Calcium Pyrophosphate Deposition Disease of the Temporomandibular Joint

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Calcium pyrophosphate dihydrate deposition disease (CPDD, tophaceous pseudogout) is a rare crystal arthropathy characterized by calcium pyrophosphate crystal deposition in joint spaces, episodes of synovitis, and radiological features of chondrocalcinosis. We present a case of a 61-year-old woman who presented with left temporomandibular joint (TMJ) pain, difficulty chewing, left facial numbness, left-sided hearing loss, and left TMJ swelling. Imaging of the temporal fossa revealed a large mass emanating from the temporal bone at the TMJ, extending into the greater wing of the sphenoid and involving the mastoid bone and air cells posteriorly. Fine needle aspiration demonstrated polarizable crystals with giant cells. Intraoperatively, the TMJ was completely eroded by the mass. Final pathology was consistent with tophaceous pseudogout. CPDD has rarely been reported involving the skull base. None of the cases originally described by McCarty had TMJ pseudogout. Symptoms are generally pain, swelling, and hearing loss. Management is nearly always surgical with many patients achieving symptomatic relief with resection. CPDD is associated with many medical problems (including renal failure, gout, and hyperparathyroidism), but our patient had none of these risk factors. This case demonstrates that CPDD can involve the skull base and is best treated with skull base surgical techniques.

Case Report

The patient is a 51-year-old right-handed Caucasian woman with a 12-month history of left ear pain that originated from the temporomandibular joint (TMJ), with some associated swelling, pain with chewing, and mild hearing loss. Her physical examination revealed mild hearing loss on the left side. Computed tomography (CT) and magnetic resonance imaging of the temporal fossa revealed a large mass emanating from the temporal bone at the TMJ (►Fig. 1A, 1B), extending into the greater wing of the sphenoid, and involving the mastoid bone and air cells posteriorly. There was erosion of the petrous carotid canal. A CT-guided biopsy was performed which diagnosed the mass as tophaceous pseudogout (tumoral CPDD).
Intraoperatively, the TMJ was completely eroded by the tumor. BrainLAB (Munich, Germany) was used to identify the tumor margins allowing for near gross total resection; a small portion of the lesion which was densely adherent to the internal carotid artery in the petrous canal was left behind. Initial frozen pathology results revealed “collagen with amorphous material and chronic inflammation.” Final pathology demonstrated numerous polarizable, rhabdoid, and rectangular crystals, consistent with tophaceous pseudogout (► Fig. 2A, 2B).

Postoperatively, the patient did develop a cerebrospinal fluid leak and was treated with antibiotics and a lumbar drain. She was discharged 6 days after surgery with no leak. On her follow-up visits, she felt a vast improvement in her symptoms with some initial hearing loss which improved on further visits.

Discussion

CPDD arthropathy was first described in 1962 by McCarty as pseudogout due to similarity of the acute episodes of arthropathy to gout.\(^2,6,7\) Previously work had been done by Zitnan and Sitaj who had described a disorder called “chondrocalcinosis polyarticularis.”\(^2\) Abnormal deposition of pyrophosphate in the joint space combines with calcium to form calcium pyrophosphate dihydrate crystals on collagen fibers; release of these crystals into the joint space results in neutrophil and monocyte-macrophage phagocytosis and release of inflammatory mediators, causing joint destruction.\(^2,8\) This becomes evident on radiological examinations as peri- and intra-articular calcifications known as chondrocalcinosis.\(^9\)

CPDD arthropathy is often associated with other medical conditions, including hyperparathyroidism, hemochromatosis,
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References


Conclusion

CPDD can be a rare cause of common symptoms and a high threshold of suspicion is necessary to arrive at the correct etiology. As CPDD often presents in many chronic medical illnesses, neurosurgeons may be the first to diagnose these medical problems by accurately diagnosing a skull base mass as CPDD.