



RESEARCH ARTICLE

RARE CASE OF SYNOVIAL CHONDROMATOSIS PRESENTING AS JAW PAIN

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ABSTRACT

Synovial chondromatosis (SC) is an uncommon disorder defined as reactive cartilaginous proliferation, characterised by synovial metaplasia (Yu, *et al.*, 2004). With intra-articular proliferation of cartilaginous nodules originating from the synovial membrane. It mainly affects large joints such as knee, hip, shoulder, and elbow. Its manifestation in the Temporomandibular joint (TMJ), is a rare finding, occurring predominantly in females. CT, MRI and arthroscopy facilitate the clinical diagnosis of SC, but pathological analysis is mandatory to confirm the disease (Martín-Granizo *et al.*, 2005). This paper reports a case of a 35 years old woman who presented with a long-standing history (1 year) of pain in right TMJ. There were no limitations to jaw opening. Initial clinical and Radiological picture done outside our hospital suggested a picture of osteosarcoma, for which she received treatment. Histological and follow-up features of this lesion diagnosed it as SC of the TMJ.

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INTRODUCTION

Synovial chondromatosis characterized by the presence of loose cartilaginous bodies, due to the abnormal proliferation of synovial membrane. These bodies can be attached or loose in the articular space. Primary occurrence in the temporomandibular joint (TMJ) is rare in a proportion of female/male 1.5:1 (Yu *et al.*, 2004; Martín-Granizo *et al.*, 2005; Koyama *et al.*, 2001; Wong *et al.*, 2001). Most of the cases are unilateral, at the right side with just a few cases of bilateral occurrence (Yu *et al.*, 2004; Wong *et al.*, 2001; Noyek *et al.*, 1977; Miller and Mertens, 2002; von Lindern *et al.*, 2002; Yokota *et al.*, 2008; Fee *et al.*, 1979; Inovay, 1962). Symptoms could mimic like that of other TMJ disorders, such as swelling on pre-auricular region, pain and limitation or deviation of mandibular movements (Koyama *et al.*, 2001; Yokota *et al.*, 2008; Schulte and Rhyne, 1969; Kusen, 1969; Trevor, 1952) Patients can also present clicking and crepitus of the joint.

Case Report

A 35-year-old female who had been experiencing pain on the right TMJ region for over a year was referred to Rajiv Gandhi Hospital and Research Centre with suspicion of malignancy by a physician.

Axial CT image (Diagnostic CT) showed multifocal calcified loose bodies (arrow) within a low-attenuated soft tissue mass medial to the right mandibular condyle. PET-CT revealed metabolically active soft tissue lesion (metabolic size – 1.9 x 1.3cm, SUV max 16.0) with calcification is seen in the right infratemporal fossa abutting the right condyle of mandible and involving right lateral pterygoid muscle. MRI was subsequently performed and showed a soft tissue mass expanding into the joint space and multiple nodules in the intra-articular soft tissue mass. The intra-articular mass had high signal intensity (SI) and the small nodules had low SI on T₁ weighted images (WI). Based on clinical and imaging findings, a presumptive diagnosis of SC of the TMJ was proposed. Histopathological assessment showed chondroid metaplasia with lobules of a chondroid lesion, individually lined by synovium in many. These lobules show sheets of chondrocytes with abundant areas of calcification and hyalinization also are seen short fascicles of spindle cells mostly at periphery, having oval to spindled nuclei dispersed chromatin, conspicuous nucleoli and moderate amount of cytoplasm, along with few scattered multinucleate giant cells. No significant nuclear atypia or pleomorphism seen in the spindled or chondroblastic component. Mitotic activity and necrosis are not seen. Therefore, we made a confirmative diagnosis of synovial chondromatosis.

DISCUSSION

SC is an uncommon, benign, monoarticular arthropathy that is characterised by the formation of cartilaginous bodies in the synovial membrane.

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This disease usually affects large joints. SC in the TMJ is rare and may show nuclear atypia, which indicates malignancy (Miller and Mertens, 2002). It occurs more often in women, which is opposite to the trend for SC in other joints, between the fourth and sixth decades of life. The clinical features of SC in the TMJ are pain, swelling, limitation of the movements of the jaw and crepitation on mouth opening. The presence of cranial nerve dysfunction indicates that the disease has reached the advanced stage (Noyek *et al.*, 1977; von Lindern *et al.*, 2002; Yokota *et al.*, 2008; Fee *et al.*, 1979). The pathogenesis of SC is unknown. Some contributing factors, such as previous trauma, degeneration, inflammatory disease and infections, have been suggested but these do not seem to be the main causes of SC. Some previous studies have suggested that fibroblast growth factor 2 and fibroblast growth factor receptor 1 play important roles in the pathogenesis of SC. The presence of bone morphogenetic proteins (BMPs) in cartilaginous nodules suggests that BMPs synthesized by the synovia and by the loose bodies are able to promote bone and cartilaginous metaplasia, contributing to the pathogenesis of SC (Martín-Granizo *et al.*, 2005; von Lindern *et al.*, 2002; Inovay, 1962; Schulte and Rhyne, 1969; Kusen, 1969). On the basis of Histopathological findings, SC can be divided into the following three stages

