Case Report
TMJ Disorders

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

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Abstract. Pigmented villonodular synovitis (PVNS) is an uncommon benign proliferative disorder of synovium that may involve joints, tendon sheaths, and bursae. It most often affects the knees, and less frequently involves other joints. It presents in the temporomandibular joints (TMJs) extremely rarely. The authors report an elderly female patient with PVNS of the TMJ with skull base extension, who had traumatic history in the same site. It was diagnosed through core-needle biopsy, which was not documented in the literature. Radical excision and follow-up for 7–8 years was recommended because of the reported malignant transformation and high recurrence rate. This case and previously reported cases in the literature are reviewed and discussed.

Keywords: pigmented villonodular synovitis (PVNS); synovitis; temporomandibular joint (TMJ).

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The term pigmented villonodular synovitis (PVNS) was introduced by Jaffe et al. in 1941 to describe a proliferative disorder of unknown aetiology involving diarthrodial joints, tendon sheaths and bursae[22]. It's an uncommon disorder, which has an annual incidence of 1.8 cases per million population[30]. It most often affects the knees, including about 80% of cases[14,30]. Other joints that may be affected by PVNS, in decreasing order of frequency, include the hips, ankles, small joints of the hands and feet, shoulders and elbows. Involvement of vertebral articular joints has also been documented[14]. It presents extremely rarely in the temporomandibular joints (TMJs). The first reported occurrence was in 1973 by Lapayowker et al.[25]. To the authors' knowledge, only 37 cases of PVNS affecting the TMJ have been reported in the English literature and 1 case in the Chinese literature; 34 of these were reported in detail (Table 1). The authors present an additional case of PVNS affecting the TMJ, and review previous cases.

Case report

A 59-year-old Chinese female was referred in February 2010 with the chief complaint of a left preauricular mass that she had noticed for about 2 years. She reported a history of left TMJ dislocation during yawning about 5 years ago, which she reduced by hand by herself immediately. Although it did not recur, she occasionally felt preauricular pain. About 2 years ago, a pea-sized preauricular mass with pain was noticed, and the symptoms disappeared after using anti-inflammatory drug. About 1.5 years ago, she underwent root canal treatment and crown restoration for the left first molar, that required several visits and mouth-opening for a long time during the operation. Left preauricular swelling and pain occurred after the dental treatment, with progressive diminution of hearing in the same ear. The swelling gradually increased in size, but the pain reduced after using anti-inflammatory drug. The patient had no complaint of mouth opening restrict or biting pain. Physical examination revealed a preauricular mass measuring approximately 3.5 cm × 3.5 cm, spherical, firm, immovable, nontender on palpation, with a somewhat detectable boundary, localized over the left TMJ region (Fig. 1). Maximum mouth opening was 3.7 cm. There was slight deviation to the left when the mouth was opened, with slight deviation to the right when the mandible was moved forward, with a notable decrease in left lateral excursion. The occlusion was normal. The patient's medical history was

Table 1. Summary of reported cases of PVNS involving the TMJ.

