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Abbreviated Reports

Sturge-Weber Syndrome with Extensive Intracranial Calcifications Contralateral to the Bulk of the Facial Nevus, Normal Intelligence, and Absent Seizure Disorder

Sturge-Weber syndrome is a rare condition first described by Sturge in 1879 in a case report of an epileptic patient [1]. The association of intracranial calcifications was subsequently described by Weber in 1922, giving this condition its familiar eponym [2]. The classical findings include gyriform intracranial calcifications, congenital and usually unilateral capillary facial nevus, convulsive disorder, glaucoma, some degree of mental deficiency, and hemiparesis and hemiatrophy.

Case Report

The subject of this report is a 57-year-old black woman with an extensive facial nevus affecting the left cheek, nares, and supraorbital area extending across the midline. This was present at birth and became roughened and pigmented with age. It had been partially resected for repeated bleeding and was reported to be an angioma. There was a long-standing history of glaucoma in the left eye with minimal light perception and marked corneal opacity. Vision on the right was correctable with lenses. She had hemiparesis and hemiatropy of the left hand and foot, which were evident since childhood. The right side was normal in size and strength.

Since childhood, the patient knew of her diagnosis of Sturge-Weber syndrome, which was based on physical appearance and skull radiographs. There was no history of seizure disorder. She complained of occasional left-sided headaches.

CT scans of the brain with and without contrast (Figs. 1 and 2) revealed bilateral gyriform calcifications in the parietal and occipital regions, more extensive on the right. The right cerebral hemisphere was smaller than the left, and the choroid plexus of the right lateral ventricle was larger with prominent contrast enhancement. Soft-tissue prominence in the left frontal region conformed to the facial nevus. There was calcification in the left globe, which was slightly smaller than the right.

Discussion

The occurrence of bilateral calcification in Sturge-Weber syndrome is well known and has been postulated to be as high as 15% [3]. Apart from one exception known to us, all cases have been associated with mental deficiency and history of seizure disorder. This exception was noted by Parnitzke in 1956 [4] and involved a 39-year-old man with bilateral gyriform intracranial calcifications who, in addition to lacking evidence of mental retardation or history of seizure disorder, had no facial nevus. Our case had the pathognomonic facial nevus on the left and extensive intracranial calcifications on the right; however, she had no evidence of mental retardation or seizure disorder.

Glaucoma and choroidal angiomas are known to occur in Sturge-Weber syndrome, and calcified choroidal angioma has also been reported [5]. However, it has not been well documented in the radiological literature.

On the CT scan, the enlarged choroid plexus of the right lateral ventricle was ipsilateral to the side of cranial hemiatrophy and extensive calcifications. Angiomatous malformation of the choroid plexus appears to be common in Sturge-Weber syndrome and may be the identifiable intracranial abnormality [6].

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REFERENCES

1. Sturge W. A case of partial epilepsy, apparently due to a lesion of one of the vasomotor centers of the brain. *Trans Clin Soc Lond* **1879**;12: 162–167
2. Weber FP. Right-sided hemi-hypertrophy resulting from right-sided congenital spastic hemiplegia, with morbid condition of the left side of the brain revealed by radiograms. *J Neurol Psychopathol* **1922**;3:134–139
3. Boltshauser E, Wilson J, Moore RD. Sturge-Weber syndrome with bilateral intracranial calcification. *J Neurol Neurosurg Psychiatry* **1976**;39:429–435
4. Parnitzke KH. Symptomwert und Symtomverteilung bei der Sturge-Weberschen Krankhest. *Zentralbl Neurochir* **1956**;16:92–109
5. Garrow A, Lowenstein A. A case of monocular hydropthalmia with special reference to its possible relation to the Sturge-Weber syndrome. *Br J Opthal* **1943**;27:335–354

