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JUVENILE FIBROMATOSIS: A REVIEW AND REPORT OF TWO CASES

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ABSTRACT

Juvenile fibromatosis is a rare, benign but locally aggressive growth of fibrous tissue that mainly affects children. It rarely involves the jaws and it can be difficult to diagnose because it looks like odontogenic tumor or cyst on examination and X-rays. This paper describes two unusual cases in young children. The first was a 9-year-old girl with a painless swelling in mandible. The second was a 3-year-old girl with a lesion in maxilla. X-rays didn't give clear clues, so doctors first thought it might be an odontogenic tumor. But tissue samples showed a mix of fibrous and cellular tissue with spindle-shaped cells growing invasively—classic signs of juvenile fibromatosis. The paper also includes a quick literature review highlighting how rare this is, and the diagnostic headaches it causes. Spotting it early and confirming with pathology is crucial for the right treatment and avoid recurrence.

KEYWORDS: Juvenile fibromatosis; Aggressive fibromatosis; Desmoplastic fibroma; Mandible; Maxilla; Pediatric jaw tumors.

INTRODUCTION

Juvenile Mandibular Fibromatosis (JMF), is a rare, benign fibroblastic proliferation thought to be arise from the fascia, musculoaponeurotic, or the periosteum structures in the mandible.[1] Lesions occurring in the head and neck region are classified as deep, extra-abdominal fibromatoses.[2] Lesions within the bone are termed Desmoplastic Fibromas (DF).[3] DF shows a distinct predilection for young individuals with an annual incidence of 2 to 4 per million population.[4,5]

JMF often poses a diagnostic challenge because is rare, easily misinterpreted, and its histologic features.[6]

We took cases from 1994 to 2025 and our rare and challenging cases of aggressive fibromatosis.

CASE HISTORY

Case report1- A 09-year-old girl reported to the Department of Oral Pathology and Microbiology with a chief complaint of painless hard swelling in the lower left angle and body of the mandible since 1 year. She was asymptomatic, medical and familial history was non-contributory. Dental history indicated a tooth extraction performed 9 months ago, the details of which were not disclosed by the patient.

Corresponding extraoral examination revealed a diffuse solitary swelling of size 3 cm × 2 cm on the left side of the face. The swelling was extending Antero posteriorly from the corner of mouth to angle of mandible and supero inferiorly from malar region to the inferior border of the mandible. Overlying skin shows no discoloration. [Figures 1.1 and 1.2].

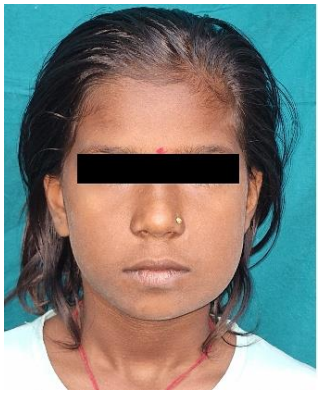


Figure 1.1: Clinical image showing extra oral view depicting swelling on the left side of the face with asymmetry.

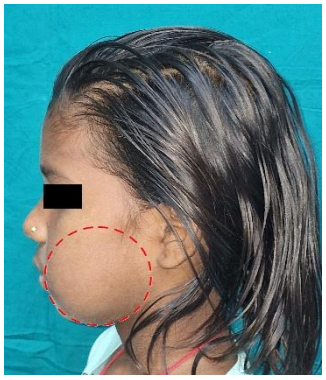


Figure 1.2: Lateral view showing a single diffuse swelling on left side of the face.

On intraoral examination, a single diffuse swelling measuring 1 cm × 2 cm in size extending from distal to 34 region with obliteration of buccal vestibule was noted [Figure 1.3].

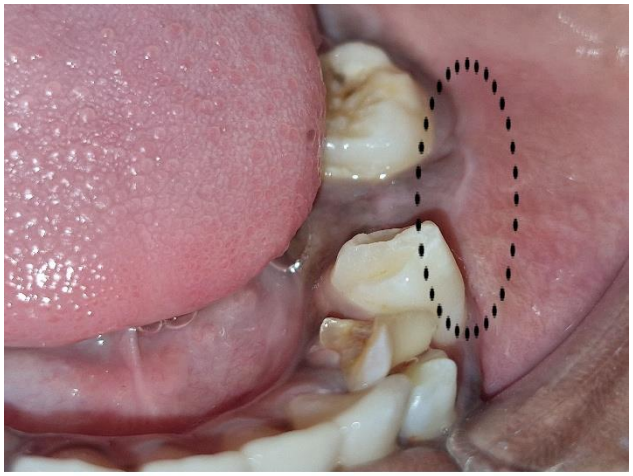


Figure 1.3: Clinical image shows intra oral view with obliteration of the buccal vestibule.

