Abstract
Fibromatosis is a rare benign mesenchymal neoplasm which primarily originates in the muscle, connective tissue, fascial sheaths, and musculoaponeurotic structures. It is commonly seen as abdominal tumour but in maxillofacial region, the occurrence of these tumours is very rare and exceedingly rare in infra-temporal space. Often misdiagnosed due to its varied clinical behaviour, fibromatosis is benign, slow-growing, infiltrative tumour without any metastatic potential, but is locally aggressive causing organ dysfunction along with high recurrence rate. We report a case of fibromatosis involving the left infra-temporal space in a 35-year-old female who presented with chief complaint of limited mouth opening for the preceding 4 years.

Keywords: Aggressive fibromatosis, Infra-temporal, Benign, Infiltrative.

Introduction
Aggressive fibromatosis (AF) or extra-abdominal desmoid tumours are rare tumours of fibroblastic origin involving the proliferation of cytologically benign fibrocytes. AF accounts for about 0.03% of all neoplasms and >3% of all soft tissue tumours with a reported incidence of 2 to 4 cases per million population per year.2 The characteristic features of AF are slow growth, benign but locally aggressive behaviour and high recurrence rate without metastasising capacity. Moreover, the tumour can damage nearby structures causing dysfunction of vital organs and may recur even after wide resection owing to its infiltrative nature.3,4 Because of its varied clinical presentation, it is often misdiagnosed as benign fibrous lesions, fibrosarcoma or reactive inflammatory processes.3,5 In histopathological review, small bundles of spindle cells, with rare mitoses, can be seen on a background of abundant stroma.6 The treatment of choice is surgical resection with tumour-free negative surgical margins.7 However, in many cases, an attempt to achieve tumour-free margins ends up as permanent disfigurement after surgery due to the involvement or proximity of vital structures.3 Adjuvant radiotherapy and chemotherapy or non-cytotoxic drugs are also considerable modalities for AF management, to avoid sacrificing functional integrity as a price of attaining tumour-free margins.3

Case Report
A 35-year-old female visited Oral and Maxillofacial Department of Mayo Hospital on October 5, 2014. Written informed consent was obtained from the patient for publication of this report and accompanying images. Her chief complaint was of progressive reduction in mouth opening gradually for the preceding 4 years. Medical history revealed that she had extra pulmonary tuberculous lesion in left side of her neck which was treated 10 years earlier. Clinical examination revealed slight thickening and fibrosis of left cheek with zero mouth opening. Overlying skin was normal. There was no...
tenderness on palpation and no associated cervical lymphadenopathy but a scar mark on the left side of neck in muscular triangle region was evidence that the area had been surgically explored previously.

On magnetic resonance imaging (MRI) there was a soft tissue density mass measuring 45×27×31 mm in CC×T×AP dimensions occupying left infratemporal fossa and left maxillary antrum, hypointense on T1, mixed signal intensity on T2 with significant post contrast enhancement. Medially the lesion was limited by the medial wall of maxillary antrum with no extension into nasal cavity. Superiorly it was limited by the floor of orbit. Inferiorly, it extended up to alveolar process of maxilla. Anteriorly it was limited by maxilla. Posterolaterally it was extended into infratemporal fossa involving muscles of mastication. Laterally ramus of mandible appeared intact (Figure-1).

Incisional biopsy under the clinical diagnosis of tuberculous lesion was performed from antrum region through Caldwell-Luc approach, but no definitive diagnosis was obtained. As such, treatment plan was modified to surgical exploration of the area and excisional biopsy of the lesion under general anaesthesia.

After fiberoptic intubation, surgical exploration was done using Weber Furgoson's incision with left infraorbital extension along with osteoplastic flap of left zygomatic bone. The lesion was found to be involving of infratemporal space extensively with an erosion of lateral and anterior walls of the maxillary sinus. All muscles of mastication of left side were also fibrosed (Figure-2). Surgical resection of the lesion with excision of involved muscles of mastication was performed and 30mm intraoperative mouth opening was obtained. Tissue was submitted for histopathological examination in two specimens. Specimen #1 maxillary sinus lesion and specimen #2 infratemporal lesion. Histopathology of specimen #1 revealed fragments of benign respiratory mucosa with mild chronic inflammation. While microscopy of specimen #2 revealed a hypocellular spindle cells lesion composed of bland spindle cells arranged in a densely collagenised background. No atypia or increased mitotic activity was noted. Immunohistochemical stain B-catenin was positive. All these findings were suggestive of fibromatosis of infratemporal space.

