

Intraosseous Desmoplastic Fibroblastoma of the Mandible: Report of a Rare Case Mimicking Aggressive Jaw Lesions

*Shruti Singh¹, Anju Devi¹, Mala Kamboj¹, Anjali Narwal¹, Virendra Singh¹

Abstract

Introduction:

Desmoplastic fibroblastoma is an exceptionally rare benign fibroblastic neoplasm that predominantly affects soft tissues, whereas primary intraosseous occurrence is exceedingly uncommon. Its variable clinicoradiological presentation often mimics aggressive odontogenic and mesenchymal tumors, rendering preoperative diagnosis particularly challenging.

Case Report:

A 20-year-old male presented with a progressively enlarging swelling of the left mandible. Computed tomography revealed a well-defined, expansile, mixed radiolucent-radiopaque intraosseous lesion with cortical expansion and focal breach. Although the incisional biopsy favoured a benign spindle-cell mesenchymal lesion, the extensive osseous involvement warranted hemimandibulectomy. Histopathological examination demonstrated a well-circumscribed, hypocellular fibroblastic neoplasm composed of bland spindle- and stellate-shaped cells with eosinophilic cytoplasm dispersed within an abundant, densely collagenised stroma. Immunohistochemical analysis showed diffuse vimentin positivity, supporting the diagnosis of primary intraosseous desmoplastic fibroblastoma. At 8-month follow-up, the patient remains free of recurrence.

Conclusion:

As the fifth reported primary intraosseous case and the fourth involving the jaws, this case expands the limited literature on this rare entity. It highlights the indispensable value of meticulous clinicoradiological, histopathological, and immunohistochemical correlation in distinguishing this benign neoplasm from more aggressive jaw lesions, thereby facilitating accurate diagnosis and appropriate surgical management.

Keywords: Desmoplastic fibroblastoma, Collagenous fibroma, Mandible

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¹Department of Oral & Maxillofacial Pathology, Pandit Bhagwat Dayal Sharma University of Health Sciences (Postgraduate Institute of Dental Sciences), Rohtak, Haryana-124001, India.

*Corresponding author: Email: shruti1995777@gmail.com

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Introduction

Desmoplastic fibroblastoma, also known as collagenous fibroma, is a rare benign fibroblastic tumor characterised by sparse fibroblastic cells embedded within a dense collagenous matrix (1). It commonly arises in the subcutaneous tissue or skeletal muscle of the extremities, shoulder girdle, and posterior neck and is exceedingly rare in the oral cavity (2,3). Clinically, the lesions present as slow-growing swellings and, in the mandible, expansion may cause facial asymmetry, tooth displacement, or cortical thinning (4,5). Radiographic features are nonspecific, with radiolucent or mixed radiolucent-radiopaque expansile patterns (6,7). Consequently, it can mimic several benign and low-grade malignant tumors of the jaws, making histopathological evaluation essential for diagnosis, supported by immunohistochemistry. Histologically, the lesion is characterised by spindle- to stellate-shaped fibroblastic cells distributed within a collagen-rich stroma with minimal mitotic activity (8).

Primary intraosseous desmoplastic fibroblastoma is exceedingly rare. A review of the literature revealed only four reported cases of primary bone involvement to date (9). The present report describes the fifth documented case of primary intraosseous desmoplastic fibroblastoma and the fourth involving the jaws, along with radiopathological and immunohistochemical correlation.

Case Report

A 20-year-old male was referred to the dental college with a swelling in the lower left posterior tooth region, which had gradually increased to its present size (5×6 cm) and caused facial asymmetry on the left side (Fig. 1a).



Fig 1a: Diffuse swelling of the left mandible.

The swelling was associated with throbbing pain and extended anteroposteriorly from the symphysis to the left ramus of the mandible and superoinferiorly from the left corner of the mouth to the lower border of the mandible, extending further to the level of the hyoid bone. The overlying skin was normal in colour, with no visible scar, sinus opening, or ulceration. The swelling was hard in consistency, non-tender, non-reducible, non-compressible, and non-fluctuant, and caused obliteration of the left buccal vestibule. The patient's medical history was unremarkable. Computed tomography (CT) images (axial, coronal, and 3D reconstruction) revealed an expansile lesion involving the left body and angle of the mandible, with well-defined to partially ill-defined margins and heterogeneous density (Fig. 1b–d).

