Introduction

There are several forms of calcium pyrophosphate dihydrate deposition disease (CPDD); the rarest form is tophaceous pseudogout. Tophaceous pseudogout involves massive crystal deposition and usually involves a solitary joint—most commonly the temporomandibular joint (TMJ).

The objective of this brief discussion is to present a case of an unusual location of a tophaceous calcium pyrophosphate mass—the infratemporal fossa— including a discussion of the presentation, radiology, pathology, and the surgical approach for resection.

Methods & Patient Presentation

The current investigation is a retrospective case review and review of the literature. The review included the patient’s medical record, radiology, and pathology as well as a PubMed Literature Search.

A 57-year-old otherwise healthy male presented with a four year history of slowly increasing left preauricular edema, trismus, and recent onset ipsilateral hearing loss. Preoperative CT scan showed a lobulated, calcified mass in the left infratemporal fossa involving the anterior margin of the sphenoid along with erosion of the zygoma. There was mass effect on the anterior external auditory canal but no obvious involvement of the TMJ (See Fig. 1a-c). The mass was contrast enhancing on post-gadolinium T₁-weighted MRI (See Fig. 2a-c).

The differential diagnosis included chondroblastoma, chondrosarcoma, chondromatosis, osteochondroma, uric acid crystals (gout), synovial chondromatosis, or calcium pyrophosphate deposition disease (CPPD). In order to guide treatment and rule-out a malignant process, an open biopsy of the mass was taken and was consistent with CPPD.

Surgical Approach and Pathology

The patient underwent a preauricular infratemporal fossa approach for removal of the tophaceous mass. The preauricular infratemporal approach is a versatile approach with many permutations that can be tailored to individual pathology. It has been referred to in the literature by various names, including the transzygomatic, preauricular subtemporal, and Type D infratemporal fossa approach.

A pretragal incision was carried up into the temporal hairline and then anteriorly toward the orbit. A temporoparietal scalp flap was elevated followed by a subfascial dissection of the temporalis muscle. The temporalis muscle was elevated off of the squamosal temporal bone with the deep temporal fascia removed off the zygoma. Zygomatic osteotomies were made allowing further inferior retraction of the temporalis muscle and exposure of the infratemporal fossa. A 5x5 cm craniotomy was made and the limits of the dissection were the external auditory canal posteriorly, the temporalis muscle inferiorly, and V₃ anteriorly. The temporomandibular joint was not entered. The entire mass was extirpated (Fig. 3 & 4). Pathology on the mass showed needle and rhomboid-shaped, polarizable birefringent crystals along with large histiocytic-appearing mononuclear cells and multinucleated cells (Fig. 5 & 6).

Summary

Tophaceous pseudogout, or calcium pyrophosphate deposition disease is a rare mass of the infratemporal fossa. It can masquerade as other benign or malignant lesions, necessitating preoperative biopsy to avoid a potentially more invasive and morbid surgical intervention. Surgical extirpation is indicated for symptomatic lesions and for curative purposes. The preauricular infratemporal fossa approach is effective for removal of benign infratemporal fossa masses.

References