Calcium Pyrophosphate Deposition Disease of the Temporomandibular Joint*

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Abstract

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 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.

Keywords
- temporomandibular joint
- calcium pyrophosphate
- CPDD
- chondrocalcinosis
- tophaceous pseudogout

Calcium pyrophosphate dihydrate deposition disease (CPDD) is a rare crystal arthropathy characterized by the deposition of calcium pyrophosphate crystals in joint spaces, episodes of synovitis, and radiological features of chondrocalcinosis.[1,2] While the disease is most prevalent in the shoulders, pelvis, knees, and joints of the hands, the axial skeleton can be affected, but rarely involves the skull base.[2–4] When involved, CPDD can cause symptoms mimicking more common neurological and neurosurgical pathologies.[2,5] We present a patient who was recently treated at our institution whose pathology demonstrated CPDD.

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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).

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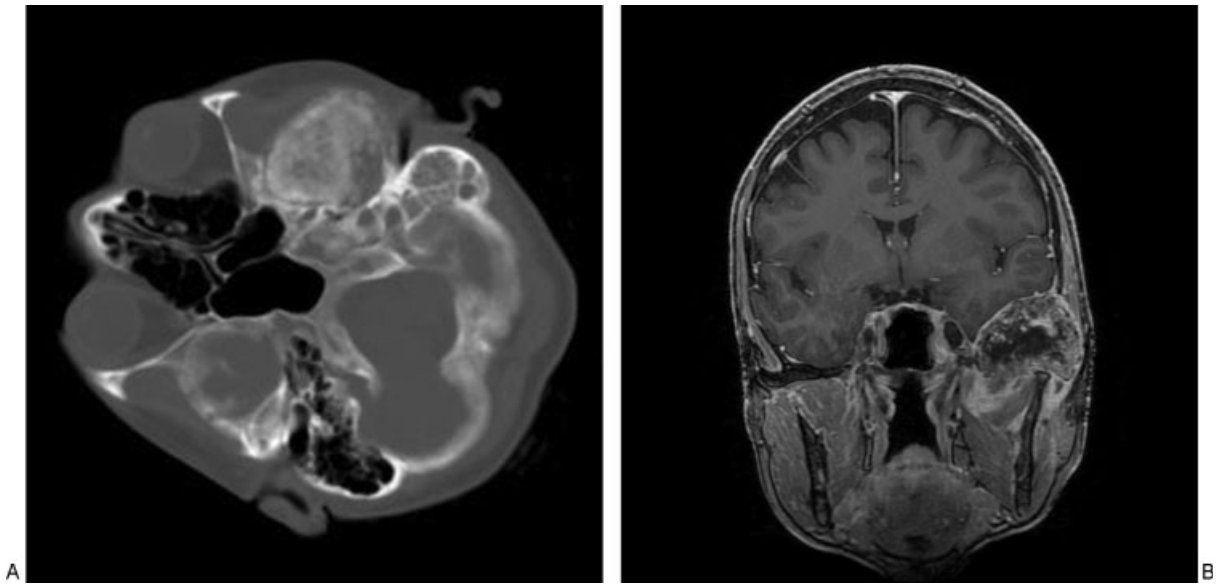

Figure 1 (A) Axial computed tomography scan demonstrating left temporomandibular joint (TMJ) erosion by a calcified lesion. (B) Coronal postcontrast T1-weighted magnetic resonance imaging demonstrating erosive lesion at the left TMJ.

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,

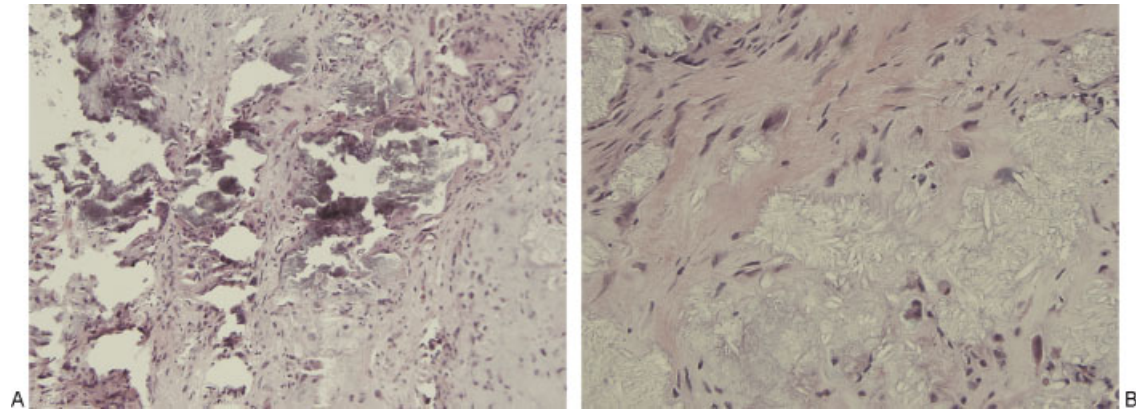

Figure 2 (A) Myxoid material with calcifications and reactive changes (H&E: original magnification 200×). (B) Myxoid material with numerous rhomboid elongated crystals (H&E: original magnification 400×).

chronic gout, renal failure, hypomagnesemia, and hypophosphatemia.[2] However, our patient had none of these conditions. It may also occur on a hereditary basis but is usually sporadic; our patient has no family history of CPDD. The prevalence increases with age, occurring in some form in 10 to 15% of 65 to 75 year olds and 30 to 50% of those >85 years old.[9,10] The disease appears to favor women 2:1[8,10] but a clear mechanism for this disparity has not been discovered. The joints most commonly affected are the knee, shoulder, hip, elbow, and metacarpophalangeal joints, while axial skeleton involvement (both cranial and spinal) is less common, but reported in the literature.[1–3,5,9–14] The origin is unknown but in general, destruction of the joint is required for crystal deposition.[2] Damage with increasing age predisposes to deposition due to changes in joint physiology (phosphate turnover, changes in synovial fluid components).[2] However, reports also exist of postsurgical CPDD.[13]

CPDD involvement of the skull specifically has also been reported usually causing severe bony destruction.[1,9,10,12,14] Involvement of the TMJ has been described in the literature in the form of case reports[10,14,15] but was not described in any of the cases originally described by McCarty.[7,12] Disease involving the TMJ typically presents with pain, swelling, and hearing loss.[9,14,15] In some reports, tumoral calcinosis has been documented but this is generally in the form of hydroxyapatite crystals.[1,2,15]

Management of tophaceous pseudogout is surgical. While some cases have been managed conservatively, symptomatic relief in most published reports occurred only after resection of the lesion.[14,15]

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.

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