Angiographic Features of Sturge-Weber Disease:
Report of a Case

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ENCEPHALOTRIGEMINAL ANGIOMATOSIS or Sturge-Weber Disease, in its complete form, consists of a facial naevus in the distribution of the trigeminal nerve and at least one of two other major features of the syndrome, namely, an intracranial angioma or an angioma of the choroid of the eye (Normal 1963).

Other associated clinical manifestations include convulsions, hemiparesis, hemiatrophy, mental retardation, bulpthalmos and congenital glaucoma. The pathologic-anatomical change intracranially is in essence a capillary venous angiomatosis of the leptomeninges. In all cases, the affected area becomes atrophic, with deposition of calcium and/or ferruginous material in the cortex and sub-cortical tissues (Lichtenstein 1954). The role of radiology lies in the demonstration of the abnormalities using plain skull X-rays, pneumoencephalography and cerebral angiography.

Allan Sturge (1879) was credited with being the first to describe the condition clinically, but in 1922, Weber published a radiograph of the characteristic double curvi-linear shadows outlining the cerebral sulci which have since become the hallmark in radiological diagnosis. Pneumoencephalography is used primarily to show or confirm cerebral atrophy. The changes include ventricular enlargement and dilated cerebral sulci on the affected side. In advanced cerebral atrophy, the basal cisterns are also widened.

Although Moniz and Lima had used cerebral angiography to investigate this condition in 1935, relatively few cases have since been studied by this technique, primarily because of the reluctance on the part of investigators to subject children to angiography.

However, Poser and Taveras (1955) stressed that this procedure was equally safe in children and adults. The same authors later (1957) collected 35 cases studied by cerebral angiography from the literature and added 15 cases of their own. From that combined series they concluded that angiographic findings in encephalotrigeminal angiomatosis were not exclusively confined to the presence of venous angiomata although such vascular anomalies were most frequently seen, accounting for 46% of positive findings of the 50 cases reviewed.

Other angiographic features, which may be demonstrable in this syndrome, include arteriovenous malformations, cerebral arterial thromboses, cerebral atrophy and anomalies of the cerebral veins and dural sinuses. It is primarily in connection with the latter appearances that the author wishes to present the following case.

Case Report

C.Y.C., a 14-year-old male Chinese patient, was admitted to the medical ward of the University Hos-

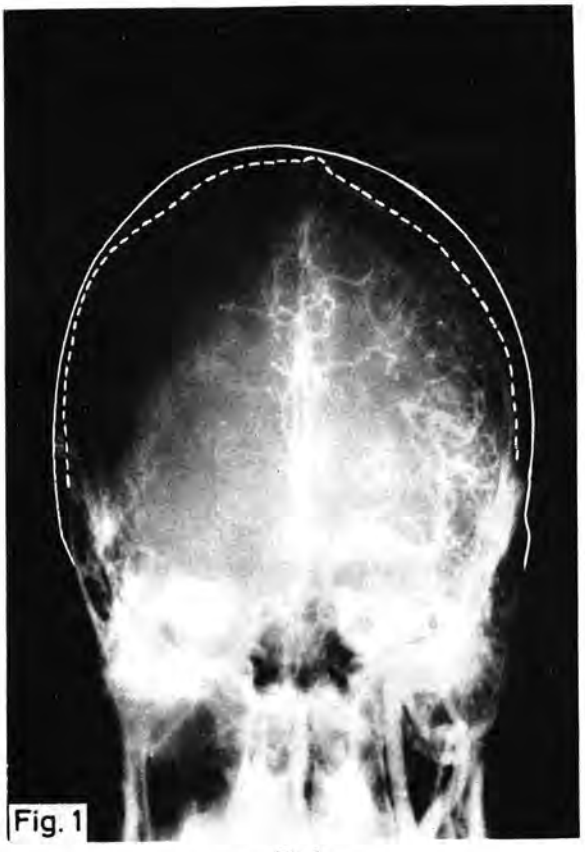

Fig 1
Minor shift of anterior cerebral artery to the left. Compensatory vault thickening indicated by continuous and dotted lines.

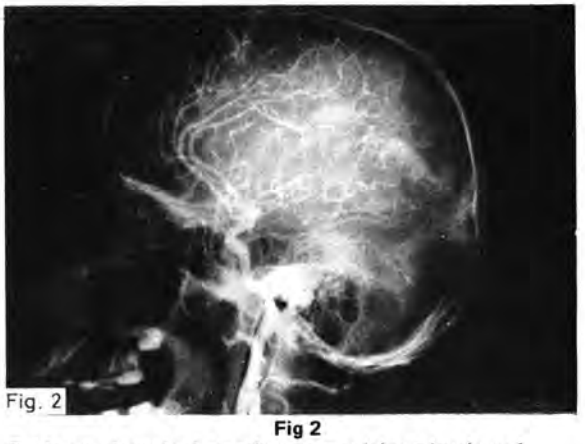

Fig 2
Evidence of cerebral atrophy: large sylvian triangle and rather low position of sylvian point. Slight increase in curvature of anterior cerebral artery.

