected limb, while in Klippel-Trenaunay syndrome the arteriovenous fistulae are numerous but very small. As there is no clear nosological difference between the two conditions the term “Klippel-Trenaunay-Weber syndrome” is generally used.

Hemangioma is a rare benign primary tumor of the vertebral column. As a clinical entity it is extremely uncommon, but it is more frequently observed as a pathological finding. In two large series of bone tumors, vertebral hemangiomas were reported in 15 of 1650 cases, and in 47 of 3987 cases, although in routine autopsy material the incidence is 7% to 12%. No clinical symptoms or radiological abnormalities were seen in any of these latter cases. It has been emphatically stated by Jaffe that clinically
silent lesions are invariably small. When such cases are excluded, symptomatic hemangio-
matous lesions of vertebrae are a rarity. Vertebral hemangioma when occurring in
association with cutaneous hemangioma has been found always to be symptomatic,
whereas a large majority of vertebral hemangiomas occurring alone are asympto-
matic. It follows that in the former situation the hemangiomas are likely to be larger.

The association of vascular nevus in
Klippel-Trenaunay-Weber syndrome with vertebral hemangioma can be explained on a
developmental basis. In the fifth week of the development of the human embryo, the
anterior appendage bud takes shape at the level of the seventh dorsal intersegmental
vessel, which arises from the dorsal aorta in relation to cervical somites. This vessel sub-
sequently becomes enlarged to form the subclavian artery. Spinal branches arising from
the dorsal intersegmental vessel supply the vertebral canal. It may be postulated that the
spinal branches are also involved in the developmental vascular anomaly affecting the
cutaneous branches of the main dorsal inter-
segmental vessel giving rise to vertebral and epidural hemangioma in association with the
large cutaneous vascular lesion in Klippel-
Trenaunay-Weber syndrome.

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