Case Report
Journal of Pierre Fauchard Academy (India Section), *Vol 34(2), DOI: 10.18311/jpfa/2020/25347, June 2020, p. 70-75*
ISSN (Print) : 0970-2199
ISSN (Online) : 2405-772X

Bilateral Temporomandibular Joint Ankylosis with Rare Finding of Odontogenic Keratocyst Extending into Ankylosed Segment: An Anomaly within Anomaly

Neelam Shakya[1]*, Deepak Agarwal[2], Vilas Niwaskar[2] and Vidhya Balke[3]

[1]Associate Professor, Department of Oral and Maxillofacial Surgery, Government College of Dentistry, Indore – 452001, Madhya Pradesh, India; dr.neelam2009@gmail.com
[2]Professor, Department of Oral and Maxillofacial Surgery, Government College of Dentistry, Indore – 452001, Madhya Pradesh, India
[3]Assistant Professor, Department of Oral and Maxillofacial Surgery, Government College of Dentistry, Indore – 452001, Madhya Pradesh, India

Abstract

Temporomandibular joint ankylosis is a pathological condition where the mandible fused to the glenoid fossa by bony and fibrotic tissues, which leads to a restriction of the mouth opening from partial reduction to complete immobility of the jaw. It is most commonly associated with trauma followed by local or systemic infection, tumors or systemic disease such as ankylosing spondylitis, rheumatoid arthritis, or psoriasis. This is a case report of an unusual presentation of bilateral Temporomandibular Joint (TMJ) ankylosis with rare finding of odontogenic keratocyst extending into the ankylosed segment. The CT scan revealed a well-defined with multicystic radiolucency appearance replaced the usual picture of TMJ ankylosis of a radio-opaque mass obliterating the normal architecture of the joint. Surgical exploration revealed cystic cavity filled with cheesy material within the ankylosed mass on right side of TMJ.

Keywords: Temporomandibular Joint, Odontogenic Keratocyst, Keratocystic Odontogenic Tumor

1. Clinical Representation

A 35 year old male patient, reported to department of oral and maxillofacial surgery, government college of dentistry Indore (India) with chief complaint of inability to open mouth since one and half years. Patient's history revealed that he had undergone unilateral surgical procedures of the right temporomandibular joint 10 years ago. On examination he had incision scar on right preauricular region ,and retruded chin (figure 1,2). His mouth opening started reducing gradually over a period of 7 years, now presented with complete Type IV, bilateral temporomandibular joint ankylosis. He developed swelling in front of right ear since one year. Patient was on liquid diet since one year. CT examination of TMJ ankylosis revealed an uncommon picture, well-defined multicystic radiolucency was seen within the usual picture of TMJ ankylosis of a radio-opaque mass obliterating the normal architecture of the joint, giving an appearance of an anomaly within anomaly (figure 3,4,5).

2. Differential Diagnosis

Neoplasms and pseudotumors of the temporomandibular joint (TMJ) are quite uncommon[1], Rare TMJ lesions include synovial chondromatosis, osteochondroma, osteoma, osteoblastoma, synovial cyst, simple bone cyst, aneurysmal bone cyst, epidermal inclusion cyst, hemangioma, nonossifying fibroma, Langerhans cell

*Author for correspondence

histiocytosis, plasma cell myeloma, and sarcoma[2]. Intraosseous lesions of the TMJ are rare, and their diagnosis is quite difficult without careful imaging. The radiologic findings often show an osteolytic lesion within the mandibular condyle and/or neck that may be caused by (1) primary bone cysts and tumors, (2) metastatic lesions of bone, (3) degenerative changes, and (4) metabolic lesions[3].

Traumatic bone cysts, aneurysmal bone cysts, giant cell lesions of the bone, Langerhans cell histiocytosis, and nonossifying fibroma are primary bone cysts and tumors. All show similar radiologic findings so that the final diagnosis can be made only by means of histologic examination.

