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Pathology in Focus

Tumours and pseudotumorous lesions of the temporomandibular joint: a diagnostic challenge

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Abstract
Tumours and pseudotumorous lesions originating from the synovial membrane of the temporomandibular joint are rare. We report a series of six cases of such disorders. There were two cases of synovial chondromatosis, two of calcium pyrophosphate dihydrate crystal deposition disease, one nodular synovitis and one synovial sarcoma. Three patients were female and three were male. Their ages ranged from 36 to 70 years. All had atypical clinical and radiographical presentation. The prevalence, clinical and radiographical findings and pathological features of each disease entity are discussed and a review of the literature is made concerning all tumours and pseudotumours arising from the temporomandibular joint.

Key words: Temporomandibular joint; Synovial membrane; Chondromatosis, synovial; Synovitis, nodular; Sarcoma, synovial; calcium pyrophosphate

Introduction

The number of medical and dental practitioners diagnosing and treating temporomandibular joint (TMJ) disorders has recently increased. This phenomenon is due not only to a greater interest and better understanding of this joint, but also to the high-quality imaging techniques (Christiansen and Thompson, 1990) now available as computed tomography (CT) or magnetic resonance imaging (MRI). Yet, despite all this progress, many patients present with tumours or tumour-like lesions of the TMJ without any correct diagnosis until histological examination. The subsequent reported cases had atypical clinical and radiographical features. Such a presentation is not uncommon for this kind of lesion as shown by the review of the literature. The purpose of this article is to illustrate the rarity of tumours or pseudotumourous lesions and to highlight difficulties in diagnosis, and the requirement for a multidisciplinary approach.

Materials and methods

Since 1988 six cases have been available for study in the files of the Department of Pathology at Rangueil Hospital, Toulouse. Clinical information and radiographs were obtained by contacting clinicians and radiologists. All the specimens were fixed in formaldehyde and embedded in paraffin. The sections were routinely processed and stained with haematoxylin and eosin. In *Case 3* immunohistochemical studies were performed on formalin-fixed paraffin-embedded tissue sections with monoclonal antibodies against vimentin (Dako), cytokeratin (Immunotech) and epithelial membrane antigen (Dako) using the streptavidin-biotin-peroxidase method (StrepABComplex/HRP Duet, Dako).

Results

Clinical and radiographical findings

The clinical and radiographical features of the six cases are summarized in Table I. Clinically, three patients presented with chronic TMJ dysfunction, one with acute pain, one with external ear canal obstruction and one with a pre-auricular swelling. Physical examination found limited mouth opening and mandibular deviation in four cases, clicking and crepitus in three cases.

Plain radiographs were generally non-contributory. Computed tomography (CT) scans showed intra-articular loose bodies in three cases (Figure 1), and peri-articular calcification and extensive destruction of the condyle in *Case 4*. Two soft tissue proliferations were visualized by CT or MRI scans (Figure 2).

Microscopic findings

Case 1 and Case 2

The firm, glossy, yellow to greyish white loose bodies (Figure 3) found in the joint consisted of cartilaginous nodules covered by a thin synovial layer (Figure 4). The chondrocytes were sometimes atypical with enlarged and hyperchromatic nuclei, but there were no mitotic figures. There was no inflammation and no chondroid metaplasia of the articular synovium.

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TABLE I
CLINICAL AND RADIOGRAPHICAL DATA OF TUMOURS AND PSEUDOTUMOROUS LESIONS OF THE TMJ

