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Case Report

CASE REPORT

Fibrous dysplasia of occipital and temporal bone. A case report

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Abstract

Fibrous dysplasia is a rare non-malignant condition where fibrous tissue replaces the normal bone architecture. Involvement of temporal and occipital bones is exceptionally rare and is associated with unique complications. A 10-year-old boy presented with right retroauricular enlargement and pain. Imaging studies and biopsy revealed fibrous dysplasia of the temporal and occipital bones. There was no hearing loss or sequelae arising from posterior fossa compression. The patient was discharged with follow-up instructions. Only 10 cases of occipital bone fibrous dysplasia have been reported in the medical literature. Occipital bone fibrous dysplasia can be complicated with Chiari malformation and syringomyelia while temporal bone involvement is associated with hearing loss. These potential developments require close follow-up that includes detailed neurologic examination, imaging and audiology.

INTRODUCTION

Fibrous dysplasia is a non-neoplastic condition affecting mostly children and adolescents where normal bone tissue is substituted by fibrous connective tissue. Fibrous dysplasia can affect a single bone (monostotic), multiple bones (polyostotic) or exhibit extaosseous manifestations with either café au lait spots and endocrinologic disturbances (McCune–Albright syndrome [1]) or soft tissue myxomas (Mazabraud syndrome [2]).

Fibrous dysplasia is most commonly diagnosed on imaging studies where it usually shows as ground glass, with alternating dense sclerotic and radiolucent fibrotic areas (pagetoid fibrous dysplasia). Alternate but less usual radiographic presentations are sclerotic fibrous dysplasia, with dense homogenous areas, and cystic fibrous dysplasia, with radiolucent areas contained within dense bone [3]. Tissue biopsy can confirm the diagnosis [4].

Fibrous dysplasia in itself is a rare condition affecting about one in 30 000 people [5] and monostotic fibrous dysplasia involving temporal and occipital calvarium is even rarer [6]. In this report we describe such a case of fibrous dysplasia affecting the temporal and occipital bones.

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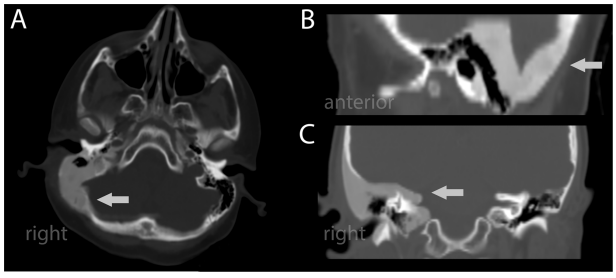

Figure 1: CT scan showcasing the typical ground glass appearance of fibrous dysplasia in the right temporal and occipital bones. White arrows point at the lesion. a. Transverse plane. b. Sagittal plane. c. Coronal plane.

Case report

A 10-year-old boy presented to a primary care physician complaining for painful enlargement in the right retroauricular region in the vicinity of the right mastoid process for the last 3 months. An ultrasound was performed, which indicated asymmetry of the retroauricular osseous anatomy. As the enlargement persisted, the patient visited the outpatient clinic of our ENT department. A head CT was arranged, which revealed considerable thickening of the right temporal and occipital bones with replacement of the normal bone by ground glass looking material, findings highly suggestive of fibrous dysplasia (Fig. 1). We noted no hydrocephalus or hindbrain herniation, which could have developed from the compression of the posterior fossa. Subsequently a tissue biopsy was performed. The sample exhibited curvilinear and disorganized trabeculae embedded in a bland fibrous stroma thereby confirming the diagnosis of fibrous dysplasia. Bone scintigraphy revealed a singular site of increased radioisotope accumulation, and therefore established the diagnosis of the monoostotic form of the disorder. Full endocrine workup showed no endocrine disruption and audiology testing did not document any hearing loss. The patient was discharged with instructions for his follow-up.

