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# Case Reports

## Fibrous Dysplasia of the Skull, with Seizures and Focal Electroencephalographic Findings

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### Introduction

Fibrous dysplasia is a non-hereditary disease of childhood, of unknown etiology, in which fibrous tissue replaces normal bone. There is a wide spectrum of involvement, from asymptomatic isolated bone-lesions to severe debilitation and the rare Albright's syndrome of osteitis fibrosa disseminata, hyperpigmented skin lesions and endocrine dysfunction (Albright *et al.* 1937). The following case demonstrates how closely fibrous dysplasia may resemble an intracranial malignancy, both clinically and electroencephalographically. Although seizures have been noted in prior reports (*e.g.* Sassin and Rosenberg 1968), electroencephalographic features of fibrous dysplasia have not been described previously.

### Case Report

This 17-year-old, right-handed Negro girl had had intermittent left parietal headaches for about 20 months and had two grand mal convulsions during a four-month period prior to her admission to hospital. Menarche had occurred at age 12 and secondary sex characteristics were normal.

The general physical examination revealed a slightly tender, firm fixed mass (6 × 8cm) over the left parietal bone.

Neurological examination revealed questionably increased reflexes in the right upper extremity but no weakness or abnormal reflexes.

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Blood studies, including alkaline phosphatase (63 and 73IU), calcium (10·3 and 9·7mg/100 ml) and phosphorus (3.8 and 3.7mg/100ml) were within normal limits. Roentgenograms demonstrated a 6 × 7cm lesion in the left parietal bone (Fig. 1), with normal location and contour of the inner table but marked bulging of the outer table as a result of an expanding process within the bone. A $^{99m}$Tc pertechnetate brain-scan showed a definitely abnormal increased uptake, about 5cm in diameter, over the left parietal bone but apparently not extending into the cerebral hemisphere.

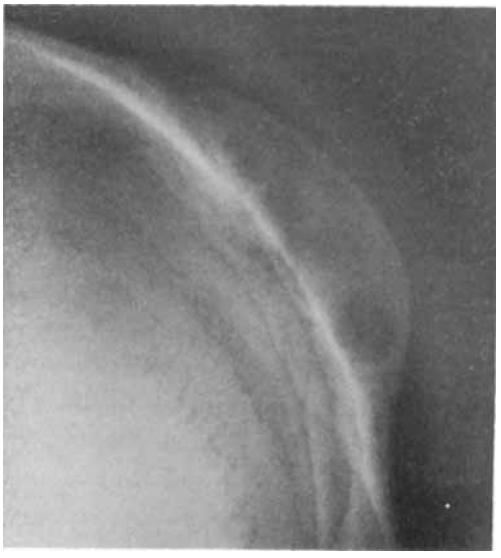

**Fig. 1.** Highlight of irregularly radiolucent and radiopaque left parietal bony lesion. Note intact inner table and outward bulging of outer table to a thickness of approximately 2cm.

A 16-channel electroencephalogram (EEG) and recording of computer-averaged visual evoked potentials were performed. The EEG revealed augmentation of voltage of intrinsic patterns in the left central region and intermittent sharp theta-frequency discharges in the left centro-parietal area. The most marked changes were at the anterior extent of the palpable lesion (Fig. 2).

The visual evoked responses are shown in Figure 3. The findings are focal to the area of the bony lesion. There is voltage depression in the left parietal region ($P_3$) of the two components designated by arrows. These two waves also show a latency shift, peaking earlier than their counterparts on the right side. Similar but distinctly lesser changes may be observed in the occipital area.

A percutaneous transfemoral selective angiographic study of the left external and internal carotid arteries, the right internal carotid artery, and the right vertebral artery revealed only slightly enlarged anterior and posterior branches of the left middle meningeal artery, and faint staining within the mass.

Craniectomy and removal of the left parietal bone mass were performed under general anesthesia, followed by an acrylic cranioplasty. No indentation of the dura was seen. The abnormal tissue was diagnosed as sclerotic fibrous dysplasia.

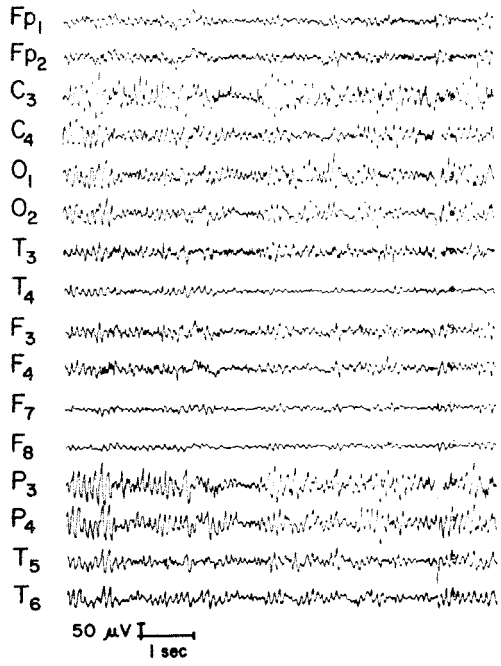


**Fig. 2.** Preoperative 16-channel electroencephalogram. Reference is joined ears.

The patient made an uneventful recovery and has been followed up with standard anticonvulsant therapy in the outpatient department. She has had rare generalized seizures.

