Case Report

Infiltrating lipomatosis, an etiology for TMJ ankylosis?

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Abstract Infiltrating lipomatosis is a rare benign condition causing diffuse fatty infiltration into the surrounding soft tissue and in rare cases causes hyperplasia of the adjacent bone. We report a case with clinical and radiological evidence of a 34-year-old female patient who reported a swelling in the left middle third of the face with exophytic temporomandibular joint (TMJ) ankylosis of the left side resulting in restricted mouth opening and facial asymmetry since 21 years. The number of cases reported in the literature is rare. Surgery is the treatment of choice.

Keywords: Infiltrating lipomatosis, maxillofacial pathology, TMJ ankylosis

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INTRODUCTION

Temporomandibular joint (TMJ) ankylosis has multiple etiologic factors. It can range from 1) trauma, 2) local or systemic infections, 3) systemic diseases, etc., Mandibular hypomobility and trismus can be due to various disorders affecting the TMJ and surrounding structures. Infiltrating lipomatosis has rarely been accounted as an etiology of TMJ ankylosis. Infiltrating lipomatosis represents a distinct clinicopathologic entity characterized by collections of nonencapsulated, mature lipocytes that infiltrate local tissues and tend to recur after surgery. It was first described by Slavin in 1983.[1] The etiology and clinical behavior of the entity remain unclear. Infiltrating lipomatosis can infiltrate facial muscles, associated soft tissue, and underlying facial skeleton causing gross facial asymmetry and dental abnormalities.[2]

Possible etiologies for the lipomatous change and infiltration include somatic mutation. Multipotential cells of embryogenic origin under the influence of hormones, trauma, chronic irradiation, muscle metaplasia, or congenital cytomegalovirus infection could precipitate the somatic defect.[3,4] Facial osseous changes in infiltrating lipomatosis ranging from hyperplasia of bony components of TMJ, osteophytosis, exophytic new bone formation, elongation of the coronoid process, etc. have been accounted in the literature.[3,5]

This is a case report of a 34-year-old female who reported to our institution with the complaint of limited mouth opening and facial asymmetry. This report includes clinical and CT findings of an infiltrating lipomatous lesion in the left malar region with secondary TMJ ankylosis of the left side.

CASE REPORT

This study was approved by the institutional review board. A 34-year-old female patient reported to the Department of Oral and Maxillofacial Surgery, Vydehi Institute of

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Dental Sciences and Research Centre with the chief complaint of swelling and reduction in mouth opening since 21 years. The patient started noticing swelling in the left middle third of the face which was initially small in size and gradually increased to the present size. The patient also started noticing a gradual reduction in mouth opening over the same period. On maxillofacial examination, a diffuse swelling, roughly oval, and approximately 2 × 3 cm in the left middle third of the face was found. The swelling was soft in consistency, non-compressible, non-reducible, non-fluctuant, and non-tender [Figure 1]. On TMJ examination, restricted TMJ movements were noted on the left side. The patient had nil mouth opening when she presented to the department [Figure 2].

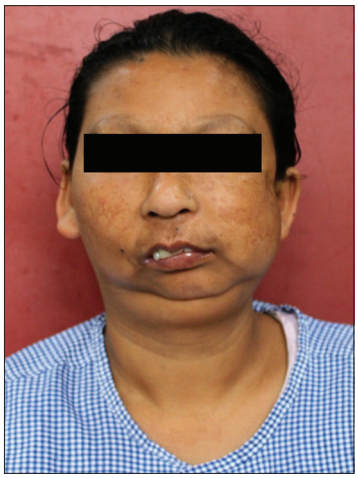

Figure 1: Preoperative frontal view

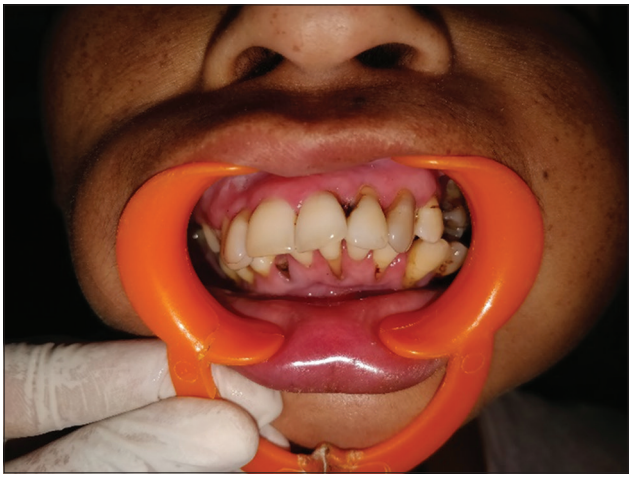

Figure 2: Preoperative mouth opening

Fine needle aspiration cytology (FNAC) was done in the left middle third of the face and was suggestive of lipoma with mature adipocytes. Panoramic, computed tomography (CT), and magnetic resonance imaging (MRI) imaging were done and were suggestive of lipomatous lesion of the left side of the face, TMJ ankylosis of the left side, and elongated coronoid process [Figures 3 and 4].