A working diagnosis of ameloblastoma was made, and the patient was advised for radiological and haematological investigations. Haematological investigations were within the normal limits. Orthopantomogram revealed a single well defined radiolucent appearance extending from distal to 34 region to till mesial to 36 region [Figure 1.4]. An incisional biopsy was done and received for histopathological evaluation.

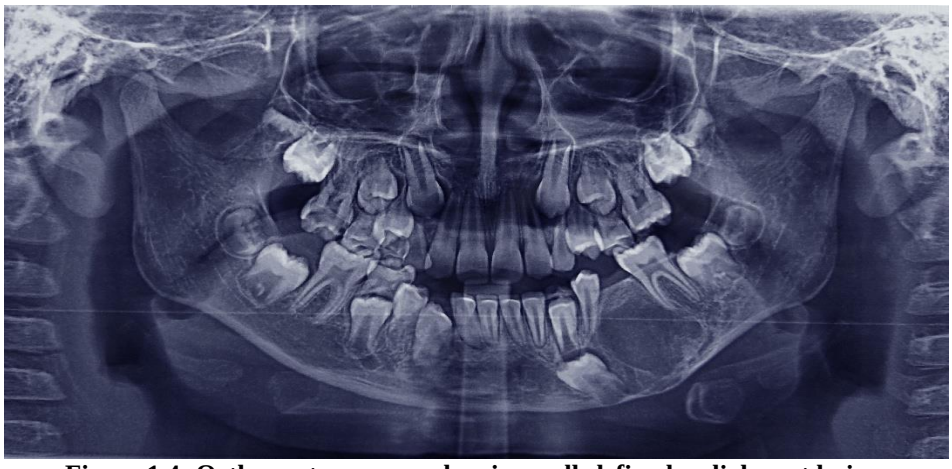


Figure 1.4: Orthopantomogram showing well-defined radiolucent lesion.

Microscopic examination of the incisional biopsy showed that the lesional tissue was devoid of epithelium and composed of a fibro-cellular connective tissue stroma. This stroma consisted of proliferating spindle cells with thin, wavy, serpentine nuclei and pointed ends, intermingled wire-like collagen fibrils. Fibroblasts were interspersed throughout, along with small and large endothelial-lined blood vessels, and a few chronic inflammatory cells, chiefly lymphocytes and plasma cells [Figure 1.6]. In this case, positive expression IHC marker vimentin [Figure 1.7] and negative expression IHC markers of SATB2 [Figure 1.8].

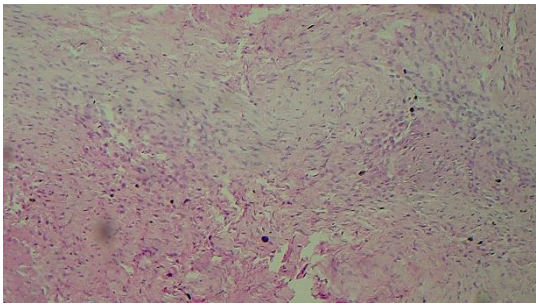
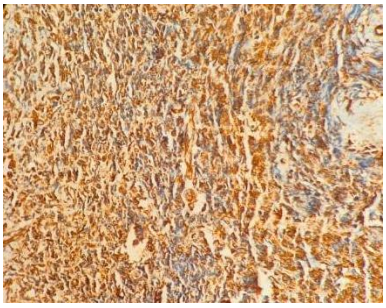
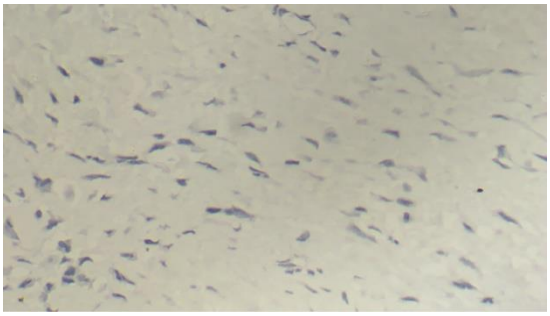


Figure 1.6: Histopathological image showing proliferating spindle cells with thin wavy serpentine nuclei and pointed ends intermingled with nerve fibers. (H&E, ×100).



