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Sturge-Weber Syndrome : A Case with Choroid Plexus Hemangioma

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Abstract **Sturge-Weber syndrome (encephalotrigeminal angiomatosis) is an uncommon neurocutaneous disorder. Not many cases of this syndrome have been under the care of neurosurgeons. The syndrome is characterized principally by facial and leptomeningeal angiomas. Hemorrhage from intracranial angiomas in this syndrome is rare. In contrast, epilepsy occurred in nearly every published case. The only known indication for neurosurgical management is to control intractable seizures. This report describes a case of Sturge-Weber syndrome who presented with clinical subarachnoid hemorrhage. The angioma involving the choroid plexus, which was a probable cause of leakage, was demonstrated by computed tomography and surgical removal was performed. Histologic examination revealed a hemangioma of the cavernous type. The literature was reviewed.**

In 1879 Sturge[1] described a patient with a facial nevus flammeus who developed contralateral partial seizures and suggested a cortical vascular lesion as the epileptic source. Sturge's expectation was proved by Kalischer (1897) who demonstrated cortical angiomas in similar cases. Weber[2] in 1922 reported a radiographic finding of intracranial calcification associated with this condition and believed that the calcification was located in pial angiomas. In 1923, Dimitri described the characteristic double contoured (tramline) radiographic shadows in this syndrome. In 1934 Krabb[3] showed conclusively that the calcification was deposited in the second and third layers of cerebral cortex. Sturge-Weber syndrome (encephalotrigeminal angiomatosis) is one of the neurocutaneous disorders traditionally known as the fourth phakomatoses. Classic features of Sturge-Weber syndrome include facial port wine stain, contralateral seizure and ipsilateral cortical calcification. Principal pathology is angiomas involving both facial dermis and leptomeninges. The ocular choroid is another site commonly involved by angiomas resulting in glaucoma and buphthalmos.[4] Choroid plexus angioma in Sturge-Weber syndrome has seldom been reported in the literature.[5] Although intracranial angioma is the essential finding in Sturge-Weber syndrome, intracranial bleeding is rare.[4,6] We report a case of Sturge-Weber syndrome that had choroid plexus hemangioma as a probable cause of bleeding.

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Case Report

A 12-year-old child with port wine colored nevi located on the left side of his face (mainly in the distribution of the first division of trigeminal nerve) was admitted to our service. (Figure 1) He had a normal intellectual level. From two years of age, he had suffered from occasional partial motor seizure on the right side. There was no family history of a similar disorder. Three days before this admission, he had complained of a sudden severe headache and frequent vomiting and he was treated in a local hospital. On the day of admission, he had a brief period of unconsciousness with decerebrate posturing and opisthotonus. After regaining consciousness, he remained drowsy. Physical examination showed low grade fever, nuchal rigidity and early papilledema. Skull radiography showed no abnormal calcification. Computerized tomography demonstrated calvarial hemiatrophy, mild atrophy of parietal, temporal and occipital lobes, occipital gyral calcification, enlargement and increased enhancement of the glomus of the choroid plexus with prominent subependymal veins on the left side. (Figure 2) The patient was considered to have Sturge-Weber syndrome with choroid plexus angioma and was suffering from subarachnoid hemorrhage. Operation aimed at removal of choroid plexus angioma was performed on the 12th day of admission. At the time of left parieto-occipital craniotomy, findings included cloudy and thickened arachnoid membrane, and numerous tortuous dilated vessels over the atrophic parieto-occipital cortex. Large abnormal vessels were obliterated by cauterization. The trigone of the lateral ventricle was then approached by superior parietal occipital incision and a cerebrospinal fluid sample was collected. After that, a well circumscribed, dark red, grape-like mass (two centimeter in diameter) was removed from the glomus. Ventricular CSF examination revealed xanthochromic fluid containing 15 erythrocytes per cubic millimeter and 65 milligram of protein per deciliter (normal ventricular CSF protein is 6-12 mg/dl). Light microscopic examination showed a vascular tumor with dilated, blood-filled lumina. Walls were lined with flattened epithelium. There was little intervening