DISCUSSION

Fibrous dysplasia of the occipital bone is exceptionally rare, as only 10 cases have been reported in medical literature. Some unique complications can arise from this rare localization. Chandy *et al.* [7] report a case of a 29-year-old man who was observed for fibrous dysplasia of the occipital bone and developed hand clumsiness and lower limb stiffness. Follow-up imaging revealed that the osseous deformity had started compressing the posterior fossa, causing hindbrain herniation and subsequently syringomyelia. This complication affected the Iseri *et al.* [8] patient, a 35-year-old woman with fibrous dysplasia of the occipital, temporal bones and skull base, accompanied with aneurysmal bone cyst. This patient too exhibited symptomatic hindbrain hernation and syringomyelia. Her symptoms consisted of sensory loss in the C4 to C8 dermotomes, hand wasting and lower limb pyramidal signs. Moreover, hearing loss, hoarseness and gag reflex attenuation were emerging, as the deformity extended to the temporal bone and the skull base. Chiari malformation appeared also in the Urgun *et al.* [9] patient, a 14-year-old girl with polyostotic fibrous dysplasia, McCune–Albright syndrome and bilateral occipital aneurysmal bone cysts. The Pan *et al.* [10] study clearly documents the propensity of the patients with craniofacial fibrous dysplasia toward basilar invagination and Chiari I malformation. Their detailed craniomorphometric evaluation suggests that Chiari I malformation stems from restriction of the posterior fossa volume.

In our case the patient presented with minimal symptoms: local enlargement and slight tenderness. So did the 19-year-old patient of Itshayek *et al.* [11], who had concomitant aneurysmal bone cyst, the 15-year-old girl of Basaran *et al.* [12] and the single patient with occipital bone involvement of the Gupta *et al.* case series [13]. It is noteworthy that the Chandy *et al.* [7] patient initially presented with no symptomatology and neurologic disturbances appeared during the follow-up period. Three more cases have been described in non-English language journals [14–16]. Various common or less common pathologies can also present as a local mass. This includes ectodermal inclusion cysts, Langerhans cell histiocytosis, multiple myeloma, osteomas, osteosarcomas or angiosarcomas, osseous hemangiomas, meningiomas and [17]. Calvarial osseous thickening is also seen in osteitis deformans, osteosclerotic conditions (e.g. osteopetrosis), acromegaly, hyperparathyroidism, anemias or phenytoin use [18, 19].

In our patient fibrous dysplasia involved the temporal bone too. Temporal bone involvement isn't common either; Mierzwinski *et al.* [3] review 66 cases that have been reported in the literature while the Boyce *et al.* [20] longitudinal study adds 116 more fibrous dysplasia/McCune-Albright Syndrome patients with temporal bone involvement. While our patient did not exhibit any hearing deficit, fibrous dysplasia of the temporal bone is associated with hearing loss, most often conductive, primarily because of crowding of the ossicular chain and less frequently due to external auditory canal stenosis [20]. Temporal bone fibrous dysplasia can also cause cholosteatoma, tinnitus, vertigo, sensorineural hearing loss and facial nerve palsy [3]. As hearing loss is a quite common event in these patients, close audiology follow-up is prudent.

Workup of these patients should also include bone scintigraphy, to evaluate the possibility of polyostotic involvement or soft tissue myxomas, a manifestation of Mazabraud syndrome [2]. Full endocrinologic workup is also needed, as the most severe form of fibrous dysplasia, the McCune–Albright syndrome, is linked with multiple endocrinologic disturbances involving pituitary, thyroid, adrenal glands and phosphate levels dysregulation [1].

Management of fibrous dysplasia can be conservative with observation and patient education. Bisphosphonates have been employed as well, especially for patients with polyostotic disease [21]. The decision for surgical intervention is dependent on the location, size and the biological effects of the lesion [22]. Hearing loss, cranial nerve impingement and posterior fossa mass effect can be reasons to consider surgical intervention.

In conclusion, fibrous dysplasia extending into the occipital bone is a very rare condition, which may present unique challenges due to posterior fossa compression. Therefore, even asymptomatic patients who are handled conservatively ought to be closely followed up with regular imaging and careful neurologic examination to observe for the development of Chiari malformation and syringomyelia. These sequelae may necessitate surgical posterior fossa decompression.

CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interest.

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ETHICAL APPROVAL

No ethical approval is required.

CONSENT

Patient consent form has been obtained.

GUARANTOR

Eleftherios Neromyliotis is the guarantor of the integrity of this manuscript.

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