INTRODUCTION

Aggressive fibromatosis is a soft-tissue neoplasm arising from fascial or musculoaponeurotic structures. It has also been termed extra-abdominal desmoid, musculoaponeurotic fibromatosis or desmoma.[1] Although the tumor was described in 1832, it was Sanger in 1864 who defined the characteristics that distinguish this tumor, including fibroblastic origin, local invasion of muscle and fascia and lack of a capsule.[1] It is also known as Grade I fibrosarcomas and non-metastasizing fibrosarcomas.

Aggressive fibromatosis is rare, reported to occur in between two and four cases per million inhabitants per year, representing 0.03-0.1% of all tumors.[1] The age range of affected persons spans from newborns to the elderly, with most cases appearing in the third and fourth decades.[2] Patients affected are usually pre-teens or teenagers with a poorly circumscribed, painless fibrous growth apparently arising from the periosteum or from the fascia of muscles attached to the jaw or mastoid.

Fibromatosis, also known as desmoid tumor, is a slow-growing and locally infiltrative disease caused by uncontrolled proliferation of fibrous tissue arising from deep musculoaponeurotic structures. Despite having benign histology, its aggressive local growth pattern can cause significant morbidity.[1]

Fibromatosis is a rare entity, representing only 0.03% of all neoplasms.[2] It is classified according to its anatomical position into abdominal, intra-abdominal and extra-abdominal subtypes.[1] Fibromatosis of the head and neck region comprises about 10-25% of all cases of extra-abdominal fibromatosis.[3] Attempts have been made to separate it from that arising from other parts of the body because of the unique clinical problems it can cause in relation to its critical anatomical position.[3-5] Oral and maxillofacial region is uncommon location for fibromatosis. Most occur in shoulder (22%), chest and back (17%), thigh (13%) and mesentery (10%); oral is around 2%. Oral and maxillofacial region (mostly the mandible, maxilla and the mastoid areas) shows younger peak age range (5-20 years). It is more infiltrative with its faster growth pattern. The treatment of choice is surgical resection, but its aggressive recurrence can be challenging.
growth rate and has much greater propensity into invading bone or to arise seemingly within the bone.

Fibromatosis may also attain large size and cause compression, infiltration and destruction of the adjacent structures. Such growth behavior presents severe management problems, especially in the head and neck region, where the presence of many vital structures within a small space renders the patient susceptible to the effects of the fibromatosis and also making complete excision difficult.

This article reports the clinical pathologies that affect this joint, discusses their management according to the diagnosis and presents our experience of the same.

**CASE REPORT**

**Initial presentation**
A 3-year-old boy came to our department with facial asymmetry. The history revealed that the child developed a swelling of 1.5 cm × 1 cm, oval in shape on left temporomandibular joint (TMJ) region noticed by mother just after few months after birth. The swelling gradually increased to the present size [Figure 1], which made the mother curious to report to our department.

The swelling was non-tender, asymptomatic and firm in consistency. The swelling was slightly mobile, but mouth opening was normal.

**Investigations**
A computed tomographic scan was advised, which showed a large soft lobulated tissue mass over the left side of the TMJ, anterior to the coronoid process without a clear fat plane to the surrounding tissues [Figures 2 and 3]. Blood test results were within normal limits. Audiometry was carried out to assess the functioning of the ear.

**Surgery**
The patient underwent wide excision of the mass under general anesthesia. Under standard skin preparation, local anesthesia with adrenaline infiltrated, preauricular incision [Figure 4] was placed and the lesion was excised completely. Care was taken that the extent of incision does not cross the tragal notch, keeping the facial nerve in mind. Under hypotensive anesthesia, soft-tissue dissection was done to reach the tumor mass. Tumor mass appeared to be firm soft-tissue mass with a glistering shiny surface. Soft-tissue dissection was carried out around the tumor mass to reach the origin of tumor mass. It appeared to be attached to the anterior surface of TMJ, apparently to the peristeum. Tumor was excised in total [Figure 5], rough bony surface was smoothened with a bone file. Betadine wash was given and the wound

**Figure 1:** Oval-shaped swelling at left side temporomandibular joint

**Figure 2:** Computed tomographic scan with tissue mass over left side of temporomandibular joint

**Figure 3:** Computed tomographic scan showing non-clear fat plane
was closed in layers using 4-0 vicryl (polyglactin). Skin was sutured with 5-0 monofilament polyamide. Pressure dressing was placed. I.V antibiotics were continued for next 48 h. Patient was discharged after 48 h. Patient was given paracetamol orally. Pain was negligible post-operatively. Sutures were removed after 7 days [Figure 6].

