Pigmented villonodular synovitis of the temporomandibular joint: case report and review of the literature

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Pigmented villonodular synovitis (PVNS) is an aggressive proliferative lesion that usually involves the synovial tissues of big joints. To date, there are ~52 cases of PVNS affecting the temporomandibular joint reported in the English-language literature, about one-third of them exhibiting intracranial involvement. We herein describe an additional case of PVNS of the temporomandibular joint with skull base invasion affecting a 26-year-old male patient and discuss its clinicopathologic features considering previously published cases. Histopathology and imaging evaluation are important for the diagnosis of PVNS, which should be included in the differential diagnosis of preauricular aggressive swellings. **(Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2011;111:e17-e28)**

Tumors and pseudotumors of the temporomandibular joint (TMJ) are rare. The most common group include arthritic changes (e.g., osteoarthritis and rheumatoid arthritis), whereas neoplasms (e.g., osteochondroma, chondroma, and osteoma) and synovial disease (e.g., synovial chondromatosis and pigmented villonodular synovitis [PVNS]) are uncommon.[1-3] PVNS is a rare proliferative benign lesion with an incidence of 1.8 annual cases per million individuals, and which preferentially affects the synovial tissue of the large joints, such as the knee and hip of adult patients in their third to fifth decades of life.[4] It can involve any articular site, including the ankle, shoulder, and elbow, and rarely involves the joints of the hands, feet, spine, and TMJ.[5]

The most common features seen in patients with PVNS affecting the TMJ region (PVNS-TMJ) include preauricular swelling with progressive pain and mouth opening restriction. Imaging examinations show important bone destruction, and about one-third of the cases present skull base involvement.[6,7] Microscopically, PVNS shows mononuclear and osteoclast-like multinucleated giant cells distributed in a vascular fibrous stroma, presenting villonodular proliferations and hemosiderin deposition.[8]

The first report of PVNS-TMJ is attributed to Lapayowker et al. in 1973, and to the best of our knowledge, 52 cases have been published in the English-language literature (Table I), and at least 4 additional cases in non-English languages.[9-12] Herein, we report an additional case of PVNS-TMJ with skull base involvement and review the literature.

CASE REPORT

A 26-year-old male patient was referred to the Oral and Maxillofacial Surgery service in the city of Aracaju (Sergipe, Brazil), complaining of difficulties in opening his mouth. It was painless, but the patient reported a progressive loss of hearing in his left ear, which started suddenly 2 months before without any association with trauma or previous treatment. His medical history was noncontributory. Extraoral examination revealed a painless diffuse swelling in the left preauricular region measuring 0.5 cm and a slight facial asymmetry on the left body of the mandible compared with the opposite side (Fig. 1, *A*). There was an opening mouth restriction of 20 mm, without mandibular deviation.

Computerized tomography (CT) showed extensive bony destruction of the left condyle and a mass measuring about 1.5 cm in diameter located in the left infratemporal fossa and middle cranial fossa (Fig. 2). Magnetic resonance images (MRI) were requested for better soft tissue evaluation, which showed homogenous hypointensity throughout the lesion on both T1- and T2-weighted sequences. The MRI analysis also showed displacement of adjacent structures, such as the left medial pterigoid muscle, masseter muscle, and left temporal lobe, confirming the infiltrative nature of the lesion (Fig. 3). Considering the imaging features of infiltrative and destructive growth, the main clinical differential diagnosis was of osteosarcoma, chondrosarcoma, or metastatic dis-

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Table I. Clinical features of 52 cases of pigmented villonodular synovitis (PVNS) of the temporomandibular joint (TMJ) reported in the English-language literature and the present case

