Dental Research Journal

Case Report

Desmoplastic fibroma of the lower jaw in a 2-year-old patient; report of a rare case

Farnoosh Mohammadi[1], Gholamreza Shirani[2], Samira Derakhshan[3], Taraneh Faghihi[4]

Departments of [1]Oral and Maxillofacial Surgery and [3]Oral and Maxillofacial Pathology, Faculty of Dentistry, Tehran University of Medical Sciences, [2]Department of Oral and Maxillofacial Surgery, Dental Research Center, School of Dentistry, Tehran University of Medical Sciences, Tehran, [4]Department of Pedodontics, School of Dentistry, Ardabil University of Medical Science, Ardabil, Iran

Received: February 2019
Accepted: April 2019

Address for correspondence:
Dr. Taraneh Faghihi, Department of Pedodontics, School of Dentistry, Ardabil University of Medical Science, Ardabil, Iran.
E-mail: trnfaghihi@gmail.com

ABSTRACT

Desmoplastic fibroma (DF) is a rare benign but aggressive fibrous lesion with an unknown etiology. It has an affinity for the mandible. DF has a high rate of recurrence after surgical resection. In this study, we report the therapeutic management of a recurrence of DF in the lower jaw in a 2-year-old boy. He responded well to second surgical intervention with wide resection and immediate reconstruction with plate. The purpose of this case report is to emphasize the particularity of the case, treatment modalities, and differential diagnosis in DF.

Key Words: Desmoplastic fibroma, mandibulectomy, pediatric patient

INTRODUCTION

Desmoplastic fibroma (DF) is a rare benign fibrous lesion.[1] It is considered a bony counterpart of fibromatosis of the soft tissue.[2] DF is an aggressive tumor with a high rate of local recurrence after surgical resection.[3] It has a predilection for craniofacial and long bones including tibia, pelvis, femur, and mandible.[4] Complete resection is the treatment of choice in order to decrease recurrence and morbidity.[5,6] According to our research, at this age, no case of DF has been reported as a recurrence due to incomplete treatment.[1,2,4] Here, we report the clinical course and therapy of a huge mandibular DF occurring in the right side of the mandible of a 2-year-old boy with 18-month follow-up.

CASE REPORT

A 2-year-old boy was referred to the Department of Oral and Maxillofacial Surgery of Children's Medical Center, Tehran, Iran, with a history of a painless mass in the right side of his lower jaw before 2 months [Figure 1a]. On extraoral examination, no associated lymphadenopathy was observed, and the mass was non tender. On intraoral examination, swelling in the right side of the mandible with firm consistency was evident. Computed tomography (CT) demonstrated a lesion extending from the first left primary molar to the right ascending ramus [Figure 1b]. The patient`s parents reported that he underwent a previous surgical treatment for a lesion in the same area about 1 year ago [Figure 1c]. The previous surgeon with an initial diagnosis of benign spindle cell tumor enucleated the lesion and used a titanium plate for reconstruction. The patient after less than 1 year presented with the recurrence of the lesion around the plate in the same anatomical site. The differential diagnoses based on

Access this article online

Website: www.drj.ir
www.drjjournal.net
www.ncbi.nlm.nih.gov/pmc/journals/1480

How to cite this article: Mohammadi F, Shirani G, Derakhshan S, Faghihi T. Desmoplastic fibroma of the lower jaw in a 2-year-old patient; report of a rare case. Dent Res J 2020;17:231-4.

perforation of the mandibular cortex and aggressive behavior of the lesion were primary bone sarcomas such as osteosarcoma or chondrosarcoma and other intraosseous soft-tissue sarcomas. Aggressive benign spindle cell tumors such as DF should be ruled out. Under local anesthesia, an incisional biopsy of the lesion was obtained. Histologically, a nonencapsulated hypocellular lesion composed of proliferation of oval-to-spindle-shaped cells with minimal atypia in the fibromyxoid background which infiltrated the muscular bundles and regional trabecular bone was observed [Figure 2a and b]. Based on histopathologic features, the differential diagnosis included neurofibroma, DF, and low-grade sarcomas such as low-grade leiomyosarcoma or fibrosarcoma. Immunohistochemically, the tumor cells showed strong immunoreactivity for vimentin and B-catenin but were nonreactive with S-100 protein and desmin, and the MIB-1 (Ki67) index was low (1%) [Figure 2c and d]. Based on the clinical, radiological, and histopathological features, a diagnosis of recurrence of the previous pathologic lesion, DF, was rendered. The patient underwent segmental mandibulectomy [Figure 3a and b]. Simultaneously, reconstruction of the defect was performed with a titanium plate (Synthes: Uiv.Fracture Plate 2.4, angled, DePutySynthes, Switzerland). At 18-month clinical and radiographical follow-up, the patient showed no evidence of recurrence with satisfactory functional and esthetic outcome [Figure 4a-c]. Prior

