# Monostotic Fibrous Dysplasia of the Temporal Bone: A Case Report

## Temporal Kemiğin Monostotic Fibröz Displazisi: Olgu Sunumu

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**Abstract:** Fibrous dysplasia is a congenital, nonfamilial, metabolic disturbance that produces 2.5% of all osseous tumors, and more than 7% of all nonmalignant tumors in bone. Involvement of the temporal bone, however, is relatively rare. An 8-year-old girl presented with progressive hearing loss. She had a mass in the left external acoustic meatus and hearing loss in her left ear. Radiological studies revealed a temporal mass. The patient underwent two surgeries in 2 months, and the mass was totally excised. A postoperative neurologic examination revealed left facial paralysis. Histological study identified the mass as fibrous dysplasia of bone. After 5 years of follow-up, there were no signs of residual tumor or recurrence of the disease. When indicated, total excision with extensive reconstruction is the treatment of choice for this type of bone neoplasm.

**Key Words:** Fibrous dysplasia; temporal bone; tumor; acoustic meatus

**Özet:** Fibröz displazi, kemiğin bütün benign tümörlerinin %7'sinden fazlasında ve bütün kemik tümörlerinin %2.5'inde görülen, ailevi olmayan, konjenital ve metabolik bir hastalıktır. Temporal kemiğin tutulumu oldukça nadirdir. 8 yaşında kız çocuğu, ilerleyici işitme kaybı ile başvurdu. Sol kulakta işitme kaybı ve eksternal akustik kanalda kitle tespit edildi. Radyolojik incelemeler temporal kitle gösterdi. Hasta 2 ay içinde iki kez opere edilerek kitle total olarak çıkarıldı. Postoperatif muayenede solda fasiyal paralizisi vardı. Histopatolojik tanı fibröz displazi olarak alındı. 5 yıllık takipte rezidü veya rekürrens tespit edilmedi. Fibröz displazi gibi kemik tümörlerinde, endike olduğu zaman, geniş rekonstrüksiyon ile birlikte total rezeksiyon seçilecek tedavidir.

**Anahtar kelimeler:** Fibröz displazi; temporal kemik; tümör; akustik kanal

### INTRODUCTION

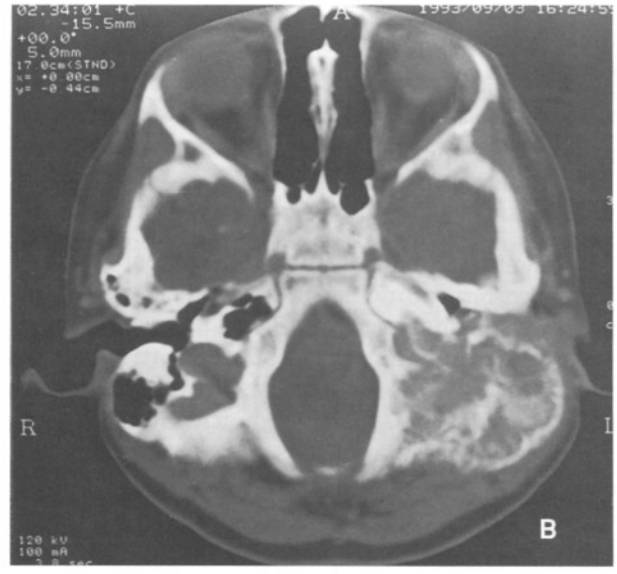
In 1938, Liechtenstein coined the term "fibrous dysplasia" to describe a condition characterized by the progressive replacement of normal bone elements with fibrous tissue (9). Fibrous dysplasia is a congenital, nonfamilial, metabolic disorder that produces 2.5% of all osseous tumors, and more than 7% of all nonmalignant bone tumors (4,5,10,15). The average age at onset is 10 years (5). Sometimes this neoplastic process is associated with abnormal skin pigmentation or endocrine abnormalities. The lesion

