Recurrent pigmented villonodular synovitis of the temporomandibular joint

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Abstract

Pigmented villonodular synovitis is a benign but locally aggressive extra-articular tumor arising from the synovial membrane of tendons and bursae occurring near a joint space. Rarely, pigmented villonodular synovitis can involve the temporomandibular joint, which is emphasized in this paper. Diffuse and localized types have been described in the literature. The diffuse type involves the entire synovial membrane and infiltrates adjacent structures, which tend to be more aggressive and associated with a higher rate of recurrence when compared with the localized type.

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Case report

A 38-year-old woman initially presented to the otolaryngology clinic with complaint of a left-sided jaw mass. Contrast-enhanced computed tomography (CT) of the neck was notable for a large soft tissue mass involving the left masticator space partially invading and displacing the masseter, lateral pterygoid, and temporalis muscles. There was no evidence of erosion or destruction of the mandibular condyle or the left temporomandibular joint (TMJ) (Fig. 1). Further characterization with contrast-enhanced magnetic resonance imaging (MRI) was recommended, which showed a T1 iso- to hypointense mass in the left masticator space with heterogeneous enhancement measuring 7.5 × 6.4 × 5.5 cm in craniocaudal, transverse, and anteroposterior dimensions. There were multiple regions of T2 hypointensity within the central aspect of the mass (Figs. 2 and 3). The mass effect on the parapharyngeal space demonstrated medial displacement with infiltration and enhancement of all the muscles of mastication. High T2 signal was noted in the mandible but without cortical destruction or evidence of invasion. Additionally, there was suspicion for involvement of the trigeminal nerve with expansion of the foramen ovale. After biopsy and surgical excision of the soft tissue mass, pathology demonstrated large nodules of solid sheets of tumor cells with hemosiderin pigment admixed with xanthoma cells and a few multinucleated giant cells (Figs. 4 and 5). Immunohistochemical staining with CD68 was diffusely positive, confirming histiocytic origin (Fig. 6). No cellular atypia was reported. Findings were consistent with pigmented villonodular synovitis (PVNS), also known as giant cell tumor of the tendon sheath-diffuse type.

The patient was then referred to our institution 8 years later with complaint of left neck swelling and jaw pain for about 4 months. Computed tomography (CT) of the neck showed a recurrent mass measuring 7.3 × 5.1 × 4.8 cm in craniocaudal, transverse, and anteroposterior dimensions, respectively. MRI demonstrated a T1 isointense mass measuring 7.3 × 5.1 × 4.8 cm with heterogeneous enhancement. The mass effect was noted on the parapharyngeal space and the left masticator space, respectively. The patient underwent surgical resection of the mass and postoperative CT showed a resection cavity measuring 7.1 × 4.3 × 3.6 cm. The patient is currently 12 months after surgery with no evidence of recurrence.

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months. Physical exam demonstrated a hard, firm, mobile 3-cm mass near the left parotid gland. Repeat contrast-enhanced MRI demonstrated a heterogenous mass with central hypointensity on T1- and T2-weighted images involving the left masticator space and wrapping around the mandibular ramus measuring $2.8 \times 4.4 \times 4.2$ cm in anteroposterior, transverse, and craniocaudal dimensions without appreciable enhancement (Figs. 7-9). There was minimal signal change in the mandible itself without enhancement. A fine needle aspiration was then obtained showing multinucleated giant cells with hemosiderin-type pigment and stromal cells highly suggestive of recurrent PVNS. A complete resection with TMJ reconstruction and anterolateral thigh free flap reconstruction is currently preoperatively planned.

Discussion

PVNS involving the TMJ is a rare entity and is difficult to diagnose clinically. The reported incidence is 1.8 annual cases per million individuals in which most lesions affect the knee, hip, or shoulder [1]. Patients usually present in the third and fourth decades with symptoms of TMJ dysfunction [2]. Imaging, particularly utilizing MRI, is helpful as it can demonstrate a characteristic hemosiderin pattern and outlines the extent of the mass to aid in treatment and prognosis. When the TMJ is involved, most cases are of the diffuse histologic type. The etiology of this disease process is unclear, with histopathology characterized by hyperplasia of the synovium in joints and tendinous sheaths. There is accentuated proliferation of the stroma cells, and a significant quantity of intracellular and extracellular hemosiderin.

Fig. 1 – Contrast-enhanced coronal computed tomography shows a solid enhancing mass in the left masticator space surrounding the mandible and displacing the parapharyngeal space.

Fig. 2 – Axial T2-weighted image shows multifocal regions of low signal intensity within the central mass involving the left masticator space and temporomandibular joint.

Fig. 3 – Coronal postcontrast T1-weighted fat-saturated image shows a solid mass in the left masticator space with areas of low signal intensity and mild heterogeneous enhancement.

Fig. 4 – Hematoxylin and eosin of the tumor demonstrating solid sheets of cells separated by fibrous stroma. Scattered histiocytes and multinucleated giant cells contain intracytoplasmic hemosiderin.
hemosiderotic pigments, as well as accumulation in multinucleated giant cells [1,3]. It should be noted that giant cell tumors of the tendon sheath are considered synonymous with PVNS microscopically [4]. The inconsistent terminology has provided sources of confusion between clinicians, radiologists, and pathologists in the past. Therefore, a unifying terminology is becoming more established.

Classically, MRI depicts scattered foci of low signal on T1- and T2-weighted images because of hemosiderin deposition with or without an incomplete fibrous capsule [5]. A reactive joint effusion may be present and often demonstrates intermediate signal on T1-weighted imaging and high signal on T2-weighted imaging. If enhancement is observed, it is usually caused by inflammation and increased vascularity of the synovium. More often, delayed enhancement of the solid component is observed [5,6]. Although these features are classic for PVNS on imaging, it should be noted that cases present with variable signal intensity depending on the degree of hemosiderin deposition. CT is useful in assessing for erosion of the mandibular condyle and the adjacent temporal bone. Additionally, CT can help differentiate PVNS from other synovial processes such as synovial chondromatosis or chondrosarcoma, which can involve the same anatomic space.

Fig. 5 – High-power hematoxylin and eosin of the tumor again demonstrating histiocytes and multinucleated giant cells engulfing hemosiderin particles.

Fig. 7 – Coronal precontrast fat-saturated T1-weighted sequence demonstrates a heterogeneous mass in the left masticator space near the temporomandibular joint with central hypointensity.

Fig. 6 – Immunohistochemical stain with CD68 is diffusely positive, which confirms histiocytic cell origin.

Fig. 8 – Coronal gadolinium-enhanced fat-saturated T1-weighted sequence showing a predominantly hypointense mass in the left masticator space and temporomandibular joint with little enhancement.
Treatment involves complete surgical excision of the soft tissue lesion and involved bones. Special attention must be given to the diffuse type as often it is difficult to completely remove because of the infiltrative nature. As a result, the diffuse type is associated with increased risk of recurrence [2–4,7].

In conclusion, we present a rare clinical entity and emphasize the utility of preoperative MRI. PVNS can be confidently diagnosed with this modality, given the presence of hemosiderin, which produces a characteristic appearance on T1- and T2-weighted sequences.

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Fig. 9 – Coronal T2-weighted image demonstrating a heterogeneous soft tissue mass in the left masticator space with central hypointensity.