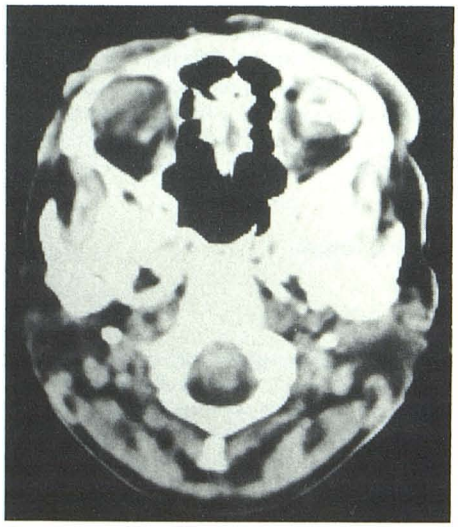

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Fig. 1.—Noncontrast CT scan near skull base showing extensive calcification of smaller left globe.

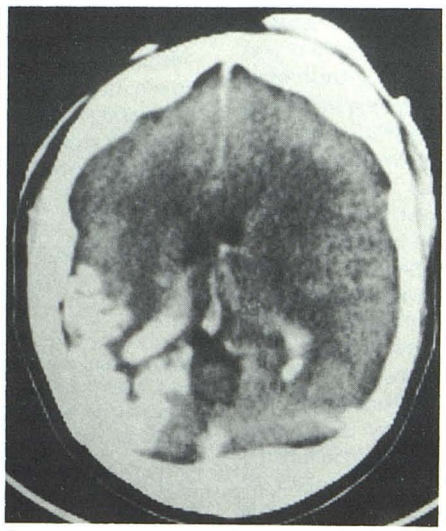

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Fig. 2.—Contrast-enhanced CT scan at level of lateral ventricle bodies showing right parietal gyriform calcifications and prominent contrast-enhancing glomus of right choroid plexus. Contrast enhancement in left frontal soft tissues and asymmetry in size of two cerebral hemispheres are also shown.

6. Stimac GK, Solomon MA, Newton TH. CT and MR of angiomatous malformations of the choroid plexus in patients with Sturge-Weber disease. *AJNR* **1986**;7:623–627

Pseudoforamina of the Skull Base: A Normal Variant

Radiologic evaluation of basal foramina of the skull is frequently a critical aspect in the diagnosis of patients with deficits referable to cranial nerves. Commonly encountered normal asymmetries and individual variations often make interpretation difficult. A pseudoforamen in the skull base was initially observed in skull radiographs of several patients and correlated with images of a dried skull. We stress the importance of recognizing pseudoforamina to avoid diagnostic confusion and error.

Bilateral, rounded lucent areas with sclerotic margins may be visualized on submentovertex or Water's views of the skull base. Located anteromedially to the hypoglossal canals and directly medial to the jugular foramen, these structures may be mistaken for the hypoglossal canal, erosion of the jugular foramen, or other normal structures (Fig. 1). The posterior margins are well defined while the anterior margins of pseudoforamina are indistinct, suggestive of bony erosion. Asymmetry between the two sides is frequent. Pseudoforamina may also be seen on CT scans of the skull base.

Using a dried skull for anatomic correlation, a pointer was placed adjacent to the medial border of the hypoglossal canal outlining the posterior margin of the pseudoforamen while the tip of a second pointer in the anterior condylar fossa lay in the center of the pseudoforamen (Fig. 2). The hypoglossal canal begins on the anteromedial aspect of the foramen magnum and runs anterolaterally and inferiorly, exiting the skull base lateral to the occipital condyles. The thicker medial wall of the hypoglossal canal forms the posterior wall of the pseudoforamen. The longus capitis muscle originates at the C6 vertebral level and inserts on a bony ridge just anterior to the anterior condylar fossa [1]. This ridge forms the anterior border of the foramen. The size and thickness of this ridge is subject to considerable individual variation, presumably reflecting muscle bulk and use patterns. This may also account for the variation in clarity with which the anterior margin of the foramen is seen. The anterior condylar fossa is located posterior to the bony insertion of the longus capitis muscle; its relative lucency accounts for the apparent opening of the pseudoforamen.

CT of the region of the foramen magnum in a dried skull also demonstrates the presence of the pseudoforamina (Fig. 3), but only at certain gantry angulations, which vary for each patient.