	Case 1	*Case 2*	*Case 3*	*Case 4*	*Case 5*	*Case 6*
Age (years)/Sex	36/Male	70/Female	52/Male	46/Female	39/Female	39/Male
Previous medical history	Bruxism	None	None	Pain of both wrists about 10 years ago	None	Viral hepatitis
Main complaints	Chronic pain of the right TMJ with recent aggravation	Chronic pain of the right TMJ	Chronic pain of the left TMJ	Acute attack of pain in the left TMJ	External ear canal mass recently increasing in size	Pre-auricular swelling
Symptoms' duration	1 year	6 years	4 years	A few days Same episode 3 years ago	3 months	2 years
Biological profile	Normal	Normal	Normal	Normal	Normal	Normal
Physical examination	Clicking Mandibular deviation Limited mouth opening	Clicking Cracking	Clicking Limited mouth opening Mandibular deviation Pre-auricular swelling	Pre-auricular swelling	Firm mass about 1 cm in size in the anterior wall of the meatus canal	Limited mouth opening Mandibular deviation Pre-auricular firm mass measuring about 3 cm
Radiographical findings	Widened joint space, ankylosis. Intra-articular loose bodies on CT scans	Intra-articular calcifications	Erosion of the condyle Peri-articular calcifications	Ankylosis Extensive destruction of the mandibular condyle		Well limited expansive process invading the temporal muscle on CT scans
Macroscopic findings	Numerous firm grey-white bodies 1 to 8 mm in size	Numerous firm greyish bodies 1 to 7 mm in size	Numerous chalky particles	Numerous chalky particles	Well limited yellow to brownish mass emerging from the joint capsule	Bilobulated white firm tumour (2×2, 5×5 cm) Evident involvement of the TMJ
Pre-operative diagnosis	None	CPPD crystal deposition disease	None	Infectious arthritis or malignant tumour	Sebaceous cyst	Lipoma before CT scans
Final histological diagnosis	Synovial chondromatosis	Synovial chondromatosis	Simultaneous CPPD crystal deposition disease and synovial chondromatosis	CPPD crystal deposition disease	Localized nodular synovitis	Synovial sarcoma
Current status	Alive No recurrence	Alive No recurrence	Alive Acute pseudogoutous attack of the left knee 6 months later	Alive No recurrence	Alive No recurrence	Alive No recurrence
Length of follow-up	3 years	7 years	5 years	4 years	7 months	5 months

In both cases, these histological findings were consistent with the diagnosis of stage III synovial chondromatosis.

Case 3

The loose bodies found in the capsule corresponded microscopically to nodules of cartilaginous tissue containing foci of microcrystalline material (Figure 5). A foreign body giant cell reaction was found close to the crystal deposits. These crystals were rhomboid to rectangular and appeared slightly birefringent under polarized light microscopy as are CPPD crystals. A synovial biopsy specimen demonstrated the same changes: CPPD crystal deposition within nodules of chondroid metaplasia.

The final diagnosis was CPPD crystal deposition disease in association with synovial chondromatosis.

Case 4

Histological examination of the mandibular condyle and temporal fossa exhibited major arthritic changes.

The fragments lying within the joint space were identified as dystrophic bone, cartilage and synovial tissue. We noticed numerous scattered microcrystalline deposits surrounded by a granulomatous giant cell reaction (Figure 6). Under polarized light, these crystals were typical calcium pyrophosphate dihydrate as previously described. This destructive lesion could be ascribed to a pseudotumorous form of CPPD crystal deposition disease.

Case 5

Microscopic examination disclosed a well-demarcated nodular lesion without any connection with the overlying squamous epithelium. The major component of the

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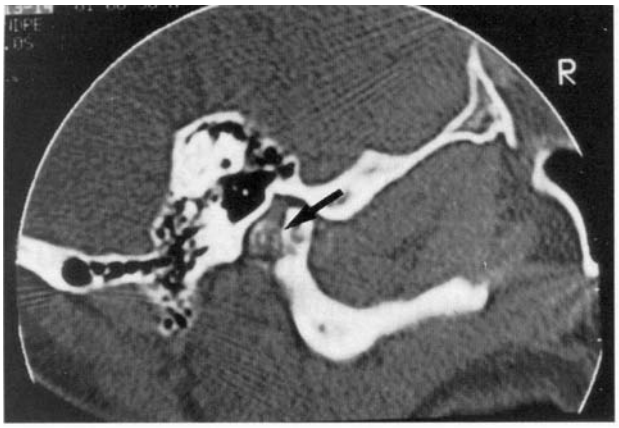

FIG. 1

CT scans of *Case 1*. Enlargement of the TMJ with irregular intra-articular radiopaque structures (arrow).

proliferation was spindle-shaped cells arranged in fascicles. Numerous polymorphous inflammatory cells consisting of histiocytes, multinucleated giant cells, lymphocytes and plasma cells were scattered throughout the lesion. Xanthoma cells were rare. There was no nuclear atypia but inconspicious mitotic figures were noted within the spindle-shaped cells. Intra- and extracellular deposits of haemosiderin were present as shown on colloidal iron staining (Figure 7). The final diagnosis of this external ear canal mass was a pigmented nodular synovitis of the temporomandibular joint.

