Desmoid Fibromatosis – A Rare Case Report

Dr. Amol Karagir*, Dr. Shridevi Adaki, Dr. Shrivardhan Kalghatgi***, Dr. Tanushree Dalvi******

*Assistant Professor, **Associate Professor, Dept Of Oral Medicine And Radiology, ***Associate Professor, ****Assistant Professor, Dept Of Public Health And Community Dentistry, B. V. D. U. Dental College, Sangli – 416414

Abstract: Desmoid fibromatosis is a rare tumor affecting bone. This tumor histologically and biologically mimics the extra-abdominal desmoid tumor of soft tissue. This is locally infiltrative and aggressive in nature. The reported incidence of such cases is around 2–4 per million population which accounts for 0.03% of all neoplasms. In maxillofacial region, incidence is less than 3% of all cases. Treatment of such tumors is surgical excision and chances of recurrence are more. We hereby report a case of a 23-year-old female patient with desmoid fibromatosis in the mandibular posterior region. [Karagir A Natl J Integr Res Med, 2021; 12(2):69-73]

Key Words: Desmoid tumor, Desmoplastic fibroma, Desmoid-like tumor

Author for correspondence: Dr. Shridevi Adaki, Associate Professor, Department Of Oral Medicine & Radiology, BVDU Dental College, Sangli- 416414 E-Mail: doc_shridevi@yahoo.com

Introduction: The fibromatosis is a generic term which is used to describe a group of benign fibroblastic lesions with similar microscopic features but with diverse biological behavior[1]. Fibromatosis is classified as superficial and deep fibromatosis[2]. Deep fibromatosis is further classified based on location into abdominal, extra-abdominal and intra-abdominal desmoids[2].

Desmoid fibromatosis (DF) is a rare tumor. The reported incidence of such cases is around 2–4 per million population which accounts for 0.03% of all neoplasms[3, 4]. DF is a connective tissue malignancy which is deep seated and locally aggressive in nature, developing in musculoaponeurotic tissues[5].

Synonyms of this lesion are aggressive fibromatosis, deep fibromatosis, musculoapo neurotic fibromatosis and desmoid tumor. DF tends to be more common in females and most common age of occurance is between 15 and 60 years[6]. DF is commonly seen in the extremities, abdominal wall and abdominal mesentery[7]. DF has a high propensity for recurrence but, it lacks metastatic potential. Therefore, DF has now been classified as “intermediate, locally aggressive” tumor in the WHO classification of soft tissue tumors. DF in the oral and maxillofacial regions is rare, representing less than 3% of all the cases. The pathogenesis of DF is multifactorial.

Suspected causes include trauma and endocrine factors leading to dysregulation of the connective tissue, along with this, there is possibility of genetic predisposition.

DF does not seem to have familiar tendency, still its incidence is higher in case of Gardner syndrome, familial desmoid tumour and familial adenomatous polyposis[8]. The mainstay of treatment is still surgical excision, although the chances of local recurrence rates after surgical resection range from 10% to 80%[9]. Chemotherapy and non-cytotoxic agents are sometimes considered as alternative strategies[9].

We hereby report a case of a 23-year-old female patient with desmoid fibromatosis in the mandibular posterior region.

Case Report: 23 years old female patient reported to the department of Oral Medicine and Radiology with the complaint of swelling in the left lower back teeth region since 2 yrs and an associated pain in the region of swelling since one month. The patient realized a slow growing swelling since 2 yrs. There was no history of trauma preceding the appearance of swelling.

The swelling was insidious in its onset. The patient gives history of biting on hard food that initiated the pain.