A. Clinical features and preoperative diagnosis

No.	Authors/date	Age (y);sex;site	Clinical features	Duration	Bone destruction	Preoperative diagnosis
1	Lapayowker et al. 1973[25]	22;M;L	PS, masticatory pain	18m	TMJ	Malignant PT
2	Lapayowker et al. 1973[25] Dinerman and Myers 1977[13]	58;F;L	Clicking; tinnitus; hearing diminution	1y	TMJ, ramus, SBICE, EAC	Biopsy: fibroadipose tissue Open biopsy: PVNS
3	Barnard 1975[2]	37;M;L	PS, pain, LMO, MD to L	3w	TMJ	Infection arthritis or neoplasia
4	Miyamoto et al. 1977[28]	34;M;R	PS	1.5y	TMJ	PT
5	Takagi and Ishikawa 1981[42]	36;M;R	Oppressive pain, clicking, LMO	3y	TMJ	NR
6	Rickert and Shapiro 1982[33]	39;F;L	Parotid mass	1m	TMJ	PT; FNA: giant cell granuloma, 'brown tumour', neoplasm
7	Gallia et al. 1982[18] Curtin et al. 1983[10]	47;F;L	PS, LMO, masticatory pain, MD to L, otalgia	2y	TMJ	PVNS
8	O'Sullivan et al. 1984[31] Bertoni et al. 1997[4]	61;F;L	Swelling in parotid area	NR	TMJ, coronoid process, ramus, zygomatic arch, EAC, SBICE	Benigh PT
9	Dawiskiba et al. 1989[11]	32;M;R	TMJ swelling, pain, LMO	NR	No	FNA: PVNS
10	Eisig et al. 1992[15]	50;F;R	HL, mass in ear canal	2m	TMJ, SBICE, EAC, mastoid, middle ear ossicles	Central giant cell granuloma
11	Syed et al. 1993[41]	10;F;L	PS	1y	No	PT
12	Franchi et al. 1994[17]	59;F;L	Swelling in parotid area, pain	6m	No	PT.
13	Youssef et al. 1994[46]	41;F;L	TMJ swelling, pain, LMO, decreased lateral excursion to L	6m	No	Incisional biopsy: PVNS
14	Tanaka et al. 1997[43]	47;M;R	PS, masticatory pain, LMO, MD to R	8m	TMJ	Giant cell-containing lesion of TMJ
15	Renaga et al. 1997[32]	70;F;L	TMJ swelling, intermittent pain, LMO, clicking	7m	TMJ	PVNS
16	Yu et al. 1997[47]	48;M;R	PS, pain when wide mouth opening	7m	TMJ	PT
17	Chow et al. 1998[8]	42;F;R	Zygomatic mass	6m	TMJ	Giant cell tumour, chondroblastoma
18	Song et al. 1999[38]	57;F;R	TMJ swelling	5y	TMJ	PT
19	Bemporad et al. 1999[3] Stryjakowska et al. 2005[40]	37;M;R	PS, pain, trismus, tinnitus, HL	4y	TMJ, SBICE, maxilla, maxillary sinus, carotid canal, EAC	PT; Incisional biopsy: 'brown tumour'; FNA: PVNS
20	Lee et al. 2000[26]	59;F;R	PS, decreased salivary flow	6y	TMJ	PT; Punch biopsy: PVNS
21	Kisnisci et al. 2001[24]	45;F;L	Cheek swelling, pain, LMO, MD to L	NR	No	NR
22	Shapiro et al. 2002[34]	36;M;R	Temporal mass, clicking, some HL, popping sensation in ear	NR	SBICE	Malignant process; CT guided FNA: benign giant cell-containing lesion
23	Church et al. 2003[9]	42;M;R	PS, clicking, HL, mass nearly occluded EAC	4m	TMJ, zygomatic arch, SBICE, EAC	Biopsy: PVNS
24	Church et al. 2003[9]	33;M;R	PS, deep pain, trismus	2y	TMJ	FNA: PVNS
25	Tosun et al. 2004[44]	60;M;L	TMJ mass, V2/V3 numbness/paresthesia, LMO, MD to L, HL, otitis media	10y	TMJ, ramus, SBICE, carotid canal, foramen ovale, EAC, sphenoid bone	Otitis, PT; FNA: PVNS
26	Aoyama et al. 2004[1]	33;M;R	PS, clicking > 20y, LMO	NR	TMJ, skull base	Malignant tumour, giant cell tumour; Biopsy: PVNS
27	Cascone et al. 2005[7]	38;F;L	PS, TMJ pain	NR	No	FNA: non-specific arthritis

Table 1 (*Continued*)

A. Clinical features and preoperative diagnosis

No.	Authors/date	Age (y);sex;site	Clinical features	Duration	Bone destruction	Preoperative diagnosis
28	Stryjakowska et al. 2005[40]	36;F;R	Painful parotid lesion, radiating pain	4m	No	PT
29	Fang et al. 2007[16]	44;M;R	PS	2m	TMJ, zygomatic arch	Pleomorphic adenoma; FNA: PT
30	Cascone et al. 2008[6]	78;M;R	PS, orofacial pain, LMO, MD to R	NR	No	NR
31	Day et al. 2008[12]	38;M;L	Mass in zygomatic process area, pain	1m	SBICE, foramen ovale, vidian canal, foramen rotundum	Cholesterol cyst, vascular bone tumour
32	Shkoukani et al. 2009[36]	74;F;R	PS, pain, LMO	Few m	TMJ	CT-guided FNA: PVNS
33	Cai et al. 2009[5]	21;F;L	TMJ pain, LMO, clicking, MD	3m	No	Anterior disc displacement without reduction
34	Herman et al. 2009[21]	36;M;L	PS, pain, numbness, LMO, MD to L, otalgia	12–18m	Zygomatic arch, glenoid fossa, SBICE	Biopsy: PVNS

