CASE REPORT

Synovial chondromatosis of the temporomandibular joint with calcium pyrophosphate dihydrate crystal deposition disease (pseudogout)

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This report describes a very rare case of synovial chondromatosis with deposition of calcium pyrophosphate dihydrate (CPPD) crystals (pseudogout) in the temporomandibular joint (TMJ) of a 46-year-old male patient. Synovial chondromatosis is a non-neoplastic disease characterized by metaplasia of the connective tissue leading to chondrogenesis in the synovial membrane. Pseudogout is an inflammatory disease of the joints caused by the deposition of CPPD, producing similar symptoms to those observed in gout but not hyperuricaemia. Both diseases commonly affect the knee, hip and elbow joints, but rarely affect the TMJ. *Dentomaxillofacial Radiology* (2012) **41**, 703–707. doi: 10.1259/dmfr/24183821

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Case report

A 46-year-old man presented to a dental clinic on 19 May 2007 with the chief complaint of right temporomandibular pain. A tumour was suspected owing to the persistence of the symptoms. The patient was then referred on 24 May 2007 to the Department of Oral and Maxillofacial Surgery at Mie University Hospital, Japan, for examination and treatment. Mild depression had been diagnosed and the patient had been prescribed oral antidepressants from the age of 45 years. He had no significant family history or history of trauma or hyperparathyroidism before the onset of the present illness. Physical examination showed a moderate physique with good nutritional status, symmetrical facial appearance and no skin lesions, such as bulges, on the skin surface around the right temporomandibular joint (TMJ). He had a painless mouth opening of 24 mm, at which painful clicking occurred, and a maximum mouth opening of 36 mm. Laboratory findings were all normal, except for a positive hepatitis virus Type B infection. Panoramic radiography revealed a small

radio-opaque lesion located anterior to the right TMJ (Figure 1). An axial view of the CT scan revealed small calcific substances surrounding the mandibular condylar head in a ring (Figure 2a). A coronal CT image also revealed small calcific substances in the condylar head in the mandibular fossa (Figure 2b). MRI revealed small mass-like low signal intensity lesions scattered in a ring-like high-signal intensity area in the same locations as on CT in the axial and coronal T_2 weighted images (Figure 3a,b). On sagittal T_1 weighted images, the lesions appeared in a tear-like shape in the anterior part of the right mandibular head corresponding to the superior joint cavity and a thick, low-signal intensity area in the posterior part of the mandibular head. T_2 weighted images showed low-signal intensity, small mass-like lesions scattered in a high signal intensity area in the same locations as on T_1 weighted images, with some small masses including high signal intensity areas. In addition, contrast-enhanced T_1 weighted images showed substantial enhancement of the region corresponding to the synovial membrane comprising the superior joint cavity with almost no enhancement inside the cavity. No abnormal findings were observed in terms of the signal intensity of the bone marrow in the mandibular head (Figure 4a,b,c). ^{99m}Tc -methyl diphosphonate bone scintigraphy revealed

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Figure 1 Orthopantomography at the first visit. A small mass-like calcific substance is observed in a region anterior to the right temporomandibular joint (arrowhead)

a weak tracer uptake in the region corresponding to the right TMJ (Figure 5). On 13 June 2008, an aspiration biopsy was performed at a site anterior to the right ear and subsequent polarization light microscopy showed a weak positive double refraction (Figure 6). Although these findings strongly suggested synovial chondromatosis with deposition of calcium pyrophosphate dihydrate (CPPD) crystals, the possibility of a tumour could not be completely ruled out. Therefore, on 24 October 2008, surgery was performed under general anaesthesia for the suspected right TMJ tumour. A temporal incision was made in a region anterior to the right ear followed by the opening of the superior joint cavity from the outside of the joint capsule and the mass was removed from the anterior part to the outer side of the mandibular head.

a

b

Figure 2 (a) An axial CT image. (b) A coronal CT image. A small mass-like calcific substance is observed surrounding the right mandibular head (arrowheads)

a

b

Figure 3 (a) An axial MR image. (b) A coronal MR image. T_2 weighted fast spin echo (time of repetition/time of echo/acquisition: 3000 ms/22 ms/2) image (a and b). Multiple small mass-like, low-intensity areas are observed in the same region as on CT

Figure 4 (a) MR T_1 weighted image. (b) MR T_2 weighted image. (c) MR gadolinium (Gd)-enhanced T_1 weighted image. (a) T_1 weighted spin echo [time of repetition (TR)/time of echo (TE)/acquisition: 500 ms/14 ms/3] image (left). (b) T_2 weighted fast spin echo (TR/TE/acquisition: 3000 ms/22 ms/2) image. (c) T_1 weighted spin echo (TR/TE/acquisition: 500 ms/14 ms/3) image with Gd-diethylenetriamine penta-acetic acid (right)

The removed mass was an elastic, soft mass measuring $10 \times 10 \times 8$ mm in size with a clear border and a smooth, pale yellow surface. Pathologically, the mass consisted of cartilage tissue with a slightly high cell density and was encapsulated in a fibrous tissue, leading to a diagnosis of synovial chondromatosis (Figure 7). With all these findings taken together, the final diagnosis of synovial osteochondromatosis of the right TMJ with deposition of CPPD crystals was made. After surgery, no recurrence of symptoms was observed and a painless mouth opening of 43 mm was achieved.