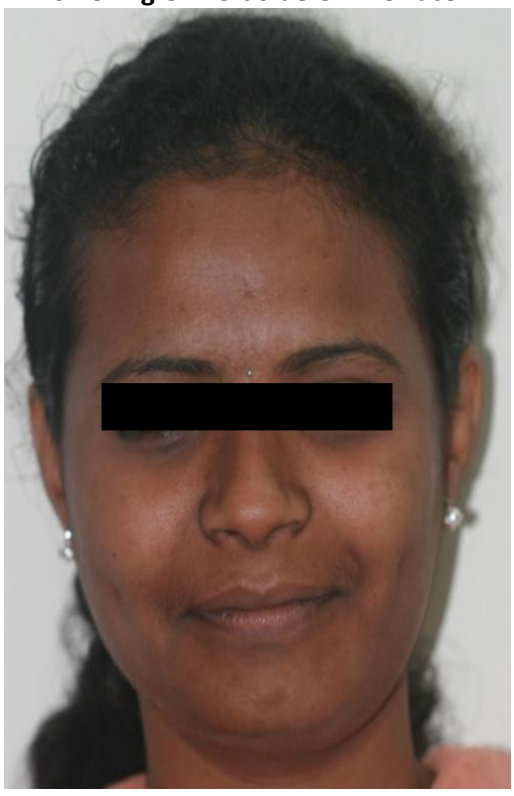
The swelling grew very slowly to attain the present size and shape. Patient gives history of pain, which was dull in nature localized at the site of intra – oral swelling since one week.

There was no history of restriction or difficulty in and / or mouth opening or movements, intra – oral discharge, no history of paraesthesia or loss of body weight.

Past dental history revealed history of restoration and endodontic treatment with mandibular right 2nd molar and left 2nd molars 2yrs back. There was no relevant medical history and family history. General physical examination revealed the patient was conscious, co-operative, well oriented and afebrile with normal gait, built and posture.

Extraoral examination (Fig 1) revealed facial asymmetry which was due to swelling on left side of mandible. Swelling was dome shaped at the mandibular body region measuring 4cm X 3cm, extending anteriorly till corner of mouth, posteriorly reaching angle of mandible, superiorly alveolar region while inferiorly extending into submandibular area. On palpation swelling was hard, firm, non fluctuant, non compressible, non pulsatile, and nontender. The overlying skin was of normal in color, and there was no localized increase in temperature.

Fig 1: Extraoral Photograph Showing Diffuse Swelling On Left Side Of The Face



Lymph nodes were not palpable and there was no abnormality on temporomandibular joint examination. Intra-oral examination (Fig 2) revealed restoration with 37 and 47 and dental caries was seen with 36. There was no intra oral swelling seen but left buccal vestibular obliteration was palpated, suggestive of cortical plate expansion.

Fig 2: Intraoral Photograph Showing Buccal Vestibular Obliteration In Relation To 36.

Based on history and clinical examination periapical cyst in relation to 36 was given as provisional diagnosis and central gaint cell granuloma, ameloblastoma, benign cyst or tumor were considered for differential diagnosis.

Radiographic investigations were planned. Panoramic radiographs (Fig 3) revealed normal condylar morphology in relation to the articular eminence on both right and left side with normal bone pattern throughout the mandible. A solitary large mixed radiopaque and radiolucent lesion covering entire left body of mandible extending anteriorly till distal aspect of 33, posteriorly till mesial aspect of 37, superiorly reaching till alveolar process and inferiorly expanding inferior border of mandible surrounded by well defined corticated border. Mandibular Occlusal radiograph (Fig 4) revealed a solitary large mixed radiopaque and radiolucent lesion covering entire left body of mandible and buccolingual cortical plate expansion seen.

Fig 3: A Solitary Large Mixed Radiopaque And Radiolucent Lesion On Left Body Of Mandible Surrounded By Corticated Border.