It looked like an encapsulated mass as we cut the mass in two halves. It was then sent for histopathological investigation.

**Histopathology**

Histopathological examination revealed fibromatosis characterized by proliferating fibroblasts and was very aggressive in nature, but non-encapsulated.

**Follow-up**

The child’s recovery was uneventful and there were no neurological deficits. Follow-up at 1 year showed no recurrence and the patient remains under careful follow-up. The mandible of the child was growing fine.

**DISCUSSION**

Aggressive fibromatosis generally presents as a painless swelling that has been present for less than a year, often fixed to the underlying muscle or bone but not to the skin.\(^2,4\) The long axis of this tumor is usually oriented in the direction of the muscle bundle in which it is found; there is a tendency to infiltrate the surrounding structures, often encasing adjacent vessels and nerves without apparent invasion, possibly accounting for the clinical lack of pain.\(^2,3\)

Pathologically, these tumors lack a true capsule, although they may appear well-circumscribed on imaging studies.\(^6\) Elongated, spindle-shaped cells of uniform appearance surrounded and separated by dense bands of collagen are noted.\(^6\) Mitoses are infrequent, with some variation in cellularity, but no cells with atypical or hyperchromatic nuclei.\(^6\) The primary histologic differential diagnoses are those of a low-grade fibrosarcoma or reactive fibrosis.\(^6\)

Although aggressive fibromatosis is histologically benign, the biologic behavior of the lesion is similar to that of a low-grade malignant neoplasm, with local invasion and a high rate of recurrence. Recurrence is more common when the head and neck are involved (in as many as 70% of cases) than when a similar tumor is found in other locations; it is uncertain whether this is because of a more aggressive tumor or the technical difficulty of excision in this region.\(^2\)

If a recurrence is to develop, it usually becomes apparent with in the 1\(^{st}\) year. The recurrence rate is inversely proportional to the attainment of surgically clear margins of clinically uninvolved tissues.
Fibromatosis of the head and neck is a rare disease. Patients usually present with a painless mass or less commonly, pain or neurological symptoms. Upper airway obstruction is a potentially fatal clinical presentation that requires urgent attention. As with fibromatosis arising from other parts of the body, surgical resection is the mainstay of treatment. The primary goal is to achieve a clear resection with wide margins, but preservation of function is of equal importance. Due to the complex anatomy and frequent entrapment of neurovascular structures in the head and neck region, these aims are often difficult to achieve. Resection often leads to injury of important surrounding tissues, for example, the brachial plexus and is associated with recurrence requiring repeated excisions. The reported recurrence rate ranges between 46% and 62%. The etiology of fibromatosis remains unclear. Physical, endocrine and genetic factors have all been hypothesized to play a role in the origin of fibromatosis; however, no compelling evidence supports any single factor. Many physicians have reported patients with a history of local trauma preceding the development of fibromatosis, such as previous surgery or blunt force injury. This has led to the hypothesis that immature fibroblasts engaged in healing begin uncontrolled proliferation that leads to tumor formation. However, there have not been enough patients with head and neck fibromatosis after local trauma to confirm this theory.

At present, the mainstay of treatment remains surgical excision, together with a cuff of normal tissue. Although fibromatosis may assume a nodular appearance, the infiltrative nature of the margin renders complete excision difficult, especially in the head and neck region. Reports of some centers suggest good results with initial chemotherapy protocols using agents often used for sarcomas, such as actinomycin D, cyclophosphamide and daunorubicin, followed by surgery. This approach may be advantageous in those with large tumors that approach the base of skull, making complete recession difficult or those tumors associated with a significant functional loss and morbidity.

In addition, radiotherapy may have a role to play in some large unresectable tumors and in recurrent or incompletely excised tumors. Response to radiation therapy in such situations has been documented, but it is slower than it is for epithelial malignancies. Radiation induced sarcomas have not been reported.

**CONCLUSION**

Although rare, aggressive fibromatosis should be considered in the differential diagnosis of any soft-tissue mass of the head and neck. In addition, once the diagnosis is suspected, the excellent anatomic detail provided by magnetic resonance can aid in pre-operative planning because complete primary resection carries the highest likelihood of cure. Due to the high incidence of recurrence, close follow-up is essential.

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