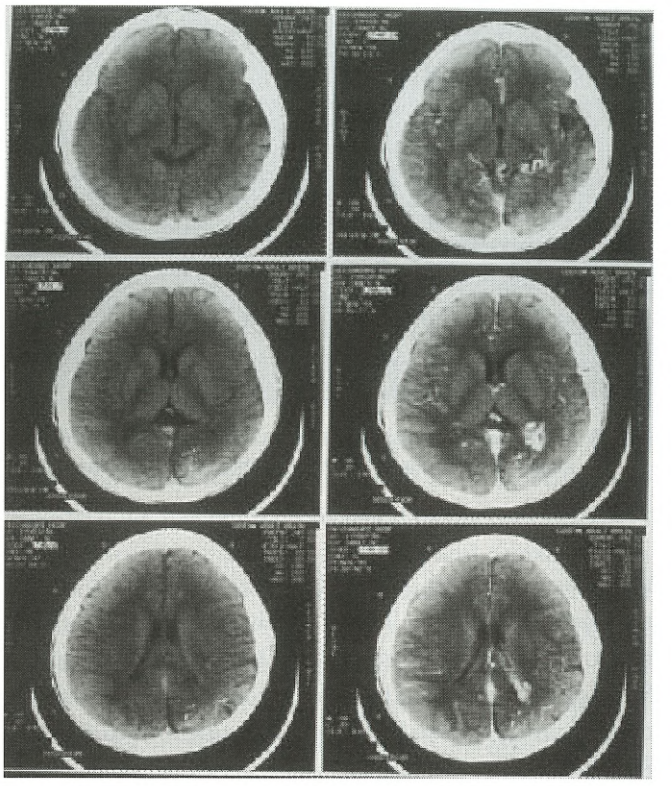

Fig. 2 Computerized tomography (left-plain CT, right-contrast CT) demonstrated calvarial hemiatrophy, atrophy of parietal, temporal and occipital lobes, occipital gyral calcification, enlargement and increased enhancement of the glomus of the choroid plexus with prominent subependymal veins on the left side.

Fig. 1 Facial port wine nevus located on the left side of the face, mainly in the distribution of the first division of trigeminal nerve.

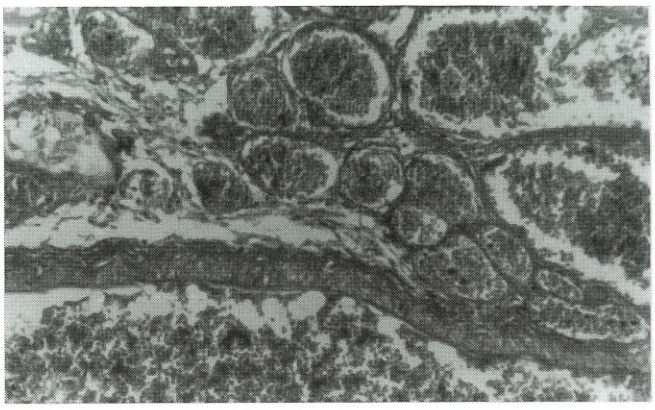

Fig. 3 Light microscopy showed a vascular mass with dilated, blood-filled lumina. There was little intervascular tissue.

intervascular tissue. Calcification was round in shape and intravascular. The histologic diagnosis was cavernous hemangioma with calcospherules. (Figure 3) The patient was discharged from the hospital without complications and he was free of symptoms during the 8-month follow-up period.

Discussion

Sturge[1] originally described the syndrome as consisting of facial port wine nevus associated with partial epilepsy and buphthalmos. Basic pathology of Sturge-Weber syndrome is angioma involving a few essential areas (facial dermis, leptomeninges and ocular choroid). Alexander, referring to Streeter's embryologic studies, suggested that pathogenesis of Sturge-Weber syndrome was due to vascular anomalies occurring during Streeter's third stage of cerebral vascularization at 4th to 8th weeks of gestation. At this early stage, the ectoderm that is to form the forehead skin lies directly over the part of the neural tube destined to form the posterior part (occipital lobe) of the cerebral hemisphere[7]. Based on new knowledge about cephalic embryogenesis, Enjolras[8] proposed a new hypothesis that a morphogenetic error, arising in a limited part of the cephalic neural crest could explain the syndrome. It is noteworthy that the three involved areas of the Sturge-Weber syndrome (i.e. facial dermis, leptomeninges and ocular choroid) are all mesenchymal derivatives of the neurectodermal germ layer, called the mesectoderm (the term ectomesenchyme is synonymous).[9,10] Sturge-Weber syndrome is also classified in the category of phakomatoses because both the skin and the nervous system are involved. Most phakomatoses are inherited, but not Sturge-Weber syndrome. The syndrome is usually sporadic in occurrence.[11] Recently it has been proposed that several sporadic syndromes are due to the action of an autosomal dominant lethal gene surviving by mosaicism.[11] The disorders cannot be transmitted to another individual because the underlying gene, when present in the zygote, leads to death of the embryo in an early stage of development. Cell bearing the mutation can survive only in a mosaic state, in close proximity with normal cells. A port wine nevus (nevus flammeus) is a dermal capillary hemangioma, and by definition, it is present at birth and remains fairly static in its extent. Involvement of the ophthalmic trigeminal area by port wine stains carries the risk of Sturge-Weber syndrome.[8] Ocular abnormalities due to choroidal angioma (glaucoma and buphthalmos) are found in approximately 30-40% of patients with this syndrome.[12] The risk of ocular problems is highest in patients with both ophthalmic and maxillary involvement by port wine lesions.[8,13] In our case, there was port wine stain overspread mainly on the ophthalmic area and over only a very small part of the maxillary area. No ocular abnormality was detected in this case. Ipsilateral to the facial nevus, abnormal leptomeningeal venous plexi are most commonly located over the posterior part of the cerebrum.[14] These vessels are nonfunctional because of their greatly delayed and prolonged filling. Anoxia and impairment of metabolism due to stasis might well be able to produce regional sclerotic atrophy with calcification in the cortical layers.[15,16] As the choroid plexus is a rich network of blood vessels of the pia mater (part of leptomeninges) which projects into each ventricular cavity, leptomeningeal angiomatosis might affect the choroid plexus as well. Choroid plexus hemangioma in Sturge-Weber syndrome has been previously reported in the literature.[5] Although subarachnoid hemorrhage in Sturge-Weber syndrome was noted in 1906 by Cushing but the first to prove subarachnoid hemorrhage with this entity was Anderson[6] in 1974. It is a very rare episode. Clinical findings (sudden severe headache, vomiting, brief loss of consciousness, nuchal rigidity and papilledema) along with operative and laboratory findings (xanthochromic ventricular CSF with erythrocyte and high protein content) in our case indicated that the patient had subarachnoid hemorrhage as a result of