Case	*Author*	*Age (y)/gender*	*Clinical features/duration*	*BD*	*IC*	*Treatment*	*Recurrence/ follow-up*
1-2	Lapayowker et al. (1973)[41]	22/M	Preauricular swelling, pain when chewing/18 mo	Yes	No	SE, partial capsulectomy	NR
		58/F	Preauricular swelling, clicking, hearing decrease/1 y	Yes	No	NR	NR
3	Barnard (1975)[61]	37/M	Painful preauricular swelling, trismus/3 wk	Yes	NR	Conservative approach	NR
4	Dinerman and Myers (1977)[62]	58/F	Preauricular swelling, clicking, tinnitus, hearing decrease/10 mo	Yes	Yes	SE, dura mater was excised	No/4 y
5	Miyamoto et al. (1977)[42]	34/M	Preauricular swelling, trismus/NR	Yes	No	SE	No/2 y
6	Raibley (1977)[43]*	62/F	Painful preauricular swelling/2 y	No	No	SE	No/5 mo
7	Makek and Drommer (1978)[17]	55/F	Painful preauricular swelling, trismus/18 mo	Yes	No	SE, capsulectomy	No/9 mo
8	Takagi and Ishikawa (1981)[50]*	36/M	Clicking, trismus/3 y	NR	No	SE, partial condylectomy	Yes/16 y
9	Gallia et al. (1982)[63]	47/F	Preauricular swelling, trismus, masticatory pain/3 wks	Yes	No	SE	No/2 y
10	Rickert and Shapiro (1982)[64]	39/F	Asymptomatic parotid mass/1 mo	No	No	SE, partial capsulectomy	NR
11	Curtin et al. (1983)[35]	47/F	Facial painful swelling, trismus/18 mo	Yes	No	SE	NR
12	O'Sullivan et al. (1984)[56]	61/F	Preauricular swelling/NR	Yes	Yes	SE, radiotherapy	Yes/3 y
13	Dawiskiba et al. (1989)[47]	32/M	Painful preauricular swelling, trismus/NR	No	No	NR	NR
14	Eisig et al. (1992)[19]	50/F	Ear canal mass, hearing loss/2 mo	Yes	Yes	SE, condylectomy	No/1 y
15	Syed et al. (1993)[59]	10/F	Preauricular swelling/1 y	No	No	SE, partial capsulectomy	NR
16	Franchi et al. (1994)[65]	59/F	Painful preauricular swelling/6 mo	No	No	SE, partial capsulectomy	No/1 y
17	Shapiro et al. (1996)[66]	36/M	Painful preauricular swelling, clicking, hearing loss/ NR	Yes	Yes	SE	No/18 mo
18	Youssef et al. (1996)[67]	41/F	Painful preauricular swelling/6 mo	Yes	No	SE, synovectomy	No/14 mo
19	Allis-Montmayeur et al. (1997)[1]	39/F	External ear canal mass/3 mo	NR	NR	NR	No/7 mo
20	Reñaga Rubin et al. (1997)[27]	70/F	Painful preauricular swelling, clicking, trismus/7 mo	Yes	No	SE, synovectomy	No/3 y
21	Tanaka et al. (1997)[25]	47/M	Painful preauricular swelling, trismus/8 mo	Yes	Yes	SE, condylectomy, temporal craniectomy	No/2 y
22	Yu et al. (1997)[48]	48/M	Painful preauricular swelling/7 mo	No	No	SE	NR
23	Chow et al. (1998)[68]	42/F	Zygomatic region swelling/6 mo	Yes	No	SE, condylectomy, capsulectomy	No/2 y
24	Omura et al. (1998)[69]	18/M	Painful preauricular swelling, trismus/10 mo	Yes	No	SE	No/2 y
25	Bemporad et al. (1999)[24]	37/M	Preauricular swelling, trismus, hearing loss/4 y	Yes	Yes	SE, temporal craniectomy	NR
26	Song et al. (1999)[23]	57/F	Preauricular swelling/2 mo	Yes	No	SE, condylectomy	NR
27	Stojadinovic et al. (1999)[26]	63/M	Painful preauricular swelling, trismus/2 y	Yes	Yes	SE, condylectomy	No/20 mo
28	Lee et al. (2000)[18]	59/F	Preauricular swelling, salivary flow decreased/3 y	Yes	No	SE, condylectomy	No/2 y
29	Kişnişçi et al. (2001)[28]	45/F	Painful preauricular swelling, trismus/NR	No	No	SE, partial capsulectomy	No/1 y
30	Klenoff et al. (2001)[32]	35/NR	Painful preauricular swelling, trismus, tinnitus, hearing loss/4 mo	Yes	Yes	SE, craniectomy, mandibulectomy	NR
31	Shapiro et al. (2002)[46]	36/M	Temporal mass, clicking, hearing loss/NR	Yes	Yes	SE, craniectomy	No/7 y
32-33	Church et al. (2003)[36]	42/M	Preauricular swelling, clicking, hearing loss/4 mo	Yes	Yes	SE, partial mandibulectomy	No/3 y
		33/M	Painful preauricular mass, trismus/2 y	Yes	No	SE, partial mandibulectomy, TMJ excision	No/2 y
34	Heo et al. (2003)[37]	45/M	Painful preauricular swelling, tinnitus, clicking, trismus/17 mo	Yes	No	NR	NR

Table I. Continued

Case	*Author*	*Age (y)/gender*	*Clinical features/duration*	*BD*	*IC*	*Treatment*	*Recurrence/ follow-up*
35	Aoyama et al. (2004)[31]	33/M	Painless preauricular swelling, clicking, trismus/20 y	Yes	Yes	SE, condylectomy, middle cranial fossa resection	No/2 y
36-39	Kim et al. (2004)[6]	28/F	NR	Yes	Yes	NR	NR
		22/M	NR	Yes	Yes	NR	NR
		47/M	NR	Yes	No	NR	NR
		58/F	NR	Yes	No	NR	NR
40	Tosun et al. (2004)[38]	60/M	Zygomatic painful swelling, trismus, chronic otitis media/10 y	Yes	Yes	SE, condylectomy, craniectomy	No/5 y
41	Cascone et al. (2005)[39]	38/F	Preauricular swelling/NR	No	No	SE, parotidectomy	No/1 y
42-43	Stryjakowska et al. (2005)[33]	36/M	Painful preauricular swelling, trismus, tinnitus, hearing loss/6 mo	Yes	Yes	SE, intradural resection, mandibulectomy	No/3 y
		36/F	Painful parotid swelling/4 mo	Yes	No	SE, partial capsulectomy	Yes/1 y
44-45	Oda et al. (2007)[34†]	52/M	Tightness in his ear/10 mo	Yes	No	SE	No/11 mo
		67/M	Painful swelling of the cheek/3 mo	Yes	Yes	SE	No/7 y 3 mo
46	Cascone et al. (2008)[70]	78/M	Painless preauricular mass/NR	No	No	SE, partial synovectomy	No/2 y
47	Chen et al. (2008)[29]	56/F	Progressive hearing loss, temporal headache/4 mo	Yes	Yes	SE, craniectomy, radiotherapy	No/9 mo
48	Day et al. (2008)[60]	38/M	Painful zygomatic mass, discomfort with mastication/2 mo	Yes	Yes	SE, partial synovectomy	No/3 mo
49	Cai et al. (2009)[51*]	21/F	Painful preauricular swelling, clicking, trismus/3 mo	No	No	SE	No/13 mo
50	Herman et al. (2009)[7]	36/M	Painful preauricular mass/12-18 mo	Yes	Yes	SE, craniectomy	No/11 y
51	Shkoukani et al. (2009)[40]	74/F	Painful preauricular mass, moderate trismus/few mo	Yes	Yes	SE, condylectomy, partial mandibulectomy	No/NR
52	Leiggener et al. (2010)[71]	22/F	Pain, trismus/6 mo	Yes	No	SE, condylectomy	No/3 y
53	Present case (2011)	26/M	Painless preauricular swelling, hearing loss, trismus/ 2 mo	Yes	Yes	SE, synovectomy, middle cranial fossa resection, radiotherapy	No/18 mo

