

## **Juvenile aggressive fibromatosis of the oral cavity treated conservatively: A case report.**

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### **Abstract**

**Fibromatosis comprises a broad group of benign fibrous tissue proliferations having similar microscopic picture and whose biologic behavior lies between that of benign fibrous lesions and fibrosarcoma. It is characterized by infiltrative growth and a tendency toward recurrence; however, these lesions never metastasize. Fibromatosis are classified as superficial and deep. Superficial are slow growing and of small size. The deep type is aggressive, grows quickly, involves deep structures and penetrates extensively mimicking malignancy. Head and neck lesions are considered as deep fibromatosis and often called as juvenile aggressive fibromatosis. Pertaining to the high recurrence rate wide surgical excision is the favored choice of management. Here, we report a case of Juvenile aggressive fibromatosis of the oral cavity in a 10 year old boy involving right side of the mandible affecting the eruption and normal positioning of the permanent tooth bud. Lesion was operated with conservative surgical approach. Two years follow up exhibited positive outcome without any local recurrence.**

**Keywords:** Extra-abdominal desmoids, Fibromatosis, Juvenile aggressive fibromatosis.

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### **Introduction**

Fibromatosis comprises a broad group of benign fibrous tissue proliferations having similar microscopic picture and whose biologic behavior lies between that of benign fibrous lesions and fibrosarcoma. Like fibrosarcoma, the fibromatosis are characterized by infiltrative growth and a tendency toward recurrence; however, these lesions never metastasize [1]. The term fibromatosis is not applicable to nonspecific reactive fibrous proliferations which do not show infiltrative growth or recurrence [2]. A number of different forms of fibromatosis are recognized throughout the body and they often are named based on their particular clinicopathologic features. Fibromatosis are classified as superficial and deep. Superficial are slow growing and of small size. The deep type is aggressive, grows quickly, involves deep structures and penetrates extensively mimicking malignancy [3]. Head and neck lesions are considered deep fibromatosis and often called as juvenile aggressive fibromatosis or extra-abdominal desmoids [3,4]. Bony counterpart is called as desmoplastic fibromatosis [4]. Considering the aggressive nature of the

lesion and complete surgical excision is suggested as the preferred choice; however, conservative surgical approach also not overlooked. This article reports a case of juvenile aggressive fibromatosis involving the oral cavity in a 10 year old boy which was treated conservatively.

### **Case Report**

A 10 year old boy approached our hospital with a chief complaint of growth in the oral cavity since 6 months. Patient was well oriented, conscious, afebrile with normal gait and attitude. The history revealed that the lesion was small initially and has grown to the current size slowly. It was a painless growth throughout. Medical history and family history was insignificant. Extraoral examination showed normal facial symmetry. Lymph node examination revealed no lymphadenopathy. Intraoral examination showed an exophytic, sessile growth in the mandibular posterior region which was firm on palpation. The lesion was of size approximately  $1.5 \times 1.5 \times 1.0$  cm and was extending from distal of 84 to mesial of 46 antero-posteriorly (Figure 1). The lesion had grown in the space of 85 and had displaced 46 distally. Part of root piece

of 85 was visible beneath the lesion. Overlying mucosa was of normal color and texture. Radiographically Orthopantomograph showed superficial bony erosion by the lesion. Also, developing tooth bud of 45 was pushed mesially (Figure 2). Based upon clinical and radiographic finding a provisional diagnosis of benign fibrous lesion was set and incisional biopsy was carried out. Incisional biopsy was reported as angiomatous hyperplasia.

Considering the young age and growing tooth bud of 45 the conservative surgical approach was taken. The whole lesion was removed along with root piece of 75 sparing the tooth bud of 45. Surrounding tissue was thoroughly curetted (Figure 3). The whole tissue was sent for histopathological examination. Histopathology revealed poorly circumscribed tissue comprised of interlacing and streaming fascicles of collagen fibres and fibroblasts. Nuclear atypia and mitotic figures were absent. Based upon the findings the diagnosis was rendered as fibromatosis. Two years follow up was uneventful without any recurrence. Eruption of 45 is noticed which

is the positive outcome of the management of lesion (Figures 4-6).

## Discussion

Aggressive fibromatosis, first identified by Stout in 1954, is a benign non-encapsulated lesion of mesenchymal origin, with a tendency for local spread along fascial planes [5]. Fibromatosis are twice as common in children as adults and 25% cases occur in children less than 15 years old [6]. Extra-abdominal type is predominantly seen in the second and third decades. Only 5% of all cases affect the head and neck [7]. Soft tissue fibromatosis of head and neck exhibit rapid or insidious growth, and is presented as firm, painless mass. Intraorally the lesion may occur at any site; however, paramandibular soft tissue region is the preferred site. Lesion can grow to a considerable size to cause facial disfigurement and destruction of adjacent bone as well [3].