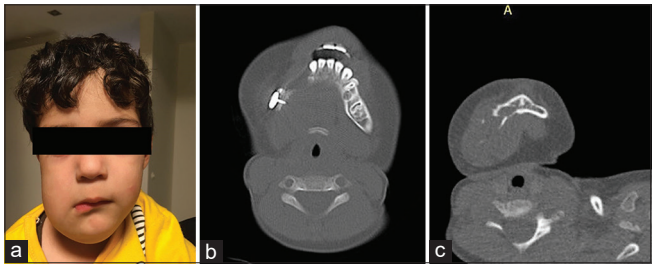

Figure 1: Clinical and paraclinical features of the patient. (a) Clinical view. Note facial asymmetry due to right mandibular swelling, (b) Axial view of computed tomography scan demonstrating radiolucent mandibular lesion with bone perforation and soft-tissue component, (c) The radiographic feature of the primary lesion about 1 year ago before any surgical intervention.

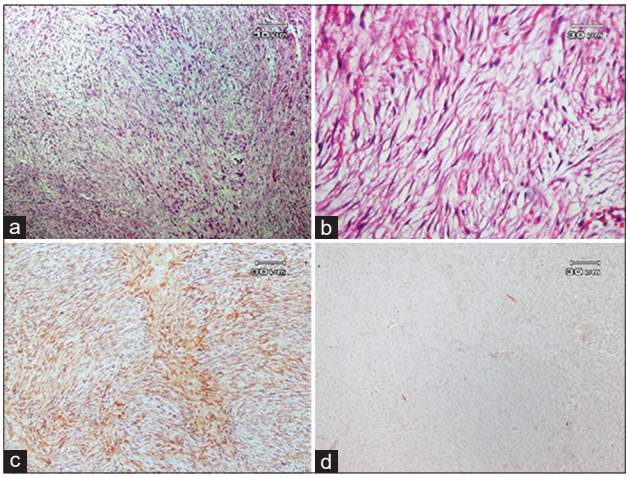

Figure 2: Histopathological and immunohistochemical examination of the lesion (H and E), (a) Proliferation of spindle cells in a fibromyxoid background (×100), (b) High-power field of hypocellular tumor reveals minimal atypia (×400), (c) Diffuse immunopositivity for B-catenin in the tumoral cells (×100), (d) Low immunoreactivity for Ki67 (>1%) in the tumoral cells (×100).

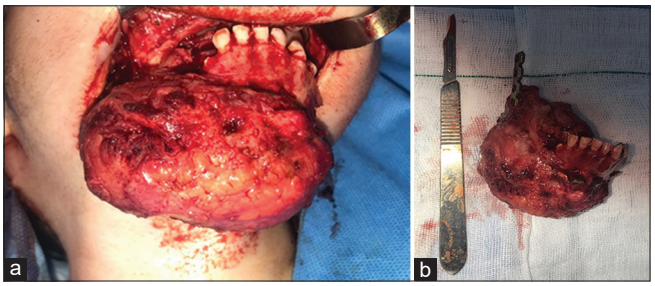

Figure 3: Operation images. (a) Intraoperative picture showing the lesion, (b) Whole lesion excision with previously applied plate.