B. Treatment and prognosis

No.	Treatment	Complications and prognosis	Rec. (FU-period) [Rec. time]
1	CE	Uneventful	No
2	CE, Condy., partial Parot., craniectomy, reconstructed with free graft of fascia temporalis	NR	No (4y)
3	Incisional biopsy	Some persistent MD to L	NR (6m)
4	CE	Temporary tinnitus, EAC narrowing	No (2y)
5	1st–2nd: CE, Condy.	Uneventful	Yes [5y]; no Rec. (11y)
6	Amputated tumour from TMJ, superficial Parot.	Temporary FP, limited jaw motion	No (3m)
7	CE	Temporary FP, occlusal discrepancy	No (2y)
8	1st–3rd: mass excision, Parot., Mand., craniectomy; 4st: RT; 5th: surgery	Dead 20d after last operation with malignant transformation, intracranial extension and lung metastasis	Yes [1.5y, 2.5y, 2y, 4y]
9	Surgery (not specified)	NR	NR
10	CE, craniectomy, Condy., radical mastoidectomy, repaired dura with temporal fascia graft	Minimal MD	No (1y)
11	CE, superficial Parot.	Temporary FP	NR (8w)
12	CE	NR	No (1y)
13	CE, superficial Parot.	Uneventful	No (14m)
14	CE, craniectomy, Condy.	Temporary FP	No (2y)
15	CE	Uneventful	No (3y)
16	1st: superficial Parot.; 2st: surgical exploration	NR	NR
17	CE, Condy.	Uneventful	No (2y)
18	CE, total Parot., Condy.	NR	NR
19	Embolization, intradural resection, Mand., total Parot., facial nerve grafting, suprahyoid neck dissection, repaired with pericranial graft and rectus myocutaneous free pedicle flap	FP	No (3y)

20	Remove mass and involved facial nerve, sural nerve grafting, Condy., reconstructed with costochondral graft	Minimal MD; FP	No (2y)
21	CE	Uneventful	No (1y)
22	Embolizaiton, CE, craniotomy	Uneventful	No (7y)
23	Wide excision, partial Mand, craniectomy	Uneventful	No (3y)
24	Wide excision	Uneventful	No (2y)
25	CE, Condy., temporal craniotomy, partial Parot.	MD to R	No (6y)
26	CE, Condy., craniotomy, covered dura by temporal muscle pedicle flap	Preauricular concavity; temporary FP; MD to R	No (2y)
27	CE, superficial Parot.	Uneventful	No (1y)
28	1st–2nd: Parot., CE	Uneventful	Yes [15m]; no Rec. (1y)
29	CE	NR	No (20m)
30	CE	Uneventful	No (2y)
31	CE, repaired temporal bone defect and tegmen tympani with titanium mesh	Temporary HL	NR (3m)
32	CE, tracheostomy tube placement, superficial Parot., Condy., partial Mand.	Temporary FP; MD	No
33	Removed lesions with arthroscopy	Uneventful	No (13m)
34	CE, craniectomy; repaired with cranial bone, lateral pterygoid and temporalis muscles	Uneventful	No (11y)

Abbreviations: F, female; M, male; L, left; R, right; y, year(s); m, month(s); w, week(s); PS, preauricular swelling or mass; LMO, Limitation of mouth opening; MD, mandibular deviation; HL, hearing loss; EAC, external auditory canal; FNA, fine-needle aspiration; NR, not reported; SBICE, skull base with intracranial extension; PT, parotid tumour; CE, complete excision; Condy., condylectomy; Parot., parotidectomy; Mand., mandibulectomy; RT, radiation therapy; FP, facial paralysis; FU, follow-up; Rec., recurrence.

significant for hypertension, diabetes mellitus, hepatitis C and appendectomy.

Computed tomograms (CT) showed a large mass with heterogeneous soft-tissue density in the left infratemporal fossa with the condyle as a centre. Bony destruction of the mandibular condyle, temporal skull base, the root of zygomatic arch and the anterior wall of external auditory meatus was noted (Fig. 2A and B). Magnetic resonance imaging (MRI) showed a 3.8 cm × 3.5 cm × 3.5 cm nodular mass in the region of the TMJ, presented as an intermediate heterogeneous signal on T1-weighted SE sequences (Fig. 2C) and low signal on T2-weighted TSE sequences (Fig. 2D). The mass extended superiorly into the middle cranial fossa. The smooth margin between the mass and the temporal lobe suggested that the lesion was intracranial but extradural.