The surgical plan consisted of two stages, the initial stage was resection of the ankylotic mass and interpositional arthroplasty using temporalis myofascial flap and the second stage was Distraction osteogenesis for the correction of Ramal Condylar unit height of the left mandible along with excision of the lipomatous lesion for aesthetic correction. However, the patient wanted only functional improvement and deferred the second stage treatment for correction of facial asymmetry.

The pathology was exposed via an extraoral preauricular approach (Alkayat Bramley). Ankylotic mass was resected along with the impacted molar, and interpositional arthroplasty using temporalis myofascial flap was done along with ipsilateral coronoidectomy [Figures 5 and 6]. Intraoperatively, a mouth opening of about 35 mm was achieved. The postoperative course was uneventful, and improvement of mouth opening was noted and was advised physiotherapy [Figure 7].

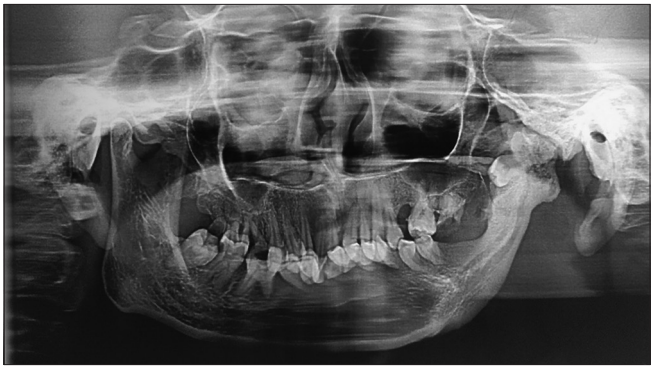

Figure 3: Preoperative radiograph: TMJ ankylosis of the left side with impacted molar within the ankylotic mass

DISCUSSION

The term "lipomatosis" refers to a poorly defined, infiltrating proliferation of mature adipose tissue. Facial infiltrating lipomatosis (IL-F) is a rare disorder characterized by a painless fatty mass with ill-defined borders, usually located in the cheek, on one side of the face.[6,7] It can occur congenital or in the early period of life. Infiltrating facial lipomatosis was first described by Slavin *et al.* (1983).[1] They described six parameters of the lesion to be as follows: (1) nonencapsulated tumors containing mature fat cells, (2) infiltration of adjacent muscle and soft tissues, (3) absence of malignant characteristics, (4) absence of lipoblasts, (5) presence of fibrous elements in conjunction with increased numbers of nerve bundles and vessels, and (6) hypertrophy of the adjacent bone.[1] It can

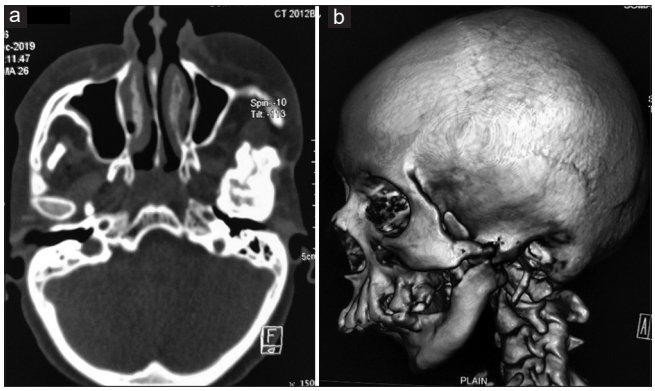

Figure 4: (a) CT axial view (b) 3D CT elongated coronoid process can be appreciated decreased Ramal Condylar unit height

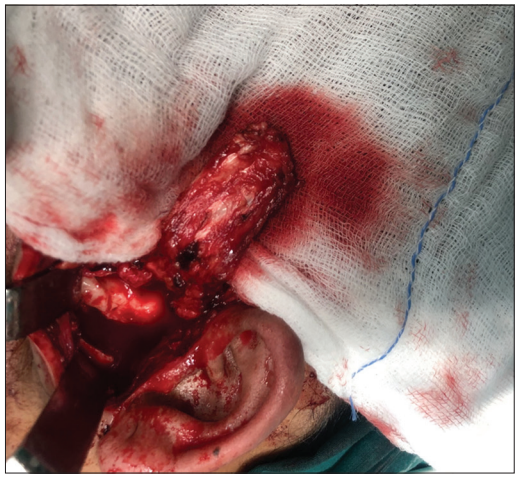

Figure 6: Resection of ankylotic mass and interpositional arthroplasty using temporalis myofascial flap