BD, bone destruction; IC, intracranial extension; SE, surgical excision; NR, not reported.
*PVNS associated with synovial chondromatosis.
†PVNS presenting chondroid metaplasia.

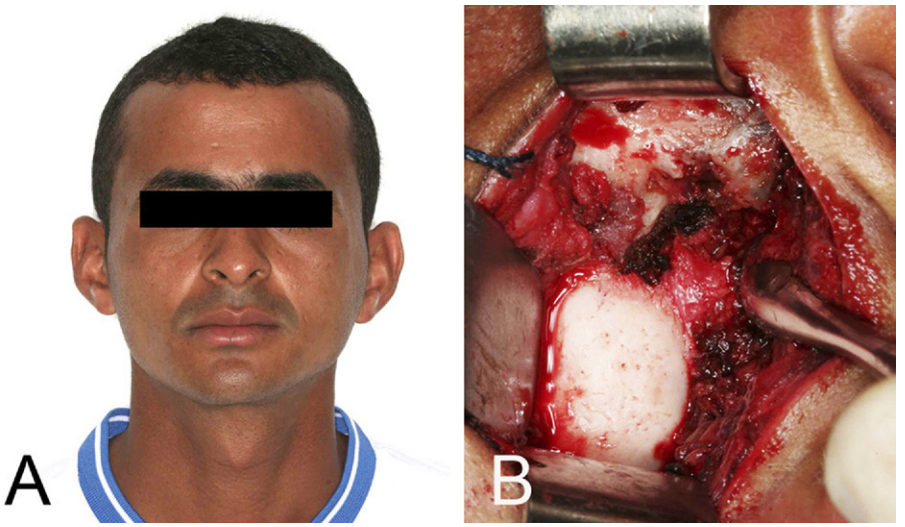

Fig. 1. **A,** Patient with pigmented villonodular synovitis of the temporomandibular joint showing discrete preauricular swelling and a slight asymmetry of the left mandible body and ramus. **B,** Transoperative view showing condylar destruction and presence of fibrous tissue with a spongiotic appearance in its central portion.

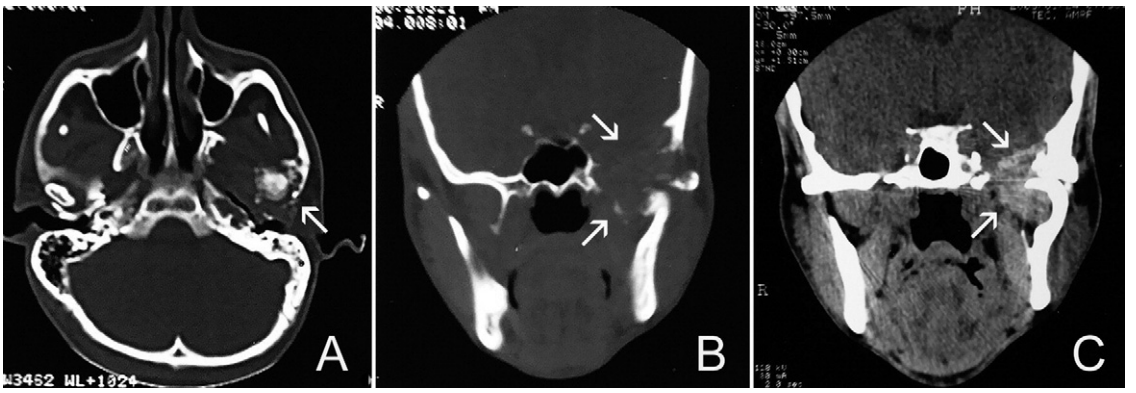

Fig. 2. Computerized tomography. **A,** Partial destruction of the left condyle and a hypodense mass in the left TMJ region (*arrow*, axial cut, bone window). **B,** Involvement of the temporal and sphenoid bones (*arrows*, coronal cut, bone window). **C,** Hyperdense mass causing erosion of the internal surface of the left condyle and the petrous part of the temporal bone, infiltrating the medial cranial fossa (*arrows*, coronal cut, soft tissue window).

ease. A biopsy was performed through a preauricular access, under general anesthesia. The intraoperative view showed partial condylar destruction and a brown-reddish fibrous mass presenting a spongiotic appearance in its central area (Fig. 1, *B*).

