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 retinas. Other features include congenital cysts of the pancreas and kidney and capillary naevi of the skin.

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

Newman was able to collect 6 such cases from the literature published previously. He cited the cases of Cobb, Wyburn-Mason (Case 6), Silverman, Gilbert, and Henson and Croft (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 semicompressible; 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 4 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.
ANGIOMA OF CORD WITH ANGIOMATA OF SKIN AND FOREARM

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 iliac 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 120/80 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 D4. 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. 3. Showing operative incision and relation of naevus situated posteriorly. Note similar lesions on left arm.
right forearm. Numerous small arteries were seen to supply these angiomata and the contrast medium passed quickly into dilated veins (Fig. 4).

DISCUSSION

In reviewing the patient's previous history, the earlier complaint of sudden intense backache accompanied by symptoms of meningeal irritation and pain and paresthesiae of her legs is strongly suggestive of spinal variety of spontaneous subarachnoid haemorrhagc.

It is well known that racemose angiomas of the cord may be the source of bleeding into the subarachnoid space and the various aspects of this syndrome were discussed by Henson and Croft. The haemorrhachis may arise on a background of pre-existent symptoms of disease of the spinal cord or nerve roots, or it may come unannounced. The combination of sudden backache and root pain with signs of spinal meningeal irritation is pathognomonic of the spinal variety of haemorrhage.

Another feature of interest is the onset of weakness of her legs in the last trimester of her first pregnancy which was followed by a remission after delivery. Although relapse and remission of symptoms in relation to pregnancy in cases of spinal cord angiomata have been reported by Delmas-Marsalet, Guthkelch, and Newman (Case 16), the explanation for this happening is obscure.

Lam et al. suggested that the symptoms are caused by mechanical obstruction to venous drainage as a result of the enlarged pregnant uterus, and by possible oestrogenic factors giving rise to further venous engorgement of the angiomatous malformation.

Evidence that hormonal factors mainly are responsible for the disturbances in these and other vascular tumours has recently been presented in a paper by Newquist and Mayfield, who drew the analogy of the appearance during pregnancy of spider naevi and palmar erythema; both these conditions are accepted as being hormonal in nature. They reinforced their argument with the fact that symptoms occur not uncommonly during the menstrual period, which supports further the theory that hormones of pregnancy have an adverse effect on the walls of the vessels comprising the angioma.

Why these symptoms did not exacerbate with her second pregnancy is uncertain. It may be argued that the decompressive laminectomy had allowed for a greater degree of vascular engorgement, so that compressive symptoms were less likely to occur.

However, the absence of symptoms during pregnancies preceding, intervening between and following pregnancies in which symptoms have occurred has been recorded by Wyburn-Mason, Epstein et al., and Newman.

The finding of a bruit on auscultation over the...
cervicodorsal spine, at the site of the previous operative incision, is of significance, and had this sign been sought on her earlier admission before surgical intervention occurred it would surely have led to the correct pre-operative diagnosis. Although the value of cranial bruit in the diagnosis of an intracerebral angioma is well recognised, little attention has been paid to the possibility of a comparable spinal bruit until the recent paper of Matthews. It is suggested that this sign may be of diagnostic value in the elucidation of a problem of unexplained subarachnoid haemorrhage or spinal cord symptoms.

It is well known that the myelographic diagnosis of arteriovenous malformations of the spinal cord may prove to be difficult; nevertheless, the appearances of a central filling defect seen in this case should be regarded as quite atypical.

The usually recognised picture is one of curved tortuous worm-like filling defects in the column of contrast medium. However, myelography reveals only the superficial extent of the lesion, and that sometimes incompletely, because not all the pial vessels are seen in the one projection or because at times the majority of the vessels that comprise the angioma ramify within the substance of the cord and thus cannot be visualised by this technique.

Hence the lesion shown at operation or necropsy is, as a rule, more developed than that seen at myelography. It is possible that the central filling defect here was caused by the deposition of Myodil around the large vascular loops, observed at operation, which because of confinement gave rise to this type of defect.

Recently, Höök and Lindvall diagnosed spectacularly 2 cases of angioma of the cervical cord by vertebral angiography and, according to these authors, the vertebral artery supplies not only the cervical cord but the upper four dorsal segments as well. It would thus appear that vertebral angiography is a useful method of diagnosing vascular anomalies in these regions of the spinal cord.

Regarding the significance of the associated vascular anomalies of the skin and forearm, it was Baersensprung (1863) who originally described the metameric distribution of the vascular naevi. He drew attention to their topographical distribution and pointed out that their development was connected directly to areas of cutaneous innervation. He suggested that as it had been demonstrated that the changes in the skin consisted of hypertrophy of elements in which peripheral nerves terminated, such as the pigmented layer of skin and the blood vessels of the papillary layer, it seemed to point to the conclusion that the naevus represented a congenital lesion of the spinal ganglia.

Cobb also concluded that these vascular malformations arose from a developmental fault of the central nervous system so that the lesions could occur in any of the organs innervated by filaments from that neuromere.

He further remarked that the naevi of the skin may at times be of diagnostic value when segmental phenomena referable to the central nervous system are present.

SUMMARY

A case is described of an angioma racemosum venosum of the lower cervical and upper dorsal spinal cord associated with angiomata of the skin and forearm of the same metameres. Various clinical, radiological and pathological features of importance are discussed.

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