The patient underwent ultrasound-guided core-needle biopsy with an 18 G cutting needle. A histopathological diagnosis of PVNS was made (Fig. 3).

In March 2010, the patient underwent complete excision of the mass through a temporofrontal approach by a multidisciplinary team, consisting of members of the Maxillofacial Surgery, TMJ, and Neurosurgery Departments. A coronal incision was made with preauricular and frontal extension (Fig. 1B). The frontotemporal scalp flap was elevated. The temporal branches of the facial nerve were dissected and protected. The parotid gland was retracted, and the mass was exposed in the infratemporal fossa, with erosion to the inferior aspect of the left temporal bone (Fig. 1C). The root of the zygomatic arch was resected to expose the mass, then temporal craniectomy and condylectomy was performed to resect the mass completely.

The middle cranial fossa floor, the glenoid fossa, and the portion of zygomatic arch involved were resected. The dura mater was intact, and the cranial bone defect was covered by a temporal muscle pedicle flap rotated inferiorly. Grossly, the mass was red-brown with pliable texture, shaggy, not encapsulated, with a red-brown and yellow-brown cut surface and firmly adhering to the TMJ capsule (Fig. 1D). The final diagnosis, based on histopathological examination, was PVNS.

The patient presented with hearing loss in the left ear, facial expression weakness in the region of the upper division of the facial nerve, slight deviation of the mandible to the left immediately after the operation. The occlusion was normal, and the maximum mouth opening was 23 mm when she was discharged from the hospi-

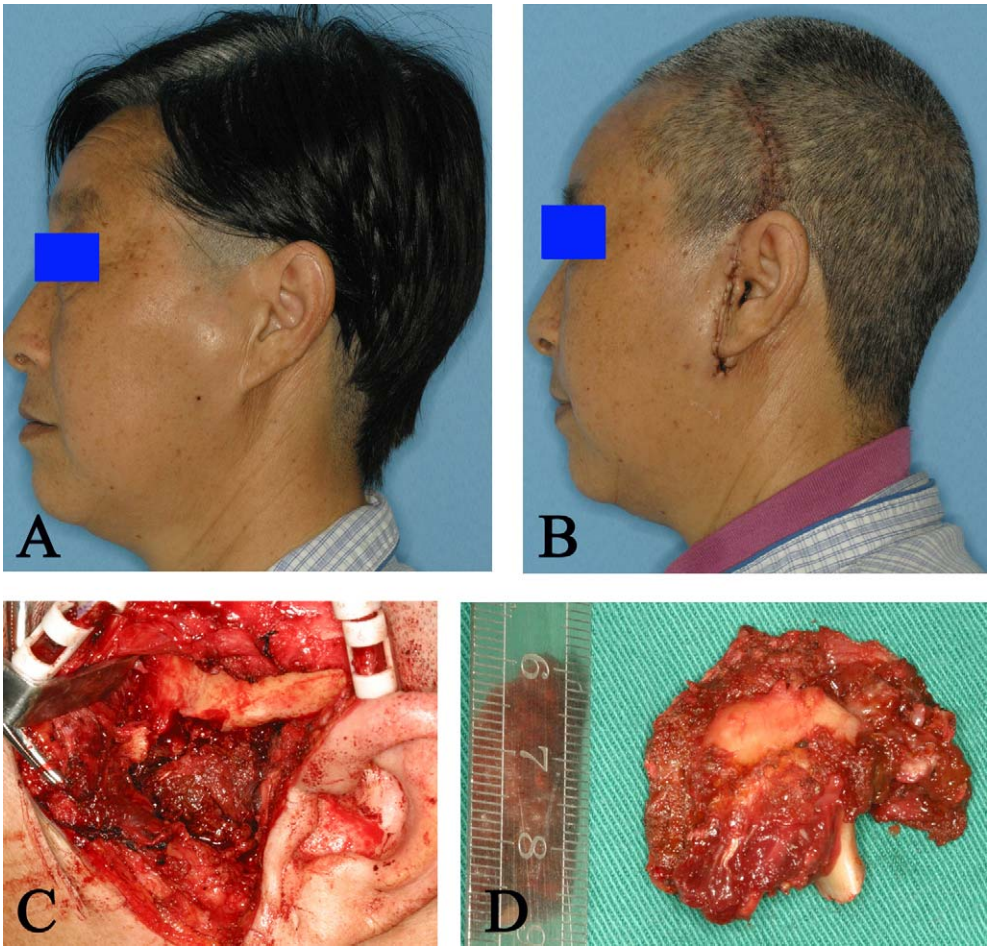

Fig. 1. (A) Before operation; (B) 10 days postoperation; (C) the mass exposed during operation; (D) the mass resected.