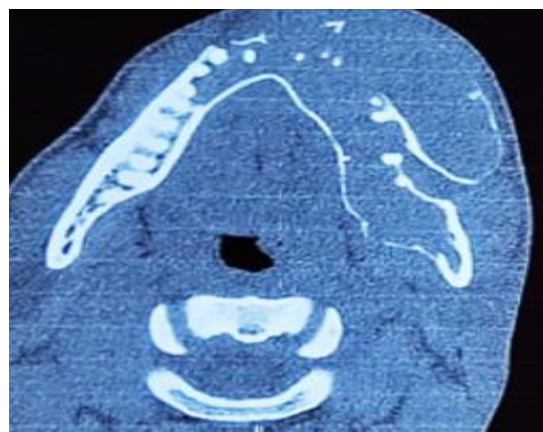


Fig 1b: Axial CT shows an expansile lesion involving the anterior mandible, predominantly hypodense (radiolucent), with well-defined to partially indistinct cortical margins.

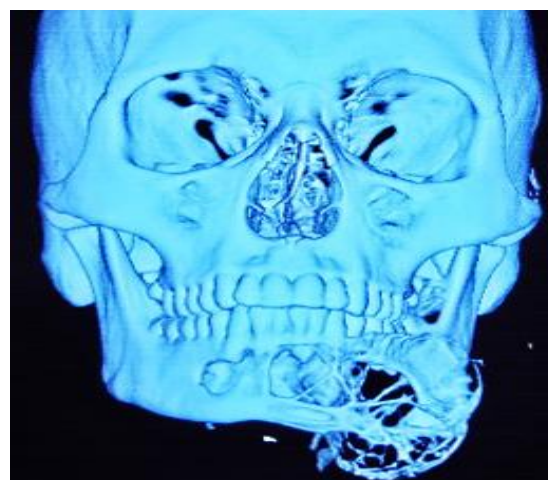


Fig 1c: CT images show marked cortical expansion with thinning and focal areas of cortical breach and displacement of adjacent soft tissues, without obvious invasion into surrounding vital structures.

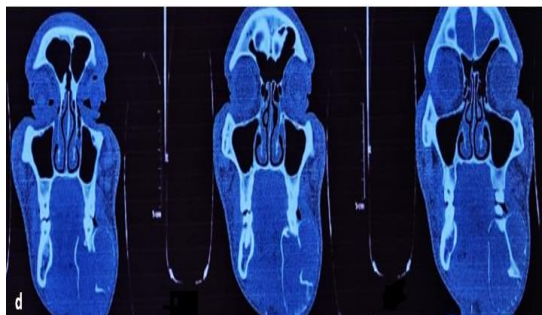


Fig 1d: Coronal CT images demonstrate a well-defined expansile osteolytic lesion involving the anterior mandible with marked buccolingual expansion, cortical thinning, and focal cortical perforation.

The lesion demonstrated mixed radiopaque-radiolucent areas with marked buccolingual expansion and focal cortical perforation, but showed no radiological evidence of periosteal

reaction, adjacent soft-tissue invasion, or pathological cervical lymphadenopathy. Incisional biopsy revealed a benign mesenchymal tumor. Considering the marked buccolingual cortical expansion with focal cortical perforation and the potential for locally infiltrative behaviour, the multidisciplinary team elected to perform hemimandibulectomy. Reconstruction was performed using a free fibula osteocutaneous flap. On gross examination, the lesion was circumscribed, pale-white in colour, and firm in consistency. Histopathology revealed a benign, lobulated, well-encapsulated mesenchymal tumor consisting of dense collagenous stroma associated with spindle and stellate cells with eosinophilic cytoplasm, along with a few binucleated fibroblastic cells (Fig. 2. a-d).