[Figure 1.7]: Positive expression of IHC markers of Vimentin.



[Figure 1.8]: Negative expression IHC markers of SATB2.

CASE REPORT-2

A 03-year-old girl reported to the Department of Oral Pathology and Microbiology with a chief complaint of painless hard swelling in the upper left maxilla since 6 months. The girl was asymptomatic, medical, familial and dental history was non-contributory.

Corresponding extraoral examination revealed a diffuse solitary swelling of size 3 cm × 2 cm on the left side of the face. The swelling was extending anteroposteriorly from the corner of mouth to zygomatic process and superoinferiorly from lateral nasal bone region to the inferior border of cheek. Overlying skin shows no discoloration. [Figures 2.1 and 2.2]. On intraoral examination, a single diffuse swelling measuring 1 cm × 2 cm in size extending from distal to 62 and involvement of 63 with palatal swelling was noted [Figure 2.3].

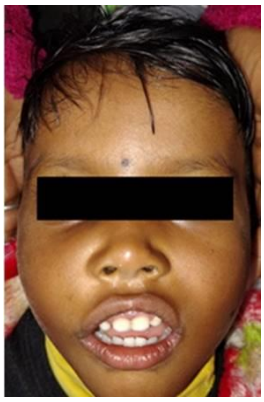


Figure 2.1: Clinical image showing extra oral view depicting swelling on the left side of the face with asymmetry.

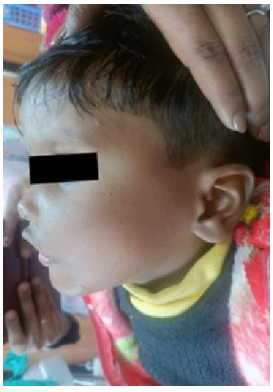


Figure 2.2: Lateral view showing a single diffuse swelling on left side of the face.

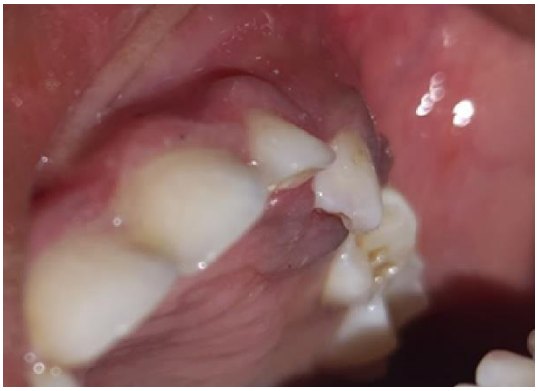


Figure 2.3: Clinical image shows intra oral view with palatal swelling with respect to #63.

A working diagnosis of adenomatoid odontogenic tumor was mad, and the patient was advised for radiological and haematological investigations. Haematological investigations were within the normal limits. Occlusal radiograph revealed a floating canine and root resorption in left maxillary region [Figure 2.4]. An incisional biopsy was done and received for histopathological evaluation.

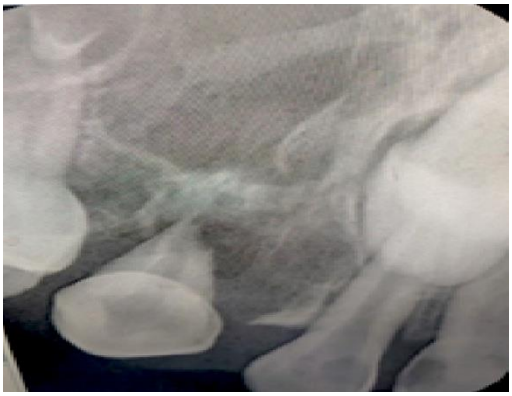


Figure 2.4: Occlusal Radiograph showing floating canine.