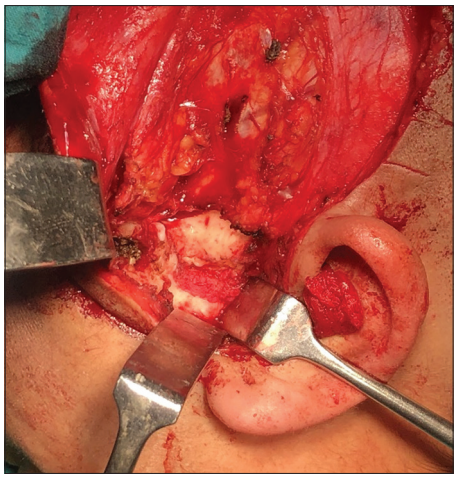

Figure 5: Intraoperative: Bony ankylotic mass in left TMJ space

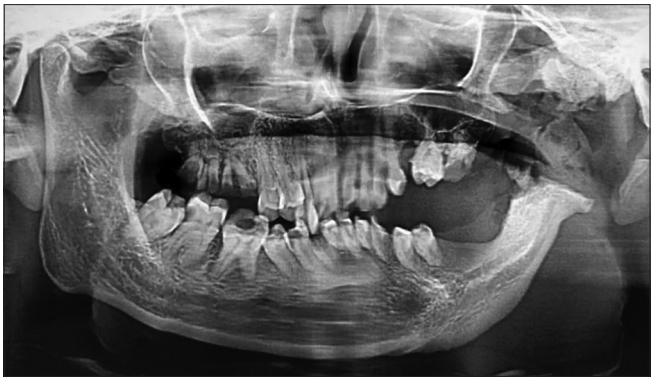

Figure 7: Postoperative radiograph

also affect other parts such as ribs and cervical vertebrae as reported by Malik *et al*.[8]

Enzinger and Weiss classified lipomatosis into three different entities: symmetrical lipomatosis (usually in the neck), pelvic lipomatosis, and diffuse lipomatosis of the limbs and trunk.[7]

The diagnosis is usually carried out by history, clinical examination, and radiological assessment. CT and MRI are the imaging tools that are important in the evaluation of lipodensity and the plane of extension and infiltration. Classic lipomas have CT and MRI signal intensity characteristics similar to those of subcutaneous fat. On CT scans, lipomas appear as homogenous hypoattenuated masses (apart from thin and wispy soft tissue attenuated septa, although occasionally, septa may be thick and nodular). They have a CT number ranging from - 60 to - 120 HU and do not typically show contrast enhancement. On MR images, fat has typical signal intensity. On T1-weighted images, lipomas tend to have a high signal intensity that diminishes with progressive T2 weighting. MRI provides better delineation because it has superior soft-tissue contrast resolution and a clear definition of the location and longitudinal extent of the mass.[9] In the present case, MRI showed poorly defined lipomatous lesion.

Other fat tissue infiltrations should also be ruled out, mainly well-differentiated liposarcoma and lipoblastomatosis. Lipoblastomatosis is characterized by the presence of fetal adipose tissue and liposarcoma by lipoblastic proliferation, pleomorphism, and a large number of cell mitoses, all absent in histopathologic examination ofCongenital Infiltrating Lipomatosis of the face (CILF) tissue.[8] Histopathological features of CIL-F include adipose infiltration of adjacent muscle and soft tissue without malignant characteristics, absence of lipoblasts, fibrous elements, increased numbers of vessels with unifocal thickened muscular walls, and increased numbers of nerve bundles of variable size with focal fibrosis.[1]

Underlying facial bone hypertrophy is a common feature in IL-F, and multiple cases have been reported in the literature. IL-F with the involvement of the mandible and the condyle on the affected side, having the clinical and

radiologic characteristics of hemi mandibular elongation, and hypertrophy was defined by Obwegeser.[10] Similar clinical and radiological features were noted in our patient. These bony changes can result from increased vascularity, periosteal irritation, or regional malformation of mesenchyme.[8] Sahai *et al*.[3] have reported clinical and CT findings in an unusual case of IL-F presenting with progressive hemifacial asymmetry, manifesting as severely restricted mouth opening owing to exophytic TMJ ankylosis.

The treatment ranges from liposuction to excision. The authors believe that the treatment plan varies according to the patient and should depend on the size of the pathology and the extent of infiltration to the underlying muscles or bone. In lipoma causing gross facial asymmetry, the treatment can be difficult to plan as excision can damage vital structures like the facial nerve. Conservative options like liposuction can be done in such cases. The timing of the surgery remains controversial. If there is no gross facial asymmetry, an option of waiting until the end of growth, the inclusion of both soft and hard tissues in the surgical plan once the growth ceases can also be considered.[11] A postresection recurrence rate of up to 62.5% has been reported during a period of up to 20 years.[12] However, the literature shows contradictory reports about the prognosis of this lesion. Although lipoma is a well-defined pathology, the etiology and clinical behavior of infiltrating lipomatosis remain unclear. Definitive management is not certain.

CONCLUSION

IL-F of the face is a rare congenital disorder, which is characterized by the presence of unencapsulated collections of lipocytes that infiltrate the surrounding facial soft tissues. The result is hyperplasia and facial asymmetry. Occasionally, the underlying bony structures are also affected. Surgery is the treatment of choice. However, relapse is common due to the diffuse nature of ILF, and the timing of surgery remains controversial.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Ethical approval

The current research was approved by the ethics committee of the Vydehi Institute of Dental Sciences and Research Centre.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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