Pre-auricular swelling and malocclusion

Bruce R Pynn, Jonathan Irish, Simon Weinberg

A 58-year-old man was referred to the Head and Neck Clinic for the diagnosis and treatment of a painless swelling in the left pre-auricular region of approximately 18 months duration. The swelling had appeared suddenly and had remained the same size until one month prior to his referral when the swelling abruptly increased in size and then slowly decreased to its original size following two courses of nonsteroidal anti-inflammatory drug (NSAID) therapy. There was no history of joint trauma, pain or joint locking. Loading of the joint during eating and talking did not produce any joint symptoms, but the patient noted that his ‘bite’ did not feel perfect. The medical history was unremarkable and specifically, there was no history of disease involving any other joints.

Physical examination showed a firm, non-tender 3.0 × 3.0 cm swelling anterior to the left ear. There was moderately limited translation of the left condyle and a slight deviation of the mandible to the left during mouth opening. The patient had an active mouth opening of 40 mm intercidentally and a mild crepitus was heard during auscultation of the left temporomandibular joint. The patient had a mild malocclusion characterised by a slight left posterior open bite with a mild anterior shift of the left mandible.

A computed tomography (CT) scan indicated that the lesion originated within the left intra-articular space and showed a poorly defined peripheral soft tissue component completely encircling the head of the condyle. The mass had displaced the condylar head anteriorly and inferiorly (figure 1). There was no involvement of the parotid gland.

The patient was taken to the operating theatre where the left temporomandibular joint was explored using a pre-auricular approach. The joint capsule was markedly distended and a thick synovial fluid was apparent as the superior joint space was entered. A white, gritty, dough-like mass measuring 3.0 × 2.0 cm was removed from the anteromedial aspect of the upper joint space. The inferior joint space was carefully examined and no additional masses were found. The condylar head was covered with normal looking fibrocartilage. The articular surface was rippled and smooth but markedly flattened. There was no evidence of erosive disease or osteophyte formation. The meniscus appeared normal in colour and texture. There was no evidence of perforation.

Histologic examination of the specimen by polarized light microscopy demonstrated positive birefringent, parallel lipid crystals (figure 2). Scanning electron microscopic (SEM) analysis demonstrated rhomboid and rod-like crystals ranging in size from 1 to 5 μm (figure 3). Infrared-spectrophotometric analysis (IRSA) was also carried out.

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**Figure 1** CT scan of the temporomandibular joint

**Figure 2** Histological appearance
The patient's postoperative course was uneventful. His occlusion reverted to the pre-morbid state very shortly after surgery. At the three-year follow-up examination, there were no signs or symptoms of disease in any joints.

Questions

1. What is your diagnosis?
2. What other factors are associated with the development of this condition?

Figure 3  SEM photomicrograph
Answers

QUESTION 1
Calcium pyrophosphate dihydrate (CPPD) deposition disease. The characteristic clinical and radiographic features along with the unique histologic and SEM photomicrograph confirm the diagnosis.

QUESTION 2
Multiple cofactors are known to be linked to the development of CPPD deposition disease (see box). The pathogenesis is not fully understood. The deposition of CPPD crystals seems to have a predilection for fibrocartilage rather than hyaline cartilage. It is therefore surprising that the temporomandibular joint, a joint that is predominately fibrocartilage is so rarely affected. It was first described in the temporomandibular joint by Pritzker et al in 1976. They postulated that the paucity of reports in the literature may be due to lack of clinical distinction between the arthritides and other causes of temporomandibular joint pain.

Discussion

CPPD deposition disease is one of the many forms of crystal-related arthropathies. It is also known as pseudogout, an older but still popular term, coined to describe the presence of CPPD crystals in the joints of patients with acute arthritis resembling gout. Pseudogout is sometimes called chondrocalcinosis which identifies the radiographic evidence of cartilage calcifications. However, chondrocalcinosis can be caused by any of the crystal-related arthropathies (e.g., apatite, octacalcium phosphate, CPPD, calcium oxalate). Since CPPD crystal deposition may present a large spectrum of clinical manifestations, the term CPPD deposition disease is accepted as a more accurate designation.

