Introduction

Calcium pyrophosphate dihydrate crystal deposition disease (CPPD) is a benign metabolic arthropathy first identified by McCarty et al, in 1962. Thought to occur secondary to abnormal phosphate metabolism, CPPD has a predilection for fibrocartilaginous joints such as the knee and wrists. Hyaline cartilage found in larynx and trachea is largely unaffected. The majority of CPPD cases present as inflammatory arthritis, although pseudotumoral calcifications have been reported. This rare entity has been coined tophaceous pseudogout, and is characterized by massive crystal deposition leading to formation of a calcified mass. CPPD of the TMJ is more common in women, with prevalence increasing with advancing age. Symptoms commonly include TMJ pain, preauricular swelling, otalgia, trismus, and facial pain. We describe a case of left sided TMJ pseudogout presenting as episodic pain and trismus.

Case Report

A 61-year-old male with a history of hypertension, gastric reflux, hyperlipidemia, post-traumatic stress disorder, depression, and gout presented with a 2-year history of left temporomandibular joint pain, and left aural fullness. He was evaluated by a dentist and diagnosed with TMJ dysfunction, with no response to conservative treatment. He subsequently underwent CT head which demonstrated a 4.4 cm x 3.2 cm x 2.6 cm groundglass density surrounding the left mandibular condyle with internal calcifications. It appeared to involve the temporal bone, entire glenoid fossa, floor of middle cranial fossa, and anterior wall of the middle ear cavity. Head CT obtained 6 years prior was reviewed, and demonstrated severe, chronic degenerative changes of the left temporomandibular joint.

Physical exam was significant for trismus and firm, non-tender left preauricular mass. Biopsy was performed via a preauricular approach. Intraoperatively, the mass was encapsulated in fibrous covering, and consisted of reddish-brown rubbery tissue interspersed with chalky material. The final surgical pathology was reported as calcium pyrophosphate dihydrate deposition disease/pseudogout.

Methods

Case Report and literature review. Patient medical records and imaging reviewed retrospectively.

Results

1. A multidisciplinary approach was employed for resection and reconstruction of tophaceous TMJ pseudogout. 2. Otolaryngology utilized a preauricular approach to expose the left TMJ with concurrent facial nerve identification and preservation. 3. Otorhinolaryngology provided enhanced access to the infratemporal fossa via an extra-oral vertical mandibular ramus osteotomy and disarticulation of the ramus-condyle unit. 4. Neurosurgery provided access to the involved middle fossa via a left temporal and infratemporal craniotomy. 5. The reconstruction was complex and required expertise of all three specialties and consisted of reconstruction of glenoid fossa with full thickness temporal bone graft, temporals myofascial flap, and cranioplasty.

6. Postoperatively, the patient required Physical Therapy and Speech Therapy to assist with the recovery of function.

7. To date, he has noticeable improvement in trismus and mouth opening. No further reports of pain.

Conclusion

1. Tophaceous pseudogout is the rarest variant of CPPD, and most commonly found in the TMJ.

2. Tophaceous CPPD in the TMJ is known to mimic other neoplastic or infectious conditions, and diagnosis requires a high index of suspicion.

3. The majority of tophaceous deposits are located medial to the joint, while only the lateral joint aspect is amenable to palpation upon physical exam. This results in delayed onset of palpable swelling, making diagnosis difficult.

4. Histopathologic analysis remains the most reliable diagnostic modality.

5. Upon gross examination, tophaceous pseudogout appears as friable, chalky, and “grits-like” granulomatous tissue coupled with temporomandibular joint erosion secondary to excess pressure on the condylar head.

6. Microscopically, CPPD is characterized by fibrocartilaginous degeneration, basophilic calcified matrix composed of rhomboid weakly birefringent crystals, and multinucleated giant cells.

7. Treatment of CPPD of the TMJ is dependent on severity, and aims to prevent crystal formation, dissolve existing crystals, and limit the biologic sequelae resulting from deposition.

8. In symptomatic, but nonexpansile cases, conservative management including steroids, colchicines, and NSAIIDs can provide symptomatic relief.

9. Additionally, joint lavage or injection of hyaluronic acid may help to reduce joint pressure and remove inflammatory crystalline particles.

10. Surgical excision is gold standard treatment for severe cases of tophaceous pseudogout.

11. We strongly advocate for the utilization of multidisciplinary approach to surgical resection including Otolaryngology-Head and Neck Surgery, Neurosurgery, and Otorhinolaryngology.

12. Postoperatively physical therapy and speech therapy are beneficial for optimal rehabilitation of function.

References


