A 85-year-old man was referred to the Otolaryngology-Head and Neck Surgery clinic in February 2013 for left ear pain associated with a small lesion with intermittent bleeding. He had no other systemic or neurologic symptoms. This was believed to be chondrodermatitis nodularis chronica helicis. One year later, the patient was rereferred for a preauricular mass on the ipsilateral side (Fig. 1) with an inconclusive fine needle aspirate.

From a symptomatic perspective, the patient had ear pain and intermittent headaches but denied any facial numbness or weakness, weight loss, fever, anorexia, trismus, or impaired hearing. The patient had a positive history of trauma to the area as he was struck with a large piece of lumber 7 years before. Computed tomography scan and magnetic resonance imaging of the face and skull base showed a destructive process with intracranial extension centered at the left temporomandibular joint (TMJ) with hypertrophy and enhancement of synovium. The lesion was peripherally calcified and heterogeneous (Fig. 2). The findings on imaging correlated with a differential diagnosis of synovial chondromatosis, chondrosarcoma, or pigmented villonodular synovitis (PVNS). A repeated fine needle aspirate was performed, which showed no evidence of malignancy and findings most compatible with PVNS.

This patient’s case was discussed at Head and Neck Tumor Board. Because of radiologic features not in keeping with the suspected pathologic diagnosis, it was felt that an open biopsy was the best way to proceed. The patient underwent an uneventful transfacial TMJ biopsy through a preauricular incision. Pathology from several pieces of the mass showed a lobulated mildly atypical cartilaginous proliferation associated with synovium with reactive changes, including chronic inflammation. The open biopsy pathology ruled out chondrosarcoma. This pathology coupled with the history of trauma and intracranial extension confirmed the diagnosis of PVNS.

The patient did not receive any further surgical intervention given his age and that the diagnosis was most in keeping with a PVNS of the TMJ. The patient is since doing well and is being followed by the Head

Summary: Pigmented villonodular synovitis (PVNS) is a rare and benign proliferative disorder of synovium with potentially locally aggressive growth and invasion of the bone. Occurring within the joints, tendon sheaths, and bursae, it is most commonly a monoarticular disease affecting large joints. In particular, most cases of PVNS occur in the knee. PVNS of the temporomandibular joint (TMJ) is a highly rare disorder, with approximately 60 cases reported. Herein, we present a unique case of an elderly male presenting with ear pain and subsequently diagnosed with PVNS of the TMJ with a history of trauma to the area. Initial imaging of the TMJ and the surrounding region looked concerning for invasive and/or malignant disease, but an open biopsy confirmed PVNS. (Plast Reconstr Surg Glob Open 2016;4:e674; doi: 10.1097/GOX.0000000000000658; Published online 6 April 2016.)
and Neck Surgery team; he receives magnetic resonance imagings every 6 months, and there has been no interval growth with conservative management over the last year.

**DISCUSSION**

PVNS of the temporomandibular joint (TMJ) is a highly rare disorder, with approximately 60 cases reported.\(^1\)\(^2\) A recent literature review of PVNS of the TMJ revealed that 88% of localized and 59% of diffuse cases had a positive trauma history, as was seen in this patient.\(^3\) In addition, approximately 33% of PVNS of the TMJ exhibit intracranial extension,\(^4\) whereas only 2% to 8% of synovial chondromatosis of the TMJ exhibit this aggressive feature.\(^5\)\(^6\) Interestingly, this patient presented 1 year before noticeable mass development with pain around the ear, which may highlight an early presenting feature of PVNS of the TMJ.

This case demonstrates the importance of integrating clinical, radiographic, and pathologic features of rapidly growing masses to determine the proper diagnosis. This patient’s mass appeared to grow 4 cm within 1 year from initial presentation; imaging showed a destructive/invasive mass centered around the TMJ, with suspicion of malignancy. However, a benign symptom profile and a biopsy not suggestive of malignancy led toward a diagnosis of PVNS.

**CONCLUSIONS**

PVNS is a rare proliferative disorder of synovium occurring within the joints, tendon sheaths, and bursae. Although mostly a disease of large joints, in rare cases \((N = 60)\), it can affect the TMJ as presented here. This case illustrates an uncommon presentation of PVNS of the TMJ in an elderly male with ear pain and a positive history of trauma to the area. Imaging revealed intracranial extension, and a biopsy of the mass ruled out malignancy confirming PVNS.

Kiersten Pianosi, BSc
Division of Otolaryngology-Head and Neck Surgery
Department of Surgery, Dalhousie Medical School
Dalhousie University, 1276 South Park Street
Halifax, Nova Scotia, Canada B3H 2Y9
E-mail: kiersten.pianosi@dal.ca

**PATIENT CONSENT**

The patient provided written consent for the use of his image.

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