



Synovial Chondromatosis of the Temporomandibular Joint with Extension to Skull Base: A Case Report of Multidisciplinary Treatment

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Abstract

A 62-year-old female consulted to our clinic with a five-year history of pain in the right preauricular area, edema, and progressive limitation of mouth opening that was episodic, with spontaneous resolution. These episodes were not associated with any other symptoms.

Introduction

Synovial Chondromatosis (SC) is a rare tumor that originates in the synovial membrane of joints and is characterized by the production of cartilaginous nodules into the joint space. It is usually a benign condition that does not undergo malignant transformation; however it results in pain, inflammation, and loss of function of the affected joint [1].

SC usually involves long bone joints such as the knee, hip, and shoulder. SC of the Temporomandibular Joint (TMJ) is infrequent, with few cases reported to date. It occurs most often in the fourth and fifth decade of life and has been described more commonly in men. SC of the TMJ, however, appears to have a higher prevalence in women [2,3]. The etiology is still poorly understood. Patients can be classified as primary or secondary SC. Primary SC occurs in patients with no known predisposing factors. Secondary SC occurs in patients with a history of trauma or disease of the joints, such as arthritis.

SC usually presents with joint pain, swelling, and reduced mobility. Liu et al. [3] reported 10 cases of SC of the TMJ, finding that the symptoms were comparable to those of SC in other joints. Pain was the most common symptom (90.8%), followed by swelling (67.1%), limitation of mouth opening (68.4%), deviation of the jaw upon opening (34.2%), and crepitation (31.6%). These symptoms are common to several pathologies of the TMJ, and Computer Tomography (CT) and Magnetic Resonance Imaging (MRI) are essential for the differential diagnosis [3].

The cartilaginous nodules can cause damage to the articular surface leading to osteoarthritis. The treatment consists of surgical removal of loose bodies, synovectomy, and discectomy and in some cases condylectomy [4]. Very rarely, the disease can progress and cause erosion of the bone of the condyle, glenoid fossa and extend to the skull base. This is the 17th patient reported to date with SC that extends to the skull base [5]. The patient underwent multidisciplinary surgical management through an endoscopic approach and with the use of a neuronavigational system. Complete resection of the lesions and resolution of the symptoms was achieved.

Case Presentation

A 62-year-old female consulted to our clinic with a five-year history of pain in the right preauricular area, edema, and progressive limitation of mouth opening that was episodic, with spontaneous resolution. These episodes were not associated with any other symptoms. The patient reported no other inflammatory changes and no abnormal secretions from the salivary glands. During the physical examination, a facial asymmetry was noted with a solid mass that could be palpated in the right preauricular region. Oral aperture was limited to 20 mm, and deviation to the left was observed during the jaw opening that adequately corrects while performing the closing movement of the mouth. The patient had correct dental occlusion. Otoscopy and salivary gland exam were uneventful.

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Figure 1: CT scan showing a coronal (a) and axial (b) plane in a bone density window. Several irregular hyperdense tumor lesions can be seen surrounding the right TMJ and extending to the skull base.

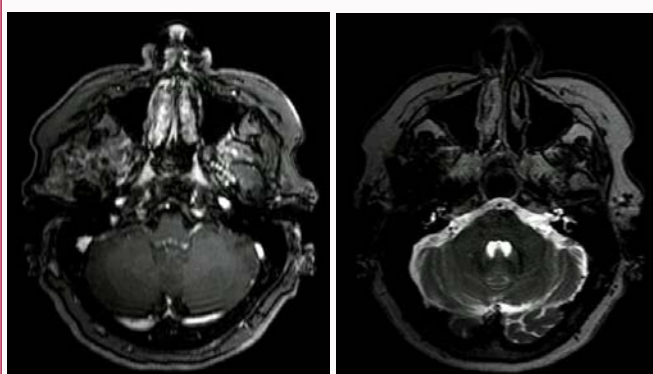


Figure 2: MRI in T1 weighted sequence with gadolinium (a) and T2 (b). The tumor shows heterogeneous enhancement with contrast and is hypointense in T2.

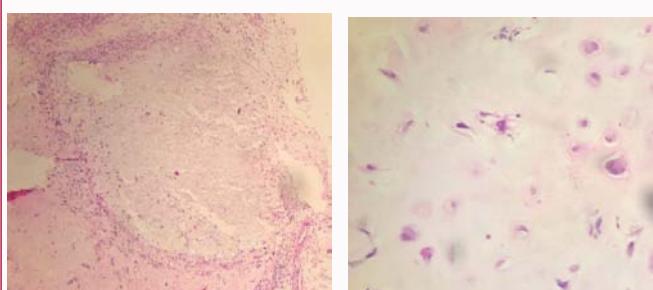


Figure 3: In the histopathological examination, a lobulated cartilaginous lesion with calcifications and chondral hypercellularity with atypia was identified.

