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HAMED JALALIAN^{1, A}, HAMED MORTAZAVI^{2, D, E}, MARYAM BAHARVAND^{2, B}

Second Report of a Mandibular Desmoplastic Fibroma with Sunray Appearance and Review of Other Imaging Features

Drugi opis włókniaka desmoplastycznego dającego odczyn poświaty słonecznej w żuchwie i przegląd innych cech w obrazie radiologicznym

¹ Department of Oral and Maxillofacial Surgery, Dental School, Zahedan University of Medical Sciences, Zahedan, Iran

² Department of Oral and Maxillofacial Medicine, Dental School, Shahid Beheshti University of Medical Sciences, Tehran, Iran

A – research concept and design; B – collection and/or assembly of data; C – data analysis and interpretation; D – writing the article; E – critical revision of the article; F – final approval of article

Abstract

Desmoplastic fibroma (DF) is a rare, non-metastasizing, intraosseous benign but locally aggressive tumor of connective tissue origin, which represents only 0.06% of all bony tumors and 0.3% of benign bony tumors. There are no specific radiological features in the diagnosis of desmoplastic fibroma. Unilocular or multilocular radiolucencies with well-defined borders were generally seen, as well as occasional destruction of the mandibular border and root resorption. One of the extremely rare radiographic patterns in desmoplastic fibroma is a sunray appearance (as a periosteal reaction). According to the data available to us, this radiographic pattern of desmoplastic fibroma was first found by Iwai in a 4-year-old girl. The aim of this article was to present the second report of a pediatric mandibular desmoplastic fibroma with sunray appearance. Meanwhile, a review of literature regarding the different imaging features of desmoplastic fibroma is presented. The key message of this case report is that sunray appearance should be considered a radiographic pattern in benign central tumors such as desmoplastic fibroma by clinicians (*Dent. Med. Probl.* 2015, 52, 1, 107–111).

Key words: desmoplastic fibroma, mandible, sunray appearance.

Słowa kluczowe: włókniak desmoplastyczny, żuchwa, poświata słoneczna.

Desmoplastic fibroma (DF) of bone is a rare, non-metastasizing, intraosseous benign but locally aggressive tumor of connective tissue origin [1]. This lesion represents only 0.06% of all bone tumors and 0.3% of benign bone tumors [2]. It closely resembles soft tissue desmoid tumors of the abdominal or extra-abdominal regions, which were first described in 1832 by Macfarlane [3]. Jaffe [4] reported the first case series of DF as a benign bone tumor in the tibia, femur and scapula in 1985. In 1965, Griffith and Ibry [5] found the first case of DF involving oral structures. Earlier, Burch

and Woodward [6] had reported a case of mandibular central fibroma in 1960. The age range of patients with DF is reported to be from birth to 71 years, but 75% of patients are younger than 30 [2, 7]. DF is more common in long bones (85%), followed by the mandible and pelvis [7, 8]. In cases of lower jaw involvement, the posterior aspect of the mandible, the angle, and the ascending ramus were affected in 80% of cases [2, 9, 10]. There is no apparent sex predilection [11]. Clinically, a jaw lesion usually manifests as an asymptomatic, slowly growing swelling, but if symptomatic, it may

mimic toothache [7]. According to some authors, DF can be seen as a locally aggressive tumor when the growth is rapid and destructive [2, 12]. Desmoplastic fibroma does not have any specific radiological features. Unilocular or multilocular radiolucencies with well-defined borders were generally seen, as well as occasional destruction of the mandibular border and root resorption [2]. One of the extremely rare radiographic patterns in DF is a sunray appearance (as a periosteal reaction). According to the data available to us, Iwai [2] first found this radiographic pattern of DF in a 4-year-old girl. In contrast, Bakaeen [7] pointed out that the periosteal reaction is lacking. Various surgical procedures such as local curettage, wide excision, resection, or mandibulectomy have been suggested in managing DF [12].

The aim of this paper was to report a mandibular DF with sunray appearance mimicking osteogenic sarcoma in a 5-year-old boy. We also reviewed the literature about imaging findings in DF of bone.

Case Report

A 5-year-old boy was referred to the Department of Oral and Maxillofacial surgery at Zahedan Dental School, Zahedan, Iran, with the chief complaint of a painful swelling of the left mandible, for about 20 days. The patient had a history of trauma during a car accident in this site one month before admission.