In the present case during CT examination an unusual picture was revealed. It elicited expansile radiolucent osteolytic lesion within the affected joint (figure 3 and 4). The above CT examination oriented differential diagnosis to following interpretations, one that of to aneurysmal bone cyst and second of a traumatic bone cyst and third of Langerhans cell histiocytosis and fourth of mastoid air cell within the ankylotic mass. Aneurysmal bone cyst is a condition that develops secondarily within another lesion of bone, most frequently it is associated with central giant cell tumors. Usually occurs in the 2nd & 3rd decade of life and with history of trauma. Aneurysmal bone cyst will classically present as a multilocular radiolucent lesion that giving the appearance of soap bubble[4].

Traumatic bone cyst is a benign, non-neoplastic lesion, predominantly among younger age group that is below 20 years. It comprises generally single lesion without any epithelial lining, surrounded by bony walls and either lacking contents or containing liquid or connective tissue. Classically radiolucent image with irregular or scalloped but well defined margins. The size is variable and a fine sclerotic margin is sometimes seen. Now it is called idiopathic bone cavity because it persists as a cavity in bone containing no specific diagnostic elements[5]. Eosinophilic granuloma is the least severe and localized form of all Langerhans cell histiocytosis it should be considered while making diagnosis, especially when a young child or even an infant with otitis media



Figure 1&2. Extra oral frontal & lateral photograph showing retruded chin and preexisting scar over right preauricular region.

or a swelling in the mastoid region that is refractory to medical treatment. On CT, eosinophilic granuloma of the skull is typically a lytic lesion without reactive sclerosis and it is diffusely destructive[6].

Odontogenic cysts and tumors such as odontogenic keratocysts or ameloblastomas may involve these areas, but they are often associated with an impacted tooth and are usually located in the mandibular angle or body from where they may expand into the mandibular condyle. Although in the present case histopathology report came out OKC.

3. Management

The surgical treatment plan included bilateral interpositional gap arthroplasty with a sling of rotated temporalis muscle as the interpositioning material and resection of the osteolytic lesion of the right side of the joint.

Surgery was performed under general anesthesia with fiber optic nasoendotracheal intubation. after aseptic part preparation TM joint was approached by Alkayat-Bramley incision, (figure 6), after reflection of flap and exposing the temporalis fascia, the dissection proceeded from a point that is 1 cm below the zygomatic arch and 1 cm to 2 cm within the tragus, to avoid injury to the zygomatic branch of the facial nerve the dissection proceeded in this plane till the joint was reached. The fused ankylotic mass when reached was found out to be cystic cavity filled with cheesy material obliterating the ankylotic mass at the site. Surgery was continued with enucleation of cyst with lining and resection of the ankylotic mass along with coronoid process and interpositioning of temporalis muscle sling on right side. While on the left side Alkayat and Bramley incision was employed (figure 6). A 45° incision given over zygomatic arch region and the arch and ankylotic mass was exposed. 1.5 cm gap was created by removing the osseous tissue along with coronoid process followed by interpositioning of temporalis muscle (figure 7). A maximum incisal opening of 50 mm was obtained after the osteotomies.

The post-operative course was uneventful. There was no motor deficit on either side of the face (figure 8,9). The histopathologic report of enucleated cyst was suggestive of Odontogenic Keratocyst.

4. Discussion

Temporomandibular joint ankylosis is a pathological condition where the mandible fused to the glenoid fossa by bony and fibrotic tissues, which leads to a restriction of the mouth opening from partial reduction to complete immobility of the jaw. There are multiple factors that can result in TMJ ankylosis, such as trauma, local or systemic infection, previous TMJ surgery, iatrogenic causes, tumors or systemic disease, such as ankylosing spondylitis, rheumatoid arthritis, or psoriasis. Trauma is the most common cause of TMJ ankylosis, followed by infection. Clinical manifestations of TMJ ankylosis are very according to severity of ankylosis, time of onset of ankylosis and duration of ankylosis. if ankylosis develop in patients who is less than 15 years of age or patient in their growing age result in severe facial deformity and loss of function but if Ankylosis develop in patients who is more than 15 years of age or after completion of growth result in marginal or nil facial deformity but loss of function severe. Facial asymmetry is the classic feature in unilateral cases. The chin deviates towards

