Fibrous Dysplasia of the Parietal Bone with Focal Motor Seizures: A Case Report

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Patient: Female, 18-year-old

Final Diagnosis: Fibrous dysplasia

Symptoms: Seizure

Clinical Procedure: —

Specialty: Neurology • Neurosurgery

Objective: Rare disease

Background: Monostotic fibrous dysplasia is a benign proliferation of fibrous and osseous tissues that expand medullary bone to cause symptoms due to compression of adjacent organs and anatomical structures. Focal seizures are rarely the first sign of this kind of lesion. This report describes a young female patient with left-sided focal motor seizures associated with fibrous dysplasia presenting as a mass in the right parietal bone.

Case Report: An 18-year-old female student with left-sided focal motor seizures presented with a mass in the right parietal bone. Computed tomography revealed an expansile mixed-density lesion on the right parietal bone, a relatively homogeneous ground-glass appearance in the outer circumferential portion, and a lucent eccentric area with thinned but sclerotic borders. Magnetic resonance imaging revealed a homogeneously hypointense signal on T1WI, a small hyperintense signal on T2WI, and avid enhancement signal intensity on post-contrast T1. Electroencephalogram showed inter-ictal epileptiform activities derived from the right fronto-central lobe. Surgical en bloc resection with a margin of normal bone and cranioplasty were performed. Histopathology showed features indicative of fibrous dysplasia, including osteoid trabeculae arranged haphazardly in a dense fibroblastic stroma, irregular trabeculae lacking conspicuous osteoblastic rimming, and intervening fibrous stroma containing cytologically bland spindle cells. The patient achieved seizure control and has remained neurologically intact.

Conclusions: This report has highlighted the importance of early diagnosis of fibrous dysplasia of bone to exclude primary bone malignancy or bone metastasis, to ensure rapid management and symptom control.

Keywords: Case Reports • Craniofacial Fibrous Dysplasia • Epilepsies, Partial • Bone Diseases • Seizures

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Introduction

Fibrous dysplasia is a developmental anomaly characterized by the localized arrest of normal architectural development, where abnormal differentiation of osteoblasts results in the substitution of normal marrow and cancellous bone by immature woven bone and fibrous stroma [1]. It may manifest either in a single bone (monostotic) or in multiple bones (polyostotic) [2].

Patients with fibrous dysplasia commonly present symptoms as a result of gradually and insidiously growing mass lesions due to facial deformity and distortion of adjacent skull base structures [3]. In cases involving the craniofacial bones, fibrous dysplasia is more frequently observed in the zygomatic-maxillary complex and anterior cranial base than in the calvaria [4]. Therefore, neurological symptoms such as focal seizures are rarely reported in association with fibrous dysplasia [5-9]. This report describes the case of an 18-year-old female student with left-sided focal motor seizures associated with fibrous dysplasia presenting as a mass in the right parietal bone.

Case Report

An 18-year-old female student was admitted to our hospital due to left-sided focal seizures lasting 3 months. Three months ago, she noticed a painless mass in the right parietal bone gradually increasing in size and becoming palpable prior to presentation, without any clinical signs. Two weeks prior to admission, she had the first seizure. Symptoms started with a tingling sensation over the left hand, spreading along the left arm and left face, then focal tonic and clonic on the left side. She experienced postictal left hemiplegia that completely disappeared a day later. She was admitted to the local hospital and given valproate 200 mg bid, but she still had 3 similar episodes of focal seizure during the next 2 weeks. She was then transferred to our hospital. We increased levetiracetam 500 mg bid and she had 1 more seizure.

She was evaluated at the epilepsy center in our hospital. Examination revealed a mass over the right side of the head behind the coronal suture, measuring 3 cm in diameter. It was tender but not painful or warm, had a firm bony structure, appeared to connect with the underlying bone, and showed no attachment to the overlying normal skin. No similar protrusions were found in other regions of the body, and there were no other abnormal findings on general and neurological examination.

A brain computed tomography (CT) scan revealed an expansile mixed-density lesion on the right parietal bone and discontinuity of the cortical surface on the outer and inner tables of the skull. A pattern with mixed areas of radiopacity and radiolucency was found. The outer circumferential portion had a relatively homogeneous ground-glass appearance. The lesion had a lucent eccentric area with thinned but sclerotic borders (Figure 1A). Magnetic resonance imaging (MRI) revealed homogeneously hypointense signal on T1WI. The eccentric lesion had a small hyperintense signal on T2. MRI T1 post-contrast images showed avid enhancement signal intensity (Figure 1B-1D).