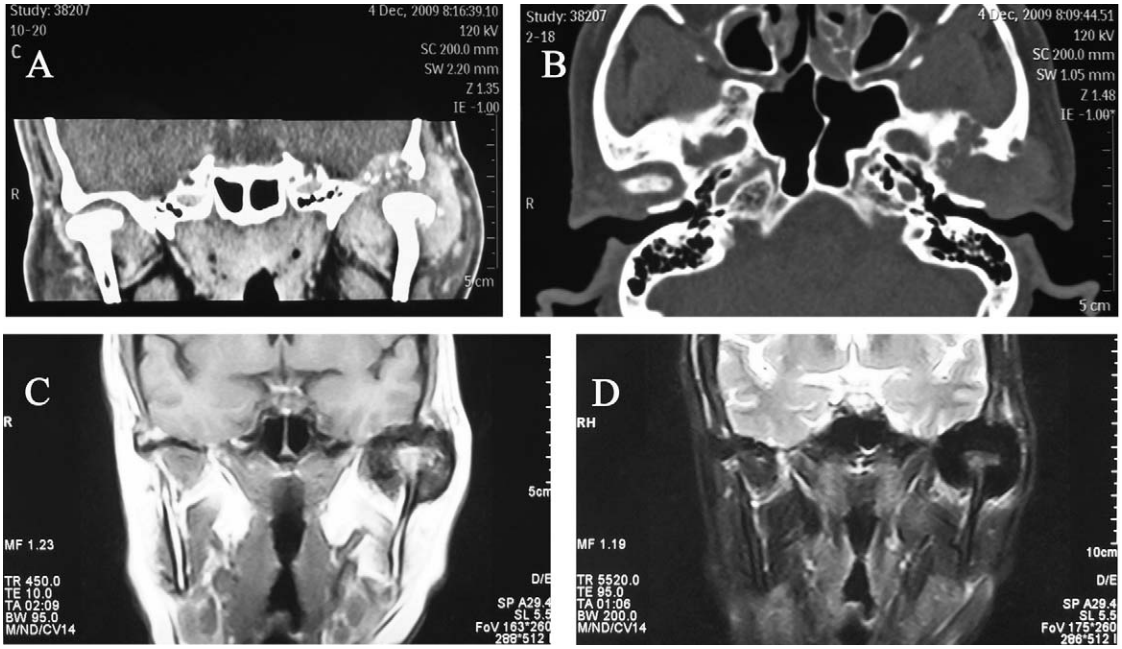

Fig. 2. Preoperative CT: soft-tissue window coronal section (A) and bone window axial section (B). Preoperative coronal MRI: T1-weighted SE sequences (C) and T2-weighted TSE sequences (D).

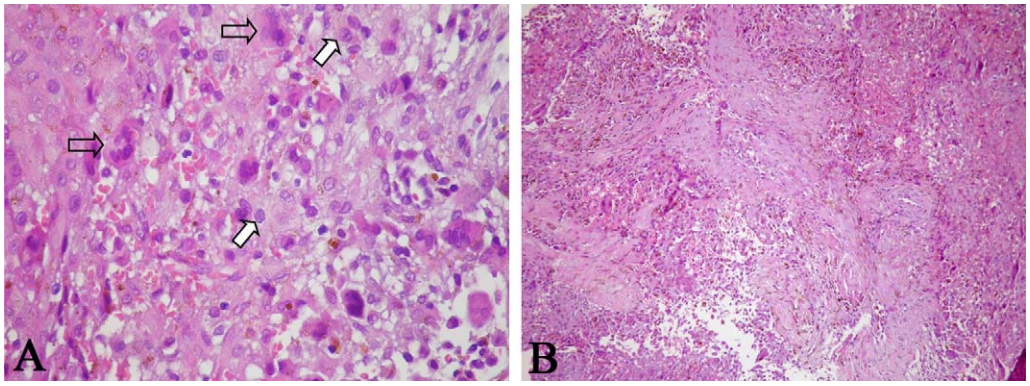

Fig. 3. Photomicrograph of a pathology specimen (haematoxylin–eosin stain). (A) High-power: showing mononuclear histiocytic cells (white arrows), multinucleated giant cells (hollow arrows), foam cells, deposits of hemosiderin, and abundant capillary vessels. (B) Low-power: showing histocytic cell infiltration into the synovial layers.