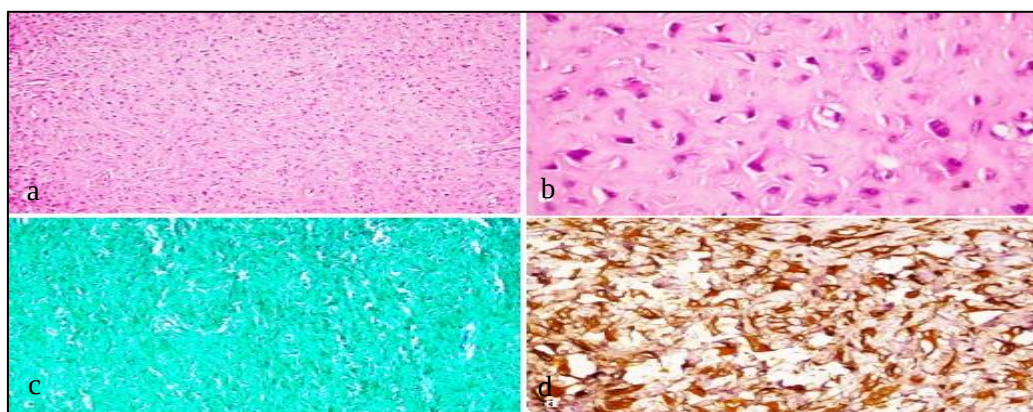


Fig 2a: Stellate and spindle-shaped fibroblasts in a densely collagenous stroma. **b:** Areas of binucleated fibroblastic cells. **c:** Masson trichrome stain positive for collagen bundles and hyalinised areas (green). **d:** Tumor cells showed diffuse positivity for vimentin.

Focal areas of basophilic dystrophic calcification and bony trabeculae at the periphery were also evident, along with blood vessels and very few inflammatory cells. Neither cellular atypia nor mitotic figures were observed. Masson trichrome staining was positive for collagen bundles and hyalinised areas, which appeared green in colour (Fig. 2c).

On immunohistochemistry, tumor cells showed diffuse vimentin positivity, while other markers were negative (beta-catenin, SMA, S100, MUC4, CD34) (Fig. 2d).

The combined histomorphological and immunohistochemical findings were consistent with intraosseous desmoplastic fibroblastoma. At the 8-month postoperative follow-up, the patient was free of recurrence.

Discussion

Desmoplastic fibroblastoma is an uncommon benign fibroblastic tumor that demonstrates distinctive clinicopathological features. Since its original description by Evans, the lesion has been recognised as a separate entity among fibrous tumors of soft tissue (1). This tumor affects mainly soft tissue; however, intraosseous involvement of the jaws remains extremely rare, with only a limited number of cases documented in the literature (9–11). Such cases often present as expansile lesions of the mandible or maxilla and may clinically resemble odontogenic tumors or fibro-osseous lesions. Among all reported cases of desmoplastic fibroblastoma, only five have been reported to be of intraosseous origin, including the present case (Table 1).

Table 1. Reported Cases of Intraosseous Desmoplastic Fibroblastoma

Case No.	References	Age (year)	Sex	Radiological features	Anatomic location	Histopathological findings
1	Gong et al., 2018 (15)	46	male	Osteolytic with thick sclerotic rim	Proximal femur	Encapsulated, with stellate-shaped tumor cells in hyalinised stroma
2	Jaafari Ashkavandi et al., 2018 (12)	58	Female	Well-defined radiolucency	Maxillary alveolar bone	Well-circumscribed, paucicellular lesion with binucleated and stellate fibroblasts in collagenous stroma
3	Garcia et al., 2018 (11)	43	Female	Well-defined radiolucency	Maxillary edentulous area	Spindle- to stellate-shaped cells scattered within a myxocollagenous stroma
4	Zaidi et al., 2024 (9)	35	Female	Osteolytic with sclerotic rim	Mandible	Well-demarcated, hypocellular lesion comprising haphazardly arranged, reactive-appearing, spindle to ovoid cells; a subset of lesional cells showed binucleation in collagenous stroma
5	Present case	20	male	Expansile osteolytic lesion with sclerotic rim	Mandible	Stellate and spindle-shaped fibroblasts in densely collagenous stroma, along with a few areas of binucleated cells

Clinical manifestations depend on the tumor size and anatomical location. It can be asymptomatic or present with nonspecific symptoms, often as a painless, slow-growing mass of generally long duration that is usually not associated with a history of trauma (12–13).