Baseline activity of awake electroencephalogram (EEG) recording in the patient showed alpha waves with a frequency of 10 Hz and amplitude of 80 μV in symmetrical parietal regions, which were inhibited as the eyes opened. Beta activity had a frequency >13 Hz, with amplitude spreading bilaterally ranging from 15-20 μV. The asleep EEG showed monophasic and biphasic slow-wave activities: reduced alpha amplitude, increased beta activity, positive occipital sharp transients of sleep (POSTs), and Vertex waves, as well as slow-wave delta theta activity in the right temporal and central-frontal regions (F4/C4/T8/T4/T6), more predominant in the frontal area, consistently extending throughout the recording. Interictal EEG recording showed some sharp activities occasionally accompanied by slow waves, recorded in the right central-frontal region (F4/C4) during sleep (Figure 2).

A right parietal craniectomy was performed with en bloc resection of the tumor, and a rim of normal bone (Figure 3A). Findings at surgery included a pinkish, solid, well-circumscribed tumor of the bone, expansile in nature and vascular, compressing the adjacent dura but not invading it. The surrounding bone was thickened in the frontal temporal region, with enlarged emissary veins. The tumor was not attached to the overlying skin (Figure 3B, 3C). We resected the dura to examine the invisible structures below. The arachnoid matter was still thin and transparent. There was no abnormal appearance in the cerebral cortex. The dura was reconstructed by watertight dural closure with an absorbable dural patch. Cranioplasty was performed with a titanium plate (Figure 3D).

The histological section showed osteoid trabeculae, which are narrow, curvilinear, or irregularly-shaped osteoid trabeculae, arranged haphazardly in a background of dense fibroblastic stroma (Figure 4A), and irregular trabeculae of bone with no conspicuous osteoblastic rimming in a fibrous background (Figure 4B, 4C). Intervening fibrous stroma contained cytologically bland spindle cells without cytologic atypia (Figure 4D). These features suggest a diagnosis of fibrous dysplasia.

On the third postoperative day, the patient had minor motor seizures only in the left hand. She was followed up and discharged uneventfully, without any further seizures. We tapered and stopped valproate because she was of reproductive age. The antiseizure medications were continued with levetiracetam 500 mg bid in the postoperative period. The patient has remained neurologically intact. She has been followed up for the last 6 months and has been seizure free.
Discussion

Fibrous dysplasia is a benign, slow-growing lesion that takes months to years before symptoms manifest, or more rarely, malignant transformation [1-3,10]. Initiation and progression of fibrous dysplasia have resulted in activating mutations of the GNAS1 gene [10]. They are reported to represent approximately 5% of benign bone tumors [1]. Common signs and symptoms of fibrous dysplasia, varying by lesion location, include pain, bony deformity, and, occasionally, pathological fractures [3,10]. Focal motor seizure disorder, as seen in our patient, is a rare manifestation of calvarial fibrous dysplasia [5]. This report has highlighted the importance of early diagnosis of fibrous dysplasia of bone to exclude primary bone malignancy or bone metastasis, to ensure rapid management and symptom control.

We have summarized previously reported cases of calvarial fibrous dysplasia with epilepsy (Table 1). Despite reports of 6 “nonfocal convulsive seizures” in a case series, there were no other supporting data describing this group of patients [9]. A few recent reports showed fibrous dysplasia of the skull with seizures, but they did not mention EEG, or gave a brief

Figure 1. Preoperative imaging. (A) Computed tomography scan revealed an expansile ground-glass lesion with well-defined borders on the right parietal bone. (B-D) Magnetic resonance imaging showed homogeneously hypointense on T1WI, a small hyperintense signal in the eccentric portion on T2WI, and avid contrast enhancement.
non-specific EEG finding [6,8]. Another report proposed that the presence of epileptiform morphology in calvarial fibrous dysplasia can manifest as a breach effect-benign EEG pattern resulting from a focal abnormality within the skull [7]. There has been only 1 previous report showing relevant focal abnormalities on the EEG that had histological confirmation and reported outcome [5]; this case presented many similar findings to our own: a young female with motor seizures and epileptiform activity on EEG. Fibrous dysplasia without intradural invasion was examined during the operation. The authors proposed theories such as mass effect or a vascular ‘steal’ phenomenon upon cerebral tissue. Our patient presented with somatosensory onset with Bravais-Jacksonian march from the left hand along the left arm and face, followed by a left-sided clonic seizure, suggesting that the seizures started over the right parietal area and spread to the motor cortex. EEG recording showed epileptiform discharges over the right central-frontal region. With the anatomo-electro-clinical correlations, we believed that the lesion over the left parietal was the epileptogenic lesion. During the multidisciplinary epilepsy team meeting, we decided to perform a lesionectomy.

