Pigmented Villonodular Synovitis of the Temporomandibular Joint: a Case Report and Literature Review
Mahdi A. Shkoukani, MD; Senja Tomovic; Kaiilash Narasimhan, MD; Lewis Clayman, MD, DMD; and Robert H. Mathog, MD
Department of Otolaryngology-Head & Neck Surgery, Wayne State University, Detroit, MI, USA

Abstract

Objective: Pigmented villonodular synovitis (PVNS) is a benign proliferation of tissues within a joint. It most frequently affects large joints but it is rarely diagnosed at the temporomandibular joint (TMJ). Radical excision with wide margins is the most accepted treatment modality. PVNS is resistant to radiotherapy. Herein we report a case involving the TMJ causing pain and trismus and its management that appears to be successful.

Study Design: A case report

Methods: A 74 year old female presented to our clinic complaining of a right pre-auricular mass that she noticed for a couple of years. The mass was growing slowly and never bothered her till recently. She started to have pain with jaw opening. Also, trismus became more pronounced. CT-neck with intravenous contrast showed an enhancing mass surrounding the right mandibular condyle with expansion of the temporomandibular joint. CT-guided biopsy of the right TMJ confirmed the diagnosis of PVNS.

Results: Patient underwent tracheostomy, right superficial parotidectomy followed by right condylectomy and partial mandibulectomy. Intraoperatively, the tumor was destroying the condyle and invading the masseter, masseter and pterygoid muscles and extending medially toward the skull base. Histologically, PVNS-diffuse type was confirmed in the specimen with negative margins. Postoperatively, patient developed a transient facial paralysis while pain and trismus disappeared.

Conclusions: PVNS is a rare lesion in the TMJ. Pre-auricular mass is the most common presenting symptom. CT-guided biopsy along with CT-neck is crucial in the workup. Resection of the lesion with wide margin is the recommended treatment. Facial paralysis is a complication of the surgical treatment but it is usually short-lived.

Introduction

Pigmented villonodular synovitis (PVNS) is a benign proliferation of synovial tissue. It can be either local or diffuse, depending on the extent of involvement of the joint space, bursae and tendon sheath. It occurs most commonly in the knees, hips, and hand joints of middle-aged people. However occurrence of PVNS in the temporomandibular joint is extremely rare, with less than 30 cases reported to date.

Patients with PVNS of the TMJ most often present with a preauricular mass that is often misdiagnosed preoperatively as a parotid tumor. Some patients may also experience local pain and trismus. The differential for this presentation is wide, and a combination of fine needle aspiration biopsy (FNA) and imaging with computerized tomography (CT) and magnetic resonance imaging (MRI) are recommended to confirm the diagnosis. Definitive treatment is with wide local excision.

Case Report

A 74 year old female presented to the university clinic complaining of a right pre-auricular mass that she noticed for few months. She admitted that the mass was painful and had been enlarging in the recent weeks. This mass was preventing her from taking big bites. On physical examination, she was noted to have 2.0 x 3.0 x 3.0 cm firm mass in the right pre-auricular area which was tender to palpation. Moderate trismus was appreciated on exam. The facial nerve was intact bilaterally.

The CT-neck with IV contrast (Figure-1) showed an enhancing mass surrounding the right mandibular condyle, with expansion of the TMJ. The MRI-neck with gadolinium (Figure-2) showed a 3.6 x 2.3 x 2.1 cm well defined mass with T1/T2 hypointense signal in the right TMJ region with subtle postcontrast enhancement. It also showed fine erosions of the mandibular condyle. The differential diagnosis included PVNS, arthritides (CPPD or gout), synovial chondromatosis, sarcoma, chronic osteomyelitis and metastasis from a distant site. A CT-guided Fine Needle Aspiration (FNA) was performed showing moderate cellularity and loosely cohesive groups of oval to spindle cells with bland nuclei, prominent nucleoli, and intracytoplasmic hemosiderin pigmentation, consistent with histiocytes. Numerous multinucleated giant cells with similar nuclear features were also present. These histologic features placed PVNS on top of the differential diagnosis list.

After obtaining an informed consent, the following procedures were performed: Tracheostomy tube placement, superficial parotidectomy, Condylotomy, partial mandibulectomy, meniscectomy and arthrocentesis. The condylectomy was done to protect the airway post-operatively. The superficial parotidectomy was performed to identify the facial nerve branches and protect them during the resection of the tumor. Intraoperatively, the tumor was destroying the condyle and invading the massicus with extension to the masseter muscle, medial pterygoid muscle, lateral pterygoid muscle and extending medially to the skull base and sphenoid spine. The final pathology report (Figure-3 and Figure-4) showed an infiltrate of dense mononuclear cells that was associated with hemosiderosis, multinucleate giant cells, and fibrosis. Proliferation replaced joint capsule, adjacent connective tissue and skeletal muscle, and occupied narrow space. This was consistent with the final diagnosis of PVNS.

Postoperatively, the patient had no new postoperative complications. Although there was a complete right facial paralysis, patient had undergone a gold weight placement and lateral canthoplasty to address incomplete right eye closure. After several months, however, movement did return in the right face, and eye closure and mouth opening did markedly improve. There was no evidence of recurrent tumor and the patient no longer has pain or trismus. Maxillofacial prosthetics were later used to improve her post-resection jaw deviation.

Discussion and Conclusions

PVNS is a benign proliferation of tissues within a joint. It most frequently affects large joints such as the knees, hips and hands.

Lapayowker et al reported the first case of PVNS in the TMJ in 1973. Jaffe et al proposed that PVNS was a chronic inflammatory process. Other theories have been set forth, including the otherwise accepted theory that describes the lesion as a benign neoplasia owing to evidence of cellular monoclonality, chromosomal abnormality, and a few reported cases of metastatic disease. Other more controversial theories describe it as a lipid metabolism disturbance, or a reaction to blood products.

Grossly, the lesion appears dark to yellow-brown in color, with a nodular and villous texture. Histologically the mass contains finger-like projections that consist of multinucleated giant cells, histiocytes and hemosiderin concentrated in stromal cells. The mononuclear histiocytes show positive immunoreactivity for CD68 and negative for S-100, thus differentiating it from chondroblastoma cells, which are positive for S-100.

Immunophenotypic staining has also suggested that the cells are of synovial origin.

The most prevalent symptom of PVNS of the TMJ is a preauricular mass. Pain and discomfort of the jaw and trismus can also occur. Less commonly patients may present with hearing loss or tinnitus. In addition to a FNA, imaging (CT and MRI) is a key component of the diagnostic work-up.

Conventional treatment for PVNS is radical excision with wide margins. The local recurrence rate has been reported to be about 9%.

There has only been one reported case of malignant PVNS of the TMJ with metastasis. A few select cases were successfully treated with radiation therapy, none of which occurred at the TMJ. The most common complication after surgical treatment is facial paralysis, which resolves in a few months.

In our case we chose to perform a superficial parotidectomy to expose and protect the facial nerve during TMJ resection, nevertheless our patient still initially suffered a complete paralysis, which resolved during subsequent follow-up visits.

References