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Case Report

Gnathic Desmoplastic Fibroma of the Pediatric Demographic: An Immunohistochemical Perspective - A Case report with Review

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Abstract

Desmoplastic fibroma is a rare, benign, locally aggressive, intra-osseous tumor that commonly involves the mandible, with a high recurrence rate. A four-year-old boy with a history of painless right mandibular expansion associated with progressive trismus over a period of 18 months was taken up for hemimandibulectomy. The excised lesion was finally diagnosed as desmoplastic fibroma upon histopathological and immunohistochemical assessment. We present the report of the case accompanied by a review of pediatric cases of gnathic desmoplastic fibroma and their immunohistochemistry.

Keywords: Desmoplastic fibroma, hemi-mandibulectomy, immunohistochemistry, intra-osseous tumor, pediatric

Introduction

Desmoplastic fibroma (ICD-O code 8823/0) is a rare, benign, albeit locally aggressive, intra-osseous neoplasm that was first described as a distinct entity from other intraosseous fibrous tumors in 1958 by Jaffe.[1,2] With an estimated incidence of 0.1% of all primary bone tumors, DF may involve any bone but is most frequent in the mandible (22%), followed by the femur (15%), pelvic bones (13%), radius (12%), and tibia (9%).[1] Woods et al[2] reviewed the literature on DF and found a total of 152 reported cases of gnathic DF in an age range of 6 months to 60 years. Gnathically, mandibular involvement is most common (84%), with a definite predilection for the posterior location in either jaw.[2]

The diagnosis of DF is based on a combination of clinical, radiological, and histopathological features. Immunohistochemical (IHC) analysis has been highlighted as a potential tool to distinguish these lesions from their differentials and in determining prognosis.[3,4] Although most gnathic lesions are painless and slow-growing, these lesions are often locally destructive, with

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a high recurrence potential.[2] Pediatric cases warrant special consideration of future growth of the face while planning the surgical extent of excision. We describe one such locally aggressive case of DF in a four-year-old child and its surgical management.

Case Report

A four-year-old boy reported to the Oral and Maxillofacial Outpatient Department with the complaint of a gradually growing, painless swelling over his right lower jaw for the past 18 months, causing progressive restriction of his mouth opening (Fig. 1). He did not report a history of any other systemic illness, hospitalization, or long-term medication. Upon clinical examination, the palpable mass was firm, non-tender, and diffuse, extending from the right submandibular region to the mid-cheek region supero-inferiorly and from the posterior border of the mandible to the angle of the mouth antero-posteriorly. Intraoral assessment was not possible as the child's mouth opening was restricted to less than a finger breadth.

Contrast-enhanced computed tomography (CECT) of the region revealed a soft tissue expansile mass in the right submandibular region measuring about 3.9 cm (AP) × 4.5 cm (TR) on axial image and 3.5 cm (SI) on coronal image (Fig. 2). The mass caused posterior displacement of the submandibular gland; however, there was a distinct fat plane between the two. Medially, it compressed the floor of the mouth. Computed tomography (CT) images showed erosion and destruction of the angle and body of the mandible and the mentum. Signs of bony remodeling of the right side of the mandible were also evident.

Segmental resection of the mandible, followed by reconstruction using reconstruction plates under general anesthesia (GA), was planned for the patient. Through a right submandibular incision, a layer-by-layer dissection was done to expose the tumor mass, and a right segmental mandibulectomy including the coronoid process while sparing the condyle was performed. A margin of about one centimeter was secured in normal tissue following excision, and reconstruction was done with a recon plate (Fig. 2c, d). The sectioned tissue was stored in 10% formaldehyde and sent for histopathological analysis.

Upon processing the excised tissue, a firm fibrous whorled mass measuring 4×3×2.5 cm was identified. Microscopically, sections showed a hypocellular spindle cell tumor with bland spindle-to-stellate cells arranged haphazardly against a collagenous background (Fig. 3a, b). These cells showed ill-defined cytoplasmic outlines, pale vesicular nuclei with mild anisonucleosis, and inconspicuous nucleoli. Mitotic activity was inconspicuous, and many capillary channels were interspersed. No atypia, necrosis, or mitosis was reported. No odontogenic epithelium, calcification, or bony trabeculae were identified, thus ruling out odontogenic fibroma and fibrous dysplasia.

