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Isolated Frontal Lobe Calcification in Sturge-Weber Syndrome

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The Sturge-Weber syndrome is a phacomatosis characterized by encephalofacial angiomatosis, seizure disorder, and cortical calcification. There are several cases of atypical or incomplete forms of Sturge-Weber syndrome in which the patient presents with a seizure disorder and cortical calcification. The cortical calcifications have been described in the occipital, parietal, and temporal areas [1-4]. Our case is unique in that the calcification was isolated to the frontal lobe. Also, the patient did not have a facial angioma. She did present with a seizure disorder. An angiogram revealed abnormalities of the superficial cortical veins, and an excisional biopsy of the frontal lobe lesion revealed changes characteristic of Sturge-Weber syndrome.

Case Report

A 9-year-old girl presented with a history of right hemispheric seizures since the age of 1 year old. She had recently noted weakness of the left upper extremity. Physical examination revealed a left central

facial nerve deficit and left upper extremity weakness. No cutaneous signs of Sturge-Weber syndrome were noted. CT scan (Fig. 1) revealed a dense cortical calcification with associated cerebral atrophy in the right frontal lobe. On a contrast-infused scan, some enhancement of the involved frontal cortex was noted. A cerebral angiogram revealed a paucity of superficial cortical veins and persistent capillary blush in the right frontal lobe (Fig. 2). The arterial phase was normal.

A right frontal craniotomy was performed and the abnormal frontal cortex was resected. Histologic sections demonstrated an absence of large veins within the subarachnoid space. Numerous thin-walled venous channels greater than 200 μ m diameter were present in the white matter just beneath the cortex. Calcifications were numerous throughout the cortex and white matter (Fig. 3).

Discussion

Sturge-Weber syndrome is characterized by a port-wine-colored angioma in the facial trigeminal distribution, cerebral cortical calcification, and a seizure disorder. Other associated

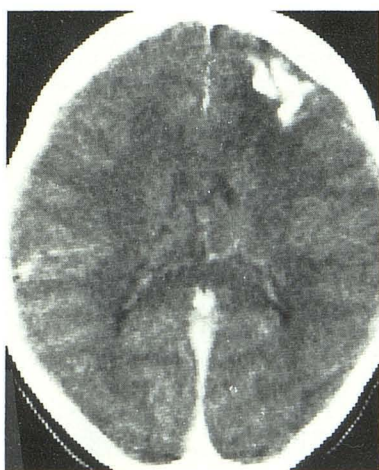
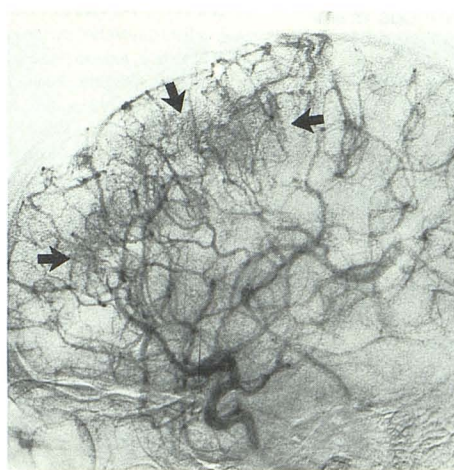
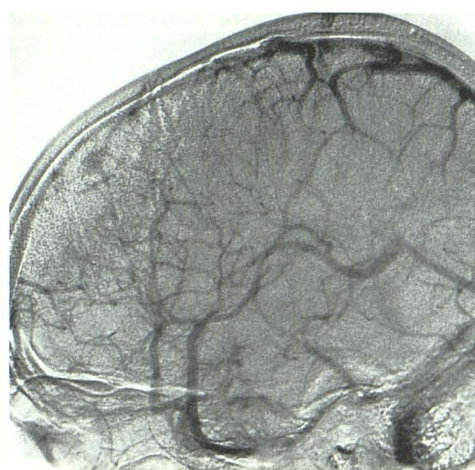


Fig. 1.—CT scan shows area of gyriform calcification in right frontal lobe.



A



B

Fig. 2.—Capillary (A) and venous (B) phases from right internal carotid injection show paucity of cortical veins in frontal lobe with dilated medullary veins. There is increased parenchymal blush (arrows) in frontal lobe.

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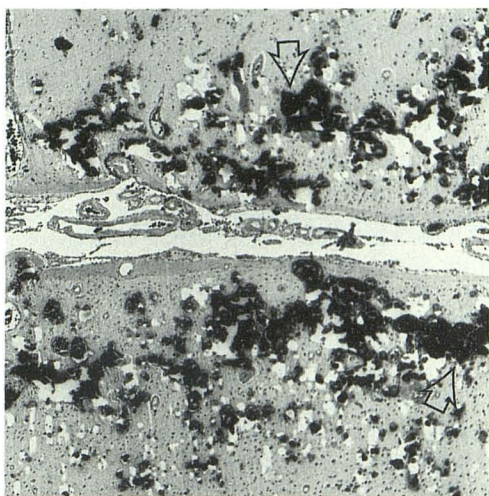


Fig. 3.—Histologic section of two cortical gyri on either side of a sulcus from resected frontal lobe lesion. Coarse spherical calcifications (arrows) are concentrated in superficial layers of cortex. Subarachnoid space contains numerous small muscular arteries and a paucity of thin-walled venous structures. (H and E $\times 45$)

features are mental retardation, hemiparesis, hemiatrophy of the body, and glaucoma [1].

There have been several documented cases of atypical or incomplete Sturge-Weber syndrome. These patients usually have a seizure disorder and cerebral cortical calcifications located in the occipital, parietal, or temporal lobes.

The present case is unusual because of the frontal lobe calcification. Several other diseases were considered to explain this finding, such as a glioma [3], ossifying meningoencephalitis [5], leukemia [6], and hemangioma calcifications [7]. Angiography revealed a paucity of cortical veins and a prolonged capillary blush, collateral superficial venous drainage, and some shunting of blood into the deep venous system. The arterial phase was normal. These findings led us to the diagnosis of Sturge-Weber syndrome. The findings are consistent with the classical description by Bentson et al. [8]. These authors believed that the basic abnormality was non-function of the superficial cortical veins with shunting of blood into the deep venous system.

The underlying cause of Sturge-Weber syndrome is un-

known. The cerebral cortical lesion is a leptomeningeal angiomas. There is some biochemical evidence of an abnormal mucopolysaccharide found in the walls of the angioma, which is believed to be the cause of the cerebral cortical calcification [9]. Others have suggested that the cause of the calcification is a defect in the blood brain barrier, which leads to the transudation of protein and calcium, which later crystallizes in the perivascular space [10]. The alteration in the blood brain barrier is thought to be caused by altered blood flow in the angioma or hypoxia from a prolonged seizure.

The CT findings in Sturge-Weber syndrome are basically cortical calcification in a gyriform pattern. The calcification can involve both the gray and superficial white matter. Gyriform and patchy cortical enhancement has been noted.

The clinical, CT, and angiographic findings of both typical and incomplete forms of Sturge-Weber syndrome have been well described. Our case is unusual because of the isolated cerebral cortical calcification located in the frontal lobe in a patient with seizures and no evidence of a facial trigeminal angioma. The cerebral angiogram and cerebral cortical biopsy are consistent with the diagnosis of Sturge-Weber syndrome.

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