This tumor is characterised by a wide anatomical distribution in the head and neck, oral cavity, upper and lower extremities, and trunk (1,3,14,15). Radiologically, intraosseous desmoplastic fibroblastomas appear as radiolucent, lobulated, and expansile tumors with well-defined to ill-defined margins, often with internal trabeculation, a non-sclerotic rim, and cortical bone expansion or destruction (9). Grossly, the tumor appears as a well-circumscribed, firm mass with a white-to-grey cut surface, without haemorrhage or necrosis. Microscopically, the tumor is more or less circumscribed and hypocellular, consisting of widely spaced, bland, spindle- to stellate-shaped cells embedded in a collagenous or hyalinised stroma, sometimes associated with a few binucleated cells. Mitotic figures are rare or absent, and necrosis is not present; blood vessels are usually inconspicuous (11,15). Degenerative changes such as dystrophic calcification and metaplastic bone formation are observed occasionally (9,12). To ascertain the rarity of this entity, an updated literature search was performed using PubMed, Scopus, Web of Science, and Google Scholar up to July 2026. The search employed the terms “intraosseous desmoplastic fibroblastoma/ collagenous fibroma of bone” and related combinations. The

reference lists of all relevant publications were additionally screened to identify any further eligible reports. Based on the available English-language literature, only four cases of primary intraosseous desmoplastic fibroblastoma have been documented to date, with three involving the jaws (Table 1). The present case therefore represents the fifth reported intraosseous case and the fourth involving the jaws, further expanding the limited clinicopathological spectrum of this exceptionally rare neoplasm. Two of the primary intraosseous desmoplastic fibroblastomas of bone occurred in the maxillary bone (11,12) and one in the mandible (9), while the other was in the proximal femur (15). The clinical presentation of these cases was an incidental finding during routine medical evaluations. Radiographically, these cases showed well- to ill-defined osteolytic lesions with a sclerotic rim. Primary desmoplastic fibroblastoma of the femur was treated with curettage, whereas the other two cases involving the maxillary bone and one involving the mandible were treated by surgical excision, as in the current case. Grossly, the lesions were associated with smooth, white-grey outer surfaces and solid cut sections with some cystic-like areas. Histological features were almost identical, with dense hyalinised stroma associated with stellate to spindle cells, along with a few areas of binucleated cells with eosinophilic cytoplasm (9,11,12,15). These findings were consistent with the present case. Immunohistochemical analysis of primary desmoplastic fibroblastoma of bone showed an

expression pattern similar to that of the soft-tissue tumor, including consistent diffuse positivity for vimentin and focal to absent reactivity for smooth muscle markers such as SMA, which was similar to the findings in this case (9,11,12,15). It may clinically and

radiographically resemble other benign or malignant jaw lesions, including fibrous dysplasia, ossifying fibroma, desmoid-type fibromatosis, and low-grade fibrosarcoma (1,9) (Table .2).

Table 2. Histopathological and Immunohistochemical Differential Diagnosis of Primary Intraosseous Desmoplastic Fibroblastoma

Lesion	Histopathological Features	Immunohistochemical Profile	Distinguishing Features
Desmoplastic fibroma (16,17)	Moderately cellular infiltrative spindle-cell proliferation with abundant collagen fibres; entrapped bony trabeculae may be present	Vimentin (+); SMA variable	Locally aggressive infiltrative growth, greater cellularity compared to desmoplastic fibroblastoma
Desmoid-type fibromatosis (17)	Long sweeping fascicles of uniform spindle cells infiltrating adjacent skeletal muscle and soft tissue with abundant collagen	β -catenin nuclear (+); SMA variable; Desmin (-)	Diffuse infiltrative margins, fascicular architecture and nuclear β -catenin positivity favour fibromatosis.
Fibrous dysplasia (17)	Irregular curvilinear trabeculae of woven bone ("Chinese character" pattern) within a moderately cellular fibrous stroma	No specific immunophenotype	Presence of woven bone trabeculae without osteoblastic rimming is diagnostic and distinguishes it from collagen-rich fibroblastic lesions.
Ossifying fibroma (17,18)	Well-demarcated fibrocellular lesion containing variable amounts of mineralised bone and cementum-like calcifications	No specific immunophenotype	Presence of mineralised material, osteoblastic rimming and a well-circumscribed lesion support ossifying fibroma.
Myofibroma (19)	Biphasic pattern comprising spindle cells and primitive round cells surrounding numerous thin-walled haemangiopericytoma-like vessels	SMA (+); Muscle-specific actin (+); Desmin (-); S100 (-)	Biphasic morphology, haemangiopericytoma-like vasculature and diffuse SMA positivity distinguish myofibroma.
Low-grade fibromyxoid sarcoma (17,20)	Bland spindle-cell neoplasm composed of alternating hypocellular collagenous and myxoid areas with a characteristic whorled growth pattern; mild cytological atypia and infrequent mitoses	MUC4 (+), Vimentin (+), SMA (-), S100 (-), CD34 (-)	Alternating fibrous and myxoid areas, infiltrative growth pattern, diffuse MUC4 positivity, and the presence of FUS::CREB3L2 or FUS::CREB3L1 gene fusion distinguish it from desmoplastic fibroblastoma.
Non-ossifying fibroma (18)	Storiform proliferation of spindle-shaped fibroblasts with scattered osteoclast-like multinucleated giant cells and foamy histiocytes	CD68 (+)	Storiform architecture with giant cells and xanthomatous histiocytes is characteristic and absent in desmoplastic fibroblastoma.

