Tophaceous pseudogout of the temporomandibular joint: a series of 3 cases

Rafik A. Abdelsayed, DDS, MS,[a] Nasser Said-Al-Naief, DDS,[b] Martin Salguerio, DMD,[c] Jon Holmes, DDS, MD,[d] and Samir K. El-Mofty, DMD, MS, PhD[e]

Georgia Regents University, Augusta, GA, USA; University of the Pacific at San Francisco, San Francisco, CA, USA; Private Oral and Maxillofacial Surgery, Birmingham, AL, USA; Washington University School of Medicine, St Louis, MO, USA

Objective. We report a series of 3 cases of tophaceous pseudogout of the temporomandibular joint (TMJ).

Study design. Three patients, two men and one woman, ranging in age between 60 and 75 years, presented with unilateral painful swelling of the TMJ area associated with limitation of mouth opening.

Results. Radiographic and computed tomographic images showed opaque masses in the supracondylar region of the TMJ. The preoperative clinical impression was a "neoplastic lesion" in the 3 cases. Microscopic examination revealed numerous deeply basophilic masses of calcified deposits, exhibiting birefringence under polarized light and morphologically consistent with calcium pyrophosphate dihydrate deposition, referred to in these cases as "tophaceous pseudogout."

Conclusion. Tophaceous pseudogout is a rare benign arthropathy that presents with clinical and radiographic features mimicking neoplastic conditions of the TMJ. Therefore, it is recommended that tophaceous pseudogout be considered in the differential diagnosis when a TMJ is involved with "neoplasm-like" lesions clinically and radiographically. (Oral Surg Oral Med Oral Pathol Oral Radiol 2014;117:369-375)

Calcium pyrophosphate dihydrate (CPPD) crystal deposition disease is a rare benign crystalline arthropathy of the articulating joints, first reported by McCarty et al. in 1962.[1] The term "chondrocalcinosis" was introduced, for the same condition, by Zitnan and Sitaj almost a year later.[2] The term "calcium pyrophosphate dihydrate crystal deposition disease [CPPDd]" was proposed by Ryan and McCarty in 1985.[3] CPPDd, by definition, is a monoarticular joint arthropathy, rarely involving two or more joints, and characterized by deposition of calcium pyrophosphate crystals, but without sodium urate, in the synovial fluid in patients with "gout-like symptoms." CPPDd is common in the aging population, with radiographic prevalence of 30% to 40% in people older than 80 years of age.[4] It is thought to result from a metabolic disturbance of the phosphate metabolism.[5] Pritzker et al.[6] in 1976 first described the condition in the temporomandibular joint (TMJ). Clinically, a wide range of symptoms has been described in association with this disorder when a TMJ is involved. CPPD may be deposited asymptomatically, or it may be associated with various chronic and acute symptoms.[7,8] These include facial pain, preauricular swelling, trismus, conductive hearing loss, otalgia, pyrophosphate arthropathy, and limitation of mouth opening.[9] The term "tophaceous pseudogout" is used when a joint is involved with massive, clinically detectable, and radiographically evident tumoral CPPD crystal deposition.[10,11]

The intent of this article is to report on the clinical, radiographic, and diagnostic pathologic features of 3 cases of tophaceous pseudogout; to discuss the differential diagnosis; and to review the relevant literature.

REPORT OF CASES

Case 1

A 60-year-old black man presented with pain and swelling of the left TMJ area and "limited mouth opening" for several

Statement of Clinical Relevance

This article highlights the clinicopathologic features of three cases of "tophaceous pseudogout" of the temporomandibular joint. This inflammatory, "tumor-like" arthropathy mimics neoplastic conditions and is often unfamiliar to clinicians of the head and neck. We report on the clinical, radiographic, and microscopic features, as well as the differential diagnosis, of this disorder. Clinicians and surgeons recognizing this pseudoneoplastic condition should help to secure accurate diagnosis and conservative patient management.

[a]Professor and Director of Diagnostic Laboratory Services, Section of Oral and Maxillofacial Pathology, Department of Oral Health and Diagnostic Sciences, College of Dental Medicine, Georgia Regents University.
[b]Professor, Oral Diagnosis, Radiology and Pathology, Loma Linda University, SOD and School of Medicine, Department of Anatomy and Pathology/Head and Neck Pathology.
[c]Assistant Professor, Department of Oral and Maxillofacial Surgery, College of Dental Medicine, Georgia Regents University.
[d]Private Oral and Maxillofacial Surgery.
[e]Head, Section of Head and Neck Pathology, Professor, Division of Anatomic and Molecular Pathology, Department of Pathology and Immunology, Washington University School of Medicine.