Microscopic examination of the incisional biopsy showed that the lesional tissue was devoid of epithelium and composed of a fibro-cellular connective tissue stroma. This stroma consisted of proliferating spindle cells with thin, wavy, serpentine nuclei and pointed ends and collagen fibrils. Fibroblasts were interspersed throughout, along with bony ossicles, small and large endothelial-lined blood vessels, and a few chronic inflammatory cells, chiefly lymphocytes and plasma cells. [Figure 2.5]

In this case, immunohistochemical marker of S100 showing negative expression [Figure 2.6]

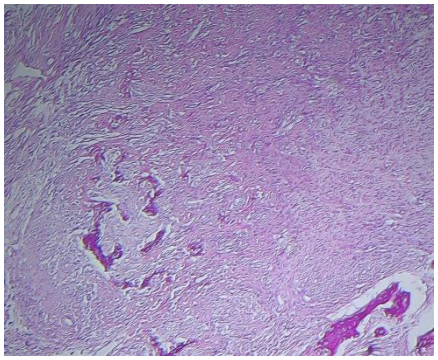
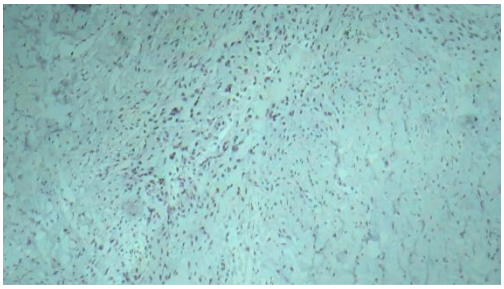


Figure 2.5: Histopathological image showing proliferating spindle cells with thin wavy serpentine nuclei, bony trabeculae at places.(H&E, ×100).



[Figure 2.6]: Negative expression IHC markers of S100.

Table 1: Juvenile Fibromatosis Case Reports from 1994-2025.

Location	Age/Sex	Clinical Features	Provisional Diagnosis	Differential Diagnosis	Histo-Pathological Diagnosis	Immuno histochemistry	Authors/years
Maxillae	4/Male	Fixed, Rapidly enlarging, ulcerative soft tissue mass of palate.	-	-	Aggressive fibromatosis	-	Sue et al 1994[10]
Right Mandible	4/Male	Painless growth of the right mandible.	-	-	Fibromatosis	-	Ahmet Kutluhan et al. 2002[11]
Right Mandible	7/Female	After injury rapidly growing right lower jaw.	-	-	Aggressive fibromatosis	-	Watzinger et al. 2005[12]
Right Mandible	8/male	Rapidly growing painless mass.	Reactive process	-	Desmoplastic Fibroma	-	Nasser Said-Al-Naief et al. 2006[13]
Left Mandible	5/Male	Painless diffuse swelling with mild facial asymmetry	Benign soft tissue tumor	Peripheral ossifying fibroma, peripheral giant cell granuloma, peripheral odontogenic fibroma	Aggressive fibromatosis	-	Keerthi Krishnankutty Nair et al. 2017[14]
Left Mandible	9/Female	Swelling without symptoms	Acute odontogenic infection	Odontogenic cyst, ameloblastoma, adenomatoid odontogenic tumor, odontogenic fibroma, aggressive fibromatosis, desmoplastic fibroma, fibrosarcoma, and malignant fibrous histiocytoma	Aggressive fibromatosis	Positivity for catenin β antibody	Dinkova et al. 2019[15]
Left Mandible	4/Male	Growing painful swelling	Odontogenic tumor, central giant cell granuloma	Neurofibroma, Neurilemmoma, Solitary fibrous tumor, Desmoplastic fibroma (Histopathological differential)	Aggressive fibromatosis	Vimentin positivity, mild positivity for CD34. Negative for S100, NSE, CD1a	Sivanandham S, et al. 2021[16]
Left Mandible	2/Male	painless firm swelling	-	-	Infantile fibromatosis	beta catenin, desmin, CD31.	Raghani MJ et al 2023[1]
Left Mandible	15/Male	Painful progressive swelling	Acute pulpitis of tooth #37	low-grade spindle cell sarcomas, schwannoma, neurofibroma	Aggressive fibromatosis		J. Yang et al. 2025[4]
Left Mandible	9/Female	Hard painless swelling	Odontogenic Cyst	Odontogenic cyst, ameloblastoma, adenomatoid odontogenic tumor.	Juvenile fibromatosis	Vimentin Positive,SATB2 negative.	Current Case 1
Left Maxilla	3/Female	Firm painless swelling	Odontogenic Tumor	Ameloblastoma, adenomatoid odontogenic tumor, dentigerous cyst.	Desmoplastic Fibroma	S100 negative	Current Case 2

DISCUSSION

Fibromatoses are a group of fibrous connective tissue lesions, morphologically classified as benign neoplasms.[7] These tumors or tumor-like growths have no clear etiology, but trauma, endocrine factors, and chromosomal abnormalities have been suggested as possible causes for the development of these tumors.[8,9] They do not metastasize to distant sites but demonstrate locally aggressive and infiltrative behaviour. The rarity of these tumors poses challenges in both diagnosis and management.

