Rare Desmoplastic Fibroma in Ramus and Mandibular Angle: Presentation of a Case Report With Review of Literature

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Abstract- Desmoplastic fibroma is a rare, benign fibrous tumor of bone which is locally aggressive. The mandible is the most prevalent affected of the facial bones. Nevertheless, reports of desmoplastic fibroma affecting the mandible are seldom in the literature. It causes bone destruction and has a high tendency for local recurrence. In this case report, we report a case of desmoplastic fibroma which is located in mandibular angle and extended to coronoid process, in 20-year-old female patient.

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Keywords: Desmoplastic fibroma; Mandible

Introduction

For the first time, Desmoplastic fibroma (DF) was introduced by Jaffe in 1958 (1) as a primarily intra-osseous fibromatosis of the femur, the tibia and also of the scapula (2). DF is a rare and benign myofibroblastic tumor that is locally invasive (3). This tumor includes less than 1% of bone tumors and 0.3% of all benign bone tumors (4,5).

The most common site of involvement in head and neck area is jaw, and the mandible is more frequently affected than the maxilla (3,6). DF usually occurs in young adults, especially during the second and third decades of life (3) and based on majority of the researches, it happens equally in men and women (7). For the first time, Griffith and Irby in 1965 reported desmoplastic fibroma in the mandible (8,9).

It presents typically a painless swelling which is growing within a few weeks or months and is firm on palpation (3). This lesion causes bone destruction and if it doesn't treat, it tends to invade the surrounding soft tissues. The recurrence depends on types of treatment plans; if inadequate treatment is done, a high tendency for local recurrence may happen (3,4).

Given the aggressive behavior of this tumor and its rare occurrence in worldwide, we report a case of desmoplastic fibroma with a relatively large expansion in a 20-year-old female with this tumor. We hope that our case report will shed new light on this special entity.

Case Report

A 20-year-old female patient referred to the Department of Oral and Maxillofacial Pathology, Qazvin University of Medical Sciences, with the chief complaint of sudden pain and swelling in the left side of her face for 40 days ago. In extra-oral examination, clear asymmetry was observed.

Intra-oral examination revealed no significant findings like teeth missing or displacement. The panoramic view showed well-defined and non-corticated radiolucency with irregular borders that are unclear in some areas, in posterior left side of mandible that extended to coronoids process (Figure 1).

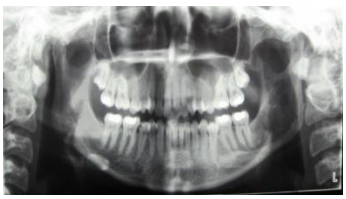

Figure 1. Panoramic view showing well-defined radiolucency in the posterior left side of mandible that extended to coronoids notch

To find out more, a Computer Tomography (CT) scan was requested. The radiologist reported a large lytic expansile eccentric bony lesion involving ramus and mandibular angle. The lesion extended medially to the medial pterygoid muscles and also filled infratemporal

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space (Figure 2). According to CT scan view, radiologists considered malignant lesions like Ewing sarcoma and Burkitt's lymphoma in differential diagnosis.

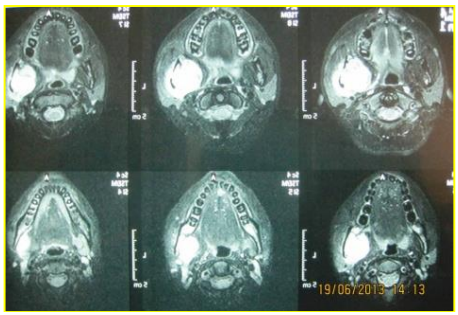

Figure 2. CT scan shows an expansile lesion that leads to cortical destruction

In MRI with multiple sections, there was an expansile lesion in left mandibular ramus with abnormal signal intensity in T1 as the low signal intensity in T1, high signal intensity in T2W images with adjacent soft tissue component that shows extension into left para pharyngeal space. No evidence of lymphadenopathy in submandibular space was seen (Figure 3 and 4).

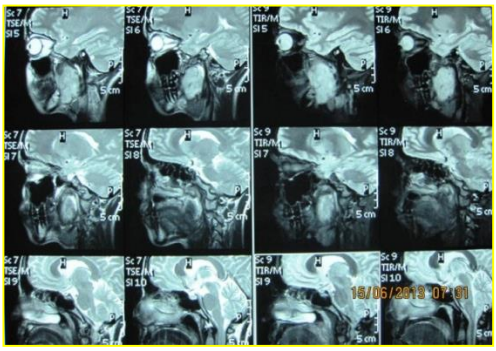

Figure 3. MRI (Sagittal view)

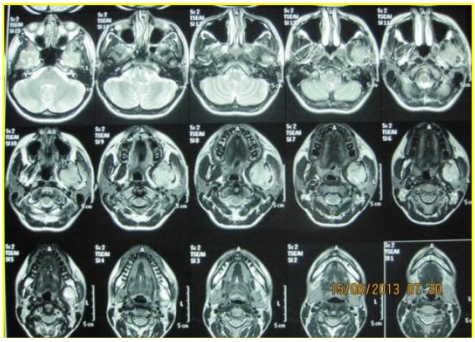

Figure 4. MRI (Axial view)

Due to the MRI view, lesions like fibrous dysplasia, aneurysmal bone cyst, eosinophilic granuloma and osteomyelitis considered in the differential diagnosis.

