# Pigmented Villonodular Synovitis of the Temporomandibular Joint

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*Pigmented villonodular synovitis (PVNS) is a relatively rare disease affecting synovium-lined joints. PVNS was first fully described in 1941 by Jaffe et al, who considered it a benign inflammatory state of the synovium of uncertain etiology. Reports of this disease in the temporomandibular joint (TMJ) are extremely rare. This report describes a case of a 78-year-old man affected by PVNS of the right TMJ. Clinical examination revealed the presence of a right-sided preauricular mass; a magnetic resonance imaging scan showed a mass located lateral to the right mandibular condyle, close to the TMJ capsule. The clinical and pathologic features of the case are described.* J OROFAC PAIN 2008;22:252–255.

**Key words:** pigmented villonodular synovitis, temporomandibular joint diseases

Benign tumors and tumorlike lesions that involve the temporomandibular joint (TMJ) are very rare. The more frequent varieties are osteochondroma, chondroma, osteoma, synovial chondromatosis, and pigmented villonodular synovitis (PVNS). PVNS is a benign, locally invasive lesion of the synovium of joints, bursae, and tendon sheaths. Chassaignac first described PVNS in 1852.[1] It was first fully described in 1941 by Jaffe et al, who considered it a benign inflammatory state of the synovium of uncertain etiology.[2] In 1973, Lapayowker et al were the first to describe a case of PVNS of the TMJ.[3] PVNS of the TMJ is a relatively rare occurrence.[4] In fact, PVNS usually affects feet, ankles, fingers, hips, elbows, wrists, and shoulder and has an incidence of 1.8 cases per million population.[5] PVNS can have a clinical presentation that might be mistaken for a temporomandibular disorder (TMD), a tumorlike parotid mass, or a simple soft tissue mass in the TMJ area. Symptoms related to TMJ dysfunction, including pain during mastication, joint noises, and limited mouth opening might be present. Nevertheless, more often, only a painless, nontender, slow-growing lump near the mandible is present.

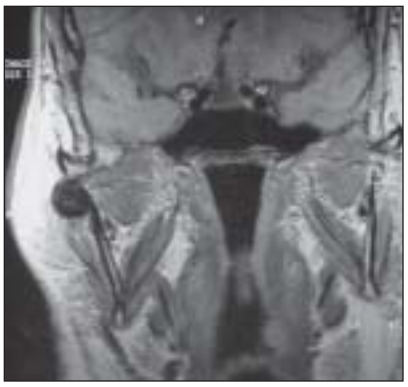

**Fig 1** MRI scan showing the lesion from its medial and lateral aspects.

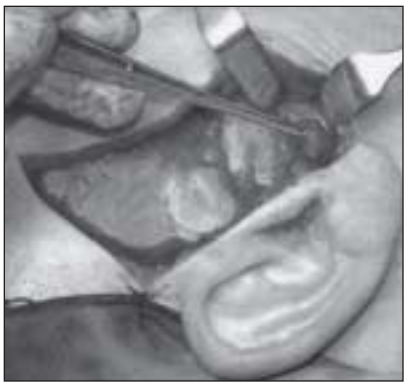

**Fig 2** Intraoperative image showing the macropathologic aspects of the tumor, a brownish, friable mass strictly adherent to articular synovia.

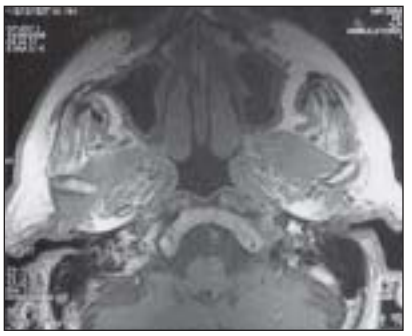

**Fig 3** MRI scan 2 years postoperatively.

The main microscopic feature of villonodular synovitis is a nonspecific inflammatory process that results in a papillary, villous, and nodular expansion of the synovial membrane.[6] Pathologic features are in fact often represented by synovial hypertrophy with hemosiderin deposition, inflammation, and, rarely, bony erosion.[7] PVNS can histologically show a nodular or a diffuse pattern.[8] The most common nodular variant (villous), including giant cell tumor, xanthoma, xanthogranuloma, and myeloplaxoma, generally affects a discrete part of the synovium. The villous pattern is generally composed mainly of histiocytelike cells and scattered osteoclastlike multinucleated giant cells, accompanied by chondroid areas with occasional lacelike calcification. The overlying cells of the synovium appear in a lobular and sheetlike arrangement of mononuclear, round, epithelioid cells and lipid-rich cells.[7,8] The diffuse variant may involve the entire synovial membrane and is usually referred to as giant cell fibrohemangioma, chronic hemorrhagic villous synovitis, or benign polymorphocellular tumor of the synovium.[7,8]

The etiology of this synovial tumor is still unknown. Because of its pathologic aspects, some authors have hypothesized that acute or repeated articular trauma might play a possible role in the development of PVNS. Other theories are based on the patient's systemic conditions.[1]

We report a case of PVNS involving the TMJ, not only for its unusual location but also to underscore how its clinical presentation can mimic a parotid mass and may also manifest pain.