Histologic differential diagnoses of solitary fibrous tumor and desmoplastic fibroma were considered, and immunohistochemical (IHC) markers were applied. IHC staining showed focal positivity for vimentin, cytoplasmic SMA (smooth muscle actin), and beta-catenin, and negativity for S100 and CD34 (Fig. 4a-d). Thus, IHC findings confirmed a diagnosis of desmoplastic fibroma.

Clinically, the surgery restored the child's mouth opening (Fig. 5), and no recurrence was noted after two years of follow-up. The parents were counseled about the need for regular follow-up and further interventions as the child grows.

Discussion

Desmoplastic fibroma (DF) is regarded as the morphological bony counterpart of desmoid-type fibromatosis and most commonly affects the mandible (~40% of all bony sites), followed by the femur and pelvis.[2] Recurrence following curettage and resection is reported as ~72% and ~17%, respectively.[2] Solitary or multiple desmoid lesions have also been

Figure 1. Pre-operative extra-oral image

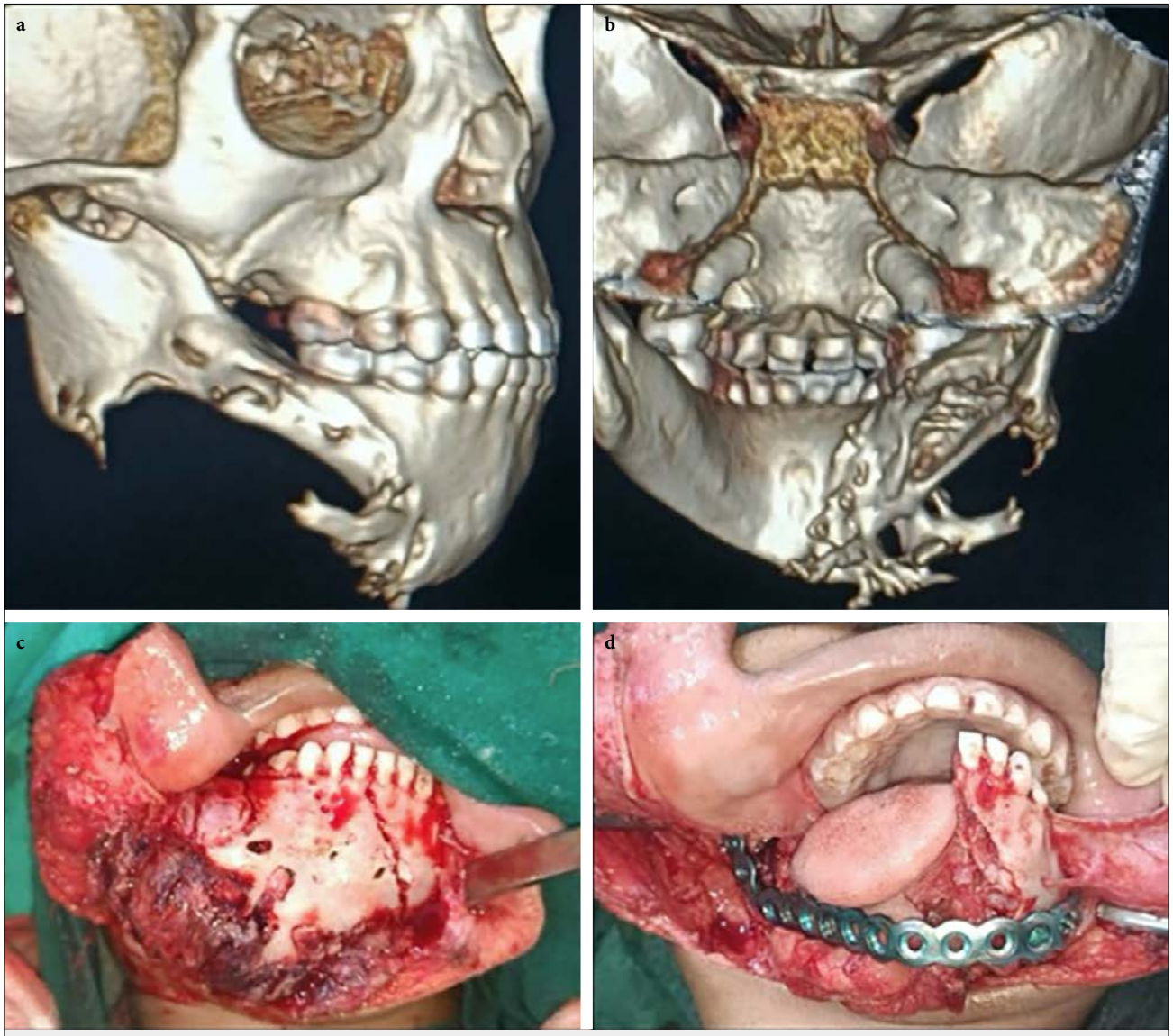

Figure 2. (a, b) Pre-operative three dimensional CBCT image of affected mandible; (c&d) Intra-operative images