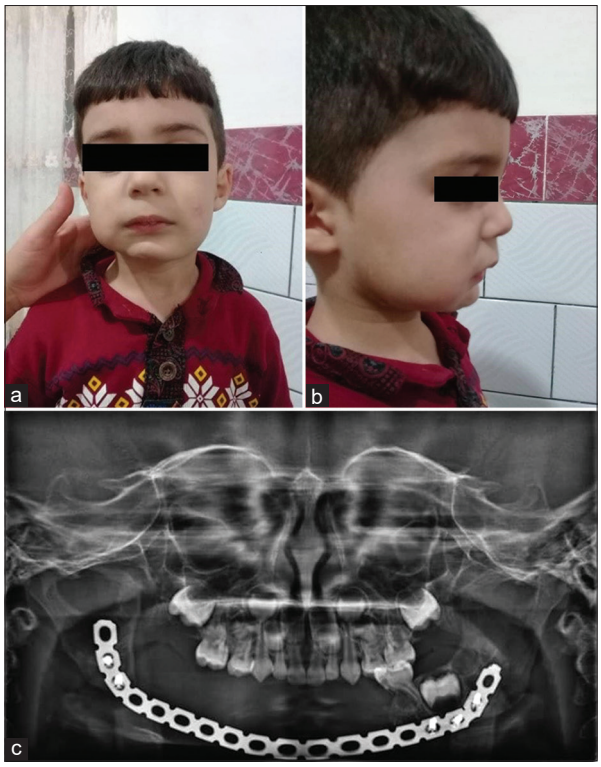

Figure 4: Clinical follow-up of the patient after 18 months. (a) Frontal view, (b) Lateral view, (c) Panoramic view.

to reporting the case, informed consent was obtained from the parents of the patient.

DISCUSSION

DF is a locally aggressive spindle-cell tumor with minimal atypia.[7] Although possible etiologic factors such as trauma[8] and endocrine[8] and genetic factors[9] have been suggested, the etiology of tumor is still unknown.[10] DF is a rare mesenchymal neoplasm which accounts for 0.3% of all benign osseous tumors.[11] Jalalian *et al*. reported approximately 100 cases of DF.[12] The mean age of patients at the time of final diagnosis is 15.1 years old,[13] while our case was far younger. Said-Al-Naief *et al*. showed a marked female predilection unlike our patient who was a male.[4] Clinically, there is no specific sign and symptom for DF; although most patients have mentioned pain and swelling, the lesion in our case was totally asymptomatic. Fractures are more frequent in long bones.[10]

Böhm *et al*. reported mandible as the most common site of DF,[14] similar to the location of DF in our case.

Radiographic features are not pathognomonic. They extend from unilocular to multilocular radiolucencies with well-defined or diffused borders with thinning and some with perforation of the buccal or lingual cortex.[15] Cortex perforation is evident in the CT of our patient, similar to the case of DF of infraorbital rim reported by Safi *et al*. which had been broken through outer cortex and extended into the overlying soft tissue.[16]

Histologically, the tumor comprised of interlacing bundles of fibroblasts in a matrix of collagen fibers.[1] It was difficult to differentiate it from low-grade fibrosarcoma.[4] Therefore, immunohistochemical evaluation was suggested. Low-grade fibrosarcoma and other sarcomas showed higher Ki67 immunoreaction versus benign spindle cell tumors such as DF. In addition, fibrosarcoma did not reveal positive reaction with β-catenin which can confirm the diagnosis. In our study, vimentin and β-catenin signal was positive and tumoral cells were nonreactive to S100 and smooth muscle actin (SMA). Jaafari-Ashkavandi *et al*. reported a periapical DF in the maxillary alveolar bone mimicking an odontogenic lesion, in which the cells were strongly positive for vimentin and negative for S-100 protein, SMA, and CD34, similar to the results of the present case.[17]

The differential diagnosis included other benign spindle cell tumors and also low-grade sarcomas, especially fibrosarcoma. As mentioned, positive immunohistochemistry reaction to β-catenin and low Ki67 can help achieve a definitive diagnosis.

Due to the aggressive nature of this benign lesion, different treatment protocols have been suggested.[3] Gersak *et al*. reported a recurrence rate of 37%–72% of the lesion, with higher recurrence rate after enucleation and curettage.[18] Analyzing the results of Böhm *et al*. showed no recurrences after resection with wide surgical margins.[14] Due to the lesion`s potential for mutagenic transformation into fibrosarcoma, radiotherapy is not suggested.[10] Therefore, complete resection was our treatment of choice. Although literature highlights the nonmetastasizing feature of the lesion, Min *et al*. reported the first case of malignant transformation of DF in the femur.[19]

Because the resection approach can result in loss of mandibular continuity and can lead to severe growth anomalies, mandibular reconstruction is mandatory. In this patient, reconstruction of the defect was carried out by a biocompatible titanium plate.

Although literature suggest that the therapy is effective after a 3-year recurrence-free follow-up,[20] our patient was reassessed 18 months after the surgical procedure. No sign of recurrence was found.