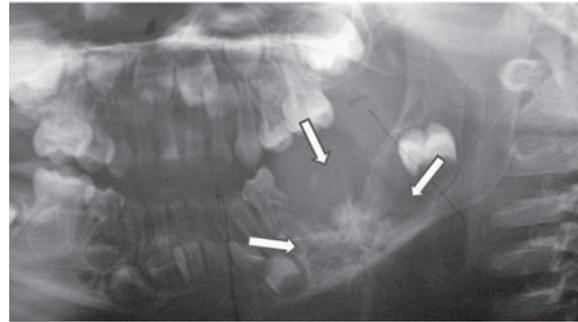


Fig. 1. Panoramic view demonstrating the sunray appearance of the lesion

He did not have any systemic disease and general examinations revealed no pathological alterations of any other body structures. Extra-orally, asymmetry was obviously observed, however the overlying skin was normal in color and texture. Intra-orally, no mucosal changes were found, and the teeth did not show mobility. Palpation revealed a tender, hard mass in the buccal vestibule at the left side of the mandible. On the panoramic radiograph, we noticed a unilocular, mixed radiolucent-radiopaque lesion with periosteal reaction of the mandibular border as a sunray appearance (Fig. 1). In addition, computed tomography (CT) showed a mixed radiolucent-radiopaque lesion with lateral and medial expansion with a sunray appearance and bone perforation (Fig. 2). A tentative diagnosis of malignant osteogenic tumor (Ewing's sarcoma and osteogenic sarcoma) was made. A biopsy specimen was obtained under local an-

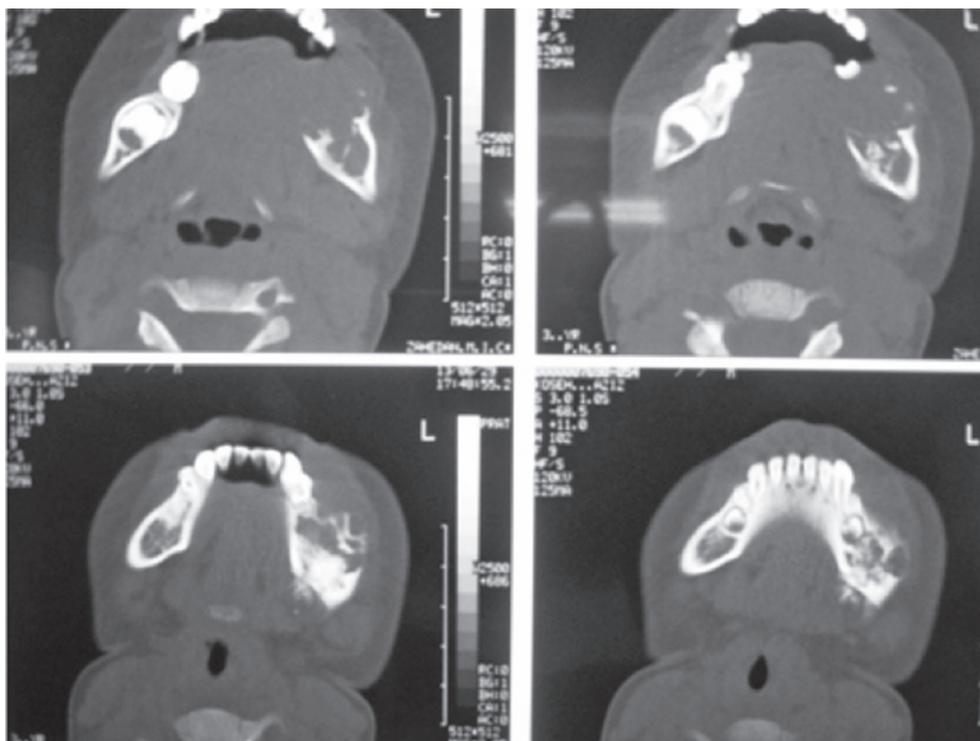


Fig. 2. Axial CT scan of the patient, which shows the expansion and perforation of mandibular bone

