## **CASE REPORT**

# A case of chronic calcium pyrophosphate dihydrate crystal disease (tophaceous pseudogout) in the temporomandibular joint

### B Meul<sup>1</sup>, K Ernestus<sup>2</sup>, J Neugebauer<sup>1</sup>, AC Kuebler<sup>1</sup>

<sup>1</sup>Department of Craniomaxillofacial and Plastic Surgery and <sup>2</sup>Institute of Pathology, University of Cologne, Cologne, Germany

Pseudogout is a rare joint disease which is characterized by the presence of calcium pyrophosphate dihydrate crystals in the intraarticular and periarticular tissue. The crystals tend to attach to fibrocartilage tissue. Pseudogout principally affects the knee and wrist joints. Involvement of the temporomandibular joint (TMJ) is very rare. There have been <20 cases reported worldwide. Both acute and chronic manifestations have been described. We present here an unusual case that necessitated a high condylectomy.

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#### **Case report**

A 54-year-old man with a 3-year history of painless swelling in the left preauricular region was referred to us with the presumptive diagnosis of a malignant tumour of the left parotid gland. The patient had a history of limited mouth opening capacity and pain in the left temporomandibular joint (TMJ), which had been progressive over a few months. His medical history showed a paraplegia following a motor-vehicle accident 25 years before, and a non-insulin dependent diabetes. The laboratory findings did not point out hypercalcaemia, haemochromatosis, hypomagnesemia, hypothyroidism, hyperparathyroidism and there was no indication of ochronosis. In addition there was no trauma to the TMJs in his medical history and there was no family history of joint diseases.

Examination of the left preauricular region revealed a painless swelling measuring  $3 \times 3$  cm. The patient was fully dentate and had no occlusal disturbance. There

was severe limitation in forward translocation and rotation in the left TMJ, and maximum mouth opening was measured at 26 mm inter-incisal distance. Lateral excursion to the right was not possible. The facial nerve function was unaffected. CT-scans (Figure 1a) showed mixed radio-opacity of the left condyle with a bony destruction of the condylar head. Articular eminence and fossa were not involved. The articular disc was atrophic. MRI investigation revealed a contrast-enhancing, well-demarcated lesion in the left TMJ (Figure 1b). Extension to the most cranial part of the condylar head and close proximity to the parotid gland could be demonstrated. Suspicious lymph-nodes were not found. All other joints were clinically free of disease and skeletal scanning showed no pathological findings.

Because of suspicion of a malignant or benign tumour of the joint based on the MRI- and CT-scan and the inaccessible anatomical region, a primary high condulectomy was performed using a preauricular and submandibular approach. White, firm gritty material was noted contained in the joint capsule. The destroyed condylar head could be dissected free from the surrounding tissue and from the parotid gland. The articular disc was removed. Reconstruction was achieved by performing an osseous-chondral graft from the seventh right rib. A stable osteosynthesis of the rib was achieved using three bicortical screws. Intermaxillary fixation was maintained for only 14 days to prevent an ankylosis of the TMJ. Even when considering that the tumour was benign, the condylar head and parts of the collum had to be resected and replaced by a graft, because these parts were destroyed osteolytically.

Pathological examination showed multiple tissue specimens with a grey-white appearance. Microscopically, the fibrous tissue demonstrated islands of basophilic crystalline material (Figure 2a). The crystalline material was aggregated and in areas with lower content of crystals they sometimes showed a needle-like and rhomboid configuration. The crystalline material was birefringent (Figure 2c, d). Surrounding these areas a

Correspondence: Dr Britta Meul, Department of Craniomaxillofacial and Plastic Surgery, Kerpener Str. 62, 50937 Cologne, Germany. Tel.: +49 221 478 5775, Fax: +49 221 478 5774, E-mail: britta.meul@ uni-koeln.de

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histiocytic reaction took place with foreign body giant cell reaction (Figure 2b).

One year after surgery the joint function is symptomfree with an inter-incisal opening of 40 mm. Clinical examination of the joint demonstrates painless rotation and translational movement of the graft.

