The Foix-Alajouanine Syndrome

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THE PURPOSE OF this paper is to report a patient with Foix-Alajouanine syndrome (cutaneous haemangioma with spinal malformations and subacute myelomalacia) not commonly found in this region. A brief review of the literature is also presented.

Case Report

A 13-year-old Chinese girl was admitted on March 1964 following sudden onset of headache and vomiting lasting for two days. Clinically, she had signs of subarachnoid haemorrhage and the diagnosis was confirmed by a bloody cerebrospinal fluid. No systemic lesions were observed at that time and radiological studies of the skull, chest and abdomen were reported as normal. On recovery bilateral carotid angiograms failed to demonstrate any cerebral pathology.

In March 1967, she was readmitted for the investigation of progressive paresis of the left leg without any other neurological symptoms. Examination then revealed a left spastic monoplegia without other neurological signs. Lumbar puncture showed a normal cerebrospinal fluid except for a high CSF protein of 140 mg%. No block of CSF flow was detected. A repeated carotid angiogram, as well as a lumbar air encephalogram, failed to demonstrate any pathological lesion. A month later, paralysis spread to the right leg, and she had difficulty in walking. The bladder and bowel functions remained normal. She had no headache, vomiting or visual changes.

On examination, she had spastic paraplegia, with the left leg more weak than the right. Tendon reflexes of both lower limbs were exaggerated and there were bilateral ankle clonus and Babinski signs. Muscle wasting was noted in the left leg but fasiculations were not observed in both limbs. Position sense was diminished from the level of the first lumbar dermatome (L1) downwards, but pain and temperature of the both legs were spared. Cerebellar signs were absent. Her mental functions and cranial nerves were not impaired. There was a thoracic scoliosis and a number of small cutaneous haemangiomata were observed on the scapular region. Bruits were not heard over the eyeballs, carotid vessels or the vertebral column.

Investigations showed normal blood and urine results. CSF was normal except for the elevated spinal fluid protein (100 mg%). Blood Kahn and L.E. tests were negative. X-rays of the thoracic and lumbar spines revealed thoracic scoliosis with small radiolucent areas in the bodies of the lower thoracic spines suggestive of spinal varices. A myelogram later confirmed the presence of a very extensive large haemangioma and varices of the whole spinal cord, extending from the cervical portion right down to the conus medullaris **(Figures 1 to 4)**.

In June 1969, she started to experience difficulty in controlling her bladder, but other neurological signs remained essentially the same. By September 1970, she was bedridden with paralysis of both legs and had bladder incontience; otherwise she was well, and had regular physiotherapy.

Comment

The syndrome of Foix-Alajouanine ("Subacute necrotizing myelitis") is characterised by myelomalacia due to the presence of spinal vascular malformations (angiomas) (Foix & Alajouanine, 1926). The vascular anomalies are large and abnormal arteriovenous angiomas found in the spinal meninges, intramedullary portion of the cord and sometimes, the nerve roots. The vessels, especially the veins, have peculiary thick walls ("arterialized veins"), and the histological picture shows marked hyperplasia and hylanosis without

Fig 1
Myelogram showing extensive spinal varices in the cervical region (A.P. view).

Fig 2
Same as Fig 1 (P.A. view).

any musculature although elastic tissues are found in the adventitia. (Cushing and Bailey, 1928; Laermittee et al., 1931; Greenfield and Turner, 1939).

The pathological changes in the veins indicate a pronounced, long-standing increase in the intravascular pressure found in conditions associated with arteriovenous shunts. The "myelitis" in this disease is due to the complications of these pre-existing angiomas which are sometimes found to bleed, thrombose and have other inflammatory changes. Thus, the patients may present as acute haematomyelia or sudden haemorrhage as in our case, or the onset of disease may be insidious from the gradual thrombophlebitis of the spinal vessels.

Clinically, the cases present with progressive amytrophic paraplegia, dissociated sensory loss, bladder disturbance and elevated spinal fluid proteins (e.g., our patient). Occasionally, cutaneous haemangiomas are observed and thus indicate a systemic involvement. The myelogram appearance is typical of the disease, showing massive spinal varices (Ransome and Mekie, 1942). The course is often progressive but long remissions, ranging from 9 to 22 years, have been reported. (Frey, 1928; Buckley, 1936; Meyer and Kohler, 1917). The syndrome has been the subject of many reviews (Jaffe & Freeman, 1943; Wyburn-Mason, 1943; Neubuerger et al., 1953; Antoni, 1962), but the pathogenesis remains obscure.

Although our patient had clinical and radiological evidence of this rare condition, the definite diagnosis can only be made at autopsy. Spinal vascular malformations seem to be extremely uncommon among Asians in Singapore. On the other hand, the intracranial form of arteriovenous malformations are not infrequently seen in this country (Tay et al., 1969; 1970).

The widespread vascular involvement in our patient precludes surgical intervention, but the smaller and more localised angiomas can be satisfactorily removed or ligated (Echols and Holcombe, 1941).

This case also illustrates the correct prediction of the spinal pathology purely by the close inspection of

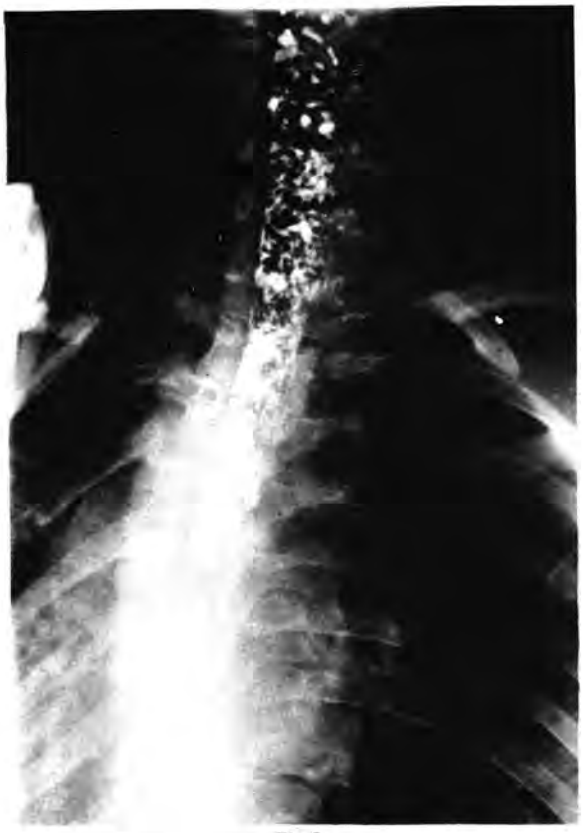

Fig 3
Varices extending into the cervo-thoracic level.

Fig 4
Varices in the thoracic and lumbar levels.

the cutaneous lesions, which in our case, was the multiple haemangiomata.

Summary

A case of Foix-Alajouanine syndrome is described and a short review of the literature presented.

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