In the next step, an incisional biopsy was done from para pharyngeal mass, and then it was sent to the Oral & maxillofacial department of dental branch, Qazvin University Of Medical Sciences for microscopic examinations. Histopathological examination (Figure 5 and Figure 6) revealed proliferation of plump and ovoid fibroblasts in a collagenous stroma which is loosely in some areas. The fibroblasts were not atypical, and mitoses were seen sparsely.

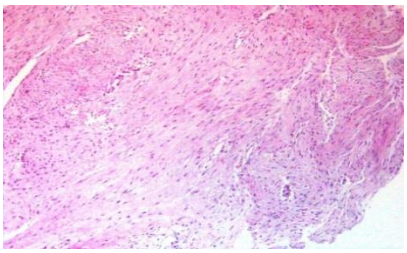

Figure 5. Hematoxylin and eosin-stained section showing proliferation of uniform–appearing spindle-shaped cells with intervening collagen (original magnification × 40)

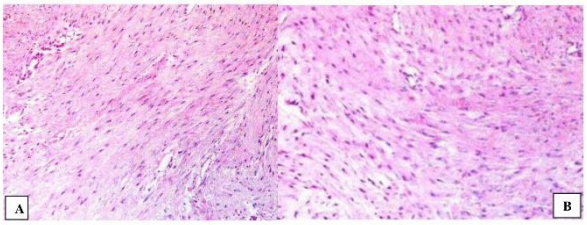

Figure 6. Hematoxylin and eosin-stained section showing interlacing fascicles of spindle-shaped cells within collagenous stroma (A) (original magnification × 40), (B) (original magnification × 100)

According to the microscopic view, to determine the nature of the cells, Immunohistochemistry (IHC) stain was done; IHC results were reported as follows:

Vimentin: positive/ Desmin: focally positive/ S100: negative/ and Ki-67: < 1%.

Finally based on clinical-radiographic and histopathologic features and immunohistochemical findings, desmoplastic fibroma was the final diagnosis.

For treatment, an extensive segmental resection in addition to removal of involved surrounding soft tissues was done. Unfortunately, the follow up has not been done due to the lack of patient collaboration.

For reviewing the international literature, a systematic search in the PubMed database of the National Library of Medicine was performed using the keywords "desmoplastic fibroma", "mandible" and "jaw".

Discussion

DF is a rare and non-metastasizing, often locally

invasive intraosseous neoplasm with normal-appearing fibroblasts (4,10). Regezi *et al*., described DF as a benign locally aggressive neoplasm of bone which has similar features with soft tissue fibromatoses and desmoid tumors (9,10). In other words, desmoplastic fibroma of the bone is now considered the intraosseous counterpart of the common soft-tissue desmoid or fibromatoses (11).

Because of its locally infiltrative nature, some authors place this entity in a borderline position with regard to malignancy. Recognition of this disease is critical because it may histologically and radiographically resemble benign fibrous lesions with conservative treatment or, more significantly, with spindle cell sarcomas with aggressive treatment (10).

The annual incidence of desmoplastic fibroma is 2-4 per 1 million individuals (5,12).

Review of the literature, despite some contradictory results, attests the rarity of these tumors and their low incidence. For example, Guruprasad *et al.,* (2015) stated that" since Jaffe original description in 1958, a number of small series and case reports have brought the total number of published cases to approximately 150" (13).

Focusing on the DF occurrence in the mandible using the keywords 'DF' and 'mandible'also showed the following results: Shekhar *et al*., (2011) reported only 14 previous cases of DF affecting the angle of the mandible in the English literature (3).

Mohamad *et al*., (2017) stated that "up to the present, about 180 of DF of mandible cases have been reported in the literature" (14). Konikath *et al*., (2019) said "up to new, fewer than 100 cases of DF in the mandible have been published to date" (10).

DF commonly occurs in the first two decades of life (reported mean age of 14 years) (9). It can affect any age group, occurs most often in the first three decades (10). The age incidence of DF is first to the third decade of life (4). DF of the jaws is most commonly discovered in the second and third decades with the mean age of 15.7 years and with slight female predilection (13). Our case happened in 20 years old patient who is similar to the results of previous studies.

The site of predilection within the jawbone is the mandible (the ramus, angle, and molar area). Mandibular involvement is reported to be approximately 40% of the various bony sites (10). The most common site of involvement in the mandible (22%), followed by pelvis (13%) and long bones such as the femur (15%), radius (12%) and tibia (9%) (15) contrary to the results found in study by Mohamad *et al*., (14). Mohamad *et al.,* present that DF usually involves the tibia, scapula and femur. The mandible represents the fourth most commonly affected site. It has a predilection to affect the posterior part of the mandible, most often in the ramus-angle region (14). In the present case, DF occurred in posterior part of mandible, a common site of involvement. Of course, it should be noted that the lesion had a large extension to the ramus and coronoid process.

