

# Desmoplastic Fibroma of Maxilla: Case Series of A Rare Entity With Review of Literature

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## Abstract

Desmoplastic fibroma (DF) is a rare non-metastatic yet infiltrating and destructive primary tumor of the bone. This fibrous benign lesion exhibits behavior similar to desmoid fibromatosis of soft tissues. It frequently affects young adults and long bones and is infrequently seen in the craniofacial region. Amongst jaws, mandible is most frequently involved. The present case series reports varied presentations of desmoplastic fibroma involving maxilla, a very rare location. The diverse clinical and radiographic features presented by the lesion have been elaborated along with its management.

**Keywords:** Desmoplastic fibroma; Maxilla; Jaw tumor; Non-odontogenic tumor.

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## INTRODUCTION

Desmoplastic fibromas (DF) are rare myofibroblastic tumors comprising fewer than 0.1% of all the bone tumors [1]. Griffith and Irby in 1965 were the first to report DF involving the jaws [2]. These central lesions have rarely been encountered in craniofacial region [1, 3]. Mandible is far more commonly involved than maxilla with neither sex at greater risk. Desmoplastic fibromas tends to occur more frequently in adolescents and young adults [4-6]. They are aggressive lesions depicting unconventional presentations from poorly defined to well defined slow growing lesions with varied clinical manifestations. Histopathologically they are composed of spindle cells (fibroblasts/myofibroblasts) on a richly collagenous, variably hyalinized background [6].

### Case Report - One

A 23 year old male patient reported with a painless swelling concerning the right upper jaw of six months duration with no other notable anamnesis. A large diffuse swelling was noted over the right zygomatico-maxillary complex region. No nasal obstruction was evident. Intraorally, swelling extended from the maxillary lateral incisor to second molar obliterating the buccal vestibule. Expansion of hard palate from the premolars to molar region was observed. Swelling was non tender, non-fluctuant with

no local rise in temperature, bony hard in consistency with bicortical expansion.

Conventional radiography revealed a poorly defined osteolytic lesion involving the right maxillary antrum, destroying the walls of antrum along with pterygomaxillary fissure and orbital floor. Contrast enhanced computed tomography (CECT) showed an enhancing soft tissue density mass measuring 4.1 x 3.2 cms (45 Hounsfield units) occupying the right maxillary antrum area. The mass had destructively invaded the alveolar process of maxilla, ethmoidal air sinuses, nasal cavity and inferior and medial walls of the right orbit. Areas of interspersed radiopacities were noted conspicuously within the soft tissue mass.

Incisional biopsy and histopathological evaluation showed interlacing spindle cell fibroblasts with benign nuclei and without mitotic activity and collagen. Areas of hyalinization and fibrosis were present. There was entrapped normal lamellar bone without osteoblastic rimming. A definitive diagnosis of desmoplastic fibroma was rendered.

Hemi-maxillectomy using Weber-Fergusson incision and reconstruction with temporalis muscle with fascia flap by hemicoronal incision was carried out. The patient showed no evidence of post operative

complications and no recurrence was noted on a 5 year follow up.

### Case Report - Two

A male patient aged 47 years reported with painless swelling over left maxillary gingiva- alveolus region for the past one year. No other contributing history was evident. On examination, a diffuse hard swelling was noted over the left side of the upper lip obliterating the nasolabial fold. Intraorally, a roughly oval solitary swelling was apparent over the labial gingivo-alveolus region extending from maxillary canine to first premolar. The swelling had obliterated the buccal vestibule, caused migration of adjacent teeth and showed no secondary surface changes. It was a non-tender swelling with no areas of decortication. The left maxillary canine and first premolar were mobile.

Conventional radiography revealed a diffuse radiolucent lesion measuring 2x1.5 cms with corticated borders in the periapical region of maxillary canine and first premolar. Advanced destruction of the interdental alveolar bone in between the canine and premolar along with migration of both teeth was observed.

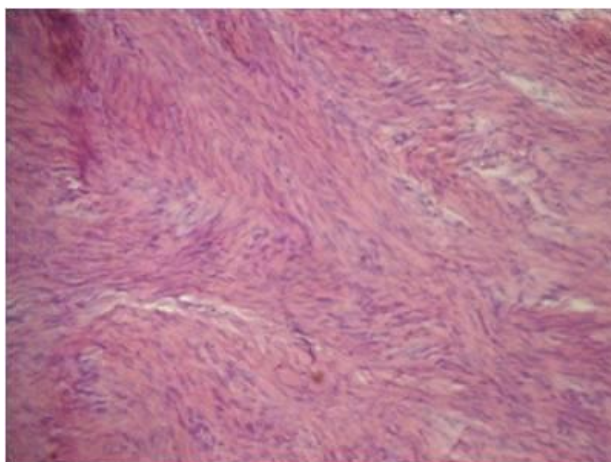
Complete excision of the mass along with buccal cortical bone and extraction of maxillary canine and first premolar was performed. Histopathology showed similar picture as in case one but without evidence of entrapped bone, suggestive of desmoplastic fibroma. No recurrence has been noted on 3 years of follow up.