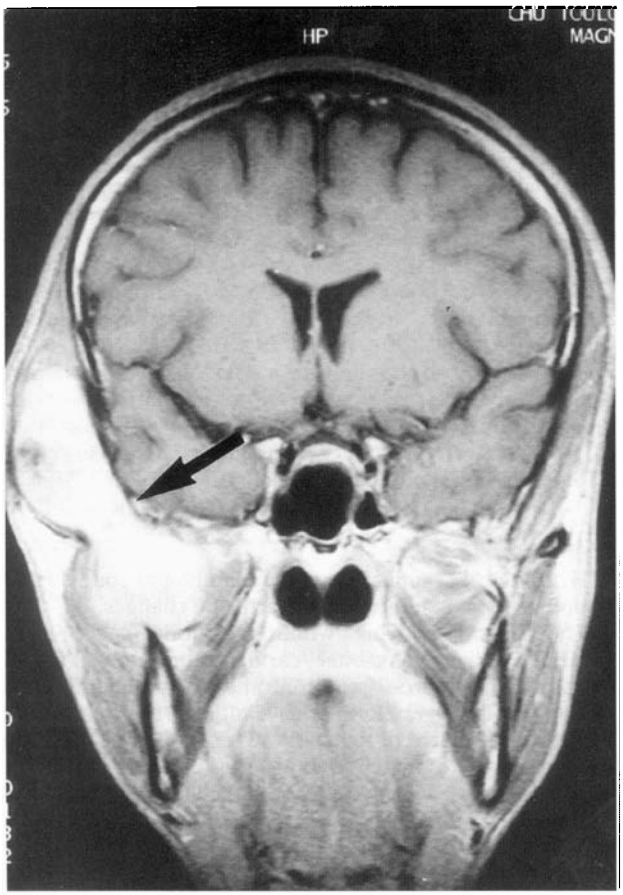

FIG. 2

MRI scan of *Case 6*. Voluminous tumoral process involving the right TMJ and invading adjacent soft tissues (arrow).

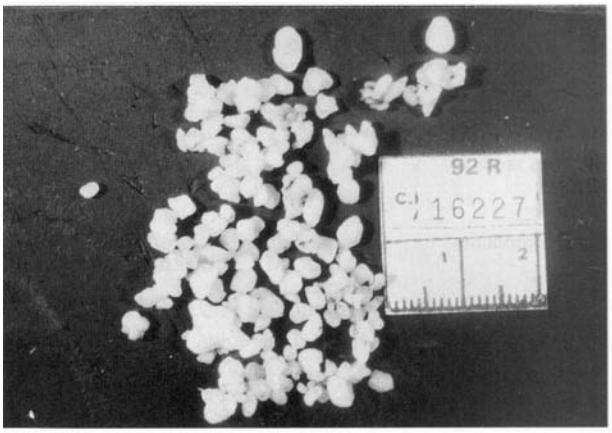

FIG. 3

Synovial chondromatosis. Macroscopic appearance of intra-articular loose bodies removed from the right TMJ in *Case 1*: about one hundred small round to oval cartilaginous nodules.

Case 6

Microscopic examination revealed that the major part of the specimen was composed of plump, spindle-shaped cells arranged in fascicles with a focal storiform pattern. Within this mesenchymal proliferation we could identify a few cyst-like spaces lined by pseudoepithelial cells (Figure 8). Mitotic figures were rare. Immunohistochemistry on paraffin-embedded formalin-fixed tissue sections had been realized. The spindle-shaped component stained with anti-Vimentin antibodies and the rare gland-like spaces demonstrated weak but clear-cut positivity with anti-cytokeratin and anti-EMA markers. These histological findings were characteristic of a biphasic grade I synovial sarcoma with a predominant mesenchymal component.

Discussion

We report here different tumours and pseudotumorous processes emerging from the TMJ synovial membrane which has the same structure as that of any other joint (Dijkgraaf *et al.*, 1996). This joint is a diarthrosis and can be the location of various disorders originating from the bone, the cartilage and the synovium. These conditions are rare, but our six cases and the review of the literature illustrate how difficult it is to make a radioclinical diagnosis without any histological examination.