tal. Following discharged, the patient underwent a course of physical therapy including a home exercise for facial function and maintenance of occlusion with a prosthetic appliance. Ten months after surgery, the patient was followed up by telephone. No facial paralysis, mandibular deviation, malocclusion or preauricular concavity were noted. The maximum mouth opening was 43 mm. Hearing was improved, approaching normal. The coronal scar was invisible and covered by hair. As the disorder often recurs, long-term close follow-up was required.

Discussion

PVNS is an uncommon benign proliferative disorder of the synovium that may involve the joints, tendon sheaths, and bursae. Two forms of PVNS are identified depending on the area involved: the diffuse form that affects the entire synovial membrane of a joint or bursa; and the localized form that affects tendon sheaths or only part of the joint lining.

The cause and pathogenesis remain controversial but several possibilities have been suggested, including disturbances of lipid metabolism, neoplasm, inflammation, haemorrhage, and trauma[19,21,38]. Prior to 1941, the diseases resembling villonodular synovitis were referred to as benign synovioma, giant cell tumour of joints, xanthoma of joints, and hemorrhagic villous synovitis[27]. In 1941, Jaffe et al. recognized the similar histologic pattern of these disorders and termed them pigmented villonodular synovitis, which has gained general acceptance[22]. He considered it to be a benign inflammatory state of the synovium of uncertain aetiology[22]. In 1954, Young and Hudacek reproduced PVNS-like lesion in dogs by repeated haemorrhage into the joint, and considered that repeated haemorrhage into the joint was essential for the development of this lesion and that trauma was often the initiating factor[45]. In 1966, Mccollum et al. repeated the experiment by intra-articular injection of blood and saline solution in dogs, suggesting that PVNS might be produced by repeated trauma to the joint[27]. They found that the presence of blood was not necessary for the development of PVNS, but that the lesion could subside after a 6-month rest period[27]. In 1969, Singh et al. repeated the experiment using iron-dextrin, blood, plasma, gum acacia, and saline in rhesus monkeys with less frequency and volume, but only the iron-dextrin and blood group presented a PVNS-like lesion, suggesting that the iron content of the inoculum was the important factor in eliciting the changes[37]. More than 80% of PVNS cases involve the knees, upper extremities are seldom involved, suggesting a predilection for weight bearing joints[14,30]. Myers and Masi reported that 88% of localized PVNS and 59% of diffused PVNS had a positive traumatic history, and all the trauma-positive localized cases and 38% of the diffused cases had suffered from acute trauma[30]. Bloody fluid was obtained in most of the cases undergoing synovial fluid aspiration, and yellow fluid was obtained in few cases[30]. In the present case there was an acute traumatic history (TMJ dislocation) about 5 years ago and chronic traumatic history (dental treatment) about 1.5 years ago, that might be associated with PVNS. The symptoms could be relieved after using anti-inflammatory drugs, suggesting that inflammation might occur in the protophase and/or in the process of PVNS. Of the 34 cases in Table 1, only 4 referred to a traumatic history, but none was positive[1,2,5,17]. None of the case reports referred to the effect of using anti-inflammatory drugs. The traumatic history and anti-inflammatory drug use was often neglected when the lesion was considered a type of tumour. The history relating to chronic trauma, such as dental treatment, oral surgery, TMJ treatment, dislocation of TMJ, bruxism and malocclusion, should be recorded as well as acute trauma. Despite that, the malignant transformation and metastases of PVNS[4] might support the theory of neoplasm. To date, the aetiology of the entity remains elusive and controversial.

The 34 cases reported in the literature are summarized in Table 1 and Fig. 4. All the cases presented monoarticular lesion, without predilection for sex or site. The age of the patients ranged from 10 to 78 years; the average was 44.1 years. The duration of initial symptom ranged from 3 weeks to 10 years, and the average was about 19.9 months. Eighty-eight per cent (30/34) of cases reported mass or swelling, 57% (17/30) of which were in the preauricular area, which might be confused with parotid tumour. Masses in the ear canal were found in two cases on physical examination. The masses were immovable, firm, nontender or mildly tender on palpation in most cases. Fifty-six per cent (19/34) of cases presented pain, that could be oppressive, masticatory, intermittent, progressive, radiating or deep pain.