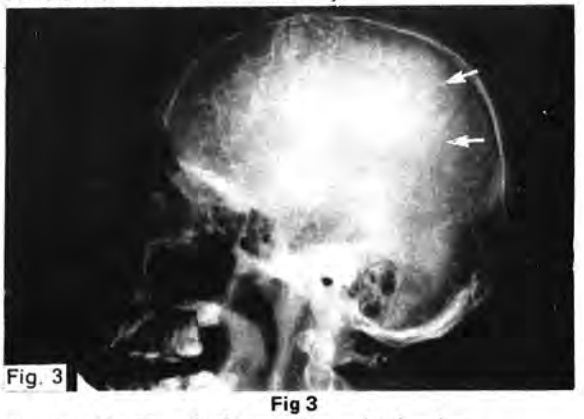

Fig 3
Non-opacification of left posterior parietal region, indicating cerebral atrophy (arrows).

pital with a history of generalised fits since the age of 1 year. The onset of the seizures was always preceded by an aura, described as a feeling of anxiety. Each fit would last about 2-5 minutes, occurring approximately once every 3-4 weeks. He had a port-wine mark on the left side of his face which had been present since birth. According to the parents, his milestones of development had been delayed. In the four months prior to admission, he was noticed to be aggressive and "destructive".

Examination showed him to be a well-built but mentally retarded patient. He had a port-wine naevus on the left temple, cheek, both lips and both eyelids.

Central Nervous System

The fundi appeared normal. The cranial nerves were intact. Motor power in the limbs was grade IV on the right side and grade V on the left. Electroencephalography suggested atrophy in the left posterior parietal area. The provisional clinical diagnosis was Sturge-Weber Disease.

Radiological Examination

Plain skull X-rays showed no sign of intracranial calcification. There was however a reduction in size of the left half of the cranial cavity with compensatory thickening of the skull vault.

A left carotid angiogram done under general anaesthesia showed the following abnormal features:-

(i) The arterial phase showed a 5 mm. shift of the anterior cerebral artery to the left side **(Fig. 1)**. Evidence of cerebral atrophy was further strengthened by the presence of a large sylvian triangle and downward displacement of the sylvian point in

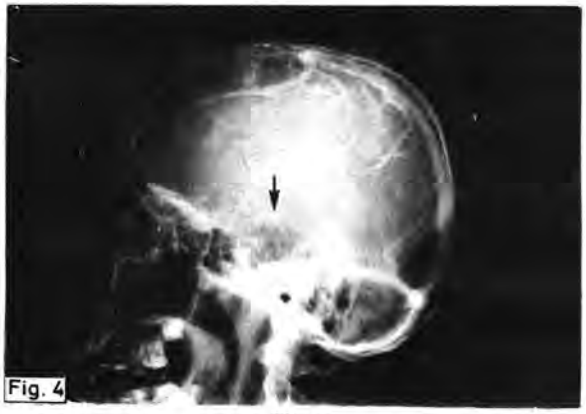

Fig 4
Small venous anomaly (arrow) and large left basilar vein. Partial agenesis of superior sagittal sinus with prominent draining veins. No evidence of filling of internal cerebral vein.

the lateral view **(Fig. 2)**. In the late arterial and capillary phase, the posterior parietal region was not opacified, indicating focal atrophy in that region. **(Fig. 3)**

(ii) The venous phase revealed a small venous angioma in the left temporal region **(Fig. 4)**. In the late venous phase, an enlarged tortuous basilar vein was seen coursing upwards and backwards to join the posterior portion of the vein of Galen. There was no evidence of filling of the internal cerebral vein or its tributaries. A partial agenesis or thrombosis of the superior sagittal sinus was noted. The superficial cortical veins draining into the superior sagittal sinus appeared numerous and prominent.

In view of the above findings, cross compression was performed to fill the venous system on the right side. The venous phase in this series showed a rather small thalamostriate vein emptying into the right internal cerebral vein **(Fig. 5)**. The left internal cerebral vein was again not visualised.

Discussion

Despite the intensive study by Poser & Taveras (1957), few recent workers have used cerebral angiography as a routine in investigating cases of Sturge-Weber Disease in its complete or incomplete forms. A reveiw of the available literature in English since 1957 shows rather disappointingly the paucity of positive findings in cases subjected to cerebral angiography