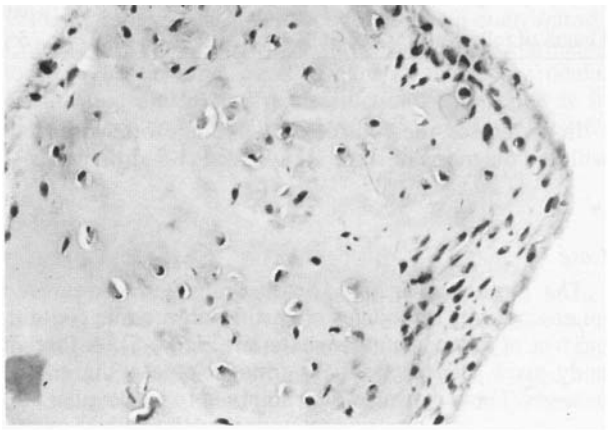

FIG. 4

Synovial chondromatosis. Intra-articular loose bodies consisted of varibly sized nodules of cartilage covered by a thin synovial epithelium. (*Case 2*: H & E; original magnification × 25).

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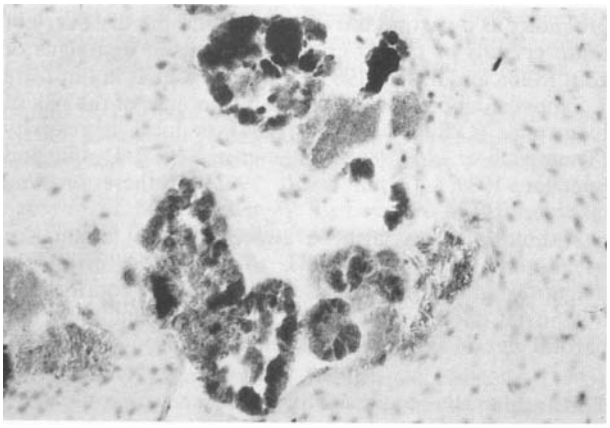

FIG. 5

Synovial chondromatosis with CPPD crystal deposition disease. Calcium-containing crystals in nodules of chondroid metaplasia. (*Case 3*: Von Kossa stain; original magnification × 25).

Synovial chondromatosis (SC) is one of the most frequent primitive disorders arising in the TMJ. It is also probably the least misleading and the easiest to diagnose without any microscopic examination. When occurring in this joint, synovial chondromatosis often presents as a pre-auricular mass (Blankestijn, 1985; Thompson *et al.*, 1986; Quinn *et al.*, 1992). The more specific radiographical features are loose bodies which are round to irregularly-shaped and variably-sized radiopaque structures. The other features are widened joint space, ankylosis, irregular condyle surface, erosion of the glenoid fossa or temporal eminence, lytic changes and absence of abnormalities (Forssell *et al.*, 1988). CT scans can be helpful to establish a diagnosis because they are able to identify loose bodies not yet visible with plain radiographs. Nevertheless, even the most sophisticated examinations, including MRI, can be negative (Holmlund *et al.*, 1992).

CPPD crystal deposition disease is a microcrystalline arthropathy sharing clinical similarities with synovial chondromatosis. The different presentations include pseudogoutous, pseudotumorous, pseudoarthritic and destructive forms. The TMJ is a rare location for CPPD crystal deposition disease, but it is the most frequent site for pseudotumorous forms (Ishida *et al.*, 1995). Fewer than twenty cases, have been reported in the literature to date. All of them displayed misleading clinical symptoms

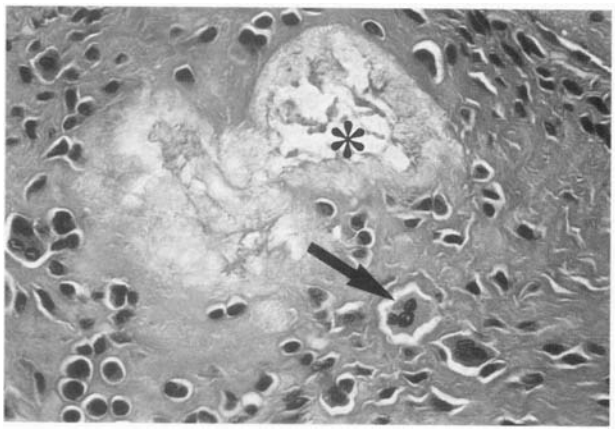

FIG. 6

CPPD crystal deposition disease. Amorphous crystalline material (asterisk) surrounded by a granulomatous giant cell reaction (arrow). (*Case 4*: H & E; original magnification × 40).

