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## Gyriform Calcifications in Tuberous Sclerosis Simulating the Appearance of Sturge-Weber Disease

G. Wilms,<sup>1</sup> E. Van Wijck,<sup>1</sup> Ph. Demaerel,<sup>1</sup> M.-H. Smet,<sup>1</sup> C. Plets,<sup>2</sup> and J. M. Brucher<sup>3</sup>

**Summary:** Two cases of tuberous sclerosis are presented. Extensive superficial occipital calcifications were found as classically described in Sturge-Weber syndrome. Other radiologic signs of tuberous sclerosis, such as subependymal calcifications in both patients and surgically proved giant cell astrocytoma in one patient, were present. At pathologic examination, the calcifications appeared to be located in extensive cortical tubers.

**Index terms:** Phakomatoses; Sclerosis, tuberous; Brain, calcification

Sturge-Weber syndrome and tuberous sclerosis are both phakomatoses. In both neurocutaneous syndromes, intracranial calcifications are classically described with a typical appearance and localization that allow radiologic diagnosis (1). In Sturge-Weber syndrome the calcifications are tram-line with parieto-occipital cortical localization (1-4). In tuberous sclerosis, nodular calcifications with subependymal localization are present (1, 5-7). We describe two patients who, in addition to the typical subependymal calcifications, had superficial parieto-occipital calcifications resembling the radiologic appearance of Sturge-Weber syndrome.

### Case 1

After an uneventful pregnancy, a baby girl was delivered by forceps extraction in 1974. At 6 weeks of age, she developed several grand mal type epileptic seizures. Clinical neurologic examination was normal. Some pigmented nevi were found on the lower limbs beside other depigmented spots.

Plain radiographs of the skull (Fig. 1A) showed extensive calcifications in the temporo-occipital region. Selective angiography of the left internal carotid artery confirmed

the infrasyllvian localization of the lesion but without evidence of neoplastic vessels. Because of the young age of the patient, no surgical intervention was planned. Differential diagnosis was proposed between a posttraumatic hematoma, secondary to the forceps extraction, and a congenital tumor, such as a teratoma. In 1976, at age 2, she was readmitted because of an increase in frequency of the epileptic seizures. A computed tomography (CT) scan was now available and confirmed the extensive calcification of the temporo-occipital area.

At surgery, the cortex in the left occipital region appeared thin, with evidence of a firm tumoral mass in the subcortical area. Biopsy of the lesion showed a mixed astrocytoma, with giant and fibrillar cells, characteristic for tuberous sclerosis. Several follow-up CT scans with higher resolution confirmed the extensive calcifications and demonstrated typical subependymal calcifications near the foramen of Monro, as typically seen in tuberous sclerosis (Fig. 1B). At the present time, this girl is an adolescent. She still suffers from frequent epileptic insults, severe psychomotor retardation, cerebral palsy, and choreoathetosis. The patient receives antiepileptic drugs, but her epilepsy is poorly controlled.

### Case 2

In 1974, a baby boy was born after uneventful pregnancy and delivery. At 5 weeks of age, the patient presented with epileptic seizures. At clinical neurologic examination, a right hemiparesis was noted with ptosis of the right eyelid. Multiple depigmentation spots were found over the abdomen and under both limbs. Angiography and air encephalography indicated a space-occupying lesion in the left temporal area. At surgery, the sylvian fissure was markedly elevated, but the temporal convolutions were preserved. One centimeter beneath the cortex, a grayish-purple tissue was found with firm and calcified consistency. An anterior temporal lobectomy was performed. Anato-

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Received February 20, 1991; accepted and revision requested May 29; revision received July 30.

<sup>1</sup> Department of Radiology, University Hospitals K. U. Leuven, Herestraat 49, B-3000 Leuven, Belgium. Address reprint requests to G. Wilms.

<sup>2</sup> Department of Neurosurgery, University Hospitals, K. U. Leuven, Leuven, Belgium.

<sup>3</sup> Department of Neuropathology, Cliniques Universitaires Saint-Luc, Brussels, Belgium.

Fig. 1. A, Patient 1: Lateral skull radiograph. Extensive parieto-occipital calcifications suggestive of Sturge-Weber disease.

B, CT scan before contrast. Confirms the location of the calcification over the left temporo-occipital cortex. Notice typical subependymal calcifications in the wall of the lateral ventricles.