Discussion

In synovial chondromatosis, cartilage tissue is generated in the synovial tissue and part of the chondrified tissue enters the joint cavity and undergoes chronic calcification, leading to formation of a joint loose body of cartilage tissue.¹ This makes imaging an important tool for diagnosis, and the effectiveness of CT and MRI has been demonstrated. MRI is particularly useful in obtaining detailed information about the lesion, such as the relationship between the lesion and surrounding tissues, and determining the surgical procedure²⁻⁴. Milgram⁵ pathologically classified synovial chondromatosis into three stages. In Stage 1, chondrogenesis occurs in the synovial membrane with no formation of a loose body. In Stage 2, chondrogenic tissue begins to enter or be released into the joint cavity with formation of oedematous synovial connective tissue and a loose body. In Stage 3, chondrogenesis in the synovial membrane disappears and only a loose body can be observed. These pathological features appear to be most accurately reflected in MRI findings. It is also important to use gadolinium (Gd)-containing contrast agents for a more accurate diagnosis. In the present case, Gd-enhanced T_1 weighted images showed a contrast-enhanced, high-signal intensity area that corresponded to synovial tissue as a result of anterior dislocation of the articular disc. This finding reflects oedema,

thickening or inflammation and corresponds to Stage 2 in Milgram's classification.⁵ If this is a characteristic finding determining the stage of synovial chondromatosis, it is considered a very significant finding, suggesting the involvement of inflammation and overload in the development and progression of the disease. Furthermore, the addition of a dynamic MRI-based evaluation may allow for a more accurate understanding of the synovial condition and thus a more accurate determination of the pathological stage.

On the other hand, pseudogout is a form of arthritis in which a response similar to that observed for uric acid in gout occurs for CPPD. More specifically, it occurs as a result of an inflammatory response to CPPD crystals deposited inside a joint capsule. Pseudogout commonly

Figure 5 Bone scintigraphy. A weak tracer uptake is observed in the right temporomandibular joint (arrowhead)

Figure 6 Polarization light microscopy. Positive double refraction indicative of calcium pyrophosphate dihydrate is observed

affects the knee joints of people aged 60 years or more, regardless of gender. However, how intra-articular deposition of CPPD crystals occurs remains unclear. It has been suggested that pseudogout is caused by abnormal increases in blood calcium concentrations due to hyperparathyroidism or in tissue iron concentrations due to haemochromatosis, or by abnormal decreases in blood magnesium concentrations in conditions such as hypomagnesaemia.⁶ However, because a majority of pseudogout patients have no history of these metabolic diseases, the involvement of genetic factors is still possible. As with synovial chondromatosis, pseudogout also commonly affects the knee joint as well as other large joints, including the elbow, wrist and hip joints,⁶⁻⁸ but rarely affects the TMJ. The present patient was positive for hepatitis virus Type B infection but it was

unclear whether it contributed to the development of the disease.

Pseudogout is diagnosed according to the following criteria as described by Kohn *et al*:⁹

- (1) identification of crystalline components by such means as X-ray analysis and chemical analysis
- (2a) identification of the appearance and shape of crystals by corrected polarization microscopy
- (2b) identification of calcification by X-ray
- (3a) in the case of large joints, the presence of findings suggestive of acute arthritis
- (3b) the presence of findings suggestive of acute aggravation of chronic arthritis, especially in the knee, hip, wrist, carpal, elbow, shoulder or metacarpophalangeal joint.

Figure 7 Pathological findings. Haematoxylin and eosin staining. (a) Original magnification $\times 10$ (left). (b) Original magnification $\times 20$ (right)

Diagnostic results are further classified into “definite” (1 or 2a and 2b), “probable” (2a or 2b) or “possible” (3a or 3b). The present case falls into the “definite” category.

Pseudogout is commonly associated with radiographic evidence of calcification. However, in the case of pseudogout affecting the TMJ, calcification may not be clearly visualized owing to the anatomical relationship with the surrounding tissues and the specific properties of rotational panoramic radiography, making MRI findings more important. If synovial fluid is observed on MRI, sampling of a small amount of synovial fluid allows for identification of CPPD crystals by polarization microscopy.¹⁰ Care should be taken to distinguish CPPD crystals from calcium oxalate, synthesized steroid crystals or ethylenediaminetetraacetic acid crystals for the proper diagnosis of temporomandibular disease. In addition, observation with an analytical electron microscope equipped with an energy dispersive X-ray analyser and quantitative analysis of crystal samples to determine the calcium:

phosphorus ratio also allows for a more accurate diagnosis.¹¹

Synovial chondromatosis is divided into primary and secondary forms. The primary form is caused by unknown factors and is highly proliferative.¹² The secondary form is more common than the primary form and is believed to be caused by inflammatory joint disease or overload on a joint.¹³ Assuming that the present case is the secondary form of synovial chondromatosis, deposition of CPPD crystals might have occurred first and then induced crystallization of pseudogout and synovial chondromatosis. This, however, remains a matter of speculation. Future studies should address issues such as identification of disease-specific genes. Guarda-Nardini *et al*'s¹⁴ article describes synovial chondromatosis in 155 cases while Takata *et al*'s¹⁵ article describes CPPD in 37 cases. However, there is no report of the simultaneous occurrence of synovial chondromatosis and CPPD with the TMJ. In conclusion, we present a very rare case of synovial chondromatosis with deposition of CPPD crystals (pseudogout) in the TMJ.

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