Microscopic analysis showed nests of large and small epithelioid mononuclear cells that had infiltrated the cortical bone of the condyle. These nests were involved with a cellular fibrous tissue and also contained a few osteoclast-like multinucleated giant cells, blood vessels, and areas of hemorrhage (Fig. 4, *A*). The mononuclear cells presented displaced ovoid or angle-shaped nuclei and an abundant eosinophilic cytoplasm, with some containing pigmented granules, which were initially interpreted as melanin or hemosiderin (Fig. 4, *F*). The mononuclear epithelioid cells were randomly disposed in a fibrous or hyalinized stroma, forming nodular areas containing an osteoid-like material (Fig. 4, *B-D*). Abundant extracellular pigmented granules were also seen in the stroma (Fig. 4, *E*). The Fontana-Masson stain was negative, but Perl's iron stained the granules deeply, confirming that the brownish extra- and intracytoplasmic granules were hemosiderin (Fig. 5, *A* and *B*). Additionally, immunohistochemistry was performed using antibodies against vimentin (Vim3B4, 1:400; Dako, Glostrup, Denmark), S-100 protein (polyclonal, 1:10.000; Dako), HMB-45 (1:200, Dako), and CD68 (PG-M1, 1:400, Dako). Osteoclast-like multinucleated giant cells and some mononuclear cells showed strong granular cytoplasmic reactivity with CD68 (Fig. 5, *C*), whereas vimentin

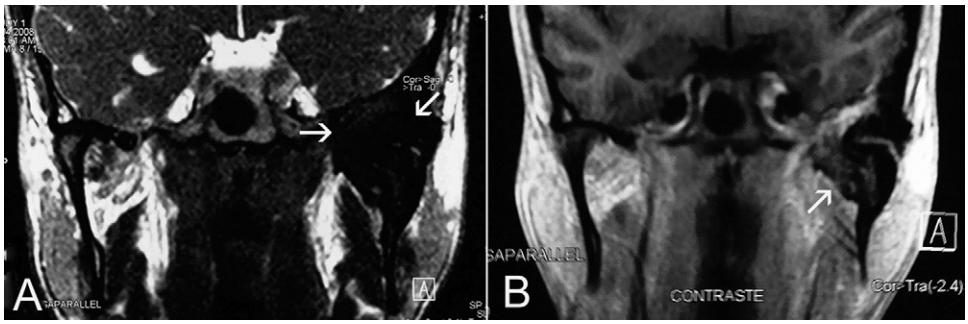

Fig. 3. Magnetic resonance imaging. **A,** Coronal T2-weighting, showing a low-signal lesion compressing the adjacent structures, including dura mater and medial and lateral pterygoid muscles (*arrows*). **B,** The lesion also displays some areas of intermediate signal in close proximity to the adjacent dura mater (*arrow*, coronal T1-weighting with contrast).

was homogeneously positive (Fig. 5, *D*). S-100 protein and HMB-45 were negative. The final diagnosis was PVNS-TMJ.

The patient was treated surgically, but during the procedure infratemporal destruction and infiltration of the dura mater was observed. Because of the diffuse growth pattern, postoperative radiotherapy was carried out using 40 Gy in 20 fractions. In addition to moderate dermatitis and eye irritation, the patient developed postsurgical facial paralysis (Fig. 6). At the time of writing, the patient exhibited preserved visual sharpness, stable and painless mandibular excursion with 30 mm mouth aperture, and improvement of the postoperative paralysis of the frontal and zygomatic branches of the facial nerve. There were no clinical or imaging signs of recurrence after 18 months of follow-up.

DISCUSSION

PVNS is a potentially aggressive proliferative lesion that involves the synovial tissues of the joints, tendon sheath, and bursa.[13] The first report of PVNS is attributed to Chassaignac in 1852, who described it as a nodular lesion of the middle finger.[5] In 1941, Jaffe et al. introduced the term PVNS for lesions showing hemosiderin and villonodular proliferation.[14] They also proposed the term pigmented villonodular bursitis/tenosynovitis for microscopically similar lesions that clearly affected the extra-articular bursa and tendons, respectively.[5,14]

In the current World Health Organization (WHO) classification,[15] the family of giant cell tumors of the tendon sheath (GCTTS) presents subtypes according to their growth pattern (localized or diffuse) and site (intra- or extra-articular). Localized GCTTS involves only part of the synovium, showing pediculated or sessile nodules, which commonly affects the fingers. Diffuse-type GCTTS involves the whole synovial membrane and adjacent structures, preferentially of the knee and hip joints of young patients.[15] Although the current WHO classification uses the term diffuse-type GCTTS as a synonym of PVNS, other authors prefer to consider PVNS as a general designation that includes the localized and diffuse subtypes, both with the possibility of intra- or extra-articular presentation.[5,13,15,16] Even though some authors have also considered the localized and diffuse subtypes as different stages of the same lesion, the distinction is important, because diffuse lesions tend to be more aggressive with a less favorable prognosis.[13,17,18]

PVNS has been associated with reactive, inflammatory, or traumatic origin, involving local hemorrhages and/or local alterations of lipids.[13,18,19] On the other hand, the continuous growth and very aggressive biologic behavior of these lesions resemble a true neoplasia. Trisomy of chromosomes 5 and 7, and a clustering of structural rearrangements in chromosome 1 (1p11-13), where the colony-stimulating factor 1 gene is located, were described in localized and diffuse-type PVNS, suggesting a neoplastic origin.[13,20-22]

To date, 52 cases of PVNS-TMJ have been reported in the English-language literature (Table I), of which 25 were male, 26 female, and 1 in which gender was not reported. Ages have ranged from 10 to 78 years old (mean age 44 years), with the majority of cases occurring in young adult patients in the fourth decade of life (Fig. 7). Clinically, PVNS-TMJ is characterized by preauricular swelling with a slow diffuse growth pattern, progressive pain, and trismus, including mouth opening restriction and discomfort in mastication.[7] A minority of patients have shown impaired hearing, clicking, and tinnitus (Fig. 8). The patient reported here was only 26 years old and presented a shorter duration of symptoms than most reported cases of PVNS-TMJ.