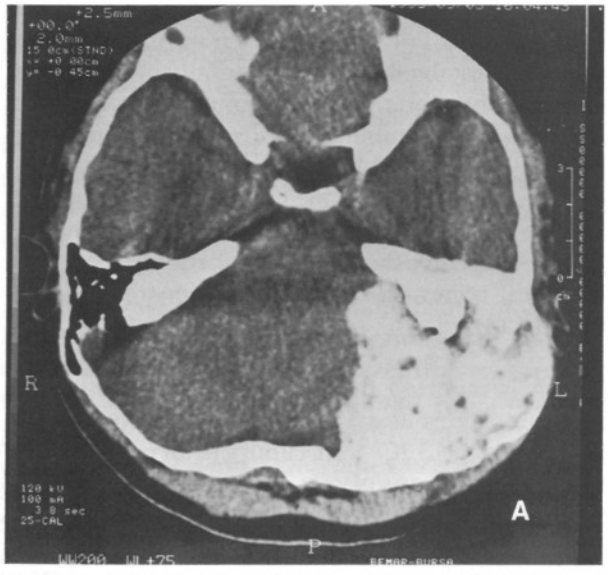
may involve one skeletal bone (monostotic) or several bones concurrently (polyostotic). The monostotic form is most common (70%) (12), with the polyostotic type accounting for 30% of fibrous dysplasia cases (11). Involvement of temporal bone is relatively rare (1). We report the case of a pediatric female patient who had fibrous dysplasia of the temporal bone, and review the relevant literature on this form of neoplasia.

## CASE REPORT

An 8-year-old girl was admitted to our department with a history of recurrent external otitis and the complaint of gradual hearing loss in the left ear. Her physical examination revealed a mass that was narrowing the external acoustic meatus of the left ear such that the tympanic membrane could not be visualized. No abnormal skin pigmentation was detected on the patient's body.

Audiologic testing confirmed that the hearing loss was of the mixed conductive type. Laboratory results indicated there were no biochemical abnormalities. Plain x-rays of the skull, including oblique views, showed a well-circumscribed protrusion of the squamous portion of the temporal bone, with curvilinear calcification. Erosion of the occipital bone was also evident. A computerized tomography (CT) scan of the region of the left temporal bone showed stenosis of the external acoustic meatus, and narrowing of the tympanic cavity and the internal ear canal. CT also revealed that the mass had invaded the bone to the level of the middle fossa and the prepontine angle (Figure 1a,b,c). Digital subtraction angiography showed that the tumor's vascular supply was the external carotid artery and its posterior auricular, middle meningeal and occipital branches. Magnetic resonance imaging (MRI) showed a lesion with low and high signal intensity on T1-weighted images, and low signal intensity on T2-weighted images. Intravenous gadolinium injection revealed a markedly enhanced extraaxial mass (Figure 2). We were not able to assess the patient with radionuclide scintigraphy.



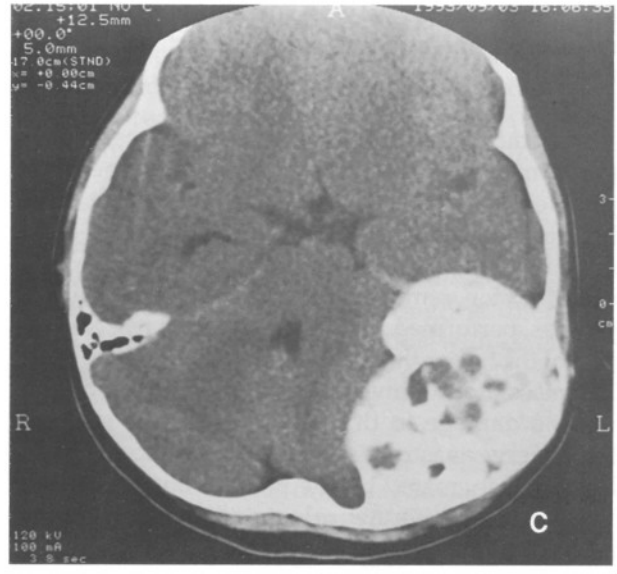




Figure 1: The CT appearance of the lesion with and without contrast demonstrates partial blockage of the external acoustic meatus and narrowing of the tympanic cavity and internal ear canal (a,b), and tumor invasion to the level of the middle fossa and prepontine angle (c).