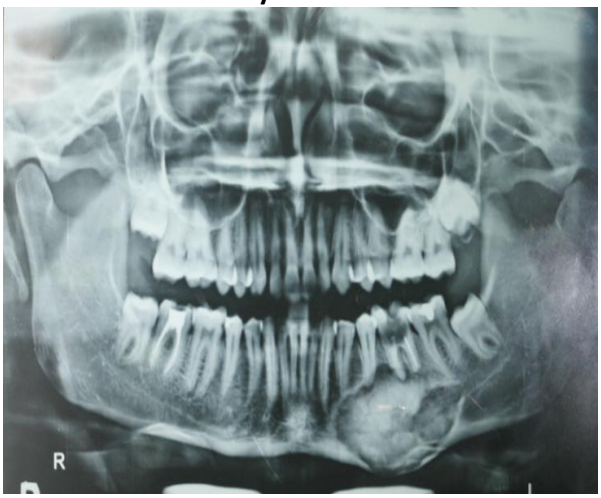
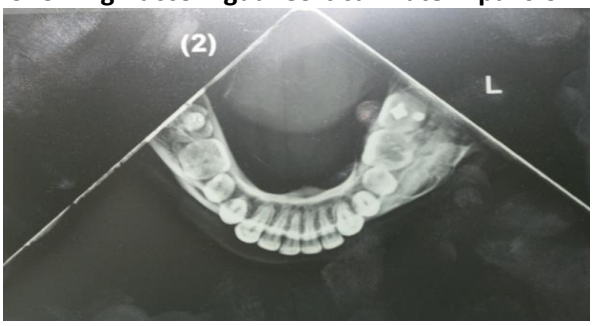
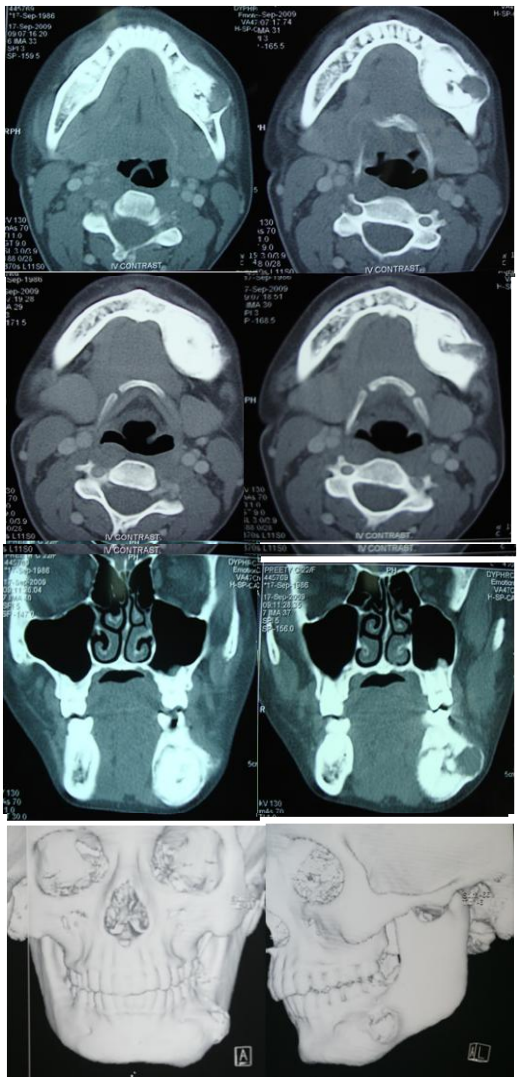


Fig 4: Mandibular Occlusal Radiograph Showing Buccolingual Cortical Plate Expansion.



Computed tomography (Fig 5) revealed well defined, lytic- sclerotic lesion measuring approximately 2.4 X 3X 4 cm in the horizontal ramus of left mandible involving the alveolar cortex in the region of the left canine, premolar, molar teeth. Lesion is causing mild expansion of the alveolar cortex and shows a well defined hypodense rim.

Fig 5: CT Showing Well Defined, Lytic- Sclerotic Lesion Causing Mild Expansion Of The Alveolar Cortex And Shows A Well Defined Hypodense Rim.