Fibrous dysplasia consists of varying amounts of cellular fibrous stroma and trabeculae of immature woven bone [11]. Imaging findings depend on the lesion stage, from early dominant fibrous elements to progressive calcification [12]. The presence

Figure 2. Electroencephalogram showing interictal epileptiform activities derived from the right fronto-central lobe (F4/C4).
of well-vascularized fibrous tissue with numerous small ves-
sels in the center and large peripheral sinusoids is a notable
histological feature of fibrous dysplasia [12]. These charac-
teristics explain a ground-glass appearance on CT, a hypoin-
tense signal on T1WI, an eccentric lesion with small hyperin-
tense signal on T2, and an avid enhancement signal intensity
on post-contrast T1 [13]. They are also indications of a meta-
bolically active lesion [12,14,15]. Fibrous dysplasia can resem-
ble other tumors of the skull, making diagnosis uncertain until
surgical excision and histology [3,10]. Calvarial lesions with a
ground-glass appearance should be considered in the differen-
tial diagnosis, which includes Paget disease, intraosseous
meningioma, osteosclerotic metastases (from prostate, breast
cancer, lymphoma), and malignant transformation in fibrous
dysplasia into sarcoma [10,13,14,16].

Patients with monostotic fibrous dysplasia are frequently with-
out symptoms and should have regular follow-up to assess
for any new symptoms and radiographic changes [1-3,10]. The
current treatment of choice for symptomatic cranial fibrous

Figure 3. Intraoperative clinical features and postoperative follow-up. (A) Intraoperative view revealed no abnormal findings in the
cerebral cortex and arachnoid mater below the bone lesion. (B, C) Gross specimen of the excised lesion showing the well-
circumscribed mass with a rim of normal bone. (D) Postoperative lateral scout view of computed tomography showing
parietal cranioplasty with a titanium plate.
dysplasia is en bloc surgical resection and removal of the rim of normal bone is recommended to prevent recurrence [3,10].

Our patient involvement of both the inner and outer tables of the parietal bone, resulting in compression of the underlying structures and probably irritation of the meninges relating to the motor cortex. This may have contributed to the seizure disorder noted in our patient. Despite extradural confinement of the mass, the visual evoked potentials were sensitive enough to allow correct localization of the lesion, but there were no abnormalities of the cerebral cortex or arachnoid mater discovered during surgery. Consequently, a biopsy in the motor cortex area was deemed unsafe for the patient. The cause of the histopathological changes in the cortex remained unclear, as a seizure involves uncontrolled, abnormal electrical activity between brain cells. Antiseizure medications become necessary in any patient like ours who develops seizures, and such patients should be monitored until the seizures subside and the medications can be safely discontinued.

**Figure 4.** Pathological findings in specimens obtained through craniectomy for the bone lesion. (A) Hematoxylin & eosin, ×40. Histology of fibrous dysplasia: Osteoid trabeculae that are narrow, curvilinear, or irregularly-shaped are arranged haphazardly on a background of dense fibroblastic stroma. (B, C) Hematoxylin & eosin, ×100. Irregular trabeculae of bone with no conspicuous osteoblastic rimming in a fibrous background. (D) Hematoxylin & eosin, ×400. Intervening fibrous stroma containing cytologically bland spindle cells, without cytologic atypia.
Table 1. Previously published cases of fibrous dysplasia associated with epilepsy.

<table>
<thead>
<tr>
<th>Author</th>
<th>Public year</th>
<th>Cases</th>
<th>Age</th>
<th>Sex</th>
<th>Type of seizures</th>
<th>Electroencephalogram</th>
<th>Imaging</th>
<th>Treatment</th>
<th>Histological confirmation</th>
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<tr>
<td>Sassin et al [9]</td>
<td>1968</td>
<td>6</td>
<td>No data</td>
<td>No data</td>
<td>Nonfocal convulsive seizures</td>
<td>No data</td>
<td>No data</td>
<td>3 Surgery 3 Nonsurgical</td>
<td>No data</td>
</tr>
<tr>
<td>Bertoni et al [5]</td>
<td>1978</td>
<td>1</td>
<td>17</td>
<td>F</td>
<td>Major motor seizures</td>
<td>Yes</td>
<td>Skull radiography, radionuclide brain scan</td>
<td>Lesionectomy, cranioplasty</td>
<td>Yes</td>
</tr>
<tr>
<td>Kanda et al [6]</td>
<td>2002</td>
<td>1</td>
<td>28</td>
<td>M</td>
<td>Convulsions</td>
<td>Yes</td>
<td>Skull radiography, computed tomography, magnetic resonance image, bone scan</td>
<td>Cranioplasty</td>
<td>Yes</td>
</tr>
<tr>
<td>Meyer [7]</td>
<td>2014</td>
<td>1</td>
<td>22</td>
<td>F</td>
<td>Generalized seizure</td>
<td>Yes</td>
<td>Computed tomography, magnetic resonance image, bone scan, Positron emission tomography</td>
<td>Nonsurgical</td>
<td>No</td>
</tr>
</tbody>
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Cranioplasty with a titanium plate yields satisfactory cosmetic results, as in our patient.

Conclusions

Neurological symptoms associated with calvarial fibrous dysplasia are rare, and focal seizures are rarely the first sign. This report emphasizes the importance of early diagnosis of fibrous dysplasia of bone to exclude primary bone malignancy or bone metastasis, ensuring prompt management and symptom control.

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