- Initial stage: in this stage, the lesions include metaplasia of the synovial membrane with proliferation of undifferentiated cells. Detached loose bodies are not observed at this stage.
- Transitional stage: this stage shows the presence of progressive metaplasia in the synovial membrane along with formation of loose bodies that contain active chondrocytes partially covered by the synovial membrane.
- Advanced stage: No intrasynovial metaplastic activity is detected in this stage; only detached loose bodies are observed (13).

SC can also be divided into two forms according to the cause of the disorder: primary and secondary. In the primary form, metaplastic changes develop in the synovium and cause cartilaginous foci to develop within the synovial membrane. These foci enlarge, detach and ossify in the synovial cavity. The secondary form occurs following trauma or other arthropathies. Fragments of cartilaginous or bony tissue dislodge and are nourished by the synovial fluid. The primary form is more aggressive and tends to recur. In contrast, the secondary form is usually slow and has a chronic presentation. Moreover, in the secondary form, the loose bodies are more uniform and show less cellular atypia than that observed in the primary form (Norman *et al.*, 1988). SC is a benign disease, but it does not respond to non-surgical treatment and does not show spontaneous resolution. Moreover, it has the potential to invade the intracranial structure when involved with the TMJ (Noyek *et al.*, 1977; Yokota *et al.*, 2008). Therefore, early and accurate diagnosis is very important for selecting an appropriate treatment and for a better prognosis (Balliu *et al.*, 2007).

Clinical findings often overlap with the symptoms presented by patients with joint disorders and/or parotid tumours. Imaging tools for diagnosis include conventional radiography, CT, MRI and arthroscopy. The clinical findings, mainly in association with intermittent symptomatology and slow growth rate of our patient condition suggests a benign process. The use of auxiliary image exams (radiographs, CT, MRI, and arthroscopy)

must be highlighted for differentiating from other conditions like osteoarthritis, osteochondroma, villonodular synovitis, and focal osteochondritis (Miller and Mertens, 2002). Panoramic X-ray (orthopantomography) is a good option for this assessment, providing good visualization of the surrounding structures with a lower radiation dose, compared with CT. However, this exam presents a limitation in the tridimensional location of the lesion (Inovay, 1962). The main imaging features of SC in the TMJ are as follows: widening of the joint space, soft tissue swelling, irregular surfaces of joints and multiple calcified loose bodies. Although many conditions present with loose bodies within the TMJ, one notable difference is that patients with SC have more than 10 loose bodies and these are packed in the soft tissue mass. This appearance resembles that of grains of rice in sushi (Yokota *et al.*, 2008; Von Arx *et al.*, 1988). MRI is useful for revealing boundaries, assessing internal derangement and detecting intracranial extension. The detection of skull-base changes is important because it indicates a more aggressive course that can potentially destroy the skull base and intrude into intracranial structures. Therefore, pre-operative CT and MRI are very useful for early detection of these changes and are beneficial in improving the prognosis of the patients (Yu *et al.*, 2004).

SC does not spontaneously resolve and may show recurrence. Therefore, it requires appropriate treatment and post-treatment follow up. The treatment options for SC are surgery and arthroscopy. Arthroscopy is a less invasive technique, but it is difficult to retrieve the intra-articular loose bodies using arthroscopy. In some cases, the lesion may show extra capsular and intracranial extension and SC of the TMJ has the potential to undergo malignant transformation. Therefore, the treatment of choice is surgical exploration (Martín-Granizo *et al.*, 2005; Noyek *et al.*, 1977; von Lindern *et al.*, 2002). Although rare, recurrences can occur (Balliu *et al.*, 2007). Surgical treatment should be followed by regular follow-up examinations and MRI can be useful in the follow-up period. The assessment of synovia as well as its removal seems to be associated with the prevention of SC recurrence. If physicians were aware of all these findings, the diagnosis of this condition would be greatly facilitated, thereby ensuring prompt and adequate therapy and improving the prognosis of the patients.

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