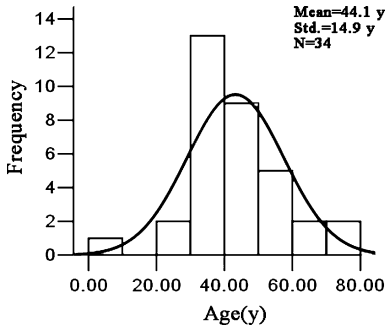

Fig. 4. Age distribution of the 34 cases.

Fifty-three per cent (18/34) of cases presented symptoms of the TMJ that mimic temporomandibular disorders. Twenty-seven per cent (9/34) of the cases presented auricular symptoms, including hearing loss, otalgia, tinnitus, a popping sensation in the ear, and otitis media. Two cases presented with numbness and/or paresthesia of the face[21,44] and in one case decreased salivary flow was noted on physical examination.

Owing to its aggressive infiltrative behaviour, 74% (25/34) of the lesions were associated with bony destruction in images and/or on intraoperative views, that might cause a diagnostic dilemma. Sixty-eight per cent (23/34) of cases eroded the TMJ. The surrounding structures including skull base (10), auditory organs (6), zygomatic arch (4), mandibular ramus (3), coronoid process (1), maxilla (1) and/or sphenoid bone (1) were destroyed. The lesions extended into the crania in eight cases, which resulted in difficulty in completely excising the mass. These lesions were of soft tissue origin, so early changes might not be detectable on plain film radiography. CT and MRI could reveal the borders and extension of the lesion, as well as local bony invasion. The most sensitive and specific method in the diagnosis of PVNS is MRI, which is the optimum radiographic method for judging the extent of the tumour before operation and recurrence afterwards. The most characteristic finding is nodular intra-articular masses of low signal intensity on T1-, T2-, and proton density-weighted sequences, that are considered to be due to a paramagnetic effect attributed to heavy hemosiderin pigmentation[23]. The appearance of PVNS on MRI is variable, depending on the relative proportion of the lipid, hemosiderin, fibrous stroma, pannus, fluid, and cellular elements[23]. Areas of high signal intensity on T2-weighted images correspond to a loculated cyst of joint fluid[3,12,23], and the accumulation of lipid in foamy macrophages may give rise to areas of high signal intensity on T1-weighted SE images, similar to subcutaneous fat[23]. CT scans are useful for determining the extent of bony destruction that often occurs.

The differential diagnoses of PVNS with tuberculosis septic arthritis, malignant synovioma, haemophilia, rheumatoid arthritis, gout, synovial chondromatosis, carcinoma of the external or middle ear, cartilagenous disruption or abnormality have been documented in areas other than the TMJ[25]. In Table 1A, 38% (13/34) of the cases had been diagnosed as parotid tumour because of the site of the mass. Other temporary diagnoses included giant cell-containing lesion (4), arthritis (2), neoplasia (2), giant cell granuloma (2), malignant tumour (2), 'brown tumour' (2), chondroblastoma (1), otitis media (1), cholesterol cyst or vascular bone tumour (1) or anterior disc displacement without reduction of TMJ (1). Only 32% (12/14) of the cases had been diagnosed as PVNS before operation. Although MRI is a sensitive and specific method for the diagnosis of PVNS, histopathology is definitive. Fine-needle aspiration (FNA) and biopsy are helpful for the diagnosis of PVNS[1,3,7,9,11,21,26,36,44,46], however misdiagnoses might occur if the specimen could not represent the lesion[3,7,16,21], especially FNA, which is not a histopathologic examination, but a cytological examination. In Table 1A, only 56% of FNA and 75% of biopsies diagnosed PVNS before a definitive operation, so multiple FNA and/or biopsy might be necessary. CT-guided FNA was carried out in two cases[1,36], but none of the 34 cases had used ultrasound-guided core-needle biopsy, which is micro-traumtic, easy to operate and locate, and sufficient for histopathological diagnosis. There were two cases with simultaneous PVNS and synovial chondromatosis of the TMJ, which made the diagnosis more elusive.