Clinical diagnosis of PVNS is difficult, but CT and MRI proved to be very helpful in the diagnosis and treatment plan, showing extension of the lesion as well as bone destruction.[23,24] Usually, CT of PVNS with

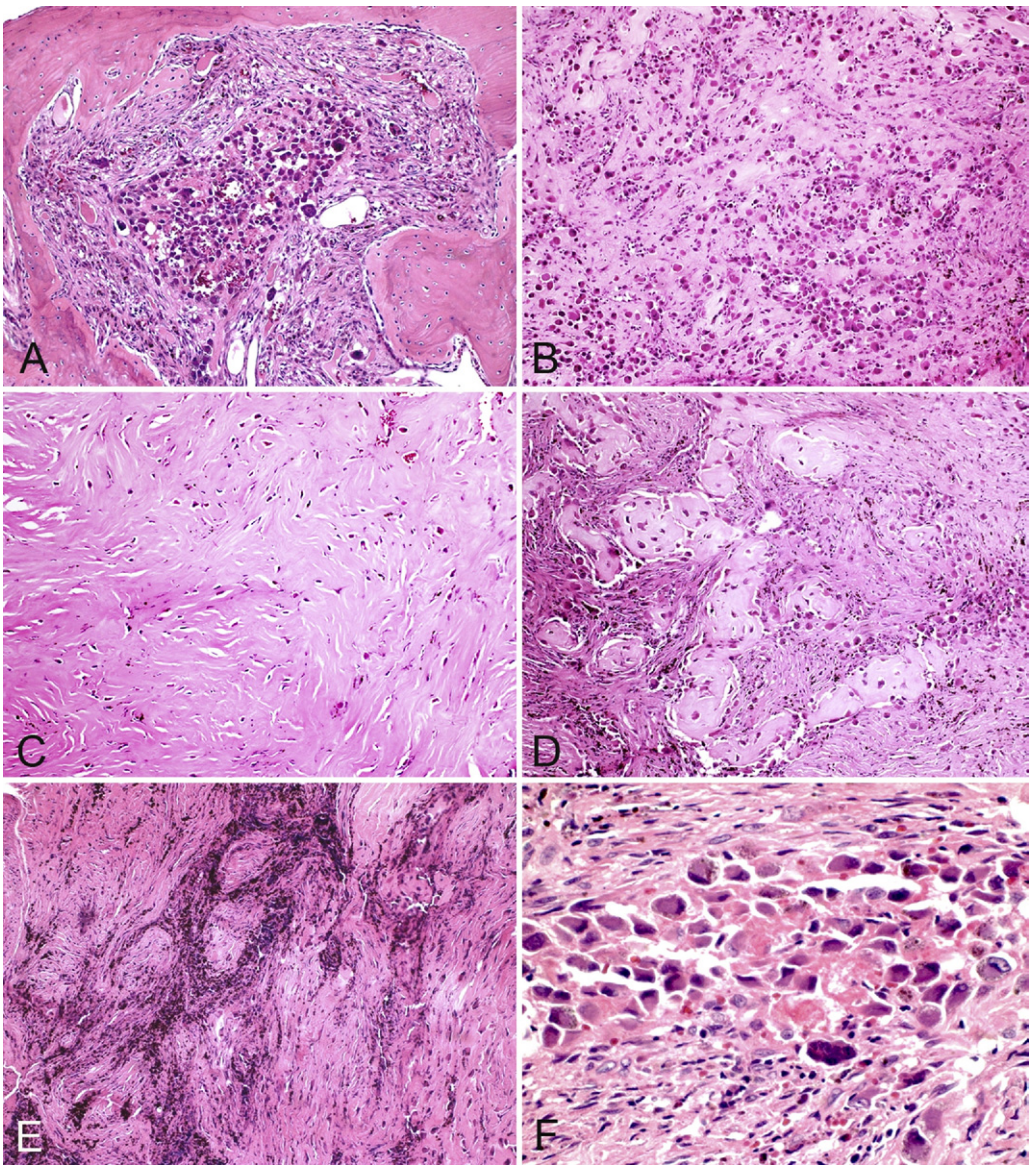

Fig. 4. Histologic aspects of pigmented villonodular synovitis of the temporomandibular joint. **A,** Mononuclear cells with epithelioid appearance and osteoclast-like multinucleated giant cells organized in nests that infiltrate the normal cortical bone (hematoxylin-eosin [HE], original magnification ×100). **B,** Mononuclear cells randomly distributed in the fibrous stroma (HE, ×100). **C,** In some areas the stroma is more fibrous, compressing the mononuclear cells (HE, ×100). **D,** Presence of fibrous nodules mimicking osteoid (HE, ×100). **E,** Diffuse pigmentation of the stroma is easily recognized, a consequence of hemosiderin deposition after hemorrhagic episodes (HE, ×100). **F,** Hemosiderin granules in the cytoplasm of mononuclear cells, which have a characteristic epithelioid appearance (HE, ×400).