The etiology of fibromatosis remains unknown. One theory has speculated that, as these tumors are characterized by high levels of estrogens and pituitary gonadotropins some endocrinologic disturbances are involved in their development. Another theory postulated that the



Figure 1. Intraoral photograph showing sessile growth

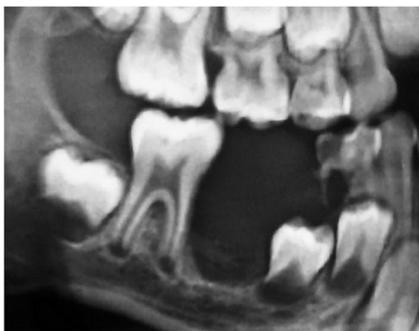


Figure 2. Panoramic radiograph showing superficial erosion of bone on right side of the mandible

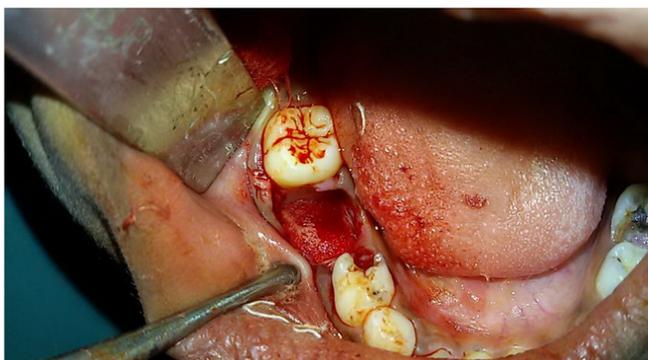


Figure 3. Intra-operative photograph after removal of the lesion



Figure 4. Panoramic radiograph after 18 months follow up



Figure 5. Clinical photograph after 2 years follow up

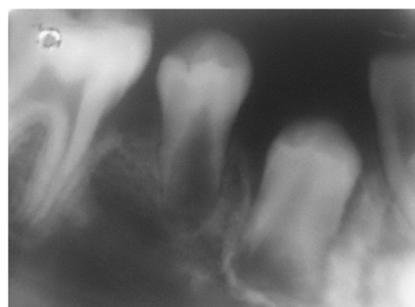


Figure 6. Radiograph after 2 years follow up

regulation of the fibroblasts activity and proliferation is carried out by steroidal hormones. However, males and females are equally affected [8]. Role of trauma or surgery in the region was also assessed by Hoos et al. and Morioka et al.; however, contradictory results were obtained [5,9]. A viral theory was also proposed, but failed to isolate the virus [10]. Aggressive fibromatosis have been associated with hereditary syndrome like Gardner's syndrome. Based on X-chromosome inactivation pattern, aggressive fibromatosis appear to be monoclonal disorder, indicative of a neoplastic rather than an inflammatory fibrous reactive process [11].

Histopathologically fibromatosis shows cellular proliferation of spindle shaped cells that are arranged in streaming fascicles and are associated with a variable amount of collagen. The lesion is usually poorly circumscribed and infiltrates the adjacent tissues. Hyperchromatism and pleomorphism of the cells is absent [2,4]. Our case also exhibited similar histopathology.

Wide and complete surgical excision is suggested as the preferred choice for treating fibromatosis to decrease the recurrence rate [12]. However, some authors have suggested a more conservative surgery in order to decrease the morbidity [13]. Other adjunctive treatment modalities are also used such as hormonal therapy, chemotherapy and postoperative radiotherapy to improve local control rate [14]. Recurrence is the serious issue for the fibromatosis. A 46% to 62% recurrence rate is reported in the head and neck region [8]. In our case considering the young age of the patient we opted for conservative surgical approach to avoid sacrifice of permanent tooth bud as these teeth are important for growth and development of jaw bones and occlusion. Two year follow up showed no recurrence clinically and radiographically as well. Eruption of 45 was the noteworthy observation.

## Conclusion

This case report emphasizes the conservative management of juvenile fibromatosis. Surgical excision with clear margins is the treatment of choice; however, in this case considering the young age of the patient conservative surgical approach was taken to avoid sacrifice of permanent tooth bud and its concomitant bad effect on the growth of the mandible. Early intervention may improve the prognosis. However, high recurrence rate of these lesions cannot be overlooked. Hence, a long term and strict follow up is advised.

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