Biopsy was done and tissue was sent for histopathological examination. Histopathology revealed mature fibrous connective tissue, hypo and hyper cellularity. Spindle-shaped fibroblasts were seen with uniform long nuclei in an abundant stroma of collagenous matrix suggestive of desmoid fibromatosis. Based on history, clinical features, radiographic and histopathologic findings, desmoids fibromatosis was given as the final diagnosis. Patient referred to department of Oral Surgery for the surgical resection of left body of mandible. Surgical resection of mandible was done (Fig 6) and segments were stabilized by placing the surgical plates and screw (Fig 7).

Fig 6: Photograph Showing Excised Specimen

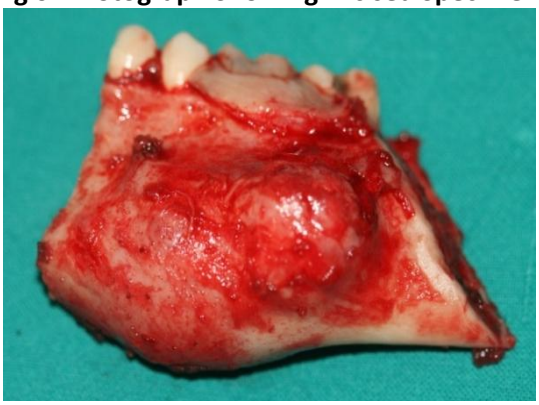
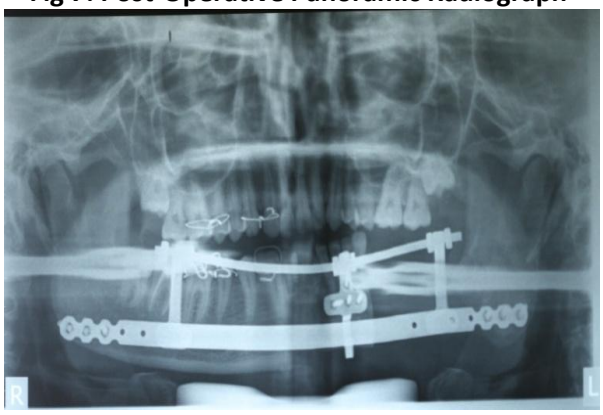


Fig 7: Post-Operative Panoramic Radiograph



Discussion: In the head and neck region, the desmoid fibromatosis may be of two types[10]. One is intraosseous (desmoplastic fibroma) and the second is more often, in soft tissue, with the highest incidence in the supraclavicular region of the neck.[11] These tumours reside in a clinical grey zone between benign fibrous lesion and malignant tumours.[12] Manifestation of desmoplastic fibroma of the jaw is in the same manner as its counterpart in the long bones.

The age of incidence is usually in the first, second, or third decade.[10] The common site of occurrence

within the jawbone is the mandible, while the maxilla is rarely affected. Within the mandible posterior part is most frequently involved like the ramus, angle and molar area, while the premolar area and the anterior segments are less commonly affected.[10] In the present case, a small diffuse swelling was seen at the body of the mandible which was persistent since two years.

The symptoms vary from no pain to painful swelling which is similar in our case, where there was no pain initially. On clinical examination, the swelling was looking like odontogenic and inflammatory.

Radiographic appearance may vary from unilocular to multilocular, with or without expansion or perforation of cortical plates according to Frick *et al*.[13]

Radiographs showed osteolytic lesions with coarsened ridge-like trabeculae in 63% of cases, osteolytic lesions in 24% of cases and mixed lytic and mildly sclerotic lesions in 13% cases.

About 53% of cases showed cortical breaching. CT revealed radiolucent (65%) or mixed radiolucent and mildly sclerotic (35%) matrix patterns. Cortical destruction was seen in 88%.

In our case radiographs revealed a solitary large mixed radiopaque and radiolucent lesion covering entire left body of mandible and buccolingual cortical plate expansion.

