Temporal bone fibrous dysplasia and cholesteatoma leading to the development of a parapharyngeal abscess

MICHELLE R. YAGODA, M.D.*, SAMUEL H. SELESNICK, M.D. (New York)

Abstract

Monostotic fibrous dysplasia of the temporal bone is a rare disease entity that may lead to progressive stenosis of the external auditory canal with resultant trapping of skin and the development of cholesteatoma. The cholesteatoma may extend widely into the temporal bone. Once cholesteatoma occupies the petrous apex, erosion out of the temporal bone can occur superiorly into the middle fossa, posteriorly into the posterior fossa, inferiorly into the neck, and infero-medially into the parapharyngeal space. We present a case in which minimal symptoms were present despite a massive temporal bone fibrous dysplasia and cholesteatoma. Eventually, cholesteatoma eroded into the parapharyngeal space, leading to airway compromise. Late presentation occurred because the otic capsule was spared, there was no external skull deformity and there was slow inferior and medial growth that was well tolerated, until the development of a parapharyngeal space abscess.

Key words: Fibrous dysplasia of bone; Temporal bone; Cholesteatoma; Abscess, parapharyngeal

Introduction

Von Recklinghausen first described the characteristic bony lesion that is now known as fibrous dysplasia (FD), in 1891. It was not until 1937, that McCune and Bruch described the clinical entity as distinct from other abnormalities of bone formation. The term 'fibrous dysplasia' was introduced by Lichtenstein in 1938. It was classified into three subgroups: (1) monostotic fibrous dysplasia (MFD) – involvement of a single bone (70 per cent); (2) polyostotic fibrous dysplasia – involvement of multiple bones (30 per cent); and (3) McCune–Albright syndrome – bony involvement associated with skin lesions and endocrinopathies (3 per cent) (Lichtenstein, 1938).

MFD comprises seven per cent of all bone tumours, and is the most frequent benign lesion affecting the skull. It usually presents in the first two decades of life. Although only 10 per cent of MFD involves craniofacial structures, it is the most prevalent form of fibrous dysplasia (FD) in the head and neck (Nager *et al.*, 1982). The maxilla and mandible are the most frequently affected cranio-maxillo-facial bones. The temporal bone is affected in only 18 per cent of all cases of FD, occurring more frequently in association with monostotic than polyostotic forms (Younis and Haleem, 1987). There have not been any reported cases of involvement of the temporal bone in McCune–Albright syndrome (Nager *et al.*, 1982).

Pathophysiology

While most agree that MFD represents a congenital anomaly of the bone-forming mesenchyme, its mode of transmission (autosomal-dominant or recessive) is still the topic of debate (Lichtenstein and Jaffe, 1942; El Deeb *et al.*, 1979). Other theories of the pathogenesis of MFD are less widely accepted. Schlumberger described FD as a disturbance in the reparative process (Schlumberger, 1947). Others have suggested that MFD occurs secondary to an arrest of bone maturation.

Pathology

Macroscopic examination of FD reveals expanded cancellous bone within a thinned cortex. The marrow and trabeculae form a soft, gritty region which may be white or red depending on the vascularity of the lesion (Lichtenstein and Jaffe, 1942). While there is no definite capsule, there is a sharp transition to normal bone. Bony suture lines are respected so that FD does not extend to articulated surfaces of neighbouring bones. There is no invasion of the periosteum (Lichtenstein and Jaffe, 1942; Batsakis, 1979).

Microscopic evaluation displays a stroma of collagen matrix with fibroblasts in a whorled pattern and trabecular of bone in 'jigsaw puzzle' or 'Chinese letter' configuration. FD displays a woven rather than lamellar pattern of bone formation which allows it to be easily differentiated from ossifying fibroma (Schlumberger, 1947; Zappia *et al.*, 1990).

Clinical presentation

MFD is a slow-growing and benign process, which often becomes quiescent after puberty. The most common otological symptom of MFD of the temporal bone is a progressive conductive hearing loss secondary to the occlusion of the eustachian tube or external auditory canal (EAC) or by direct impingement of ossicular motion (Lambert and Brackman, 1984).