CBCT: Cone-beam computed tomography

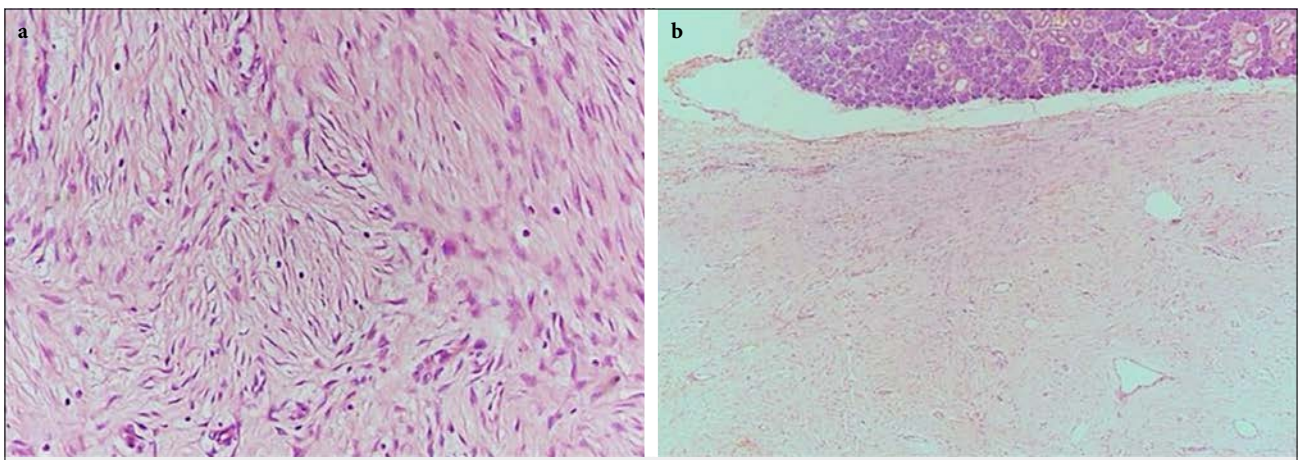

Figure 3. Histopathological images (a) H&E x40; (b) H&E x100

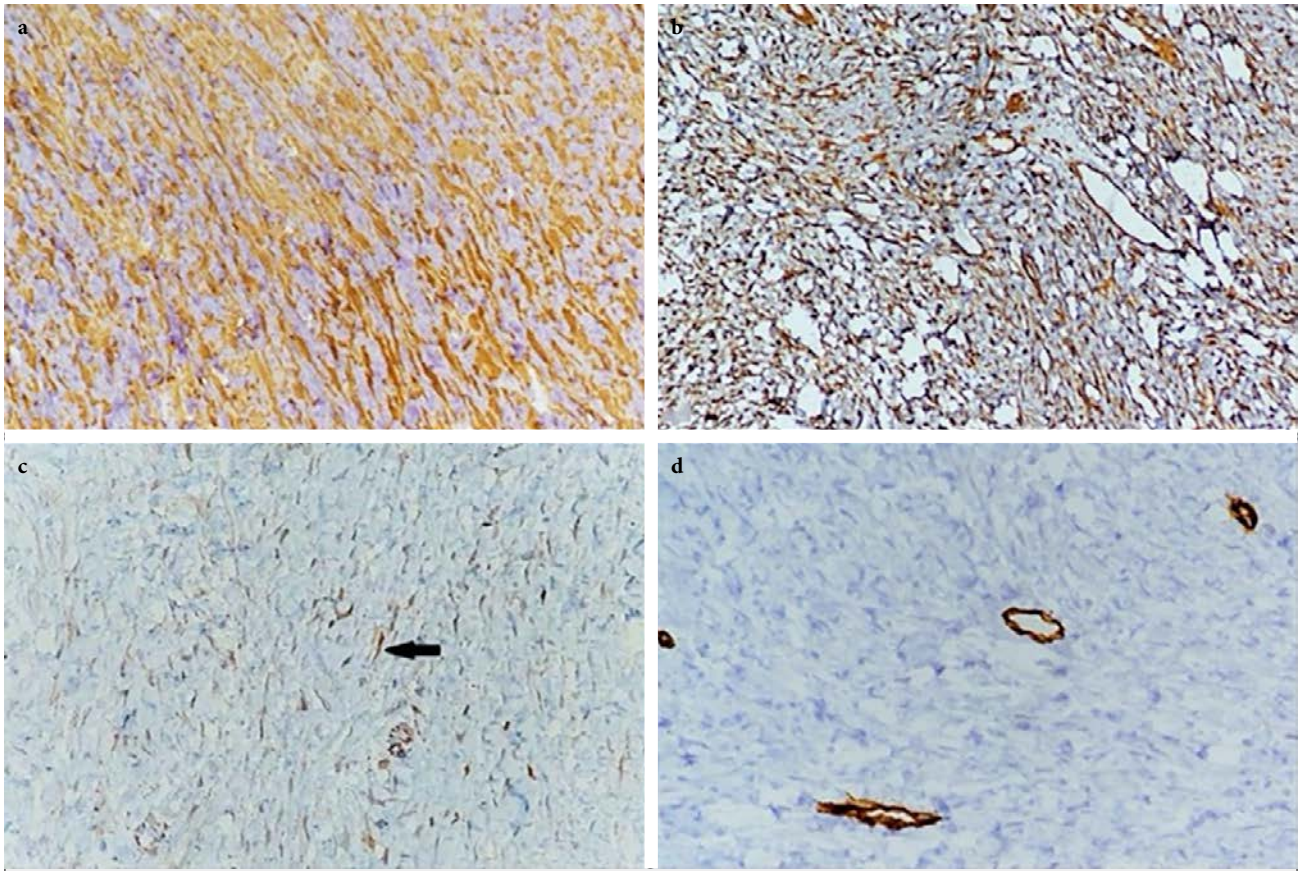

Figure 4. Immunohistochemistry staining (a) Diffuse cytoplasmic immunoreactivity for Vimentin (Vimentin×400); (b) Diffuse strong immunoreactivity for SMA (SMA×100); (c) Diffuse strong focal nuclear immunoreactivity for Beta-catenin (β-catenin×400); (d) CD 34 negative in umor cells, strong positivity in blood vessels (CD34×400)

SMA: Smooth muscle actin, CD: Cluster of differentiation

found in association with conditions like tuberous sclerosis and Gardner syndrome.[4,5] About 84% of gnathic DF cases are found in individuals aged less than 30 years, with a mean age of 16 years.[2]

Considering the vast sea of pathologies manifesting as painless, expansile swellings of the pediatric jaws, a diagnosis of DF is confirmed by correlating the clinical and radiological picture with an astute histological examination supported by immunohistochemistry of the tissue. The World Health Organization defines the histological criteria for desmoplastic fibroma as that of a benign tumor of low to variable cellularity, whose cells can be ovoid or elongated with uniform nuclei that lack atypia, pleomorphism, and mitotic activity.[2]

A literature search was made using MESH terms "Desmoplastic fibroma," "child*," "mandible OR maxilla" in the PubMed database. Twenty-six reports of gnathic DF occurring in children under 15 years were thus identified and compared (Table 1). Twenty-three of those were reported in the mandible, with the majority occurring in the posterior mandible. The youngest child reported to have a gnathic DF was an infant aged

Figure 5. One month post-operative extra-oral image with significant improvement in the mouth opening