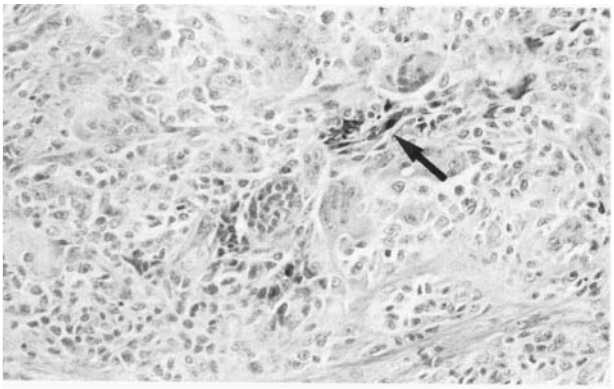

FIG. 7

Pigmented nodular synovitis. Haemosiderin deposits (arrow) within a polymorphous infiltrate of histiocytes, multinucleated giant cells, lymphocytes and plasma cells. (*Case 5*: Colloïdal iron stain; original magnification × 25).

(parotid mass, otological signs, pseudogoutous attack) (Zemplenyi and Calcaterra, 1985; Magno *et al.*, 1992) and often atypical radiographical features (extensive lysis of the condyle and degenerative changes with or without intra- or peri-articular calcifications). In this respect, we must point out, that in the TMJ, radiographical findings of CPPD crystal deposition have different, deluding and non-specific features compared to other joints: the fine dense linear border in the joint space corresponding to the calcified meniscus is absent even on CT scans. The most frequent changes are peri- or intra-articular calcified loose bodies (Figure 3), in association with major degenerative damage. That is why the positive diagnosis is usually not made until histological examination.

Compared to CPPD crystal deposition disease, gouty tophi are infrequently localized to the TMJ. Nevertheless, this microcrystalline arthropathy is also able to mimic tumours when involving the TMJ as described by Ballhaus *et al.* (1989). Radiographical findings reveal bony lysis and even possible complete destruction of the mandibular condyle. Califications in the joint space are an uncommon feature.

Pigmented villonodular synovitis (PVNS) or nodular synovitis (NS), which are variants of the same disease, can be considered as a pseudotumorous proliferation of synovial origin rather than a benign tumour. The TMJ is a very rare site for the occurrence of this lesion. Among

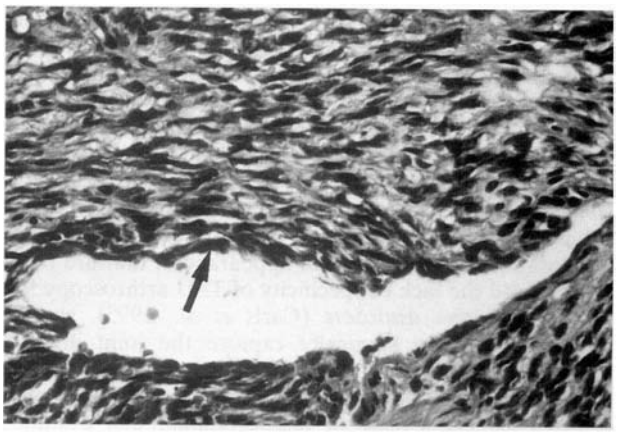

FIG. 8

Biphasic synovial sarcoma. Dense proliferation of spindle cells with formation of pseudoglandular or cyst-like spaces lined by pseudoepithelial cells (arrow). (*Case 6*: H & E; original magnification × 40).

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the reported cases, most of them had a singular way of presentation. Only few had clinical symptoms directly referable to the TMJ. The majority were thought to have a parotid tumour, some had otological signs: hearing loss (Lapayowker *et al.*, 1973), external ear canal obstruction (Eisig *et al.*, 1992; our *Case 3*). One had a first diagnosis of brain tumour (Eisig *et al.*, 1992). On radiological examination, the TMJ is frequently preserved. The most common finding is a mass near the parotid gland or in adjacent soft tissues. Erosion and even invasion of the bone is possible, this behaviour being better illustrated on CT scans.

