Fibrous dysplasia of the temporal bone presenting as a parotid abscess

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Fibrous dysplasia of the temporal bone (FDTB) is a benign, localised bone disease in which the temporal bone is progressively replaced with fibrous tissue and woven bone. This results in deformity and functional impairment.[1] A parotid abscess as the first presenting sign of FDTB is incredibly rare, and this report presents such a case with an associated literature review of the clinical presentation, histological and radiographic features and management. In lieu of prospective trials to assess treatment outcomes, we hope the evidence presented will increase clinician awareness in identifying FDTB in patients with atypical symptoms and inform surgeons on its management.

Case report

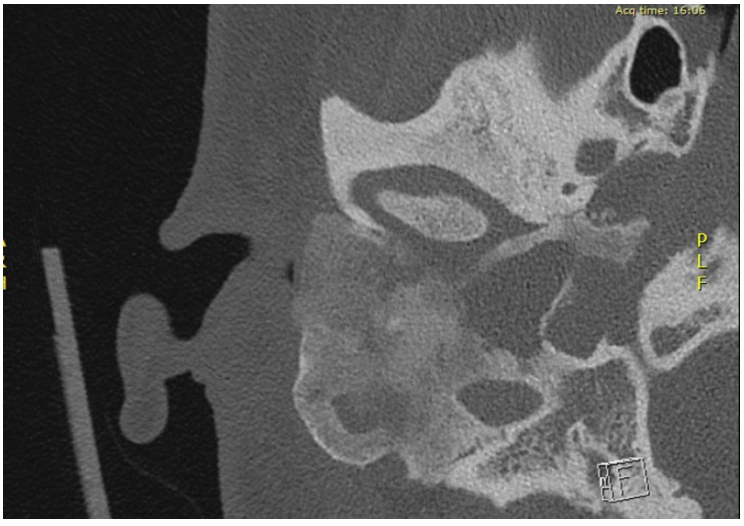
A 22-year-old Samoan male presented with a 3-day history of right-sided tender neck swelling and trismus. He was morbidly obese (body mass index 40.5) and reported years of intermittent ear discharge and right-sided hearing loss but had never consulted a general practitioner (GP). His medical history was unremarkable.

Examination revealed an enlarged, erythematous and tender right parotid gland (8x10cm) and trismus limiting mouth opening to 3cm. Saliva was expressed without pus, dentition was normal and cranial nerves were intact. Flexible nasoendoscopy showed medial displacement of the right tonsil but a normal nasopharynx and epiglottis. The vocal cords had preserved function. Otomicroscopy revealed right external auditory canal (EAC) occlusion by a firm, skin-covered mass. Severe right-sided conductive hearing loss was confirmed on audiometry. Inflammatory markers were elevated.

Computed tomography (CT) illustrated an oedematous right parotid gland with a 25mm–deep lobe fluid collection, cervical lymphadenopathy and temporal bone changes consistent with fibrous dysplasia. These included mastoid air cell obliteration, a ground-glass appearance, anterior cortical destruction and lateral EAC stenosis with no facial nerve involvement. Magnetic resonance imaging (MRI) was contraindicated due to the patient's weight.

Ultrasound-guided aspiration drained 11ml of pus. The patient underwent right subtotal petrosectomy with blind sac closure, removing cholesteatoma extending to the infratemporal

Figure 1: Computed tomography scan—coronal image showing dysplastic changes in the mastoid process, with increased bone volume and areas of lucency and ground-glass opacity, indicative of fibrous dysplasia.



fossa and mesotympanum. Histopathology confirmed monostotic fibrous dysplasia causing EAC occlusion, secondary cholesteatoma and a parotid abscess.

At 4 months post-operation, the patient was asymptomatic with a stable blind sac closure.

Discussion

FDTB, first described by Lichtenstein in 1938, has over 100 cases documented in the literature.[2–3] Temporal bone involvement in generalised fibrous dysplasia is uncommon and usually unilateral, making up an estimated 18% of all cases.[4] However, its prevalence can be under-estimated due to asymptomatic cases. FDTB that presents as a parotid abscess is extremely rare; to our knowledge, this is the first time it has been reported in the literature.

Non-inherited, somatic-activating GNAS1 point mutations have been implicated in FDTB aetiology.[5] This disease has a mean age at diagnosis of 16–25 years, which is consistent with our clinical case. It also shows a female predominance of 55–59%.[6–8] Monostotic manifestations are the most common in general fibrous dysplasia, constituting 70% of cases, followed by polyostotic forms (23%) and McCune–Albright syndrome (7%). However, there is no consensus on the most common form in the temporal bone. Our clinical case is likely monostotic because these lesions generally grow slowly and remain quiescent after puberty, whereas polyostotic forms typically manifest in late childhood.[9]

The most common presenting symptom is conductive hearing loss due to EAC stenosis or ossicular chain impingement. Canal occlusion can lead to epithelial incarceration and keratin accumulation, fostering secondary cholesteatoma formation.[9–10] Sensorineural hearing loss and infection also occur due to complications from cholesteatoma, cochlear destruction, internal auditory canal stenosis or vestibular fistulisation. Patients can also present with tinnitus, dizziness, pain, trismus and neurological signs from middle or posterior cranial fossa involvement. The first presentation as a parotid abscess is rare but possible due to the anatomy of the anterior mastoid cortex. The fibrodysplasia lesion likely expanded within a thinning shell of cortical bone, leading to skeletal destruction and invasion into neighbouring structures.[9] Given that the deep lobe of the parotid lies between the mastoid process and the mandibular ramus, the erosion of the patient's anterior mastoid cortex suggests this was the invasion pathway. The relatively asymptomatic nature of the disease was primarily due to the absence of external deformity and the slow progression, which was tolerated until the parotid abscess developed.