CONCLUSION

We reported the case of a 2-year-old boy with a mandibular DF with a rapid-happened recurrence in less than 1 year because of the incomplete previous surgical intervention. DF is a benign but aggressive tumor with a high rate of recurrence after enucleation and curettage.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parents have given consent for their child's images and other clinical information to be reported in the journal. The parents understand that their child's name and initials will not be published, and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

The authors of this manuscript declare that they have

no conflicts of interest, real or perceived, financial or non-financial, in this article.

REFERENCES

1. Khatib B, Pogrel MA. Desmoplastic fibroma of the mandible in young children – A case series. Int J Oral Maxillofac Surg 2017;46:173-80.
2. Skinner HR, Vargas A, Solar A, Foncea C, Astorga P. Desmoplastic fibroma of the mandible in a pediatric patient: A case report of resection and reconstruction with A six-year follow-up. J Oral Maxillofac Surg 2017;75:1568.e1-1568.e10.
3. Stefanidis K, Benakis S, Tsatalou E, Ouranos V, Chondros D. Computed tomography and magnetic resonance imaging of desmoplastic fibroma with simultaneous manifestation in two unusual locations: A case report. J Med Case Rep 2011;5:28.
4. Said-Al-Naief N, Fernandes R, Louis P, Bell W, Siegal GP. Desmoplastic fibroma of the jaw: A case report and review of literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2006;101:82-94.
5. Bontemps P, Brix M, Simon E. Desmoplastic fibroma of the zygoma. Rev Stomatol Chir Maxillofac Chir Orale 2014;115:169-71.
6. Taconis WK, Schütte HE, van der Heul RO. Desmoplastic fibroma of bone: A report of 18 cases. Skeletal Radiol 1994;23:283-8.
7. Fletcher CD. Pathology and genetics of tumors of soft tissue and bone. World Health Organ Classif Tumors 2002;4:35-46.
8. Reid EN, Lawoyin DO, Suresh L, Longwe E. Desmoplastic fibroma of the anterior mandible. Case report and review of literature. N Y State Dent J 2009;75:32-3.
9. Bridge JA, Swarts SJ, Buresh C, Nelson M, Degenhardt JM, Spanier S, *et al.* Trisomies 8 and 20 characterize a subgroup of benign fibrous lesions arising in both soft tissue and bone. Am J Pathol 1999;154:729-33.
10. Jamali M, Lazow SK, Solomon MP, Berger J. Desmoplastic fibroma: Report of rare lesion in unusual craniofacial location. N Y State Dent J 2013;79:43-5.
11. Herford AS, Reder P, Ducic Y. Multifocal desmoplastic fibromas of the mandible. J Oral Maxillofac Surg 2001;59:1078-81.
12. Jalalian H, Mortazavi H, Baharvand M. Second report of a mandibular desmoplastic fibroma with sunray appearance and review of other imaging features. Dent Med Probl 2015;52:107-11.
13. Ikeshima A, Utsunomiya T. Case report of intra-osseous fibroma: A study on odontogenic and desmoplastic fibromas with a review of the literature. J Oral Sci 2005;47:149-57.
14. Böhm P, Kröber S, Greschniok A, Laniado M, Kaiserling E. Desmoplastic fibroma of the bone. A report of two patients, review of the literature, and therapeutic implications. Cancer 1996;78:1011-23.
15. Inwards CY, Unni KK, Beabout JW, Sim FH. Desmoplastic fibroma of bone. Cancer 1991;68:1978-83.
16. Safi Y, Shamloo N, Heidar H, Valizadeh S, Aghdasi MM, Eslami Manouchehri M, *et al.* Desmoplastic fibroma, report of a rare case in infraorbital rim. Iran J Radiol 2015;12:e25518.
17. 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;34:247-50.
18. Gersak MM, Lenghel LM, Baciut G, Moldovan I, Mitea H, Botar-Jid C, *et al.* Ultrasonography of desmoplastic fibroma of the mandible; a case report. Med Ultrason 2015;17:407-10.
19. Min HS, Kang HG, Lee JH, Lee GK, Ro JY. Desmoplastic fibroma with malignant transformation. Ann Diagn Pathol 2010;14:50-5.
20. Makek M, Lello GE. Desmoplastic fibroma of the mandible: Literature review and report of three cases. J Oral Maxillofac Surg 1986;44:385-91.