Exuberant fibrous dysplasia growth in the region of the external auditory meatus leads to progressive stenosis with resultant trapping of keratin debris (Smouha *et al.*, 1987). Full erosion of the ossicles may occur secondary to entrapped cholesteatoma (Basek, 1967). Once cholesteatoma occupies the petrous apex, erosion from the confines of the temporal bone may occur in any of four directions: superiorly into the middle cranial fossa; posteriorly into the posterior cranial fossa; inferiorly into the neck; and, inferomedially into the parapharyngeal space. With pro-

FIG. 1

CT of the temporal bone: coronal image. Otic capsule spared; fibrous dysplasia in superior portion of temporal bone, while cholesteatoma extends inferiorly and medially.

gressive spread of disease in the temporal bone, there may be cranial nerve or central nervous system involvement (Zappia et al., 1990).

External FD growth may lead to skull asymmetry and cosmetic deformity. Displacement of the temporo-mandibular joint (TMJ) by FD may be manifest as dental malocclusion or TMJ dysfunction.

Radiographical findings

The diagnosis of FD cannot be made unequivocally on radiographical findings. Generally, FD is characterized by a homogeneous radiodensity within an expanded cortex; however, radiographical features vary with the degree of fibrosis in the disease process (Bergeron et al., 1984). Three presentations described by Fries (Fries, 1957) are noted on radiography: (1) Pagetoid (56 per cent) – alternating areas of density and radiolucency; (2) sclerotic (23 per cent) – homogeneously dense; and (3) cyst-like (21 per cent) – spherical/ovoid lucency with dense boundary. It involved temporal bones, the otic capsule is usually spared (Batsakis, 1979; Nager et al., 1982; Younis and Haleen, 1987).

Differential diagnosis

The differential diagnosis of FD includes both benign and malignant lesions. Benign lesions include solitary unicameral cyst, nonossifying fibroma, eosinophilic granuloma, cholesteatoma, meningioma, Paget's disease, osteochondroma, ossifying fibroma, giant cell reparative granuloma, exostoses, aneurysmal bone cyst, osteitis fibrosa cystica, adamantinoma, and plasma cell myeloma. Malignant lesions include sarcoma and metastatic osteoblastic lesions.

Treatment

The treatment of FD is conservative surgery with three directives – the restoration of function, the prevention of complications and the restitution of cosmesis.

Radiation therapy is to be avoided (Smouha et al., 1987). The incidence of spontaneous malignant transformation has been reported to be less than 0.5 per cent and has never been described as having occurred in the temporal bone. However, sarcomatous

transformation after radiation therapy can occur. Of the sarcomas encountered in malignant transformation, osteosarcoma is the most common variety, followed by chondrosarcoma (Schwart and Alpert, 1964; Huvos et al., 1972).

Case report

A twenty-two-year-old Asian woman was referred to The New York Hospital–Cornell University Medical Centre with otorrhea and hearing loss. Her recurrent left-sided otorrhea began at the age of six years. She had frequent episodes for a five-year period, one of which required hospitalization in Hong Kong at the age of 11 years. For the next 11 years, the patient remained asymptomatic with the exception of a progressive left-sided hearing loss which she did not investigate. At the age of 22 years, after an 11-year hiatus, the foul otorrhea recurred. She was referred by her local otolaryngologist for neurological evaluation. Her otological, neurological and endocrinological review of systems were otherwise unremarkable.

On physical examination, there was no external deformity of the contour of the patient's head, skin lesions nor stigmata of endocrine dysfunction. Otoscopic evaluation revealed a stenotic left external auditory canal. Medial to the stenosis was a massive cholesteatoma with obliteration of middle ear landmarks. Cranial nerve function was normal except that Weber and Rinne tuning fork evaluations suggested a left-sided conductive hearing loss. Cerebellar function was judged to be intact. The remainder of the head and neck evaluation was unremarkable.