Differential diagnoses encompass both benign and malignant lesions. Benign possibilities include: meningioma, aneurysmal bone cyst, unicameral cyst, fibroma, Paget's disease, osteochondroma, giant cell tumour, eosinophilic granuloma, exostosis and osteoma. Malignant options are sarcoma and metastatic osteoblastic lesions.[9] The diversity in fibrosis, calcification and varied symptom profiles can complicate pre-operative diagnosis. Therefore, imaging is essential.[3] High-resolution CT is the gold standard, and MRI is recommended in cases of suspected extensive disease. CT assesses: 1) the degree of external auditory canal stenosis, 2) the involvement of middle or inner ear structures, 3) the presence of an associated cholesteatoma, 4) the extent of possible facial nerve involvement, and 5) the dysplasia type. Our imaging revealed that the patient exhibited the most common pattern, the pagetoid or "ground glass" pattern, present in 56% of all cases (Figure 1). This pattern appears as a combination of dense and radiolucent areas indicative of fibrosis. The sclerotic pattern, observed in 23% of cases, is characterised by uniformly dense bone. The cystic pattern, seen in 21% of cases, is defined by spherical or ovoid areas of lucency surrounded by a dense boundary.[11]

Histology is not required for diagnosis as the radiological picture is pathologic. However, if histological samples are taken, the macroscopic examination should reveal expanded cancellous bone within a thinned cortex. There will be a sharp transition to normal bone but no definite capsule. It should not extend to articular surfaces of neighbouring bones or invade the periosteum. Microscopic examination should show a collagen matrix stroma with fibroblasts arranged in a whorled pattern and trabecular bone in a "jigsaw puzzle" configuration.[9]

Treatment depends on symptom profile, preventing complications and restitution of function and cosmesis.[1,6] Surgical treatment is recommended for EAC stenosis with conductive hearing loss, recurrent infections, secondary cholesteatoma, craniofacial deformity, pathological fracture, nerve compression and in cases of suspected malignancy.

Radiation therapy is contraindicated due to

a high risk of malignant transformation into sarcomatous disorders such as osteosarcoma and chondrosarcoma.[9] Malignant transformation into these disorders has not yet been reported in the temporal bone.[1,3] Regular patient follow-up is fundamental to management, with worsening pain and local swelling being suspicious clinical findings to monitor.[3]

Herein, we describe a rare presentation of FDTB in a morbidly obese Samoan male. While there is no direct evidence linking obesity or ethnicity to an increased risk of fibrous dysplasia, Pacific peoples in New Zealand experience a higher prevalence of ear disease and hearing loss.[12–13] This disparity is largely attributed to socioeconomic factors, inequitable access to healthcare and structural and systemic racism across various social institutions.[14] In our case, poor access to primary care—exacerbated by barriers such as cost, lack of transport, inflexible work schedules, closed clinic books and challenges in enrolling with a GP—potentially contributed to disease extension into the parotid gland.[15] To address access issues, GPs could adopt more patient-centred strategies, such as extending opening hours and offering same-day appointments.[15–16] Government investment in Pacific-led healthcare services in regions with high Pacific populations, such as Auckland and Porirua, is also essential, as culturally appropriate, holistic care not only enhances health outcomes but also aligns with Pacific families' preference for Pacific providers.[15–16] To support this, funding should enable continuity of care at a single location, while greater funding is required to increase the Pacific healthcare workforce.

Given the growth of the Pacific population in New Zealand and the prioritisation of Pacific health since the first *Pacific Health and Disability Action Plan*, early detection is important.[17] Routine otoscopic examinations during GP visits may aid in identifying infections or masses at an earlier stage.

This case highlights the need for increased clinical vigilance and a lower threshold for imaging in Pacific patients presenting with symptoms such as parotid swelling, trismus, unilateral hearing loss and/or masses in the ear canal. It also underscores the importance of including FDTB in the differential diagnosis of a parotid mass.

COMPETING INTERESTS
Tanja Jelicic is involved in the multi-disciplinary care of microtia/atresia patients at Starship Children's Hospital; has recently joined the Northern Cochlear Implant Programme (NCIP) as an implant surgeon; and is actively involved in the teaching and training of medical students and junior doctors, and also lectures at The University of Auckland.

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URL
https://nzmj.org.nz/journal/vol-138-no-1611/fibrous-dysplasia-of-the-temporal-bone-presenting-as-a-parotid-abscess

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