high iron content shows a high-density noncalcifying soft tissue mass that might cause erosion or extensive destruction of the condyle, temporal bone, and infratemporal fossa. In some cases, there is an expansive lesional invasion into the middle cranial fossa that compresses but does not infiltrate the dura mater.[6,7,19,25,26] MRI is the best ancillary technique to diagnose PVNS, characteristically presenting a specific finding of low to intermediate signal in both T1- and T2-weighted images.[6,27-30] The MRI low signal intensity areas of PVNS are best seen in T2-weighted images and are attributed to a paramagnetic effect produced by the reaction with iron, owing to the high quantity of hemosiderin.[4,6,13,31] The presence of hemosiderin causing the low signal intensity in MRI is also known as the "blooming effect" and is considered to be pathogno-

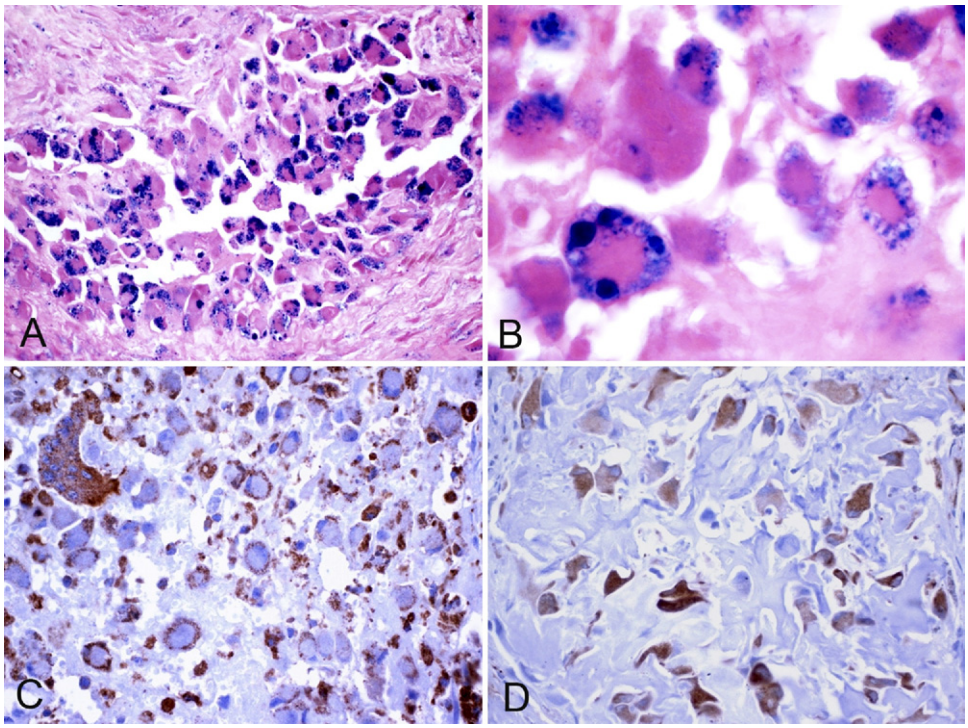

Fig. 5. **A,** Mononuclear cells presenting intracytoplasmic hemosiderin granules (Perl's stain, original magnification ×400). **B,** Hemosiderin granules characteristically disposed at the periphery of the cytoplasm (Perl's stain, ×1,000). **C,** Mononuclear and multinucleated giant cells positive for CD68 (immunohistochemistry, ×400). **D,** Mononuclear cells positive for vimentin (immunohistochemistry, ×400).

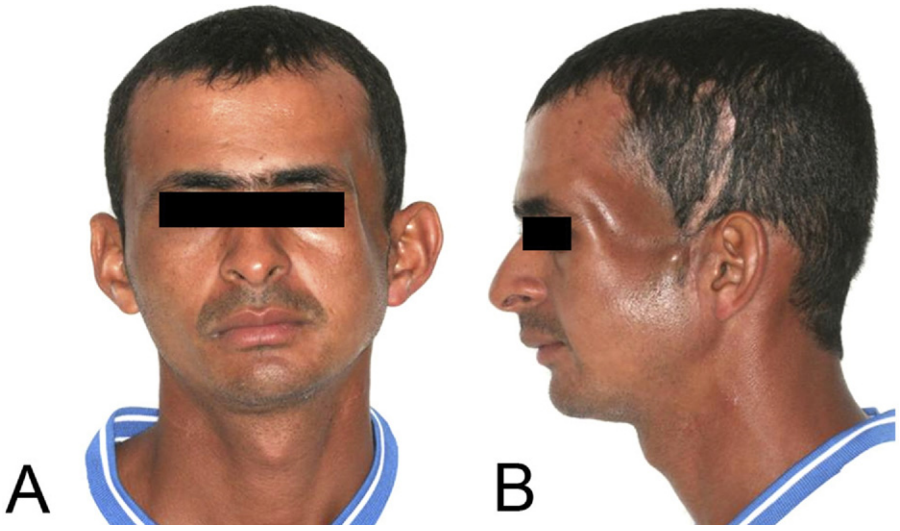

Fig. 6. Clinical aspect of the patient 6 months after surgical excision and adjuvant radiotherapy of pigmented villonodular synovitis of the temporomandibular joint, showing the temporal defect: **A,** frontal view; **B,** side view.