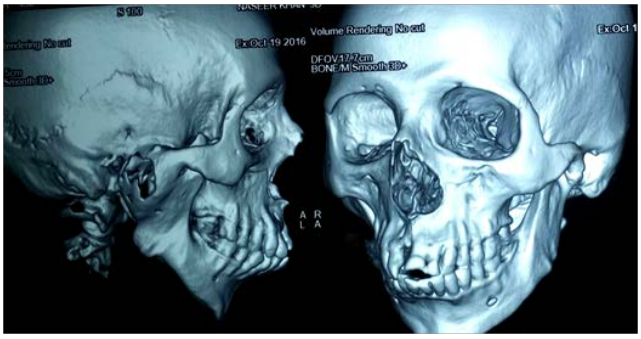


Figure 3. 3-D reconstruction showing expansile multilocular lesion on right side showing, altered morphology of coronoid and condyle and on left side ankylotic mass.



Figure 4. Coronal section showing osteolytic lesion.

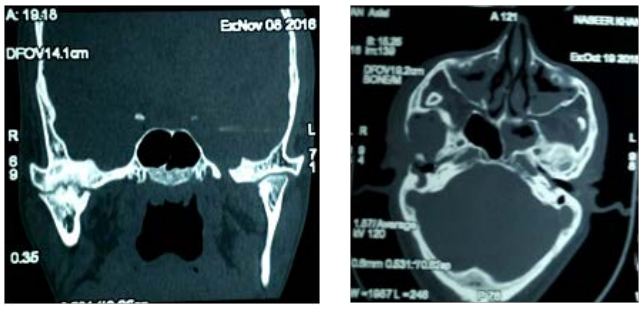


Figure 5. Coronal and axial section showing altered morphology of condyle and ankylotic mass.



Figure 6. Al Kayat Bramley incision.



Figure 7. Resection of Lytic lesion.



Figure 8. Post operative adequate mouth opening.



Figure 9. Post operative OPG.

the affected side, and the vertical height of the affected side is reduced when compared with the unaffected side. On the other hand, complete limitation of the mouth opening and symmetrical recession of the chin are typical characteristics of bilateral ankylosis.

TMJ ankylosis may be classified by a combination of location (intra- or extra-articular), type of tissue involved (bony, fibrous, or fibro-osseus) and extent of fusion (complete, incomplete). Literature classifies ankylosis as true and false. Any condition that gives rise to osseous or fibrous adhesion between the surfaces of the TMJ is a true ankylosis. False ankylosis results from pathologic conditions not directly related to the joint.

In the year 2002 (Ibrahim EL-Sayed EL Hakim and SA Metwalli)[8] proposed that ankylosed joints can be grouped to the relation of their ankylosed mass to the surrounding vital structures, especially at the base of the skull base. This new classification gives the surgeon opportunity for the careful surgical planning and achieves better surgical results with minimum operative complications.

The first goal in the management of TMJ ankylosis is the resection of the ankylosed segment and reestablishment of the mandibular movement. Additional surgical procedures are required for the reconstruction of the TMJ and atrophic ramus.

A number of surgical techniques have been reported, such as condylectomy, gap arthroplasty or interpositional arthroplasty, and reconstruction with costochondral graft, total joint prosthesis, free vascularized bone or total joint graft.

Interpositional Gap Arthroplasty is a highly effective and safe surgical management option for TMJ ankylosis with acceptable immediate and long term outcome, particularly when temporalis myofascia are used for adults and costochondral grafts with fascia interposition used for children[9,10].