The following diagnostic criteria have been established for CPPD crystal deposition disease: CPPD crystals are detected in the fibrocartilage of a joint by electron microscopy or X-ray diffraction, crystals compatible with CPPD are found using polarized light, and the patient has a diagnostic radiographic pattern. If only one of these criteria is present, a diagnosis of probable but not definitive chondrocalcinosis is made.

CPPD deposition disease occurs sporadically and appears to be more common in women, with an increasing incidence beyond the fifth decade of life. A hereditary component with autosomal dominant transmission has also been described.

It has been observed most often in the articular fibrocartilage of the menisci of the knee, the triangular fibrocartilage of the wrist and the symphysis of the pubis. It has rarely been reported in the temporomandibular joint. The synovial fluid levels of pyrophosphate are thought to be elevated in these patients, although serum and urine levels are within normal limits. However, the exact mechanism of CPPD crystal synthesis and deposition is unknown.

Pain is the most common symptom associated with CPPD arthropathy of the temporomandibular joint, followed by swelling, malocclusion, trismus and related ear problems. Pain is also the prevalent feature of temporomandibular joint internal derangement, a condition in which the aetiology may be closely related to CPPD crystal deposition disease.

The present patient had CPPD arthropathy confined to the superior joint space of the temporomandibular joint, unexpectedly sparing the lower joint space and the condyle. The temporal component, however, showed the classical radiographic signs attributed to the destructive arthropathy associated with this disease.

CPPD crystal deposition disease has been termed the great imitator for its ability to mimic other diseases. Not only can the clinical and radiographic features of CPPD crystal deposition disease be mistaken for other ailments, but its histologic characteristics may also emulate other disorders such as synovial osteochondromatosis.

For practical clinical diagnosis, the simplest and most effective method of identifying CPPD crystals, as well as distinguishing them from monosodium urate (MSU) crystals, is the use of compensated polarized light. On a wet preparation, CPPD crystals appear more pleomorphic than MSU crystals. CPPD crystals appear blue when lying parallel to the axis of the compensator but yellow when lying perpendicular to it, whereas MSU appear in the opposite configuration. CPPD crystals are weakly positively birefringent, while MSU crystals are strongly negatively birefringent. In a SEM analysis, CPPD crystals commonly appear as rod- and rhomboid-like shapes (1–6 μm) while MSU crystals appear more rod- and

CPPD crystal deposition

**Associated conditions**
- hyperparathyroidism
- hypocalciuric hypercalcaemia
- haemochromatosis
- hypomagnesaemia
- hypophosphatasa
- hypothyroidism
- aging
- trauma/surgery
- chronic renal dialysis

**Clinical and laboratory features**
- acute and chronic arthritis
- elderly (6th to 8th decade)
- usually multiple joints involved
- white blood cells in synovial fluid <50 x 10⁹/l
- normal urine/plasma levels of inorganic pyrophosphate

**Radiographic features**
- punctate calcifications
- symmetrical radio-opacities
- radiodensities in linear pattern

**Specific to temporomandibular joint**
- calcified mass anterior to condyle
- erosion and sclerosis of fossa
- misshapen condyle
Self-assessment questions


Recurrent hypoglycaemia

P H Dyer, T A Chowdhury, J Milles

A 61-year-old non-diabetic man was admitted from out-patients with a three-month history of dizziness. These would generally occur when he was hungry, and would be relieved by eating. He was on no regular medication, and had no relevant medical history. Physical examination elicited an abdominal mass in the left upper quadrant which moved with respiration. Baseline investigations showed a normal blood count, urea and electrolytes, liver function tests, thyroid function tests and a short synacthen test. Computed tomography (CT) scan showed the presence of a mass present in the mid-thorax and extending down into the abdomen, the nature of which was uncertain. During his admission, he developed fasting-induced hypoglycaemia (blood sugar 1.3 mmol/l). Insulin and C-peptide levels during the episode were undetectable. Plasma β-hydroxybutyrate levels during the episode were also suppressed (<25 µmol/l). He subsequently required regular parenteral 10% dextrose infusions to maintain normoglycaemia.

Questions

1 What are the possible causes of his hypoglycaemia?
2 What further investigations are required?
3 What is the treatment of choice?