Received for publication Oct 24, 2013; returned for revision Dec 3, 2013; accepted for publication Dec 4, 2013.
Published by Elsevier Inc.
2212-4403/$ - see front matter
http://dx.doi.org/10.1016/j.oooo.2013.12.007

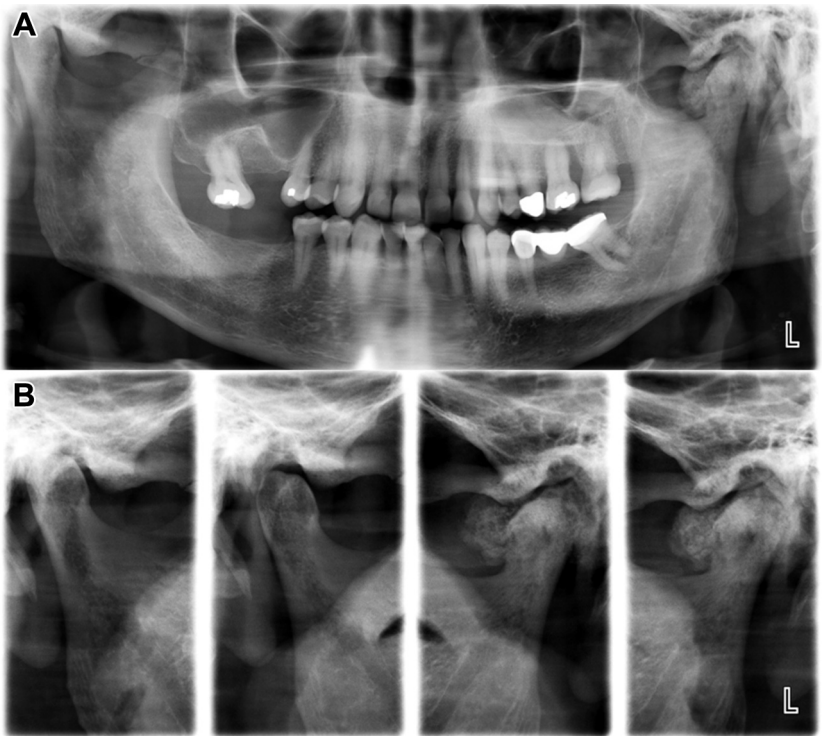

Fig. 1. Case 1. A panoramic radiograph showing a well-demarcated homogeneous opaque mass above the left condylar head (A). Left and right temporomandibular joint radiographs in centric relation and in translational movement (limited opening) positions showing the left condylar opaque mass (B).

years. The patient's medical history was insignificant, and there was no history of TMJ trauma or systemic joint disease. On physical examination, there was a unilateral facial asymmetry due to swelling in the left TMJ. The patient had limited jaw movement, with only a few millimeters interarch space when opening his mouth, accompanied by pain and tenderness. Panoramic and TMJ radiographs revealed a well-defined opaque mass with smooth outline in the left articulating space, which surrounded the condylar head (Figure 1, A, B). Axial and coronal sections of a computed tomography (CT) scan revealed pericondylar calcified deposits, with fine ground-glass matrix (Figure 2, A, B). Focal areas of bony erosions were noted in the condylar head as well as in the articulating eminence. The preoperative clinical diagnosis was "synovial chondromatosis," possibly a neoplasm. Excision of the lesion was performed with the patient under general anesthesia. During surgery, a significant amount of intracapsular calcareous material, described as chalky or "grits-like," was noted (Figure 3). The entire lesion was removed and submitted in 10% formalin for microscopic examination. Histologic examination revealed sections composed of multiple nodules of deeply basophilic calcified deposits embedded in a fibrovascular connective tissue stroma exhibiting cartilaginous metaplasia (Figure 4, A, B). Aggregates of rhomboid crystalline deposits were noted (Figure 4, C). These exhibited birefringence under polarized light (Figure 4, D). The metaplastic cartilaginous areas are composed of uniformly distributed mononuclear chondrocytes with uniform basophilic nuclei, without marked atypia, multinucleation, or mitosis. Benign, multinucleate, osteoclast-like giant cells were present. A diagnosis of "tophaceous pseudogout" was made. The surgical management consisted of an extraoral vertical ramus osteotomy to gain access to the portion of the mass medial to the mandibular condyle. The ramus-condyle segment was reimplanted and fixated with titanium miniplates. The postoperative healing was uneventful, and the patient was pain-free in his 6-month recall examination and regained almost full mouth opening.

Case 2

A 75-year-old white man presented with a history of pain in his right jaw associated with trismus for 4 years. For a period of 1 year, he had noted pain in the right ear with blockage and thumping tinnitus as well as hearing loss and episodic positional off-balance sensation. He had had right ear surgery for cholesteatoma. Physical examination revealed a swelling of the preauricular area of the right TMJ and marked swelling of the anterior wall of the external auditory canal. A high-resolution CT scan of the temporal bones, without intravenous contrast, showed a 3.8 × 4.2 × 4.7-cm dense mass centered on the right TMJ (Figure 5, A). The mass had a diffuse ground-glass matrix. There were associated smooth bony erosions of the posterior aspect of the greater wing of the sphenoid bone as well as the right mandibular condyle and proximal ramus. The mass extended into the middle ear cavity and external auditory canal and into the right middle cranial fossa. The clinical impression was of a low-grade chondroid tumor. A biopsy was planned, and exploration of the right