monic of PVNS.[5] Nevertheless, the presence of lipids in xanthomatous macrophages and cystic areas can produce high signal in T1- and T2-weighted images, respectively.[29] Fifty reported cases of PVNS-TMJ described bone destruction, 40 (80%) of them consisted of a diffuse osteolytic lesion, and, interest-

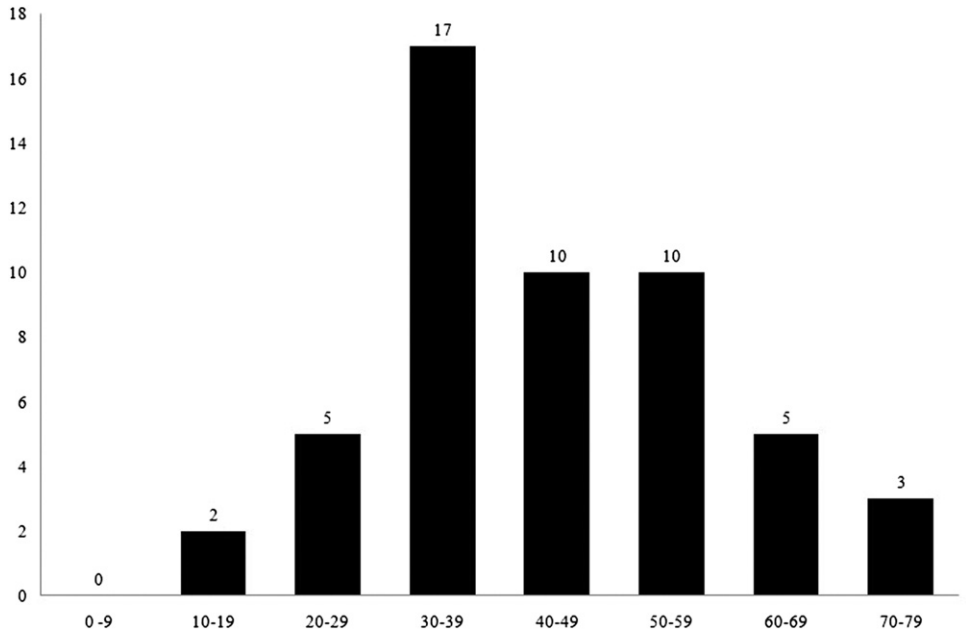

Fig. 7. Age distribution in decades of life of 52 cases of pigmented villonodular synovitis of the temporomandibular joint published in the English-language literature.

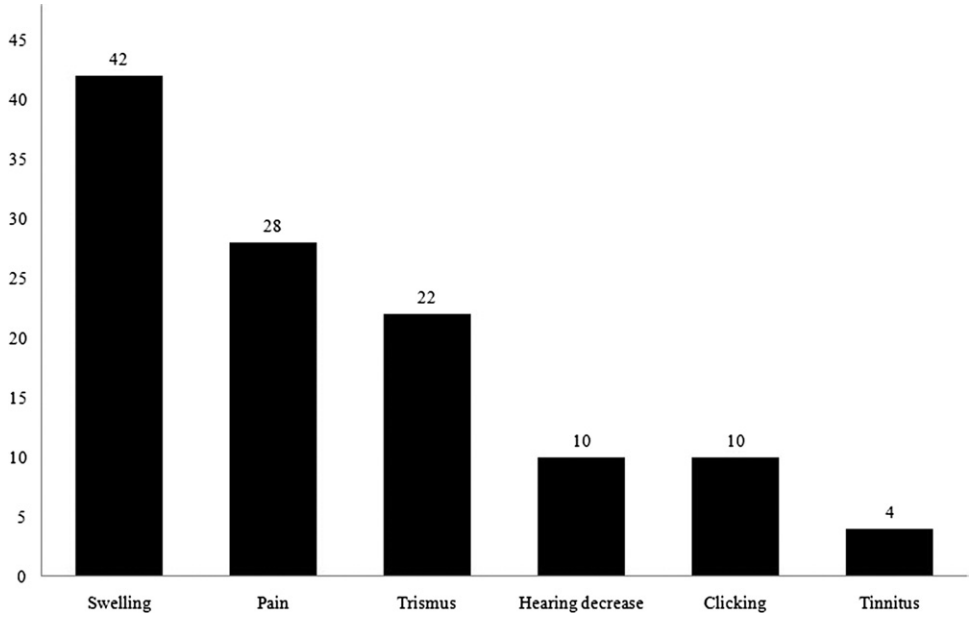

Fig. 8. Prevalence of main clinical features of 48 pigmented villonodular synovitis of the temporomandibular joint published in the English-language literature.

ingly, one-half of these cases exhibited intracranial involvement highlighted by imaging examinations. We also observed the typical imaging findings of PVNS-TMJ, including the intracranial extension and the close proximity with the dura mater.