leakage from choroid plexus hemangioma. Sturge-Weber syndrome produces a variety of characteristic changes in diagnostic imaging. The classic radiographic features (tramline gyral calcification) of this syndrome as seen on plain skull radiographs are relatively late since calcification is slight in infancy, gradually becoming denser until the maximum degree is attained probably by the second decade.[14,17] It is rare to show cortical calcification in plain radiography within the first year, and only 50-60% of patients by the age of 20 years demonstrate them on radiographs.[13,18,19] Relatively few cases of this syndrome have been studied by means of cerebral angiography. A localized increase in the size and number of veins and large capillaries (venous angioma and telangiectases) has been considered typical angiographic findings in Sturge-Weber syndrome. That only 24% of the cases are found by cerebral angiography might be explained by the slow circulation in the angiomatous area.[15,16]

Diagnostic imaging in Sturge-Weber syndrome is now based on computerized tomographic and magnetic resonance evaluation. Imaging findings include cortical calcification, cerebral lobar atrophy, calvarial hemiatrophy, prominent deep venous systems and choroid plexus enlargement.[20-22] Computerized tomography has the advantage of earlier and more precise detection of cortical calcification. Magnetic resonance imaging more sensitively demonstrates gray and white matter abnormalities and engorgement of the deep venous system.[23,24] Sturge-Weber syndrome produces major neurological problems such as epilepsy, sensorimotor deficits and intellectual deterioration. Since the syndrome is relatively rare and few neurosurgeons will encounter several examples during their professional life, so the best line of treatment still has not been developed. Surgical management is seldom, and only reluctantly, performed in various stages of the syndrome, mostly as a desperate attempt to control intractable seizures.[25,26] The basic abnormality in Sturge-Weber syndrome is non-function of the superficial cortical veins.[15,16] Blood flow is redistributed and the deep medullary veins then become overloaded, dilated and angiographically prominent. Occlusion of the deep venous drainage pathways, or their overloading and radial stasis might be the pathogenic factor for a general damaging process leading to intellectual impairment and dementia.[16,27] Surgical intervention prior to the stage of non-function of the central venous pathways could prevent such event. Frobst[16] suggested a close angiographic evaluation for this.

The choroid plexus angiomas in Sturge-Weber syndrome have been noted in a few neuropathology reports. But enlargement and increased enhancement of the choroid plexus, compatible with the presence of angiomatous malformation, showed by diagnostic imaging, appears to be a common finding in their syndrome.[20,22,24,28] The glomus, the most heavily vascularized portion of the choroid plexus, seems to be the most commonly involved site. Bleeding from choroid plexus angiomas as a cause of intraventricular hemorrhage has been mentioned in the literature.[29,30] The hemorrhage is usually sudden and massive but it may occur more slowly. The indication for excision should be established in patients with evidence of ruptured choroid plexus angiomas to prevent repeated hemorrhage. Further information is required concerning the risks of hemorrhage from asymptomatic choroid plexus angiomas in Sturge-Weber syndrome.

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