Serum chemistries were all within normal limits. Pure tone audiometry revealed a maximal left-sided conductive hearing loss with an air-line at 70 dB. Hearing in the right ear was within normal limits.

Both CT and MRI with gadolinium were obtained. Massive FD of the entire left temporal bone with sparing of the otic capsule structures was noted (Figures 1 and 2). There was severe compression of the left cerebellum, but no evidence of hydrocephalus. The carotid artery was displaced anteriorly, and the jugular–sigmoid system was partially thrombosed. The area of fibrous dysplasia was filled with a large cholesteatoma that eroded the expanded petrous apex, and extended inferiorly and medially to the parapharyngeal space. The patient refused surgical intervention at that time.
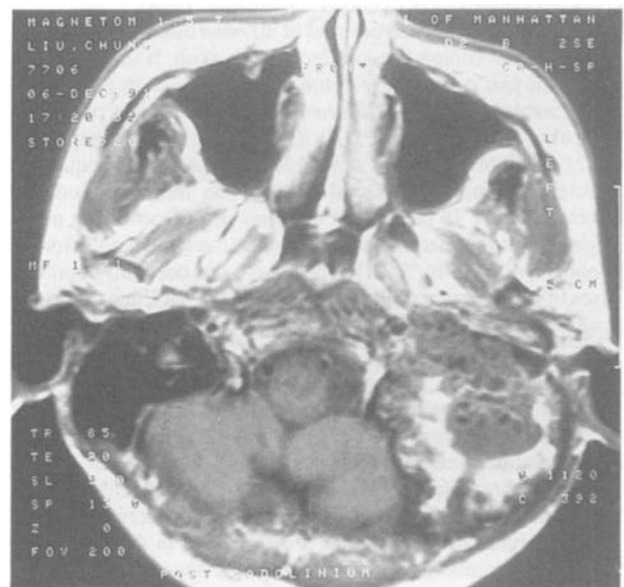

FIG. 2

MRI of the temporal bone with gadolinium, T_1 weighted image. Nonenhancing cholesteatoma that extends medially into the parapharyngeal space. There is anterior displacement of the carotid artery and compression of posterior fossa structures.

FIG. 3

CT of temporal bone: axial image. Abscess present in parapharyngeal space medial to ascending ramus of the mandible.

Within one month, the patient presented with severe trismus, odynophagia and fever to 38.6°C. On examination, the patient was able to open her mouth to 1 cm interincisor distance, and had a parapharyngeal mass displacing the left tonsil medially. An interval CT of the temporal bones and neck demonstrated a large parapharyngeal space abscess medial to the ascending ramus of the mandible (Figure 3).

The patient was taken to the operating room where a tracheotomy was performed under local anesthesia, and the left ear canal was suctioned to debride the cholesteatoma. With this, the parapharyngeal space abscess was decompressed and 50 ml of purulent debris exuded from the external auditory canal. Examination of the oropharynx revealed resolution of the abscess. A large meatoplasty was performed and a silastic stent was placed into the external auditory canal. The patient's condition rapidly improved and she was discharged on a four-week course of intravenous antibiotics.

The patient was later electively readmitted for subtotal temporal bone resection and petrous apicectomy. The internal carotid artery and the vertical portion of the facial nerve were noted to be dehiscent. Cholesteatoma and fibrous dysplasia were confirmed histologically. Post-operatively, the patient is fully recovered with normal facial nerve function.

Conclusion

We present the first reported case of FD and acquired cholesteatoma leading to the development of a parapharyngeal space abscess that was drained endaurally. Interestingly, minimal symptoms were present initially despite a massive temporal bone lesion. The relatively asymptomatic nature of the disease process was largely due to the sparing of the otic capsule, the lack of external deformity, and the slow inferomedial extension of cholesteatoma that was well tolerated until a parapharyngeal space abscess developed.

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Address for correspondence:
Samuel H. Selesnick, M.D.,
Department of Otolaryngology,
Cornell University Medical Centre,
525 East 68th Street,
Starr 541,
New York,
New York 10021,
USA.