A 7-step KABAN protocol has been developed for the treatment of TMJ ankylosis: 1) aggressive resection of the ankylotic segment, 2) ipsilateral coronoidectomy, 3) contralateral coronoidectomy when necessary, 4) lining of the joint with temporalis fascia or cartilage, 5) reconstruction of the ramus with a costochondral graft, 6) rigid fixation of the graft and 7) early mobilization and aggressive physiotherapy.

This protocol formed the basis of the treatment plan that was undertaken in this patient, except the reconstruction and rigid fixation of the graft.

The term Odontogenic Keratocyst (OKC) was 1st given by Philipsen in1956 and its characteristic features was 1st described by Pindborg and Henson in (1963)[11,12]. Over the years many researchers have been trying to understand the nature, identification and management of the OKC leading to classify and reclassify the disease. Previously classified under developmental odontogenic cyst of the jaw by WHO in 1971 and 1992. OKC has been reclassified and renamed as Keratocystic Odontogenic Tumor (KCOT) in WHO classification in 2005 due to its aggressive behavior, high recurrence rate and specific histopathological characteristics[11]. But recently WHO in 2017 reclassified KCOT back into cystic category.

OKC comprise approximately about 5 to 10 percent of odontogenic cyst of the jaw bone and seen predominantly in 2nd, 3rd and 4th decades of life, though then can occur in any age group. OKC have a slight male predilection. It can occur in any part of the jaw, but majority of the lesions are seen in the mandible, most commonly in the posterior body and ascending ramus.

The treatment modalities of OKC depend on patient's age, size and location of cyst, soft tissue involvement and histopathological variant of lesion. Treatments are usually classified as conservative like Enucleation with or without curettage and marsupialization and aggressive like peripheral ostectomy and chemical curettage with Carnoy's solution, cryotherapy, or electrocautery and resection.

The recurrence rates are significantly high and differ in various published articles, varying from 0% to 62%, depending on the site of involvement, type of surgical procedure used, and length of follow-up[14]. Successful treatment of OKCs is usually attributed to the use of techniques which address the margins of the initially presenting cyst, with the objective of removing all of the epithelial lining.

According to Taipale and colleagues[15], cyclopamine, a plant-based steroidal alkaloid, inhibits the cellular response to the SHH signal. They found that cyclopamine blocks activation of the SHH pathway caused by oncogenic mutation making it a potential "mechanism-based" therapeutic agent for human tumors whose pathogenesis involves excess SHH pathway activity. Zhang and others[16] postulate that antagonists of SHH signaling factors could effectively treat KCOTs. Their suggested strategies include the reintroduction of a wild-type form of PTCH, inhibiting the SMO molecule by synthetic antagonists and suppressing the downstream transcription factors of the SHH pathway. They suggest that intracystic injection of an SMO protein-antagonist has the greatest potential as a future treatment option[14,15,16].

In the present case, CT reveals complete loss of condylar architecture on the right side along with coronoid. Although OKC exhibit a locally aggressive behavior and spread via marrow spaces, the involvement of condyle and coronoid process is rarely reported. This may be due the high bone density and deficient marrow spaces in these regions. Due to its aggressive nature, Peripheral ostectomy was planned which is removal of few millimeters of bone beyond the visible margins of the lesion after enucleation. It can be carried alone or may be assisted by vital staining of the surgical bone defect. Staining is done with help of stains like methylene blue or crystal violet. The dye penetrates into cells that have an abnormal increase in nucleic acids. This helps to identify areas that have been incompletely excised during peripheral ostectomy[14]. Post-operative follow-up with annual radiological examinations is essential for at least five years following surgery. In our case patient was advised for follow up check up at an interval of every 3 months for at least 5 years.

5. Conclusion

Odontogenic keratocyst are usually seen in body and ramus region although in condylar region of mandible few cases have been reported. But its occurrence in temporomandibular joint ankylotic mass region is quite rare. So that it should be considered in differential diagnosis of cystic lesion in temporomandibular joint region.

In such cases where multiple pathologic findings are observed, a well planned treatment is to be performed following all basic principles of treatment.

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