Sturge-Weber Disease With Subarachnoid Hemorrhage

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Subarachnoid hemorrhage (SAH) in encephalofacial angiomatosis or Sturge-Weber disease (SWD) is rarely mentioned in the literature. The few published reports usually have been deficient in details, or the diagnosis of SAH was not fully established. The present report describes the case of a woman with previously undiagnosed Sturge-Weber disease who was in excellent health until stricken with an SAH.

Case Report
A 32-year-old white woman was admitted to the Massachusetts General Hospital on April 6, 1972, with a severe headache and stiff neck. On April 3 she first noted a midscapular backache followed by a severe headache and pain in the cervical region.

She had been in excellent health all her life with no history of hypertension, seizures, or other neurological difficulties. She graduated from high school and worked as a secretary. There was no family history of seizures, skin lesions, or mental retardation.

Examination on admission showed the patient alert and of average intelligence. Blood pressure was 118/76 mm Hg. There was an extensive port-wine nevus over much of the face (left side more than right, and involving all three divisions of the trigeminal nerve) and the left side of the neck and anterior chest. Nuchal rigidity was present. No cranial, ocular or cervical bruit was heard. There was no tenderness or bruit over the spine. Neurological examination was normal except for increased deep tendon reflexes in the right limbs. Ophthalmological examination revealed no evidence of glaucoma or hemangioma of the choroid.

LABORATORY DATA
Complete blood count, prothrombin time and partial thromboplastin time were normal. The CSF was bloody under a pressure of 140 mm H$_2$O. The unspun fluid from the second and fifth tubes contained 25,000 and 23,000 red blood cells per cubic millimeter, respectively, and the supernatant was xanthochromic. The protein was 80 and the glucose 64 mg per 100 ml.

X-ray films of the skull and the cervical, thoracic and lumbar spine were normal. Cerebral angiography was performed via the femoral route with catheterization of both common carotid arteries and the left vertebral artery. In the left cerebral hemisphere there was rapid shunting (first seen at 1 sec after injection) through a capillary angiomatous malformation in the basal ganglia and thalamic regions (fig. 1). During the late arterial phase there was early filling of the deep venous system. The proximal end of the great vein of Galen was dilated while the distal end appeared attenuated, and retrograde flow occurred along the vein of Rosenthal (fig. 2). Also, there was an abnormal collection of early filling veins over the left parietal surface (fig. 3). No angiographical abnormalities were present in the right cerebral hemisphere, the posterior fossa, or cervical spine, and there was no evidence of an intracerebral mass, aneurysm, vascular spasm, cortical atrophy or ventricular dilatation.

COURSE
The patient's course was uncomplicated and she was discharged on diphenylhydantoin. Over the next two months the headache gradually disappeared and 14 months later she was well.

Comment
There is not complete agreement on the specific criteria for the diagnosis of SWD. Alexander considered the "essential pathological components of the syndrome" to be "the facial nevus, and the leptomeningeal 'angiomatosis'," the majority of the cases demonstrating radiological, neurological or ophthalmological secondary effects of the vascular anomalies. Poser and Taveras listed the cerebral angiographical abnormalities in SWD as capillary-
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Left carotid arteriogram. Rapid shunting through a capillary angiomatous malformation in the basal ganglionic and thalamic regions (arrows).

Right carotid arteriogram. Distension of the inferior sagittal sinus with poor filling of distal vein of Galen and of straight sinus.

Left carotid arteriogram. Early filling of abnormal parietal surface veins, sphenobregmatic vein and deep venous system during arterial phase (3 sec).

venous angioma, arteriovenous malformations, arterial thromboses, anomalies of the veins and dural sinuses, and anomalies of the external carotid artery. The main clinical features of this disease in addition to the cutaneous and intracranial vascular lesions are seizures, intracranial calcifications, mental retardation, ocular disorders (congenital glaucoma, buphthalmos and retinal or choroidal hemangiomas), hemianopia and hemiparesis with hemihypertrophy or hemiatrophy.

Our patient was relatively unusual in that she did not have seizures, appeared to be of normal intelligence and had a normal skull x-ray and ophthalmological examination. She did not have hemiparesis, hemiatrophy or hemihypertrophy, but she did have asymmetrical deep tendon reflexes. The presence of nevus flammeus and the characteristic intracranial vascular abnormalities (deep capillary telangiectasis, surface venous angioma and anomalous attenuation of the great vein of Galen) were sufficient to establish the diagnosis. Furthermore, some authors feel that in the presence of the characteristic intracranial lesions even the facial nevus need not be present to classify the case as SWD.6,8

There have been several reports of SWD in which the question of SAH has been raised. Cushing in 1906 reported three cases of "spontaneous intracranial hemorrhage associated with trigeminal nevi." However, the diagnosis of intracranial hemorrhage was based on a clinical picture of seizures (cases 1, 2, and 3), the onset of infantile hemiplegia (cases 1 and 3) and hemianopia (case 1). Cushing admitted that in none of the cases was there "proof positive that the acute illness which led to the cerebral complications was due to hemorrhage." In none of the cases was there mention of nuchal rigidity or a description of the cerebrospinal fluid. Livingston et al.4 (case 5) described a two-year-old child with seizures. The terminal episode involved convulsions, fever and a stiff neck. Postmortem examination showed subarachnoid...
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blood, but in the case description the “initial lumbar puncture produced clear fluid” and “the second puncture was recorded as a bloody tap.” Wohlwill and Yakovlev\(^7\) described an epileptic woman (case 4) who was found at autopsy to have a localized SAH and another case (case 1) in which they demonstrated clumps of brownish hemosiderin pigment attached to vessel walls. In neither case was there clinical history of SAH. Matson\(^8\) stated that “intracranial hemorrhage from the surface malformation must be exceedingly rare. It has occurred only once in our clinic and successful evacuation of a subdural hematoma was carried out in this case.” No other details were given. Kanzmeier and Voigt\(^9\) reported three cases of “von Hippel-Lindau Sturge-Weber” disease presenting with the symptoms of SAH but in which the examination was normal and the cerebrospinal fluid was clear.

Thus in none of the above cases was the diagnosis of SAH unequivocal. Poser and Taveras\(^2\) had no case of SAH in their large series of angiographically demonstrated cerebral vascular abnormalities and found none in their review of the literature. In the present case with well-documented SAH and cerebral vascular malformations the course was unusually benign, quite different from that described in past reports. The presence of multiple vascular abnormalities prohibits definite localization of the bleeding site, but the deep capillary telangiectasis and the surface venous angioma are suspect because of their location in the hemisphere contralateral to the side with hyperreflexia. The initial symptom of back pain raises the possibility of a spinal vascular lesion, but no source was found on the vertebral angiogram.

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References
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