## Case Report

In June 2006, at the Department of Maxillofacial Surgery of the University in Rome "La Sapienza," we observed a 78-year-old man complaining of right-sided orofacial pain. A small mass was visible in the right preauricular area, and slightly limited and right-deviated mouth opening was also evident. The patient's medical history was significant only for arterial hypertension and diabetes.

Clinical examination revealed the presence of a spherical, painless, and well-circumscribed mass in the right preauricular region. Right facial nerve function was well-preserved. A magnetic resonance imaging (MRI) scan showed a preauricular mass of 1.5 cm located lateral to the right mandibular condyle, close to the TMJ capsule (Fig 1). Erosion of the bony structures nearby was not apparent.

The patient underwent surgical intervention through the right preauricular and right coronal access. The articular disc and the mandibular condyle were preserved, whereas the mass was removed with a partial synovectomy (Fig 2). Intraoperative examination of the mass showed a brownish, friable tumor strictly adherent to the articular synovia and close to but easily removable from either the condyle or the articular disc. Definitive histologic examination of the tumor disclosed a nodular pattern of very dense multinucleated giant cells and ovoid cells, confirming the diagnosis of PVNS. The postoperative course was uneventful and the patient was discharged 1 day after the surgical intervention.

At 2 years of follow-up, there was good mandibular function, and radiologic assessment showed no evidence of local recurrence (Fig 3).

## Discussion

PVNS is a benign proliferative disorder of the synovium. Jaffe et al in 1941 introduced the term PVNS to describe an idiopathic proliferative lesion involving tendon sheaths, bursae, and diarthrodial joints.[2] Such an entity was considered to belong to a common family of lesions called pigmented villonodular synovitis, pigmented villonodular tenosynovitis, or pigmented villonodular bursitis, depending on the area involved.

The etiology of PVNS is still unknown. There are 4 main theories: etiologic factors, disturbances of lipid metabolism, response to blood or blood products, and an inflammatory response to an unknown nontraumatic irritant, although several authors consider PVNS a simple benign neoplasm of synovial vascular or fibrohistiocytic origin.[1,3,6] Histologically, the tumor can show a diffuse or a villous pattern. Fewer than 30 cases of PVNS of the TMJ have been reported in the international literature, which reveals that PVNS can occur at any age, with a peak incidence in the fourth and fifth decades of life, and has no sex prevalence. Moreover, all reviewed cases show that PVNS has a clinical presentation that might be mistaken for TMJ disease, a tumorlike parotid mass, or a simple soft tissue mass in the TMJ area.[1–10]

Analysis of the literature reveals that symptoms related to TMJ dysfunction, including pain during mastication, joint noises, and limited mouth opening, might be present. Nevertheless, more often, only a painless, nontender, slow-growing lump near the mandible is present.[11] On first clinical presentation only 3 patients showed earache, trismus, diplopia, vomiting, and hearing loss, possibly a result of skull base involvement. A skull base mass was already evident in only 1 case.[11] In the remaining 2 cases, the preauricular mass extended into the infratemporal fossa.[11–13]

Although it is relatively rare, a PVNS should be suspected in the radiologic evaluation of all patients with preauricular swelling and bone cysts of the TMJ.[1,3,9] The radiologic aspect of TMJ PVNS is usually normal during the early stage, whereas evidence of bony erosion in the articular fossa might appear at an advanced stage.[1–3,9–16] If PVNS is suspected, a computerized tomography scan or MRI evaluation is essential to define the features, localization, and extent of the lesion.[14–17] Arthroscopy would also appear to be a useful diagnostic tool for PVNS, and fine-needle aspiration cytology might confirm preoperative diagnosis.[6,7,16]

Of the villous (nodular) and the diffuse patterns, villous is the more common. Its clinical-pathologic features resemble those of a chondroblastoma of the bone, with the exception of the villous pattern. Immunologic studies have shown that the histiocytelike cells that are a feature of the villous pattern have a positive immunoreactivity for CD68, whereas they are generally negative for S-100 protein.[15] Some previously reported cases of chondroblastoma in the temporal bone may have actually been cases of PVNS with chondroid metaplasia.[1,15] When chondroblastomalike lesions involve the temporal bone or TMJ, the possibility of PVNS with chondroid metaplasia should also be considered, in addition to chondroblastoma of the bone. The correlation between this lesion and synovial chondromatosis remains unknown.[15]

Treatment of PVNS is exclusively surgical and consists of complete removal of the mass and the nearest involved structures; since synovial lesions have a high rate of local recurrence (> 50%), synovectomy is strongly recommended.[15] Some authors have proposed postoperative radiotherapy (35 Gy in 15 fractions) in patients with a high risk of recurrence.[18,19]

Recurrence of PVNS in the TMJ was reported in 2 patients by Takagi and Ishikawa at 5 years after surgical excision.[17] O'Sullivan et al reported 3 cases of recurrence, which were treated by radiation therapy, during a 7-year follow-up period.[18]

Facial paralysis might be a major postoperative complication depending either on the tumor location or on nearby structures involved. Facial paralysis has been described in 4 cases,[12,20–22] and 1 patient complained of facial weakness.[23]

## Conclusions

PVNS is an uncommon benign tumor of joints and affects the TMJ relatively rarely. Nevertheless, despite the benign nature of the lesion, preoperative planning is mandatory for successful treatment of PVNS because of the possible extension of the lesion into the middle and infratemporal fossae or the external and middle ear, the vicinity of the carotid canal, and possible postoperative complications.

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