A previously performed ultrasound showed submandibular and parotid glands of normal appearance. Conventional radiography of the TMJ showed a radiopaque heterogeneous mass on the right side. A CT scan showed multiple irregular calcified lesions in the right TMJ and sclerosis of surrounding bone structures. These altered the contour of the joint's articular surface, displaced the surrounding muscles and the parotid gland, and extended to the zygomatic process of temporal bone and skull base (Figure 1). An additional MRI scan demonstrated multiple heterogeneous hypointense lesions in T2 weighted images with marked enhancement with gadolinium. No intracranial involvement was noted (Figure 2). Biopsy confirmed synovial chondromatosis (Figure 3).

Due to the extent and location of the lesions, a multidisciplinary

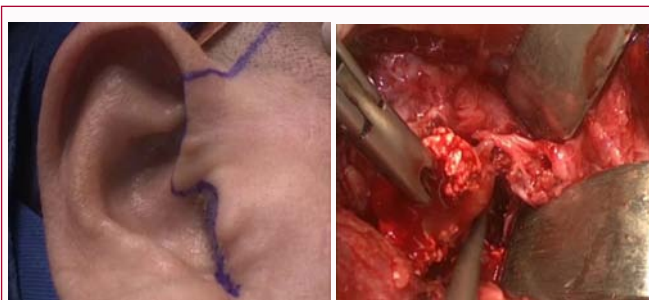


Figure 4: Modified endoaural approach (a) and endoscopic surgical vision (b).

approach was planned in conjunction with otolaryngology and skull base surgery team aided by neuronavigational system and endoscopic vision.

A modified endaural approach with a temporal extension was used to access the TMJ (Figure 4).

Extensive ossification was encountered as well as multiples cartilaginous amber colored tumors. Arrectomy, condylectomy, and meniscectomy were performed, and complete macroscopic resection of the tumor was achieved (Figure 5).

During the postoperative follow up the patient did not present complications. An outpatient control was performed at the three months, six months, and one-year mark. An increase in oral aperture up to 35 mm was observed, with absence of pain and without evidence of facial nerve involvement (Figure 6).

Discussion

Synovial chondromatosis of the TMJ is an infrequent diagnosis whose clinical manifestations of pain, swelling, and limited motion of the joint can frequently be confused with other diagnoses. Diagnostic images have an important role and can help differentiate SC from other pathological conditions. Especially CT and MRI scans detect the multiple calcified lesions within the joint and changes to the joint surface. Early detection can help patients achieve appropriate treatment and avoid damage to the TMJ. When surgical management is decided, pathological examination of the specimen will confirm the diagnosis, and is currently the gold standard. Surgery can improve pain and joint mobility. However, due to the complex anatomy of the TMJ, and the extent of the lesions, traditional approaches may be insufficient, as in the case shown above. In these situations, a multidisciplinary approach using endoscopic vision and neuronavigational systems can aid the surgeon in achieving complete tumor resection. It is vital to remove all loose bodies and synovial tissue surrounding a joint to prevent recurrence of the disease. Few studies exist on the recurrence rate of SC however; data from SC of the TMJ and other joints such as the elbow have found a recurrence rate between 1% and 18.2% [6,7]. Apart from incomplete removal of the tumor, continuing trauma can also be a risk factor for recurrence. In few cases, malignant transformation into chondrosarcoma has been described, supporting the need for appropriate, timely, and complete surgical excision of the tumor [8].

This case exemplifies the need for a high clinical suspicion, and the advantages of multidisciplinary work in the medical field. With high definition cameras, endoscopic vision, and the aid of neuronavigational technology, complete tumor removal was achieved from the TMJ and skull base.



Figure 5: Neuronavigational system allowed the surgeons to identify all of the lesions and excise the tumor in its entirety.

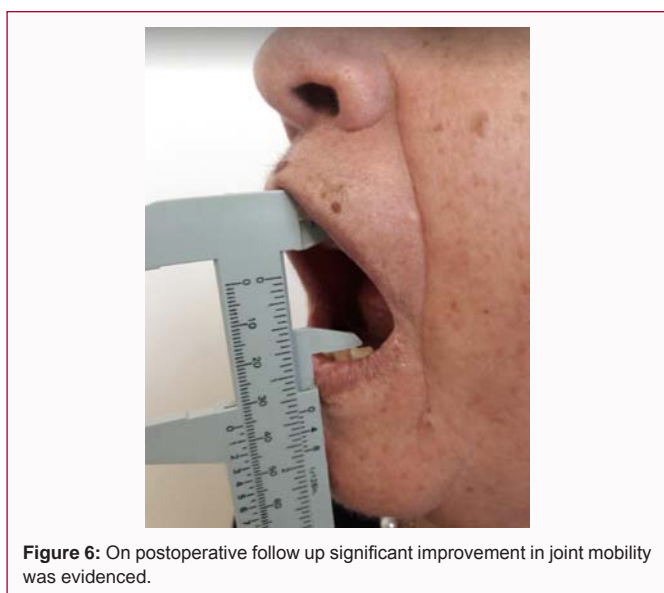


Figure 6: On postoperative follow up significant improvement in joint mobility was evidenced.

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