Imaging findings are often nonspecific; therefore, histopathological examination and immunohistochemical analysis are essential for definitive diagnosis and to avoid misdiagnosis and overtreatment.

Early identification and appropriate surgical management can ensure complete tumor removal while allowing reconstruction of mandibular continuity and restoration of oral function. The most important differential diagnostic consideration for primary intraosseous desmoplastic fibroblastoma is desmoplastic fibroma of the bone (Table.2), a rare, locally aggressive bone tumor that most commonly affects the posterior aspect of the mandible and long tubular bones (4).

Desmoplastic fibroblastoma shows low signal intensity on T1-weighted MRI images and slightly high signal intensity on T2-weighted

images, suggesting a collagen-rich tumor (1,11). Unlike intraosseous desmoplastic fibroblastoma, microscopic examination of desmoplastic fibroma typically shows interlacing, hypocellular to moderately cellular fascicles of uniform, bland spindle cells embedded in an abundant fibrillar collagenous stroma with scant vascularity, often intermingled with thin spicules of reactive bone trabeculae (11,12,15). Strong and diffuse overexpression of the nuclear transcription factor FOSL1 has been recently identified in 80% to 100% of soft-tissue desmoplastic fibroblastoma, which is regarded as a specific feature of this tumor type (4,9).

This is supported by the identification of somatic rearrangements involving FOSL1 in most tumors. This would be of great utility in rare and challenging cases, although conventional microscopy alone is considered

sufficient to reach a diagnosis of desmoplastic fibroblastoma in most cases. Desmoplastic fibroblastomas have pseudodiploid karyotypes with translocations involving bands 11q12–13 and different partner chromosomes. By FISH analysis, the breakpoint in chromosome 11 could be mapped to a 20-kb region including FOSL1, which encodes a transcription factor (TF) that can dimerise with members of the JUN family (10,12). Increased expression has been found at both the mRNA and protein levels, making immunohistochemistry for FOSL1 expression a useful diagnostic tool.

Unlike its soft-tissue counterpart, primary intraosseous desmoplastic fibroblastoma presents a significant diagnostic challenge because of its variable radiological appearance and overlap with other benign and locally aggressive spindle-cell lesions of the jaws.

Conclusion

This case highlights the importance of a multidisciplinary diagnostic approach integrating clinical, radiographic, histopathological, and immunohistochemical findings. As the molecular landscape of fibroblastic tumors continues to evolve, FOSL1 expression may emerge as a valuable diagnostic hallmark, offering greater confidence in distinguishing desmoplastic fibroblastoma from its histological mimics. As the catalogue of reported cases continues to grow, so too will our ability to unravel the biological nuances of this enigmatic neoplasm, refine its diagnostic framework, and optimise patient care through evidence-based management.