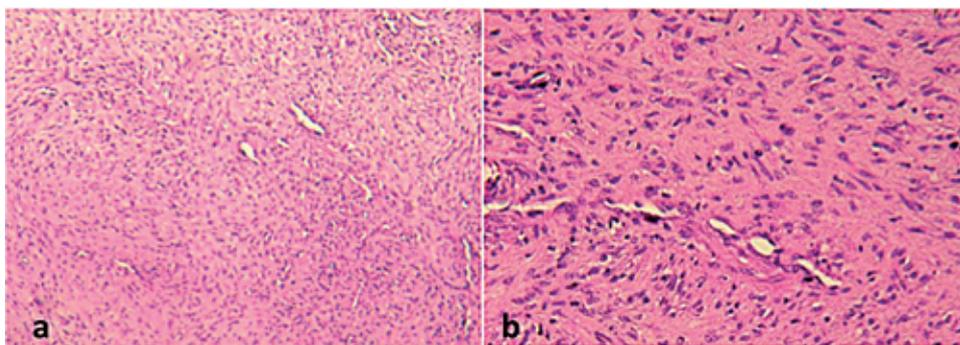


Fig. 3a–b. Histopathological sections show a loose cellular fibrous tissue containing slender trabeculae of woven bone of variable shapes as well as scattered osteoblasts, confirming the diagnosis of desmoplastic fibroma (Hematoxylin & Eosin, $\times 40$)

esthesia, and a histopathology examination established the diagnosis of DF (Fig. 3). Finally, under nasal intubation anesthesia, the site of the lesion was exposed through an extra-oral (submandibular) approach and resection of the left mandible was done from angle to symphysis with 1 cm safe margins. Reconstruction of the mandible was performed using bone graft from the fibula and 2 mini plates. The left submandibular salivary gland was removed, because it was in the field of surgery. The postoperative specimen was re-evaluated microscopically and the diagnosis of DF was confirmed. There was no sign of recurrence after 14 months of follow-up.

Discussion

Desmoplastic fibroma of bone is considered the intraosseous counterpart of common soft tissue desmoids or fibromatoses [13–15]. The main cause of this entity is unknown, although trauma, endocrine discrepancies and genetic factors have been implicated [12, 16]. The most likely precipitating factor is trauma to the affected site [5]. In long bones, pathologic fracture is usually a presenting sign of this lesion, while in the craniofacial region a gradual swelling, asymmetry or both most commonly alert the patient and physician to its presence [17–19].

In this paper we reported mandibular DF in a 5-year-old boy. In accordance with our report, Sandrini et al. [8], Shekhar et al. [9], and Rius Peris et al. [10] have documented the presence of DF in males. However, based on other reports, there is no significant sex predilection [7, 11, 20, 21]. The involvement of children with DF has been documented in case reports by Ikeshima and Utsunomiya [18], Nussbaum et al. [22], Kwon et al. [23] and Tepleton et al. [24]. Hopkins et al. [12] demonstrated that the mean age of patients with DF of the jawbone was 14 with a range of 12 months to 46 years.

In agreement with our experience, Bakaeen [7] and Ikeshima and Utsunomiya [17] noticed that the mandible is the most common affected site when DF arises in the head and neck. Gradual

swelling of the mandible in our case had a history of 20 days. This duration has been found to be between two weeks and 6 months in other studies [2, 7, 8]. Our patient had mild pain. According to Bakaeen [7], jaw lesions usually appear as asymptomatic, slowly growing swellings, but if symptomatic, it resembles toothache. As we observed, a sunray appearance of the mandibular border, a malignant tumor was first suspected. This radiographic pattern is very rare in DF. Periosteal reaction occurs when a lesion destroys cortical bone, extends to the lateral periosteum and elevates it, resulting in new bone proliferation from the inner layer of the periosteum [2]. There are several reasons why a sunray appearance was seen in our patient: the invasive character of the tumor, with possible spread under the periosteum, the relatively fast tumor growth, and the very young age of the patient all could have contributed to the vigorous periosteal new bone formation [2].