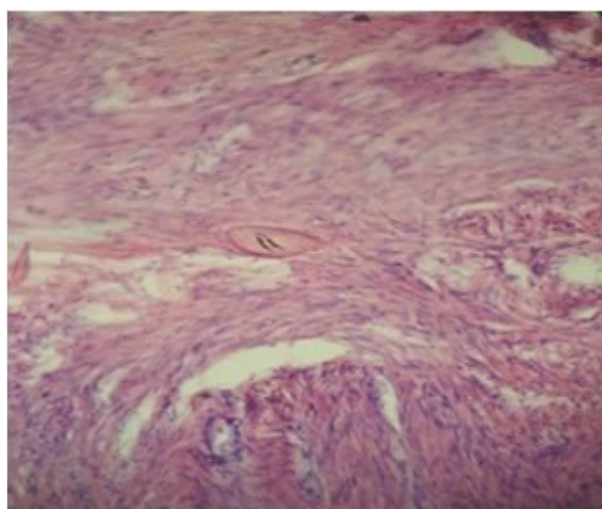
**Fig-1: Photograph of the patient showing the extraoral swelling on right middle third region of face**



**Fig-2: CT (coronal scan) showing osteolytic lesion and extension of soft tissue mass into orbit, nasal cavity and into buccal cheek region**



**Fig-3: Photomicrograph (Hematoxylin-eosin stain; x 400) showing elongated fibroblasts with abundant collagen**



**Fig-4: Photomicrograph (Hematoxylin-eosin stain; x 400) showing elongated fibroblasts with abundant collagen and ossified area without osteoblastic rim**



**Fig-5: Reconstruction with temporalis muscle and fascia flap after surgical hemi-maxillectomy**



**Fig-6: Intraoral photograph showing a well defined tumor arising from maxillary gingival region**



**Fig-7: Intraoral periapical radiograph showing diffuse radiolucency involving the interdental and periapical alveolar bone of premolar and molar**

## DISCUSSION

Desmoplastic fibroma (DF) of the jaw represents the intrabony counterpart of desmoid fibromatosis of abdomen. It may occur in a wide age range spanning from 2<sup>nd</sup> to 6<sup>th</sup> decade with a mean age of 25 years [7, 8]. Equal sex distribution has been usually seen in most reviews. However, contrasting male and female predominance has been documented in literature [10]. Occurrence of lesions exclusively in jaws have shown slight female predilection [9].

DF is a rare entity with incidence of 0.003% to 0.1 % [9, 10]. It involves the pelvic and long bones more frequently besides the mandible. Among the jaw bones maxilla is a very rare site with incidence of only 14% in contrast to 86% in the mandible [9, 11]. A silent slow growing swelling or rarely a painful swelling or combination of both may be the initial symptoms. Rarely these lesions cause pathological fractures which are more common in long bones. Both the cases presented in this series were male patients in 2<sup>nd</sup> and 4<sup>th</sup> decade respectively and presented with asymptomatic swellings in the maxilla.

A large number of lesions can be considered in differential diagnosis of such slow growing bony swellings such as fibro-osseous lesions, benign odontogenic and non-odontogenic tumors of bony origin as well as cysts.

Radiologically desmoplastic fibromas have no pathognomonic appearances. Often they are described as osteolytic lesions with destruction of cortical bone, marginal sclerosis and pseudotrabeulation. TR Woods found that among 152 reviewed cases majority (56%) of the DF were ill-defined while 42% were well defined lesions. Cortical expansion or destruction was observed in 74 % of the cases. Interspersed trabeculation may give an impression of mixed lesions further complicating the radiographic diagnosis. In larger lesions, computed tomography helps in better evaluation of the tumor extension and aids in treatment planning. MRI shows hypointense signals on both T1 and T2 images [1, 5, 6, 9, 10]. Among the two cases presented, one was a massive lesion involving whole of the maxilla in a young patient with well defined mixed



radiological features whereas the other was a localized maxillary gingival swelling in an elderly patient with a diffuse radiolucency in the alveolar bone. This highlights the contrasting clinical behavior and radiographic presentation of desmoplastic fibroma of the jaws.

Histologically these lesions consist of benign fibroblasts and abundant collagen fibres. The fibroblasts may be seen in an interlacing or fascicular growth pattern. Epithelial rests are never noted. Fibroblasts are small, thin and slender with indistinct borders and their cytoplasm blend with the supporting collagen. Multinucleated giant cells are occasionally seen. Desmoplastic fibromas do not produce any calcifications; however bony spicules may be seen if the biopsy represents the interface between the tumor and adjacent bone [5, 12, 13].