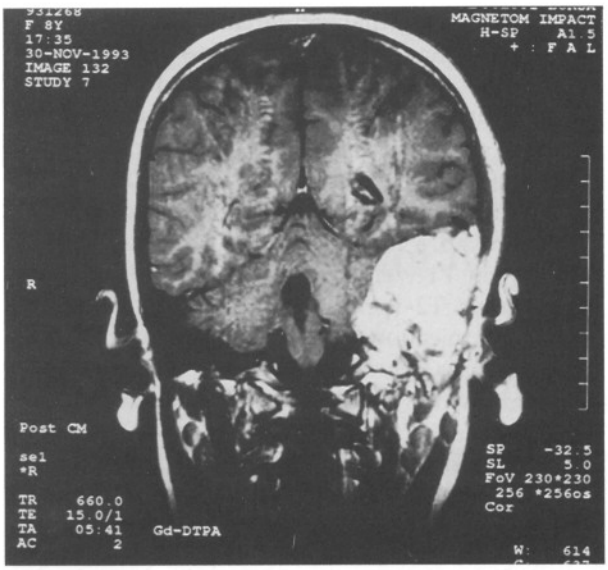



Figure 2: The appearance of the lesion on contrast-enhanced coronal MRI.

The first step in treatment was embolization of the left posterior auricular and occipital arteries using polyvinyl alcohol injection. Once this was complete, we used a left petroclival surgical approach to remove the tumor. Grossly, the lesion was a subcutaneous mass of dense sclerotic material composed of vascular fibrous substance and bone. It extended to the pyramis of the temporal bone. We did a partial mastoidectomy, and observed that the incus and stapes had been destroyed by the invasive disease in the narrowed middle ear. We also noted that the facial nerve was surrounded by fibrous dysplastic bone, and we carefully resected this tumor tissue.

In this initial surgery, only subtotal resection of the tumor was possible due to intraoperative problems with massive hemorrhage and hypotension. However, 2 months later, we did a second operation. In this procedure, we used the same incision, removed the remainder of the mastoid process, performed a petrosectomy and resected the part of the tumor that had invaded the occipital bone. The mass did not involve the dura mater, and there was no damage to this structure during surgery. Facial nerve anastomosis was not possible due to the large defect created by the petrous bone resection. The postoperative neurologic exam revealed left facial paralysis.

Histological examination confirmed that the lesion was fibrous dysplasia of bone. The mass was composed of moderately cellular fibrous stroma with uniform, benign-looking spindle cells. Throughout the stroma there were foci of irregularly shaped trabeculae of immature woven bone. There was no evidence of recurrence or residual tumor at 5 years postsurgery (Figure 3), but the patient's facial paralysis and left hearing loss remained unchanged.

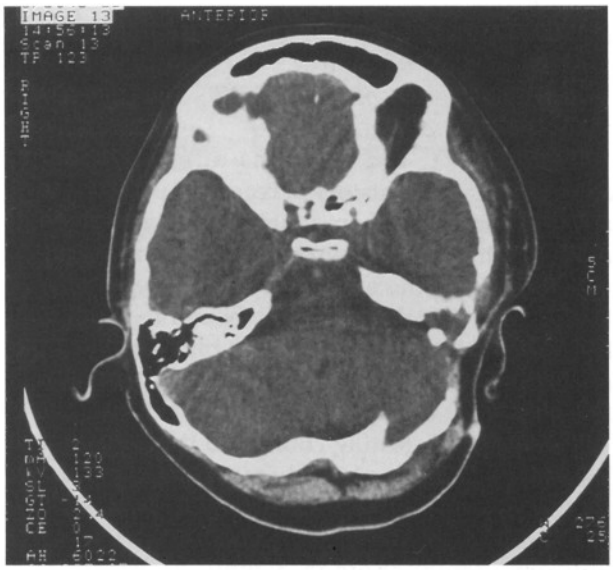



Figure 3: Postoperative CT scan of the site of interest at 5 years after the patient's final surgery.

## DISCUSSION

Fibrous or fibroosseous dysplasia occurs in three forms: a) Monostotic, when only a single bone is affected; b) Polyostotic, when several or many bones are involved, either unilaterally or bilaterally; and c) Polyostotic with endocrine abnormalities, including any combination of precocious puberty, cafe-au-lait spots (McCunne-Albright syndrome), goiter, hyperthyroidism, Cushing's disease and acromegaly (20). In the skull, fibrous dysplasia is known to involve the ethmoid, sphenoid, frontal and temporal bones, in decreasing order, respectively (5,11,15,18).