## Discussion

Cranial fibrous dysplasia presenting as convulsive seizures is not rare, although deformity and headaches are more likely to be the initial complaints. Sassin and Rosenberg (1968) mention that six of their 50 cases with central nervous system involvement had 'nonfocal convulsive seizures', but unfortunately no electroencephalographic data were presented.

Fibrous dysplasia of the skull may clinically and radiographically mimic meningiomas (Windholz 1947, Feiring *et al.* 1951, Leeds and Seaman 1962, Gass 1965,

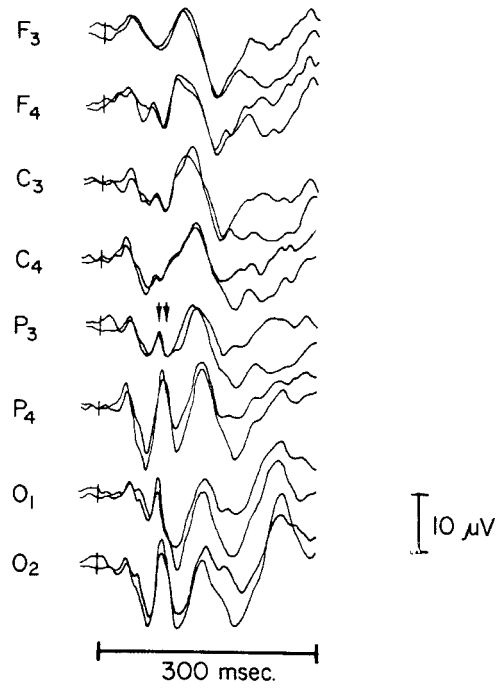


**Fig. 3.** Visual evoked responses of patient. Focal voltage depression of response, particularly of surface-negative and surface-positive components (designated by arrows) may be observed in left parietal region ($P_3$). Lead $P_3$ was situated over area of outward bulging of outer table of skull (Fig. 1). Lead placements correspond to the 10–20 system. Reference is joined ears.

Finney and Roberts 1976) and hypoparathyroidism (Leeds and Seaman 1962, Cahill 1963). The present case well illustrates the potential confusion with meningioma, as the lesion presented with convulsive seizures and there was a positive brain-scan and electrophysiological evidence of a focal cerebral lesion. Age at presentation is of diagnostic importance, since the active phase of fibrous dysplasia is during childhood, while meningiomas are relatively rare in childhood and become increasingly common with advancing age.

The sensitivity of the technique of visual evoked potentials for localizing the lesion is underscored in this case. Previous studies of visual evoked potentials have established normal limits (Kooi and Bagchi 1964, Ciganek 1969, Harmony *et al.* 1973) and an increasing amount of data is accumulating with regard to cerebral abnormality.

Precisely how fibrous dysplasia of the skull in this or other cases is associated with seizures remains an enigma. Chance occurrence of the mass lesion and the focal electroencephalographic abnormalities, while unlikely, cannot be absolutely disproved, especially in a single case. But certainly the presence of clinical seizures supports the hypothesis that the focal findings may be related to the mass lesion.

Artifact related to bony or soft tissue deformity is a recognized problem in interpretation of the EEG. In this case there was relative voltage *enhancement* of cortical potentials surrounding the lesion according to the EEG, but voltage *suppression* of the visual evoked potentials. These findings are apparently inconsistent with the supposition that the EEG and VER abnormalities are solely due to the same artifact.

The hypervascularity of the mass lesion, as evidenced by both the brain scan and angiography, suggests the possibility of relative ischemia of cerebral cortex resulting from a 'steal' phenomenon. However, the lack of anastomotic vessels in the dura at operation and angiography would make a focal 'steal' syndrome less tenable, although it is possible that the external carotid artery may have taken more than its share of common carotid flow.

One further possibility is that the clinical and electrophysiological manifestations of the patient's extradural mass were the effect of local pressure on the cerebral cortex, which presumably could lead to metabolic derangement of cerebral tissue. But since no displacement of the dura was reported at operation, this seems less likely. However, the standard technique of hyperventilating this patient during craniotomy to reduce the intracranial volume would have obscured a minor depression of the underlying cortex.

This case illustrates that pathological verification is a requisite for definitive diagnosis of fibrous dysplasia of the skull. The features of the disease, including convulsive seizures, are extremely variable and may often 'masquerade' as more ominous lesions, even after careful history-taking, clinical examination, laboratory studies, plain skull roentgenography, radioisotope studies and cerebral angiography. Focal abnormalities in electroencephalography and computer-averaged visual evoked potentials, as reported in this example, also may be seen.