The bone forming pathologies show a rim of active osteoblasts encircling the new area of ossification which is distinctively lacking in desmoplastic fibroma and fibrous dysplasia. This may be attributed to the fact that the rarely encountered ossified areas in DF are just scattered resorbing bone fragments, wherein ossifications of fibrous dysplasia are due to metaplasia. In fibrous dysplasia, multiple irregular lamellar bone trabeculae are seen arranged in a specific form which is not noticed in desmoplastic fibroma. [13]

The biologic behavior and histopathologic features of desmoplastic fibroma resembles low grade fibrosarcoma to a great extent. However characteristic 'herring bone' appearance due to fascicular arrangement of fibroblasts, pleomorphism, spindle cells, increased mitotic activity and scanty collagen are seen in fibrosarcoma which may help in distinguishing it from desmoplastic fibroma [13].

Fibrous dysplasia and low grade osteosarcoma are other closely resembling pathologies. Fibrous dysplasia shows more cellularity with longer fibroblasts and areas of calcifications which may be characteristically arranged in 'Chinese letter pattern'. Low grade osteosarcoma also has areas of bone formation and more cell atypia which aids in discriminating it from DF [5, 13]. Odontogenic fibromas show areas of odontogenic cell rests demonstrating their origin which is absent in DF [5]. Immunohistochemistry has proved to be of not much use in further diagnosis and differentiating desmoplastic fibromas from others.

Surgery has been the treatment of choice for this locally aggressive lesion. Curettage of these lesions has recurrence chances as high as 70% and enucleation upto 40%. Hence resection with wide margins/segmental resection are considered as treatment of choice. Hemisection of the jaw has also been reported as an option for larger lesions. Chemotherapy has

shown no success while radiotherapy carries the risk of mutagenic transfer and hence both the treatment modalities are not recommended [10, 12].

## CONCLUSION

Desmoplastic fibromas are rare benign lesions with hostile behavior commonly seen in long bones along with higher affiliation to mandible when seen in jaws. Present paper reports two cases occurring in maxilla with differing clinical behaviors which vindicate considering these rarities in the list of differential diagnosis. Comprehensive knowledge about the pathology, behavior and presentation of Desmoplastic fibroma is indispensable for a precise diagnosis and apt management.

## REFERENCES

1. Jamali, M. (2013). Desmoplastic fibroma: report of rare lesion in unusual craniofacial location. *New York State Dental Journal*, 79(3), 43-45.
2. Griffith, J. G., & Irby, W. B. (1965). Desmoplastic fibroma: report of a rare tumor of the oral structures. *Oral Surgery, Oral Medicine, Oral Pathology*, 20(2), 269-275.
3. Majumder, A., Gupta, S., Friedman, P., & Isaacson, B. (2015). Desmoplastic fibroma of the temporal bone. *Otology & Neurotology*, 36(7), e119-e120.
4. Guruprasad, Y., & Chauhan, D. S. (2013). Desmoplastic fibroma of mandible. *Journal Cranio Max Dis*, 2:93-95
5. Regezi, J. A. (2002). Odontogenic cysts, odontogenic tumors, fibroosseous, and giant cell lesions of the jaws. *Modern pathology*, 15(3), 331-341.
6. Fornasier, V., Pritzker, K. P. H., & Bridge, J. A. 'Desmoplastic fibroma of bone'. In: "World Health Organization Classification of Tumour - Pathology and Genetics of Tumours of Soft Tissue and Bone" Edited by Christopher D. M., Fletcher, K. Krishnan Unni Fredrik Mertens. PUB: IARC Press International Agency for Research on Cancer (IARC) 69008 Lyon, France, 288.
7. Inwards, C. Y., Krishnan Unni, K., Beabout, J. W., & Sim, F. H. (1991). Desmoplastic fibroma of bone. *Cancer*, 68(9), 1978-1983.
8. Gebhardt, M. C., Campbell, C. J., Schiller, A. L., & Mankin, H. J. (1985). Desmoplastic fibroma of bone. A report of eight cases and review of the literature. *The Journal of bone and joint surgery. American volume*, 67(5), 732-747.
9. Woods, T. R., Cohen, D. M., Islam, M. N., Rawal, Y., & Bhattacharyya, I. (2015). Desmoplastic fibroma of the mandible: a series of three cases and review of literature. *Head and neck pathology*, 9(2), 196-204.
10. Evans, S., Ramasamy, A., Jeys, L., & Grimer, R. (2014). Desmoplastic fibroma of bone: a rare bone tumour. *Journal of bone oncology*, 3(3-4), 77-79.
11. Crim, J. R., Gold, R. H., Mirra, J. M., Eckardt, J. J., & Bassett, L. W. (1989). Desmoplastic fibroma of

- bone: radiographic analysis. *Radiology*, 172(3), 827-832.
12. Shukul, V. K., Saxena, S., & Shankar, B. G. (2004). Desmoplastic Fibroma: Mandible. *MJAFI*, 60:307-309.
  13. Angela, C. C. (2014). 'Bone pathology'. In: Neville, B. W., Damm, D. D., Allen, C. M., & Bouquot, J. E. Textbook of oral and maxillofacial pathology. 3<sup>rd</sup> Edition. Philadelphia, PA, Saunders; Reprint-India, 613-677.