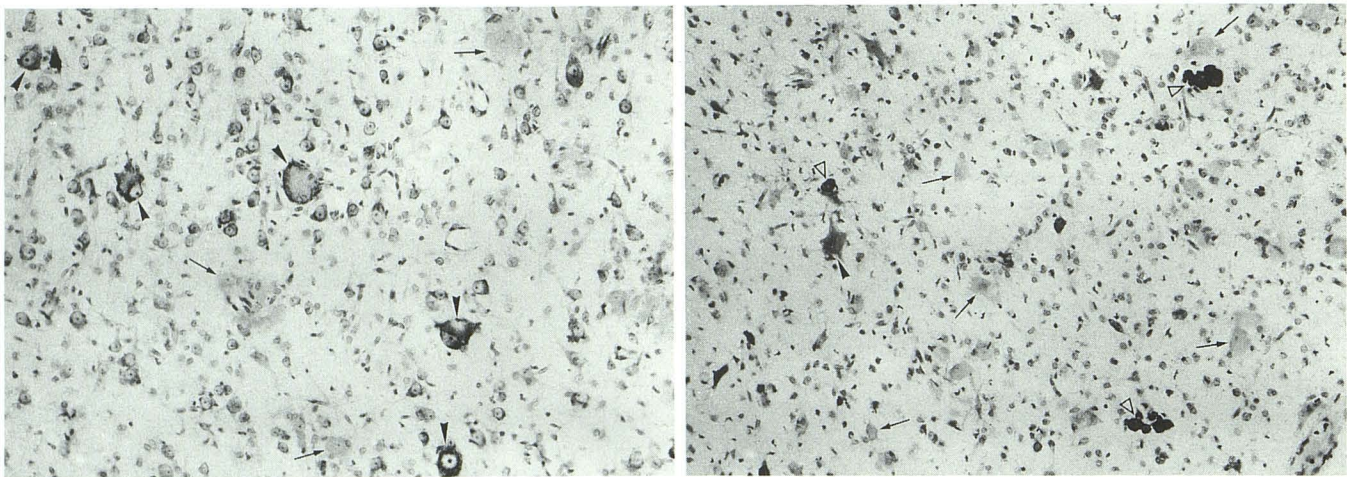
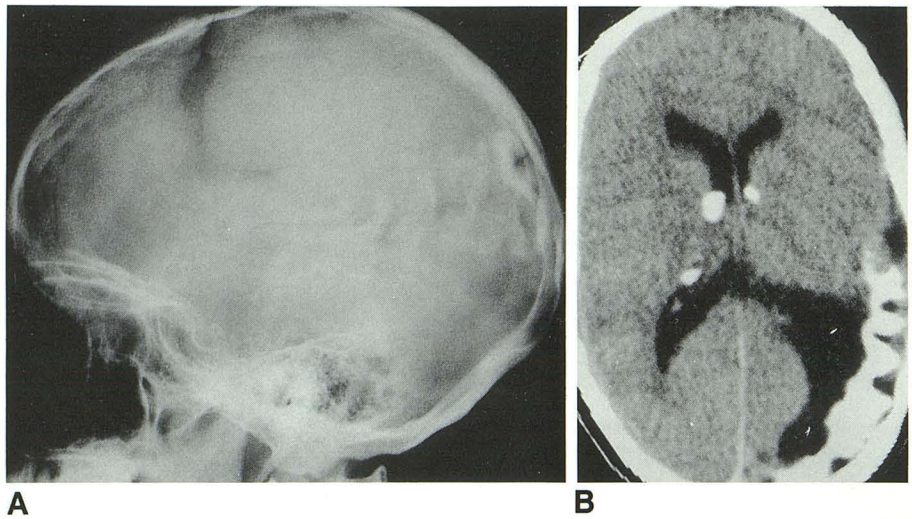


Fig. 2. Surgical biopsy of a diffuse temporo-occipital tuberosity.

A, Large, abnormal, neuronal cells (*black arrow heads*) and some hypertrophic astrocytes (*arrows*) are scattered in the fifth and sixth cortical layers.

B, Another cortical area contains numerous hypertrophic astrocytes (*arrows*), few large neuronal cells (*black arrow head*) and some darkly stained, aggregated calcospherites (*open arrow heads*). Paraffin section, cresyl violet,  $\times 189$ .

C, CT scan. Extensive cortical calcifications in right temporo-occipital region.



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mopathologic examination indicated a typical calcified cortical tuber, as seen in tuberous sclerosis (Figs. 2A and 2B).

Initial CT scan, performed in 1976, confirmed the extensive gyriform calcifications in the left temporo-occipital region, together with subependymal calcifications near the foramen of Monro.

During the clinical follow-up in 1982, on CT scan (Fig. 2C) a tumoral mass was noted near the right foramen of Monro, with obstructive hydrocephalus. At surgery, the expected giant cell astrocytoma was confirmed.

## Discussion

In tuberous sclerosis, four types of intracerebral lesions may be found: cortical tubers, white matter clusters of heterotopic giant cells, subependymal nodules, and subependymal giant cell astrocytoma (8–11). The most characteristic radiologic features of tuberous sclerosis are the subependymal calcifications, seen along the external walls of the lateral ventricles. These calcifications can be subcortical or cortical, occurring in heterotopias or cortical tubers (1, 5–7). Cortical tubers consist of gyral broadening with abnormal firmness or palpation and pachygyria-like appearance (8, 9). They are most often located in the frontal lobe but occipital occurrence is not infrequent. Calcification within cortical tubers is not uncommon, increases with increasing age, and may be seen in up to 50% of the patients by age 10 (6, 8, 9). Gyriform calcification of cortical tubers, as in our cases, has, to our knowledge, not been described.

In Sturge-Weber syndrome, serpiginous gyriform calcifications in the temporo-occipital region are the most typical radiologic feature (1–4). These calcifications are localized within the cortex and appear as curvilinear tram-line densities that follow the cerebral convolutions. Similar gyriform calcifications have been described in glioma (4–12), infarction (12, 13), purulent meningitis (14), viral encephalitis (15), ossifying meningoencephalitis (16), leukemia following intrathecal administration of methotrexate and skull irradiation (17), and subarachnoidal fat (18). Our cases add to this differential diagnosis.

In our cases, the calcifications were dense without obvious tram-line patterns, were located in the subcortical area, and there was no hemispheric hypertrophy. These features, together with the typical subependymal calcification, allowed a diagnosis of tuberous sclerosis in our patients.

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