Correlation of radiographic features of the pseudoforamina in both dried skull and clinical case material demonstrates the anatomic basis for this normal variant. Recognition of its benign nature is vital to avoid diagnostic error in evaluation of the skull base.

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REFERENCE

1. Williams PL, Warwick R, eds. *Gray's anatomy*, 36th British ed. Philadelphia: Saunders, **1980**:305

An Unusual CT Appearance of Lupus Cerebritis

Cerebritis is a common complication of systemic lupus erythematosus (SLE). It affects 14–75% of SLE patients and is a leading cause of death [1]. Diagnosis of this disease remains difficult because of its nonspecific and varied presentation. Persistent headache, alteration in mental status, seizures, psychiatric symptoms, and stroke syndromes among others may represent lupus cerebritis. Patients with such symptoms require prompt corticosteroid therapy.

CT plays an important role in diagnosing lupus cerebritis, and its patterns have been the subject of several recent reviews [2–6]. Two general CT patterns have been described, one associated with an acute clinical presentation, e.g., infarction or focal hemorrhage, and another with more chronic symptoms and signs. CT may demonstrate single or multiple infarctions or hemorrhages in areas unusual for hypertensive bleeding. Patients with a chronic or insidious presenta-

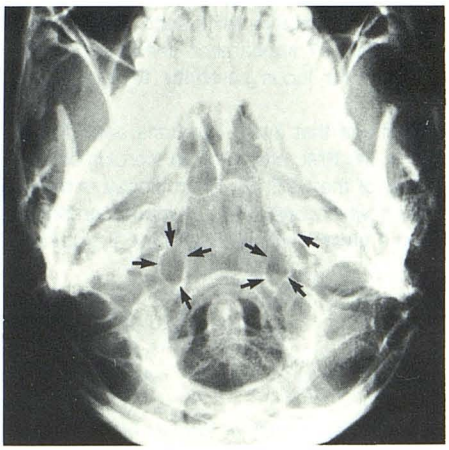

Fig. 1.—Plain film demonstration of bilateral pseudoforamina, more prominent on right (*arrows*).

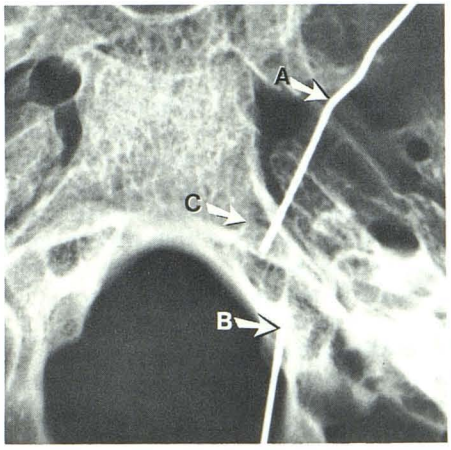

Fig. 2.—Dried skull radiograph. A = tip in anterior condylar fossa appears inside pseudoforamen; B = tip adjacent to medial border of hypoglossal canal; C = bony ridge for insertion of longus capitis muscle.

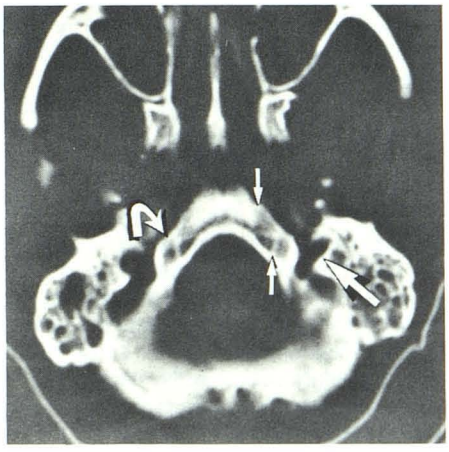

Fig. 3.—CT of dried skull demonstrates anterior and posterior borders of pseudoforamen (*small arrows*), showing relationship to hypoglossal canal (*curved arrow*) and jugular foramen (*large arrow*).