The etiology of aggressive fibromatosis is not known, but several factors have been suggested to play a important role in its development, which include physical factors such as trauma, x-ray exposure, hormonal and genetic. Hormonal changes, particularly during pregnancy or with the use of oral contraceptives, may increase the likelihood of developing aggressive fibromatosis. In addition to this genetic factors are strongly implicated, as mutations in the adenomatous polyposis coli (APC) gene lead to Familial Adenomatous Polyposis (FAP), an autosomal dominant condition that markedly increases the risk by nearly 1000 time of aggressive fibromatosis.[14]

Clinically most cases are present with painless, progressive swellings, though some of it has rapid growth, pain, ulceration, or onset following trauma, often leading to provisional diagnoses such as odontogenic infections, cysts, benign soft-tissue tumors, or even low-grade sarcomas. The differential diagnoses were broad, reflecting significant overlap with odontogenic, neurogenic, and spindle-cell lesions.

Histological distinction between Aggressive Fibromatosis (AF) and fibrosarcoma is crucial yet challenging due to their differing therapeutic approaches. Fibrosarcomas, whether infantile or adult, typically exhibit a herring bone growth pattern, increased cellular proliferation, zonal necrosis, and hemorrhage—features generally absent in fibromatoses. In the present case, the patient's age effectively rules out infantile fibrosarcoma, which is usually congenital or arises within the first year of life.

Immunohistochemistry played an essential role, with β-catenin and vimentin positivity frequently aiding diagnosis, while negative S100 staining helped exclude neural tumors (Dinkova et al., 2019; Sivanandham et al., 2021). The studies investigated various IHC markers and predominant of them showed positivity for Vimentin, β-catenin (see Table 1), and no single marker can be reliably used to confirm the diagnosis. Future studies should be carried out to identify a reliable and specific IHC marker in the detection of Aggressive fibromatosis.

In summary, histopathological identification of JMF can be difficult. Referral to a specialized histopathology centre is recommended when aggressive fibromatosis (AF) is suspected. Accurate diagnosis, requiring adequate biopsy and comprehensive clinical and histological evaluation, is crucial for optimal management. The literature indicates that surgery is the preferred treatment for JMF of the head and neck involving the mandible, with cases demonstrating that insufficient excision frequently results in recurrence.[1] Although extensive resection of a benign lesion is a difficult decision, the potential for a lethal outcome necessitates complete removal of JMF with histologically confirmed tumor-free margins (R0 resection).[9] Where bone involvement exists, surgical treatment must include the affected mandibular portion—ranging from removal of the periosteum or parts of the cortical bone to partial mandibulectomy for more widespread disease. Consequently, JMF may be misdiagnosed as desmoplastic fibroma due to their comparable histopathological characteristics.[4] If the extraosseous component is missed under the assumption of an osseous desmoplastic fibroma, incomplete excision may lead to recurrence of JMF.

In pediatric patients, surgical treatment poses additional challenges due to the need for harmonious facial growth, with the mandible being integral to this process.[4] Preserving facial development post-resection is critical, and immediate reconstruction can help minimize the functional and aesthetic impact of tumor removal. All therapeutic decisions must balance the benefits of treatment against the typically painless nature and minimal functional impairment associated with these lesions.

Fibromatosis and desmoplastic fibroma in the maxillofacial region, illustrating the rarity, variable presentation, and diagnostic challenges associated with these lesions in children.

The mention two cases included in this study further enrich the literature, particularly the rare maxillary desmoplastic fibroma, highlighting the importance of correlating clinical, radiographic, histopathological, and immunohistochemical features for accurate diagnosis and management.

CONCLUSION

Aggressive fibromatosis of the maxillofacial region, though rare, requires a high degree of clinical suspicion and a multidisciplinary approach involving both surgeons and pathologists for accurate diagnosis and effective management.

Further studies are needed to establish reliable immunohistochemical (IHC) markers for early and precise detection of aggressive fibromatosis. Regular and long-term follow-up is essential to identify any recurrence at an early stage and to ensure proper functional and facial development in growing children. Overall, this cases reinforces that timely diagnosis, adequate surgical resection, and continuous follow-up

are the cornerstones in successfully managing aggressive fibromatosis of the mandible and maxilla.

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