Synovial sarcoma is a sarcomatous tumour arising from the joint capsule, tendon sheaths and bursae of young adults. Involvement of the TMJ is rare. We report here only the fourth synovial sarcoma of the TMJ described in the literature. Among the previously reported cases, one presented with a pre-auricular swelling interpreted as a parotid tumour (DelBalso *et al.*, 1982), and another one with no precise diagnosis before excision (White *et al.*, 1992).

Based on the review of the literature, the most frequent tumours involving the TMJ are metastases of malignant neoplasms (Bavitz and Chewning, 1990). Nevertheless, the context is generally evocative and no surgical exploration is required for diagnosis. This is not the case with primary lesions arising from intrinsic tissues of the joint. These lesions can potentially originate from bone, cartilage or synovial membrane. Bony and cartilaginous tumours are more common than synovial disorders. Osteochondroma (Gringrass and Sadeghi, 1993), osteoblastoma (Weinberg *et al.*, 1987), chondroblastoma (Bonin *et al.*, 1995), chondrosarcoma (Nitzan *et al.*, 1993) and neurofibroma (Van Damme *et al.*, 1996) of the temporomandibular region have been reported. Concerning tumours or pseudotumorous lesions directly originating from the synovial membrane, we have reported four types (synovial chondromatosis, CPPD crystal deposition disease, nodular synovitis and synovial sarcoma). This list is not exhaustive and a review of the relevant literature revealed the possibility of other synovial disorders. Pseudotumorous gouty tophus can be considered as another pseudotumorous lesion of the synovium (Ballhaus *et al.*, 1989; Suba and Szabo, 1989). Synovial chondrosarcoma either primary or secondary to synovial chondromatosis is a rare entity and has never been reported in the TMJ. Ganglion developing from the TMJ synovial membrane have been reported by some authors (Copeland and Douglas, 1988). In any of these TMJ disorders, plain radiographs are infrequently helpful because of superimposition of other anatomical structures that make interpretation difficult. CT scans allow a better approach for osseous lesions whereas MRI scans are more sensitive for non-osseous proliferations. Clinicians should not hesitate to perform these more sensitive imaging modalities. Intra-articular loose bodies, a parotid or para-articular soft tissue proliferation can then be discovered. Osteolysis is also a common feature, maybe due to the narrowness of this joint which leads to rapid ischaemic changes as soon as an intra-articular process develops. Such radiographical appearances, that are non-specific, and the lack of specificity of TMJ arthroscopy for non-degenerative disorders (Carls *et al.*, 1995), should incite clinicians to surgically explore the joint and to include tumours and pseudotumorous lesions of the TMJ in the differential diagnosis of any pre-auricular tumour, external ear canal mass or temporomandibular dysfunction, especially after failure to respond to an appropriate therapy.

A surgical approach is necessary for a correct histological diagnosis in order to arrange correct patient follow-up and therapy: a synovial sarcoma does not have the same prognosis as a parotid mixed tumour and the discovery of urate crystals or CPPD crystals can be the first signs of gout (Suba and Szabo, 1989) or pseudogout as in our *Case 3*. Synovectomy is also indispensable because of the risk of recurrence (O'Sullivan *et al.*, 1984), or local aggressivity (Thompson *et al.*, 1986; Nokes *et al.*, 1987; Daspit and Spetzler, 1989; Quinn *et al.*, 1992) of these synovial disorders.

Pathologists must also be aware of these tumours or pseudotumours of the TMJ to avoid an erroneous diagnosis: an exceptional case of histological confusion of a synovial chondromatosis with a mixed tumour of the parotid gland have been reported (Thompson *et al.*, 1986).

Conclusion

Tumours and pseudotumorous lesions of the temporomandibular joint are a challenging diagnostic problem both clinically and radiographically because of atypical and misleading presenting symptoms and absence of specific signs. Their frequency is probably underestimated due to difficulties encountered by clinicians, and a number of patients are misdiagnosed. This study was designed to make clinicians and pathologists aware of these potential disorders of the TMJ in order to optimize the likelihood of timely and appropriate diagnosis and therapy.

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