Our patient received post-op radiation therapy and was followed in the out-patient clinic regularly for 6 months with no evidence of recurrence of the disease that was assessed by MRI after 6 months post-operatively (Figure-3).

**Discussion**

Fibromatosis is a histologically benign tumour of musculoaponeurotic structures that can present in any part of the body. Commonly it involves the subcutaneous tissue, the muscles and the neurovascular structures. In our case along with these structures bone was also involved which is a rare finding.8

Different synonyms, including 'extra-articular desmoids', 'desmoids tumours', 'grade-1 fibrosarcomas', 'non-metastasising fibrosarcoma' and 'aggressive fibromatosis' are used in literature for fibromatosis of oral and maxillofacial region.6,9 Classification as superficial and deep fibromatosis are on the basis of location of the lesion anatomically. The superficial fibromatoses are small, slowly growing lesions that usually don’t involve the deep structures. While, the deep fibromatosis are rapidly growing lesions that are large in size and have a higher recurrence rate after treatment, thus known as "aggressive fibromatosis".2,5,9 Fibromatosis is extremely rare in head and neck region, especially of the infratemporal fossa. One study reported an incidence of head and neck fibromatosis as 11.1% of all extra-abdominal fibromatosis. Most common site for head and neck fibromatosis is the supraclavicular fossa (40%-85%), followed by face (about 25%).9 To date, according to our knowledge, there are only five such reported cases of fibromatosis that involve the infratemporal fossa.4,6,8,9 Another study reviewed an experience with fibromatosis of the oral and perioral region in 1994. Surprisingly, there...
was no tumour of infratemporal fossa in their series of 68 cases.6 The most common presentation is a painless slow-growing mass or swelling, but it can cause significant dysfunction through involving the important structures. Trismus is the most important sign and chief complaint of all reported cases of infratemporal fibromatosis.4,6,8,9 Aggressive fibromatosis is a diagnostic dilemma which remains in grey zone between benign and malignant lesions. Differential diagnosis include mandibular fibroma, well-differentiated fibrosarcoma, fibrosing histiocytoma, and inflammatory reactive process.5,6 The golden standard in the diagnostics of AF is the nuclear or membrane expression of beta-catenin, which is found in 90% of the cases.5 In our case also, modalities like MRI and ultrasound-guided fine-needle aspiration cytology (FNAC) failed to give any picture of problem and conclusive diagnosis was based on histopathology of excised lesion, and positive expression of beta-catenin.

Current management of aggressive fibromatosis involves a multidisciplinary approach. The treatment of choice is surgical resection with tumour-free margins that is often difficult to achieve because of the tumour’s extent and invasiveness, because of the infiltrative nature of these tumours, local recurrence rates after excision may be high. External-beam radiotherapy has been shown to improve local control of aggressive fibromatosis, both in adjuvant and primary settings.2

One retrospective analysis of patients with aggressive fibromatosis during a 5-year period (2008 to 2012) concluded that in the treatment of aggressive fibromatosis, preservation of form and function should be given greater priority in all age groups. It also revealed importance of postoperative adjuvant therapy in patients with gross or microscopic residual tumour to obtain progression-free survival.10 Others recommended aggressive surgery, followed by postoperative radiotherapy.2 One study demonstrated in many cases achieving free margins often involves disfiguring surgery. Thus, in these cases the use of chemotherapy/non-cytotoxic drugs can be an alternative.3

In our case we also did resection, preserving the function and avoiding disfigurement. We achieved function of adequate mouth opening, and administered post-op radiotherapy to avoid recurrence.

Conclusion
Aggressive fibromatosis of infratemporal region is quite rare often causing organ dysfunction i.e., trismus. Its diagnosis is exceedingly difficult due to its variable clinical behaviour and limited surgical approach to take a biopsy specimen. Preservation of form and function is more important than to achieve clear margins because it is benign and slow-growing. Furthermore, adjuvant therapies are available that have proved to be beneficial in recent literature.

References