References

1. Evans HL. Desmoplastic fibroblastoma: a report of seven cases. *The American journal of surgical pathology*. 1995 Sep 1;19(9):1077-81.
2. Nielsen GP, O'Connell JX, Dickersin GR, Rosenberg AE. Collagenous fibroma (desmoplastic fibroblastoma): a report of seven cases. *Modern pathology: an official journal of the United States and Canadian Academy of Pathology, Inc*. 1996 Jul 1;9(7):781-5.
3. Miettinen M, Fetsch JF. Collagenous fibroma (desmoplastic fibroblastoma): a clinicopathologic analysis of 63 cases of a distinctive soft tissue lesion with stellate-shaped fibroblasts. *Human pathology*. 1998 Jul 1;29(7):676-82.
4. Watanabe H, et al. Clinical features of desmoplastic fibroblastoma. *J Dermatol*. 2008;35: 561–7.
5. Nagao T, et al. Collagenous fibroma of oral cavity. *Oral Oncol*. 2002;38:278–282.
6. Yamamoto A, et al. Radiologic findings of desmoplastic fibroblastoma. *Radiol Case Rep*. 2017; 12:1234–1240.
7. Murphey MD, et al. Imaging of benign fibrous tumors. *Radiographics*. 2009;29:203–219.
8. Bernal K, Nelson M, Neff JR, Nielsen SM, Bridge JA. Translocation (2;11)(q31;q12) is recurrent in collagenous fibroma (desmoplastic fibroblastoma). *Cancer genetics and cytogenetics*. 2004 Mar 1;149(2):161-3.
9. Al-Zaidi R, AL-Zaidi RS. An unusual case of intraosseous desmoplastic fibroblastoma (collagenous fibroma) of the mandible. *Cureus*. 2024 Dec 17;16(12).
10. Mesquita RA, Okuda E, Jorge WA, de Araújo VC. Collagenous fibroma (desmoplastic fibroblastoma) of the palate: A case report. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 2001; 91(1):80-84.
11. Garcia NG, De Freitas Filho SA, Soares CT, Figueiredo CM, Oliveira DT. Intraosseous collagenous fibroma (Desmoplastic fibroblastoma) involving maxillary bone. *J Clin Diagn Res*. 2018 Feb 1;12:6.
12. Jaafari-Ashkavandi Z, Shirazi MY, Assar S. Desmoplastic fibroblastoma in maxillary alveolar bone mimicking an odontogenic lesion: a novel case report with review of literature. *Turk Patoloji Derg*. 2018 Sep 1;34(3):247-50.
13. Fong F, Odell E, Simo R: Collagenous fibroma (desmoplastic fibroblastoma) of the neck presenting with neurological symptoms. *Head Neck Pathol*. 2009, 3:47-50.
14. Hornick JL, Miettinen M, Bridge JA: WHO classification of Soft tissue and bone tumors. WHO Classification of Tumors Series, 5th Edition. WHO Classification of Tumors Editorial Board (ed): International Agency for Research on Cancer, Lyon, France; 2020. 3:
15. Gong LH, Liu WF, Ding Y, Geng YH, Sun XQ, Huang XY. Diagnosis and differential diagnosis of desmoplastic fibroblastoma by clinical, radiological, and histopathological analyses. *Chinese Medical Journal*. 2018 Jan 5;131(01):32-6.
16. Nielsen GP, Rosenberg AE. Desmoplastic fibroma of bone. In: WHO Classification of Tumors Editorial Board. *Soft Tissue and Bone Tumors*. WHO Classification of Tumors. 5th ed. Lyon: International Agency for Research on Cancer; 2020.
17. WHO Classification of Tumors Editorial Board. *Soft Tissue and Bone Tumors*. WHO Classification of Tumors. 5th ed. Lyon: International Agency for Research on Cancer; 2020.
18. WHO Classification of Tumors Editorial Board. *Head and Neck Tumors*. WHO Classification of Tumors. 5th ed. Lyon: International Agency for Research on Cancer; 2022.
19. Mentzel T, Calonje E, Nascimento AG, Fletcher CDM. Myofibroblastoma in adults: Clinicopathological analysis of a series of cases. *Am J Surg Pathol*. 1995;19:306–315.
20. Doyle LA, Möller E, Dal Cin P, Fletcher CDM, Mertens F, Hornick JL. MUC4 is a highly sensitive and specific marker for low-grade fibromyxoid sarcoma. *Am J Surg Pathol*. 2011;35:733–741.