AUTHORS' APPOINTMENTS

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SUMMARY

A 17-year-old girl presented with major motor seizures and headaches, and electroencephalography and computer-averaged visual evoked potentials showed focal abnor-

malities. Fibrous dysplasia without intradural involvement was verified at operation. Despite extradural confinement of the mass, the visual evoked potentials were sensitive enough to allow correct localization of the lesion.

Cranial fibrous dysplasia more often presents with deformity, headache or cranial nerve encroachment, but convulsive seizures may also be a presenting factor. Mass effect or a vascular 'steal' phenomenon upon cerebral tissue are possible explanations for these phenomena, and alternative explanations are discussed.

RÉSUMÉ

*Dysplasie fibreuse du crâne avec comitialité et anomalies électroencéphalographiques focales*

Chez une fille de 17 ans, présentant des crises comitiales et des maux de tête, il fut observé des anomalies focales à l'EEG et aux potentiels visuels évoqués moyennés par calculateur. La dysplasie fibreuse sans atteinte intradurale fut vérifiée à l'intervention. En dépit de la localisation extradurale de la masse, les potentiels évoqués visuels furent assez sensibles pour permettre une localisation correcte de la lésion.

La dysplasie fibreuse du crâne se présente plus souvent avec des déformations, des maux de tête ou des compressions des nerfs crâniens mais il est souligné que des crises convulsives peuvent également être observées. L'effet de la masse ou un phénomène de 'subtilisation' vasculaire sur le tissu cérébral sont des explications possibles des phénomènes et d'autres explications sont discutées.

ZUSAMMENFASSUNG

*Fibröse Dysplasie des Schädels mit Krampfanfällen und fokalen elektroencephalographischen Befunden*

Es wird von einem 17 jährigen Mädchen berichtet, das grosse Anfälle und Kopfschmerzen hatte und bei dem fokale elektroencephalographische Veränderungen sowie visuell evozierte Potentiale gefunden wurden. Bei der Operation fand sich eine fibröse Dysplasie ohne Penetration in die Dura. Obwohl der Tumor auf den Extraduralraum beschränkt war, waren die visuell evozierten Potentiale sensitiv genug, um eine genaue Lokalisation der Läsion zu erlauben.

Die cranielle fibröse Dysplasie geht häufiger mit Deformierungen, Kopfschmerzen oder Beteiligung der Hirnnerven einher, es wird jedoch darauf hingewiesen, dass auch Krampfanfälle auftreten können. Als Erklärung für dieses Phänomen kommen Tumoreffekt oder das sogenannte Gefäss 'steal' Phänomen auf das Hirngewebe in Frage; ausserdem werden Alternativerklärungen diskutiert.

RESUMEN

*Displasia fibrosa del cráneo con convulsiones y hallazgos electroencefalográficos focales*

Una muchacha de 17 años de edad presentaba crisis motoras mayores y cefalálgias y se halló que tenía anomalías focales en el electroencefalograma y unos potenciales evocados visuales computados de promedio. En la intervención quirúrgica se observó que presentaba una displasia fibrosa sin alcanzar la estructura intradural. A pesar de la limitación extradural de la masa, los potenciales evocados visuales eran suficientemente sensitivos para permitir la correcta localización de la lesión.

La displasia fibrosa craneana da lugar con mayor frecuencia a deformidad, cefalàlgia y alteración de nervios craneales, pero se pone de relieve que los ataques convulsivos pueden

también estar presentes. El efecto de masa o un fenómeno de 'captura vascular' sobre el tejido cerebral son explicaciones posibles para este fenómeno y se discuten otras posibles alternativas.

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# Transient Thrombocytopenia in a Child on Sodium Valproate

*A. P. Cole*

## Introduction

Sodium valproate has proved to be one of the most effective anticonvulsants in the treatment of intransigent epilepsy in both children and adults (Kugler *et al.* 1973, Barnes and Bower 1975). It has also been one of the best-tolerated anticonvulsant drugs. However, thrombocytopenia (Winfield *et al.* 1976) and platelet dysfunction (Voss 1976) have been reported when this drug is used alone or in combination with other anticonvulsants.

## Case Report

S.W. had been delivered with difficulty by forceps after a prolonged labour. She had a generalised convulsion at nine months, and from the age of three years was subject to non-febrile weekly grand mal attacks and daily petit mal attacks. During febrile illnesses the convulsions occurred more frequently. Her inter-ictal EEG showed centrencephalic polyspike activity and three per second complex spike and slow waves.

Five different anticonvulsants had failed to control her attacks. Her behaviour was so disturbed that schooling was impossible and she was receiving tuition at home.

At the age of six years sodium valproate was substituted for her previous medication, resulting in a 75 per cent reduction in the frequency of her convulsions. She started going to a school for the physically handicapped.

At $7\frac{1}{2}$ years she was admitted to hospital complaining of headache, sore throat and a cough, and was found to have a temperature of 39°C, with an increased respiratory rate. During the previous 24 hours she had had six grand mal seizures. The fauces were inflamed. A chest X-ray revealed a

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