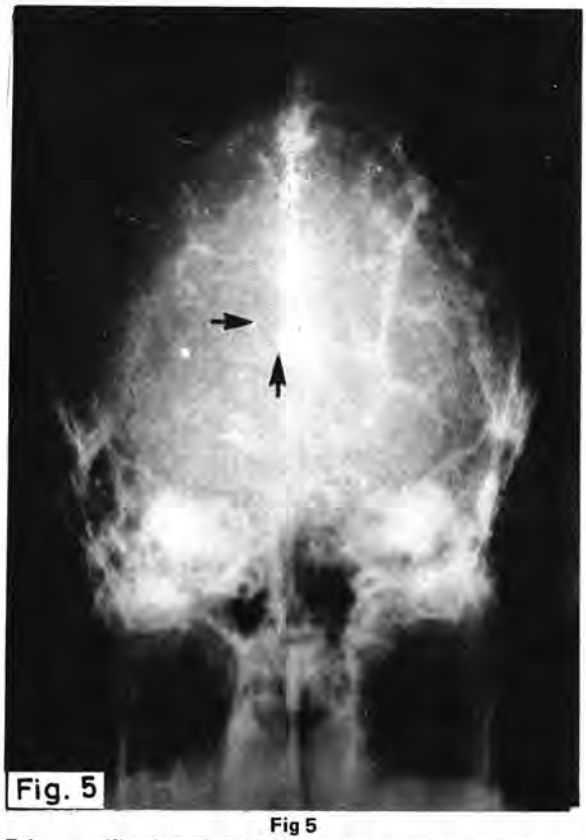

Fig 5
Faint opacification of right thalamostriate vein (arrows) and right internal cerebral vein on cross compression. Left internal cerebral vein not visualised.

(Table 1). A classic form of this condition was reported in detail by Nellhaus, Haberland and Hill (1967) who considered the angiogram to be negative although there was obvious evidence of hydrocephalus and cerebral atrophy.

In their reports, Peterman, Hayles, Dockerty and Love (1958), Falconer and Rushworth (1960) and Rimon and Katila (1966) did not state the criteria they used in classifying the angiograms as abnormal or otherwise. The technique employed was also not described. It is possible that these authors might have used stricter critera in their radiological interpretation of abnormality than those advocated by Poser and Taveras, who also emphasised the use of rapid serial films to achieve more complete visualisation of the capillary and venous circulation. Poser & Taveras also remarked on the relatively low percentage of positive angiographic findings (37%) in patients with characteristic intracranial calcification in comparison with the 65% positive findings in the group with no visible gyriform calcification.

TABLE 1

An analysis of the results obtained from radiological procedure performed on 3 different series of patients since 1957

	Number of patients in each series	Radiological Examinations					
		Typical "Weber type" calcification on plain skull X-rays		Pneumoencephalogram		Cerebral angiogram	
		No. of patients examined	No. of patients with positive findings	No. of patients examined	No. of patients with positive findings	No. of patients examined	No. of patients with positive findings
Poser and Taveras (1957)	15	15	3	5	5	15	12
Peterman et al (1958)	35	35	22	5	5	5	0
Falconer & Rushworth (1960)	5	5	4	5	5	2	0
Rimon & Katila (1966)	2	2	2	Not done	–	2	0

In both the series presented by Falconer & Rushworth (1960) and by Rimon & Katila (1966), the patients presented with extensive gyriform calcification at the time of cerebral angiography, whereas calcification was noted in only 2 of the 12 "positive" cases of Poser & Taveras (1957). The lack of positive angiographic findings could be attributed to the extensive intracranial calcification that these patients had at the time of examination. In the absence of convincing clinical evidence and characteristic intracranial calcification, however, cerebral angiography may be the procedure of choice to show intracranial anomalies.

This case is presented to illustrate some common radiological manifestations of encephalotrigeminal angiomatosis. The under-developed left hemicranium with compensatory vault thickening, combined with angiographic evidence of mid-line shift to the ipsilateral side and paucity of capillary filling in the left posterior parietal region, were indicative of underlying cerebral atrophy.

A few interesting features in the venous system were shown, amongst which was non-filling of the left internal cerebral vein. Two attempts were made to fill this vessel without success although in the series done with cross compression the contralateral internal cerebral vein was visualised in the Towne's projection. One may assume therefore that the left internal cere-

bral vein was absent in this patient. This could be part of the venous anomalies associated with this condition although so far no such reports have been recorded. The venous angioma was small and might have been missed if rapid serial films were not done. The small angioma appeared to drain into a large left basilar vein which in turn emptied directly into the vein of Galen. The latter appearance was noted by Taveras & Wood (1964) and again by Banna and Young (1970) who considered it an anatomical variant. Partial agenesis or thrombosis of the superior sagittal sinus with prominent draining veins is a well recognised appearance and was described in 3 of Poser & Taveras' 15 cases. Two of these 3 cases were associated with the complete form of encephalotrigeminal angiomatosis showing no visible intracranial calcification.

The present case can be regarded as a complete form of the disease with angiographic evidence of cerebral atrophy and venous anomaly. It illustrates the value of cerebral angiography when negative plain films of the skull cast doubt as to the exact form of the disease. If the use of this procedure can be limited to clinically complete, or incomplete forms without characteristic gyriform calcification, results could be more rewarding. It can then contribute more significantly in establishing diagnosis and in assessing those cases in whom hemispherectomy or surgical excision of the venous anomaly is being considered.

Acknowledgements

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