Table 1. Characteristics of Paediatric cases of Gnathic DF in literature

Author &year	Age/gender	Site/size	Treatment	Immunohistochemistry	Follow up; Reurrence
Said-Al-Naief et al[1]	8/M	Right mandible	Manbdibular Inferior Border resection	NA	4.5 years; Nil
Woods et al[2]	13/M	Right mandible	Marginal resection	Positive labelling: SMA, Vimentin, HHF-35/ muscle specific actin. Negative reactivity: β-cateninand S-100, Ki-67<1 %.	3.5 years; Nil
Madakshira MG et al[3]	5/M	Right mandible	Excision	Cytoplasmic positivity for vimentin and smooth muscle actin. Negative for β-catenin	5 mo; Nil
Vargas Gonzale et al[5]	14/M	Left Maxilla	Resection	NA	NA
Wippold FJ et al[6]	6mo/M	Right mandible	Hemimandibulectomy	NA	NA
Summa et al[7]	3/F	Right Mandible	Excision	NA	NA; Recurrence reported, treated with Chemotherapy
Flucke et al[8]	8/F	Mandible	Excision	Nuclear β-catenin positivity	3 years; Slow progression
	2/M	Mandible	Resection	Nuclear β-catenin positivity	13 years; Nil
Karimi A et al[9]	2/F	Right mandibular angle	Segmental Manidbulectomy	Strongly positive nuclear immunoreactivity for β-catenin. Mild positive staining for α-SMA	8 mo; Nil
Iatrou IA, et al[10]	10/M	Left mandibular angle	Peripheral ostectomy	Fibroblasts showed positive results for Vimentin and S-100 protein antibodies	5 years; Nil
Motevasseli S, et al[11]	5/M	Right Mandible	Resection	Positive for Vimentin (cytoplasmic) and β-catenin (Diffuse cytoplasmic, focal nuclear)	14 mo; Nil
Khatib B, et al[12]	8/F	Left mandible	Left mandibulectomy	NA	14 year; Nil
	4/F	Left posterior mandible	Resection	NA	13 years; Nil
	2/M	Left Mandibular angle	Segmental resection	NA	12 years; Nil
	2/F	Right Mandibular ramus extending into infratemporal fossa	Chemotherapy (discontinued after 12 mo)	NA	6 years; Tumor persisted without progression
Cupero, et al[13]	14/F	Right maxilla extending through orbital floor	Right maxillectomy with orbital preservation	NA	48 mo; None
Herford, et al[14]	11/F	Right mandibular angle	Marginal Mandibulectomy	NA	NA
Kaplan, et al[15]	3/M	Right mandible	NA	NA	NA
Sandrini, et al[16]	11/M	Left mandibular angle	Excision	NA	33mo; Nil
Ferri, et al[17]	3/F	Right mandible	Partial right mandibulectomy	NA	36 mo; Nil
	2/F	Left mandible	Partial left mandibulectomy	NA	26 mo; Nil
	2/F	Right mandible	Right mandibular resection	NA	17 years; Nil
Gersak MM, et al[18]	3/M	Right mandible	Right mandibular resection	NA	NA
Skinner HR, et al[19]	3.7/M	Right mandible	Right mandibulectomy and reconstruction with osseous free fibula flap	NA	6 years; Nil
Mohammadi, et al[20]	2/M	Right Mandible (Recurrent lesion after previous enucleation <12mo ago)	Segmental mandibulectomy	Strong immunoreactivity for Vimentin and β-catenin. Non reactive with S-100 and desmin.	18 mo; Nil

M: Male, F: Female, NA: Not applicable

6 months.[6] Recurrence or persistence of lesions was reported in three cases, all of which had received a relatively conservative initial treatment with either local excision or chemotherapy.[7–9] The majority were treated with complete or partial mandibulectomies or segmental resections. The final diagnosis of DF was established based on histological reports consistent with the criteria defined by the World Health Organization (WHO).[2]

Immunohistochemistry was reported in eight cases, and they expressed variable positivity for vimentin, SMA, desmin, cytoplasmic β-catenin, and nuclear β-catenin.[2,3,8–10] Desmoid-type soft tissue fibromatosis has been shown to express nuclear and cytoplasmic beta-catenin positivity.[4,8] However, the consistency of their expression in the intra-osseous counterparts is uncertain. Kahraman et al[4] reported positive expression of cytoplasmic β-catenin in more than half of their 22 cases, but none with nuclear β-catenin positivity. Meanwhile, Flucke et al[8] reported nuclear β-catenin positivity in all seven of the cases in their case series. Our search identified five reports of pediatric gnathic DF with positive nuclear β-catenin expression similar to our case. Unfortunately, very few authors reported the IHC characteristics of the excised lesions.

Motevasseli et al[11] discussed a periosteal reaction in the mandibular inferior border as a sign of DF in its initial stages; it evaded surgical treatment at its incipient stage due to nonspecific histological findings.

Conclusion

The case went on to grow aggressively, as is typical of DF, eventually warranting hemimandibulectomy. With nonspecific clinical and radiological presentations and the wide spectrum of histologically similar pathologies affecting the jaws, immunohistochemistry should be considered an essential tool in diagnosing these locally aggressive lesions, thus avoiding demolitive interventions at advanced stages. Therefore, standardized protocols for IHC analysis and reporting in the literature need to be developed and enforced.

The authors also feel the need for longitudinal studies highlighting patient outcomes in such conditions.

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