The main clinical differential diagnosis of PVNS-TMJ includes benign or malignant parotid tumors as well as other lesions that affect the synovium and adjacent structures, including rheumatoid arthritis, tuberculosis, synovial hemangioma, synovial osteochon-

dromatosis, hemophilic arthropathy, amyloid arthropathy, chondroma, osteochondroma, gout, tophaceous pseudogout, chronic osteomyelitis, serous otitis media, central giant cell lesion, sarcomas, and metastasis.[1,28,32-34] Only 31 out of the 52 PVNS-TMJ cases (59%) included a possible differential diagnosis, but in 7 (13%) PVNS was included in the differential diagnosis,[24,35-40] whereas 5 (9%) proposed the diagnosis of malignant or metastatic disease.[31,40-43] The clinical and imaging features prompted us to propose the diagnostic hypothesis of malignant neoplasia or metastasis in the present case. Even though the TMJ region is an uncommon site for malignant neoplasias, preauricular swellings are more commonly malignant, including adenoid cystic carcinoma, mucoepidermoid carcinoma, malignant lymphoma, melanoma, squamous cell carcinoma, chondrosarcoma, synovial sarcoma, osteosarcoma, and fibrosarcoma.[2,26,37] Metastatic tumors affecting the TMJ region are extremely rare, and the most common primary sites are breast and lung, even though prostate, rectum, stomach, uterus, and pancreas have also been reported in the literature.[26,44,45]

Fine needle aspiration cytology (FNAC) is an important approach to discard the diagnosis of a malignant neoplasia and can in fact help in the diagnosis of PVNS.[46] Only 8 PVNS-TMJ cases (15%) were reported using FNAC for diagnostic purposes, which on the cytopathologic examination showed mononuclear cells and multinucleated giant cells containing hemosiderin pigments, similarly to other lesions such as central giant cell lesion and chondroblastoma.[24,25,36,38,40,46-49] FNAC was not performed in the present case.

Macroscopically, PVNS shows increased thickness of the synovium, a nodular growth pattern with villous formation, and a rusty brownish color due to hemosiderin deposition.[29,31,41] As in the present case, the loss of the typical villous feature in PVNS-TMJ is common, probably owing to its predominantly diffuse growth pattern.[15]

Microscopically, PVNS presents synovial hyperplasia with villous projections covered by multiple layers of synovial cells. The cellular types include large and small mononuclear cells, osteoclast-like multinucleated giant cells and xanthomatous macrophages, which are found in a vascularized fibrous stroma showing fusiform to polygonal stromal cells and intra- and extracytoplasmic deposition of hemosiderin.[8,33,34] The fibrous stroma can also show cleft-like spaces and hyaline nodular formation mimicking osteoid material, which are more evident in extra-articular areas.[8] Mononuclear cells are distributed in nests or randomly, displaying hemosiderin granules, which might be shown by staining methods for iron, such as Perl's Prussian blue.[8,24,30,31,34] Although mitoses and cellular atypia are absent, mononuclear cells might predominate, suggesting a sarcoma, especially in cases with scarce osteoclast-like multinucleated giant cells.[31] In fact, our first microscopic impression was of a malignant neoplasm, most likely an osteosarcoma or a melanoma with epithelioid features. Nodular areas showed osteoid-like formation that at first glance could be malignant; on the other hand, the pigmented cells could be confused with malignant melanocytes. Melanoma was ruled out because the pigment was positive for iron and cells were negative for S-100 and HMB-45. Immunohistochemistry is not essential to diagnose PVNS, but in certain cases it helps to rule out other possible diagnoses.

It is interesting to mention reports of PVNS-TMJ cases showing simultaneous occurrence with synovial chondromatosis[43,50,51] and chondroid metaplasia.[34] There are also reports of PVNS showing features of malignancy,[52-54] but in the present case, anaplasia, mitotic figures, and necrotic areas were not detected.

The standard treatment for PVNS is surgery, with partial or complete excision of the synovium and involved bones.[18] The surgical procedure of knee lesions is usually done by arthroscopy. Some aggressive diffuse PVNS cases have been treated by surgery followed by low doses of radiotherapy.[55] Recurrences occur when surgical excision is incomplete and there is diffuse involvement of the adjacent structures. In fact, radiotherapy has been suggested for these recurrent cases as well as for extensive cases which probably could recur.[33,56,57] Adjuvant radiotherapy is administered ~2 months after surgery, in 15-25 fractions with doses ranging from 20 to 50 Gy.[5,55,57] Even though the efficacy of bisphosphonates has not been reported in any case of PVNS-TMJ so far, Namazi[58] considered bisphosphonate as a novel possibility for PVNS treatment because of its property of osteoprotegerin stimulation, which can lead to a reduction of receptor activator of nuclear factor kappa B ligand and osteoclast recruitment. Facial paralysis has been described as a common postsurgical complication,[39,59] but facial nerve function can be completely recovered a few months after surgery,[59] as seen in the present case. Follow-up should be made for 5-10 years after the surgical procedure, including periodic CT and MRI examinations.

All reported PVNS-TMJ cases have been treated by surgical excision, and in 2 cases adjuvant radiotherapy was performed.[29,56] As noted above, PVNS-TMJ can invade the cranial base and compress the dura mater, making necessary the participation of a neurosurgeon on the surgical team.[7,29,38,60] In only 2 PVNS-TMJ cases was adjuvant radiotherapy used, one for an unre-

sectable lesion[56] and another for an extensive lesion involving vital structures.[29] Recurrence data were available in 36 PVNS-TMJ cases, and only in 3 (8%) of them was recurrence reported.[33,50,56] The median follow-up period was 2 years 9 months (range 3 months to 16 years). In the present case, the association of surgery and low doses of radiotherapy has shown good results so far.

In summary, PVNS is an uncommon proliferative benign lesion of the TMJ with aggressive potential and should be included in the differential diagnosis of preauricular swellings, especially when accompanied by mouth aperture and hearing difficulties. CT and MRI are helpful for diagnosis, treatment, and follow-up. Clinicians and oral pathologists should keep in mind the possibility of PVNS when aggressive lesions involve the TMJ, causing preauricular swelling.

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