It is reported that only 10% of patients with monostotic fibrous dysplasia have craniofacial bone involvement (13,20). Van Tilburg et al. reviewed 144 cases of skull involvement in monostotic and polyostotic disease. They found that the frontal and sphenoid bones were most often affected, and that the temporal bone was affected in only 18% of cases (19). The prevalence of temporal bone involvement of fibrous dysplasia in males is double the figure

noted in females (7). The disease most often appears in late childhood or early adolescence, and is rarely seen in adults (3,10,13). Our patient was 8 years old, and presented with hearing loss, a history of recurrent external otitis and a mass that was almost totally obstructing the external acoustic meatus. The most common sign of fibrous dysplasia at this site is progressive protrusion of the mastoid or squamous portion of the temporal bone, occasionally with a preauricular protrusion that may interfere with temporomandibular joint mobility. The lesion may also result in cranial nerve (CN) palsy, with CN VII most commonly involved (11,20).

Regarding diagnosis, the three major groups in the radiographic classification of fibrous dysplasia are pagetoid, sclerotic and cystic lesions (1,6,11). The radiographic features of the disease vary depending on the stage of development and amount of bony matrix within the lesion. CT evaluation is useful for documenting the extent of bone and extraosseous involvement (1,4,10,11,14,15). We confirmed our patient's diagnosis on CT. Radionuclide scintigraphic evaluation is another important tool for detecting fibrous dysplasia. This type of scan is diagnostic when over 50% of the calcified bone at the diseased site is replaced by fibrous tissue. Radionuclide studies are more specific than CT for delineating the intra- and extraosseous extent of the lesion, and for demonstrating polyostotic or disseminated disease (8,10). Unfortunately, we were unable to perform a radionuclide examination on our patient.

The differential diagnosis for fibrous dysplasia includes giant cell granuloma, ameloblastic fibroma, osteoma, odontogenic cyst, hyperparathyroidism, juvenile Paget's disease, chronic hyperphosphatasemia tarda, the Hunter-Hurler syndrome of gargoylism, cherubism, neurofibromatosis and tuberous sclerosis (4,5,10,11,13).

Surgical management is not always indicated in these cases. Small solitary lesions will usually remain static and asymptomatic. However, surgical excision and curettage are required when marked progressive bone deformity, cranial nerve compromise or pain syndromes are manifested. Unless neurological compromise becomes evident, it is recommended that surgical intervention be delayed until adolescence or until growth is completed (10). Our case called for immediate surgery because the mass was compressing the patient's brainstem and the bulb of the jugular vein. We were able to totally excise the tumor through a petroclival approach.

Serious intraoperative vascular complications developed in both surgeries that were done in this case. The patient's preoperative angiographic work-up indicated that the mass was receiving arterial supply from branches of the external carotid and the posterior circulation; thus, our first step was to embolize the feeding vessels. All signs indicated that this procedure had been successful, but severe hypotension developed in the first surgery due to massive bleeding through blood transfusion. Faced with this urgent situation, we were only able to resect part of the tumor. We completed the excision in the second operation, but, although no embolization was performed in that session, we encountered similar hemorrhage once again.

In terms of outcome, fibrous dysplasia is a benign condition that has a good prognosis. The disease process is usually halted at puberty; however, Ramsey et al. and Harris and colleagues have reported cases where progression has continued (20). Hormone treatment is reportedly ineffective, and adjuvant irradiation is contraindicated given the high potential for malignant transformation (7,13,16,17). Chen and Fairholm reported 11 cases of malignant transformation in 13 patients who received this form of therapy (2). In patients with monostotic disease, the prevalence of malignant degeneration is highest in lesions that affect the craniofacial region (10). Overall, this translates into a 0.5% risk of malignant transformation for fibrous dysplasia lesions that are left untreated. One study has indicated that the mean interval between diagnosis of the condition and the development of malignancy is 13.5 years (5).

Finally, decision-making can be difficult in certain situations, and the finding of an aggressive form of fibrous dysplasia in a preadolescent patient presents the surgeon with the toughest dilemma. When indicated, complete resection with extensive reconstruction is the treatment of choice.

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