#### Discussion

The first case of pseudogout of the TMJ was published in 1976 by Pritzker (Pritzker *et al*, 1976). Because of the gout-like symptoms of joint-pain and swelling, the term 'pseudogout' was applied to the condition. The complex pathogenesis of pseudogout is unclear (Dijkgraaf *et al*, 1995; Vargas *et al*, 1997). There may be an association with hypercalcaemia or hyperparathyroidism (Grant *et al*, 1999), but a single aetiological factor cannot be identified on a routine basis. The disease is typically classified as idiopathic, hereditary or secondary. An association between diabetes mellitus and pseudogout – like an association between diabetes mellitus and gout known as plurimetabolic syndrome/syndrome X – statistically does not exist (Silveri *et al*, 1994). Pseudogout can appear as in a single joint or as polyarticular. Figure 1 (a) Axial CT at the level of temporomandibular joint (bone window). A considerable calcified mass arroding the left mandibular condyle (arrows). (b) MRI, frontal slice. Both the upper and the lower joint are affected by a heterogeneous tissue, the mandibular condyle is arroded. The tumour seems not to involve the infratemporal fossa. Arrows show the medial and lateral expansion

Figure 2 (a) This overview of tophaceous pseudogout showed the nodular aggregates of crystalline material. HE, 100×. (b) Higher magnification of a: the aggregates of crystals were basophilic and surrounded by a histiocytic reaction with foreign body giant cells. HE, 200×. (c) The crystals formed aggregates and were birefringent. Polarized light, 400×. (d) The crystals showed a needle-like and sometimes rhomboid configuration. Polarized light, 400×

The differential diagnosis includes rheumatoid-, osteoand suppurative arthritis (Nakagawa et al, 1999). Radiologically there are no clear distinguishing features between these different diseases and there is no known typical radiological appearance of pseudogout. The clinical and radiographic features may also lead to a provisional diagnosis of a malignant tumour of the TMJ or the parotid gland. There may be destruction of the joint and expansion into the cranial fossa (Zemplenyi and Calcaterra, 1985; Grant et al, 1999). Accordingly it is not possible to make a diagnosis using only clinical and/or radiological findings. Histopathological findings of aspirated or resected specimens are necessary to confirm the presumptive diagnosis of pseudogout. In the present case the presumptive diagnosis of a malignant tumour of the TMJ was disproved by the histopathological diagnosis.

Microscopically, the fibrous tissue demonstrates islands of basophilic crystalline material. Calcium pyrophosphate dehydrate gets lost when decalcification is performed. However, in the presented case most of the tissue does not receive decalcification and as a consequence the crystals can be identified with conventional haematoxylin eosin (HE) staining. The crystals show a

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needle-like or rhomboid configuration in areas with single layered crystals, which is typical for calcium pyrophosphate dehydrate crystals (Ishida *et al*, 1995). In addition the crystals were birefringent.

The differential diagnosis includes gout tophi. However, monosodium urate crystals disappear in formalin fixed tissue and are only preserved with alcohol fixation. In the present case the crystals were clearly visible with formalin fixation. In addition, urate crystals show a different morphology appearing as more monomorphic long slender needle-shaped crystals.

Calcium phosphate depositions are small blue amorphic masses without needle or rhomboid configuration. They are not birefringent. In addition, foreign body reaction in general is not as distinct as in pseudogout. In the present case the crystalline material showed the typical morphology and characteristic features of calcium pyrophosphate dehydrate crystals.

The acute from of pseudogout is characterized by a crystalline-induced synovitis and can be treated with colchicine or triamcenolone acetenoid while non-steroidal anti-inflammatory drugs (NSAIDs) are contraindicated. Where the disease only involves the synovium, joint lavage may prove beneficial in removing the crystalline particles (Nakagawa *et al*, 1999). Significant and bulky deposits as in the presented case should be removed by surgery. To replace the resected parts of the joint it may be necessary to use a graft or joint prosthesis. Recurrences after surgery are described in about 20 percent of the cases (Zemplenyi and Calcaterra, 1985; Dijkgraaf *et al*, 1995; Ishida *et al*, 1995).

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