According to Crim et al. [25], DF of the bone and its radiographic features have been addressed infrequently in the radiology literature. Radiography offers more information about bony lesions and remains the cornerstone for the differential diagnosis of skeletal tumors or tumor-like lesions because of its higher specificity to detect morphologic hallmarks of the tumor. The radiographic features that help the radiologist make the diagnosis or at least narrow the diagnostic possibilities include lesion margins (sclerotic rim to ill-defined margins), patterns of bone destruction (geographic, moth-eaten or permeated), internal characteristics of the lesion (non-matrix producing tumor, non-mineralized matrix producing tumor), type of host bone response (medullary or periosteal), site (metaphysis, diaphysis or epiphysis), and position (central, eccentric or periosteal) of the lesion in the skeletal system and in the individual bone, soft tissue involvement, and single or multiple lesion nature. Patterns of bone destruction, margins and reactive changes in the host bone clearly show the growth rate of a bone lesion, which reflects its biologic activity. The matrix of the lesion, as well as lesion site and position may allow a specific di-

agnosis. This information, in addition to clinical findings, helps define whether the lesion is neoplastic or non-neoplastic, malignant or benign, primary or metastatic, and will help further direct the subsequent work up as well [26].

Crim et al. [25] analyzed the radiographic findings of DF in a retrospective study and summarized them as follows: 96% of patients had a geographic pattern of bone destruction with a narrow transition zone and non-sclerotic margins. Internal pseudo-trabeculation and cortical breaching was also found in 91% and 28% of cases, respectively.

In another study by Ikeshima and Utsunomiya [17], multilocular and unilocular lesions were seen in 27.5% and 8.6% of patients, respectively. 60.8% of cases showed a radiolucent lesion, 4.3% had mixed lesions, and only one case (1.45%) was radiopaque. 2% of lesions had well-defined borders. Bone expansion, destructive periphery, and root resorption were also reported in 31.8%, 11.5%, and 5.9% of cases, respectively.

According to Frick et al. [27], radiographs revealed that DFs of the bone were centrally in 61% and eccentrically in 39% of patients. The matrix was osteolytic in 24%, osteolytic with coarsened ridge-like trabeculae in 63%, and mixed lytic and mildly sclerotic in 13% of cases. Cortical breaching was present in 53% of lesions. Well-defined, partially well-defined and ill-defined margins were also found in 34%, 61%, and 5% of cases, respectively. The margins were sclerotic in 46% of lesions [27].

Computed tomography showed that 65% of lesions were osteolytic. Mixed osteolytic and mildly sclerotic lesions were detected in 35% of cases. Cortical destruction and a soft tissue mass extending beyond the original destroyed cortex were observed in 88% and 41% of patients, respectively [27].

Mahnken et al. [28] evaluated the cross-sectional imaging patterns of DF and noticed that

CT demonstrates more accurate details of bone destruction, including cortical and articular invasion. In addition, soft tissue extension and intra-medullary tumor growth are best diagnosed with MRI. On un-enhanced T1-weighted images, DF exhibited nonspecific low signal intensity and an intermediate to high signal intensity including areas of low intensity on T2-weighted images. DF showed a disparate, heterogeneous gadolinium enhancement. Although cross-sectional imaging features of DF are not specific, some MR characteristics such as inhomogeneous contrast enhancement and the presence of low-intensity regions on T2-weighted images are helpful in establishing the differential diagnosis [28].

The radio-graphical differential diagnosis of DF includes fibrosarcoma, giant cell tumor, aneurismal and solitary bone cyst, hemangioma, fibrous dysplasia, chondromyxoid fibroma, central ossifying fibroma, traumatic bone cyst, and some odontogenic cysts and tumors such as ameloblastoma, myxoma and keratocyst [8, 12].

Varying surgical procedures, radiation therapy, and chemotherapy have been suggested in the management of DF of the bone [8]. It is noted that DF treated with a major excision or resection tends not to reoccur, but lesions treated with simple enucleation or local excision recur in 20% to 40% of cases, and tumors treated only using curettage reoccur in up to 70% of cases. Curettage can be used only in intra-osseous lesions without signs of cortical rupture or extension to the soft tissue. In lesions with aggressive behavior, resection should be considered [8]. Radiotherapy is not indicated as a primary treatment method, and can be used when the lesion is inoperable. Chemotherapy has also been mentioned as an adjunct to total resection [8, 12].

In conclusion, a sunray appearance should be considered as a radiographic pattern in benign central tumors such as desmoplastic fibroma.

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Address for correspondence

Hamed Mortazavi
Department of Oral and Maxillofacial Medicine
Dental School
Shahid Beheshti University of Medical Sciences
Chamran Highway
Tabnak St, Daneshjoo Blvd
Zip code: 1983963113
Tehran
Iran
Tel.: +89 21 29902311
Fax: +98 21 22403194
E-mail: hamedmortazavi2013@gmail.com

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