The histological features of desmoplastic fibroma and the extra-abdominal desmoid tumour are identical. They are characterized by uniform-appearing fibroblastic cells in a stroma containing various amounts of collagen fibres.[10]

DF of maxilla or mandible with extra-osseous extensions is treated with complete excision including a margin of uninvolved soft tissue.[14]

These tumors are locally aggressive and can recur with a subtotal resection. The recurrence rate of about 40-47% is seen in lesions treated by curettage or intra-lesional resection[14] making follow-up a necessity.

In our case the, resection of involved mandibular region was done and area healed uneventfully and no signs of recurrence observed even after 3 years of follow up.

Conclusion: Since desmoid fibromatosis in locally infiltrating and aggressive tumor, treatment option should be choosen carefully. In the present case, on initial examination it appeared like odontogenic swelling but on radiographic investigation there was mixed radioopaque and radiolucent lesion. Dentists should be aware of such entities, their nature, behavior so that investigations and treatment can be planned accordingly with multidisciplinary approach.

References:

1. Weiss SW, Goldblum JR (2008) Enzinger and Weiss's soft tissue tumors, 5th edn. Mosby Elsevier, St. Louis
2. Arya AN, Saravanan B, Subalakshmi K, Appadurai R, Ponniah I. Aggressive fibromatosis of the mandible in a two-month old infant. J Maxillofac Oral Surg. 2015;14(Suppl 1):235-9.
3. Sakorafas GH, Nissotakis C, Peros G. Abdominal desmoid tumors. Surg Oncol 2007; 16:131–142
4. Nieuwenhuis MH, Casparie M, Mathus-Vliegen LM, Dekkers OM, Hogendoorn PC, Vasen HF. A nation-wide study comparing sporadic and familial adenomatous polyposis-related desmoid-type fibromatoses. Int J Cancer 2011;129:256–61.
5. Ganeshan DM, Amini B, Nikolaidis P, Assing M, Vikram R. Current Update on Desmoid Fibromatosis. J Comput Assist Tomogr. 2019;43(1):29–38.
6. de Camargo VP, Keohan ML, D'Adamo DR, et al. Clinical outcomes of systemic therapy for patients with deep fibromatosis (desmoid tumor). Cancer 2010;116:2258–65.
7. Fiore M, MacNeill A, Gronchi A, Colombo C. Desmoid-Type Fibromatosis: Evolving Treatment Standards. Surg Oncol Clin N Am 2016; 25:803–26.
8. Sharma A, Ngan BY, Sàndor GK, Campisi P, Forte V. Pediatric aggressive fibromatosis of the head and neck: a 20-year retrospective review. J Pediatr Surg 2008;43:1596-604.
9. Burlini D, Conti G, Bardellini E, Amadori F. Rare case of desmoid-type fibromatosis of the mandibular region in a child: diagnosis and surgical management. European Journal of Paediatric Dentistry. 2013;14(4):333-4
10. Shukul VK, Saxena S, Shankar BG. Desmoplastic Fibroma : Mandible. Med J Armed Forces India. 2004;60(3):307-9.
11. Batsakis JG, Raslan W. Pathological consideration, extraabdominal desmoid

fibromatosis. Ann Otol Rhinol Laryngol 1994;103:331-4.
12. Ayal AG, Ro JY, Goepfert H, Cangir A, Khorsand J, Flake G. Desmoid fibromatosis : a clinicopathologic study of 25 children. Semin Diagn Pathol 1986; 3:138-50.
13. Frick MA, Sundaram M, Unni KK, Inwards CY, Fabbri N, Trentani F, et al. Imaging findings in desmoplastic fibroma of bone: Distinctive T2 characteristics. AJR Am J Roentgenol. 2005; 184:1762–7.
14. Dalili-Kajan Z, Adham-Fumani G, Dalili-kajan H. Desmoplastic fibroma with periosteal reaction. Acta Med Iran. 2007; 45:325–8.

Conflict of interest: None
Funding: None
Cite this Article as: Karagir A, Adaki S, Kalghatgi S, Dalvi T. Desmoid Fibromatosis – A Rare Case Report. Natl J Integr Res Med 2021; Vol.12(2): 69-73