Facial Palsy and Mastoiditis from Fibrous Dysplasia

This young adult man presented to ENT clinic with a complaint of left facial weakness and persistent left retro-auricular pain. High resolution CT of the mastoids was performed following clinical assessment.

In this case, there is extensive sclerotic bony expansion with a ground-glass appearance involving the left zygoma, sphenoid and petrous temporal bone.

The bony expansion is centred on the medullary bone and has an abrupt zonal transition (Figure 1). The bone involvement encompasses almost complete bony stenosis of the left external auditory meatus down to 1-2mm with consequential fluid in the external auditory canal and middle ears (Figure 2). The bony expansion involves both the tympanic and mastoid segments of the facial canal which are stenosed. The ossicular chain remains intact. The left mastoid air cells are under-pneumatised and completely occupied by fluid.

Figure 1. Axial High Resolution CT of the Mastoids: Diffusely sclerotic, expansile left temporal bone with a ground-glass appearance (**') and sharp transition to normal bone (wide black arrow). This involves the temporal component of the facial nerve canal with narrowing of the mastoid segment of the facial canal (thin black arrow). The mastoid air cells are opacified with fluid (white arrow).

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DISCUSSION

Fibrous dysplasia (FD) is a benign congenital process that typically manifests itself as a localized defect in osteoblastic differentiation and maturation. Normal bone is replaced with haphazard fibrous tissue and immature woven bone.\(^1\)

Fibrous dysplasia is predominantly a condition of children and young adults (those less than 30 years of age). Disease growth usually halts after the third decade of life. FD may be a monostotic or polyostotic in nature and in some cases is part of a syndrome such as McCune-Albright.\(^2\)

The zygomatic maxillary complex is the most commonly reported location for fibrous dysplasia. The temporal bone is a typical site in polyostotic disease in up to 70%, but less often observed in monostotic disease. Disease of the temporal bone most typically results in hearing impairment due to bony stenosis of the external auditory canal. Facial nerve involvement is a less frequent feature, resulting in facial nerve paralysis, due to involvement of the nerve as it exits through the petrous temporal bone.\(^3\) The anatomical location of the facial nerve compression is hard to access and treat surgically.\(^3\)

CT is the imaging investigation of choice giving the most exquisite bony definition.

Typical CT features (as shown in this case) are:
- A diffuse ground-glass appearance to the affected bone
- Homogeneously sclerotic bone
- Well-defined borders between the diseased and unaffected bone (abrupt zone of transition)
- Bony expansion with overlying cortical bone intact

The CT appearances apply equally to the anatomical site involved, however the combination of imaging appearances can be variable presenting a diagnostic dilemma, which may merit a confirmatory bone biopsy.

REFERENCES