ANGIOMA RACEMOSUM VENOSUM OF SPINAL CORD WITH SEGMENTALLY RELATED ANGIOMATOUS LESIONS OF SKIN AND FOREARM

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It is well recognised that vascular hamartomata of the central nervous system may co-exist with other neural or extraneural lesions of a similar dysplastic nature.

The most familiar of these associations is Sturge-Weber-Dimitri disease. The cardinal features of this syndrome are an extensive capillary-venous malformation of the leptomeninges, atrophy and calcification of the affected cerebral hemisphere and a facial port-wine naevus in the distribution of the ipsilateral trigeminal nerve.

Another is the von Hippel-Lindau syndrome, in which a capillary hemangioblastoma of the cerebellar hemispheres occurs with similar tumours affecting one or both retinae. Other features include congenital cysts of the pancreas and kidney and capillary naevi of the skin.

The analogous combination, however, of a racemose angiomatous lesion of the spinal cord with a cutaneous lesion in the corresponding segment of skin is surprisingly rare.

Newman10 was able to collect 6 such cases from the literature published previously. He cited the cases of Cobb,1 Wyburn-Mason12 (Case 6), Silverman, Gilbert, and Henson and Croft5 (Case 4).

The following case is unusual in that not only were the vascular anomalies of the spinal cord and skin related segmentally, but in addition an abnormal fistulous communication between the arteries and veins of the forearm, in the same neuromere, was demonstrated by brachial arteriography.

CASE REPORT

A 26-year-old housewife complained of difficulty in walking, of increasing severity for 4 years, and attacks of left-sided sciatic pain for 6 months.

The history of her illness is somewhat vague, but apparently, 11 years previously, when aged 15, she suddenly became ill. Whilst playing she experienced a sharp attack of pain in the middle of her back which moved to her head and neck. At the time she complained of severe headache and felt dizzy and weak. This was followed by pain and paresthesiae of her legs.

A diagnosis of influenza was made. She remained in bed for a month when the condition began to improve and after that was completely well again.

Four years prior to admission, when aged 22 years, during the last trimester of her first pregnancy, she awoke to find that her right leg had become weak and heavy so that she had difficulty in walking without support. At first the loss of power was confined to the right leg, but this was followed later by gradual weakness of her left leg.

She was delivered at term and later began to improve, slowly regaining the power in her legs, and apart from occasional attacks of left-sided sciatic pain, enjoyed a remission for nearly 2 years.

Then, following a fall, there was development of pain and paresthesiae of her left leg and buttocks. These sensory symptoms were followed, a day or two later, by weakness of her right leg and stiffness and clumsiness of her left leg.

Since then she had suffered from increasing loss of power and spasticity of both her legs, and recurrent attacks of pain down the lateral aspect of her left leg. At no time had there been any disturbance in sphincter control.

Examination (May, 1955). She was well developed and well nourished. Most of the right arm was covered with an extensive vascular naevus. The lesion was reddish-blue in colour, raised above the skin, soft and semifirm; in places it was covered with a thickened hyperkeratotic layer of skin and was more marked over the ulnar border of the arm and wrist (Fig. 1). A few small lesions of similar morphology and papular character were observed over the dorsal aspect of the left arm corresponding to C5/C8 segments. There was another naevus measuring 4½ inches by 6 inches on the back, to the right of the midline, corresponding to D2/D4 segments (Fig. 2).

The temperature of the right forearm was higher than that of the opposite side.

In the upper extremities there was no wasting or weakness. Sensory appreciation was normal. Deep reflexes were present and equal. Abdominal reflexes were all present and the abdominal muscles were powerful.

Both legs were spastic, the right more than the left and fasciculation was noted in the right quadriceps and muscles of the calf. There was 1 inch of wasting of the right thigh and ½ inch of the calf. Power was markedly reduced in flexion, abduction and adduction of the right hip; there was moderate weakness of extension of the knee and dorsiflexion of the ankle and toes. On the left there was slight weakness in abduction and flexion of the hip and extension of the knee.

Pain and temperature were not appreciated from below D2 on the left and diminished below D9 on both sides. Sensation of light touch was diminished below D10 on the left. There was impairment of vibration...
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Fig. 1. (Left) Showing extensive involvement of right forearm by vascular naevus. (Right) Involvement along ulnar border of right hand.

sense on the left below the iliae crest and loss of position sense in the left toes.

The right knee and ankle jerks were very brisk and both plantar responses were extensor.

Blood pressure was 180/80 mm. Hg. Haemoglobin was 14.9 gm./100 ml. Erythrocyte sedimentation rate was 8 mm. in 1 hour (Westergren).

Lumbar puncture produced clear cerebrospinal fluid at a pressure of 160 mm. of cerebrospinal fluid. The manometric tests showed no subarachnoid block. The fluid contained 20 mg. of protein per 100 ml. and 4 lymphocytes per c.mm.

Plain roentgenograms of the cervical, thoracic and lumbosacral spines showed no abnormality. Films of the right upper extremity and chest were normal.

Course. On the 10th hospital day myelography was performed. The subarachnoid space was well visualised from the caudal limits of the sac to the level of the cervicodorsal junction. Here a large round filling defect was noted at the C7/D1 vertebral level (Fig. 3). There was no block to the further upward flow of Myodil. The appearances were interpreted as most likely to be caused by an extramedullary neoplasm, probably an intradural neurofibroma.

In view of the progressively disabling nature of the patient's disease and the difficulties in accepting the myelographic interpretation as that of a neurofibroma, especially in the presence of abnormal content of protein in the cerebrospinal fluid, it was felt necessary to explore the lesion.

Operation, May 1955. Laminectomy of C6/D1 vertebrae was performed. An incision was made in the mid line of the cervical-dorsal junction to expose the lowest cervical and upper dorsal spines and laminae.

The spines and laminae of C6, C7, and D1 were removed first. The dura mater was felt to be tense and was pulsating in its upper part.

A mass of dilated pulsating arteries and veins was found over the cord and entering it at about the level of C5. The lower end of the mass was not visible but several vessels were seen to enter the dorsal surface of the cord at the level of D2. A radical removal of the angiomatous malformation was not feasible. However, slips of ligamentum denticulatum were divided to give the cord more room and the dura mater was left open as a decompression.

Postoperative Course. In the months following operation there was considerable subjective improvement.

Her walking improved slowly and the attacks of pain in her legs were less frequent and severe. Objectively, there was some improvement in the tone of both legs though the plantar responses remained extensor in type.

She was re-admitted in April, 1960, nearly 3 years after operation, because in the preceding few months the vascular naevus over the right wrist and forearm had ulcerated in places and bled.

Two years earlier, despite advice to the contrary, she underwent a successful and uneventful pregnancy. She was now walking with only a slight limp. There was still weakness of all groups of muscles in the right leg with spasticity, hyperreflexia and an extensor plantar response. The motor power and reflexes in the left leg were normal.

The sensory level had fallen to below D6 on the left side, and the only sensory deficit present on the right was a mild hypalgesia below L1. On auscultation of the spine over C5-D4 vertebrae, the site of the previous operative incision, a loud high-pitched murmur was heard clearly; it was rhythmic, correlating with the apex beat, but was unaccompanied by a palpable thrill.

Brachial angiography was performed as a preliminary to a proposed resection of the vascular naevus on her right forearm with plastic repair. The angiogram revealed the presence of several angiomatous lesions in the

Fig. 2. Showing operative incision and relation of naevus situated posteriorly. Note similar lesions on left arm.