The typical pathologic features of PVNS are indicated by the name[14,22,29]. Grossly, the synovium has a mossy or nodular texture, spongy cut surfaces, and a rusty, red-brown, or yellow-brown colour. Microscopically, the synovium is composed of fingerlike or rounded masses of fibrous stroma covered by hyperplastic lining cells. Large numbers of foamy macrophages in the stroma account for the yellow colouration, whilst the rusty or brown colour is due to hemosiderin deposits in the stroma, cytoplasm of macrophages and synovial lining cells. The hemosiderin is a breakdown product of haemoglobin, the presence of which signifies old haemorrhage. Multinucleate giant cells varying in number, are derived from macrophages and synovial lining cells, and may also contain hemosiderin. Mitotic figures with a normal configuration are easily found in the proliferating fibroblasts, macrophages, and synovial lining cells. If malignant PVNS develops, it would present the following histological characteristics[4]: a nodular, solid infiltrative growth pattern of the lesion; large, plump, round or oval cells with deep eosinophilic cytoplasm and indistinct borders; large nuclei and prominent nucleoli; and necrotic areas. Atypical mitoses are seen occasionally.

All 34 cases in Table 1 underwent surgery. Thirty-one cases received complete or wide excision, of which 28 cases were followed-up and 3 patients suffered recurrences (11%) (3/28), that might have been due to incomplete resection. It is difficult to assess the true recurrence rate because there are not many long follow-up reports. The recurrence rate is 17% (3/18) accounting for the cases who were followed up for no less than 2 years. Only one case underwent radiotherapy (RT) for three recurrences. The lesion regressed after RT, but recurred again with malignant transformation and lung metastasis 4 years after RT, and the patient died 20 days after the last operation. Only one case underwent arthroscopic treatment to remove the lesion. Although some patients presented postoperative symptoms, most had an uneventful recovery. Only six cases had permanent complications, including mandibular deviation, facial paralysis, decreased strength, and preauricular concavity.

Owing to the infiltrative, aggressive behaviour and high recurrence rate, the treatment recommended in most of the literature is complete excision with synovectomy and/or capsulectomy. Simple excision should result in a complete cure for the localized form of PVNS, and RT probably has no place in the treatment of these solitary lesions, and the recurrence (range 0–27% in all joints) may be due to incomplete excision[19,35]. Diffuse PVNS poses a diagnostic and therapeutic problem. Where possible, complete synovectomy is indicated but the recurrence rate after such surgery is quite high, ranging from 8% to 46% in all joints[5,19,35]. Long periods of follow-up (more than 7–8 years) and regular MRI examinations after operation are required to obtain the optimal outcome[35]. Postoperative RT for high-risk or incompletely resected PVNS should probably be considered as an effective treatment for local tumour control, complications, such as stiffness, pain, pathologic fracture of the joint, may occur[19,20,39].

Three secondary and five primary malignant PVNS cases were reported by Bertoni et al. in 1997. Sixty-three per cent (5/8) of them had local recurrence after surgery; 50% (4/8) suffered lung, inguinal, and/or spine metastases, and were dead within 3 years even though wide excision and RT were carried out[4].

In view of the infiltrative nature, high recurrence, malignant transformation and metastasis, wide or complete excision of the entity is necessary, and close observation and regular MRI examination are required for more than 7–8 years.

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Competing interests

None declared.

Ethical approval

Not required.

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Case Report
Clinical Pathology

Leiomyomatous hamartoma of the midline maxillary gingival presenting as a congenital epulis: a case report with an immunohistochemical study

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M. Zhang, K. Matsuo, Y. Yamashita, T. Takahashi: Leiomyomatous hamartoma of the midline maxillary gingival presenting as a congenital epulis: a case report with an immunohistochemical study. Int. J. Oral Maxillofac. Surg. 2011; 40: 1322–1326.

Abstract. An otherwise-healthy 2-year-old Japanese female presented with a polyp-like lesion on the palatal surface at the incisive papilla. The appearance of the lesion was similar to that of a congenital epulis. The histological findings showed proliferating mesenchymal components that contained mainly smooth muscle admixed with collagen fibres, nerve fibres, small vessels and mucous salivary glands. The immunohistochemical staining findings for α-smooth-muscle actin, desmin and S-100 protein were all positive. The histological diagnosis was therefore leiomyomatous hamartoma, based on clinical microscopic observations.

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