In the present case report, a woman who was suffering from the disease referred to our department. But in most previous studies, there was no significant tendency to men and women (4,10) and even in Frick et al study a slight predominance of men is reported (11).

The etiology of DF involves a wide range of factors including genetic predisposition, endocrine factor and trauma such as fractures and surgery (4,9,12).

Generally, there is no pathognomonic sign and symptom for DF, but pain and swelling have been declared by most patients (9). Although, some lesions can be asymptomatic. Most common signs of this lesion which is mentioned in the literatures were asymptomatic swelling (72%) followed by the facial asymmetry (20%), tooth displacement with or without roots divergence (12%), the history of trauma to the affected area (13%), trismus (7%) and teeth mobility (5%). Pathological fractures also have been reported in 9-15% of the patients (14-16). In our patient pain and swelling were the main symptoms and obvious asymmetry was observed too; but no pathological fracture or tooth displacement was found.

In the literature, the most consistent radiographic findings are well defined lytic lesions (94%), with internal pseudo-trabeculation (91%) and bone expansion (89%) (15). Radiographic features in the present case are also similar to the frequent radiographic view means lytic lesion. Radiographically, DF usually mimics other lytic lesions. DFs are characterized by bone expansion, well or ill-defined margins, benign features with pattern of destruction, multi- or unilocular and totally radiolucent or radiolucent-radiopaque lesions. The multilocularity is three times more frequent based on previous reports (9). Sometimes rapid expansion followed by cortical perforation, leads to the formation of a new bone structure from osteogenic layer of periosteum (15).

Also, Hashimoto and colleagues (1991) analyzed the radiological findings in 47 cases of desmoplastic fibroma; well-defined multilocular radiolucency was common radiographic finding in these series (17). According to radiologic concepts, this rare lesion may mimic fibro-osseous, benign and especially malignant lesions. So many lesions such as myxoma, giant cell tumors, aneurysmal and traumatic bone cyst, hemangioma, fibrous dysplasia, chondromyxoid fibroma, ossifying fibroma, eosinophilic granuloma, sarcomas and

metastatic lesions are classified as a differential diagnosis for this lesion (9,11).

Regarding different treatment plans, identification of this lesion is essential. Establishing a diagnosis is difficult by imaging studies alone. If cortical destruction and a soft tissue mass are noted, desmoplastic fibroma of bone may resemble more sinister pathology, such as fibrosarcoma, intra-osseous osteosarcoma and metastases (9,18).

Magnetic resonance imaging (MRI) is typically used to determine the tumor type and extent of it and is suitable for surgical planning (2,12). In MRI, a majority of reported cases have shown low to intermediate signal intensity foci on T2 weighted images and no restriction of the apparent diffusion coefficient, which confirms a benign but locally aggressive neoplasm without an inclination to malignancy (10,12)

A DF macroscopically has a firm consistency with well-defined advancing surfaces that may extend into surrounding soft tissue (19).

In 2002, DF was defined as a rare benign bone tumor which includes spindle cells with minimal cellular atypia and the overproduction of collagen (20). Numerous authors have subsequently attempted to clarify the histological criteria and, in 2013, the WHO described the microscopic appearance of desmoplastic fibroma as being composed of the slender, spindle to stellate cells with minimal cytological atypia and abundant collagenous matrix (18). It is characterized by low cellularity with some possible foci of hypercellularity. Lack of a capsule and infiltrative nature of the lesion are hallmarks of DF (10).

Histological analysis is the gold standard for diagnosis (12). The diagnosis is dependent on the correlation of clinico-radio-pathological data (12).

In histopathological examination, it is important to distinguish and rule out some lesions in the differential diagnosis of DF by histopathological features. For example: Low-grade fibrosarcoma and low-grade intraosseous osteosarcoma are the most difficult differential diagnoses. In some cases of low-grade fibrosarcoma, mitoses are not manifested and areas with predominant collagen tissue may be found, which makes the distinction extremely difficult (10), but herringbone pattern and osteoid formation are the helpful features distinguish these two malignancies.

Fibrous dysplasia looks likes DF in areas with fibrous connective tissue and no obvious osteoid production. So, additional sampling to recognize areas of bone formation can be helpful. Also the nuclei in fibrous dysplasia are shorter and more compact-looking than the elongated, slender nuclei seen in desmoplastic fibroma (13).

Fibrous histiocytoma exhibits a storiform pattern of collagen bundle arrangement, but DF displays the interlacing pattern of collagen bundles arrangement (3).

Apart from histopathological examination which reveals a mesenchymal tumor, composed of spindle-shaped cells with myofibroblastic differentiation, abundant collagen formation and low proliferation activity; immunohistochemical stainings can be helpful for diagnosis, because similarity of the tumor-cells to the other tumoral cell (spindle-shaped). The tumor cells showed a positive reaction for smooth muscle actin and a negative reaction for S100. With the proliferation marker Mib1 less than 5% of the tumor cells proved to be positive (2).

Immunophenotypic analysis showed that DF shared features identical to those of aggressive fibromatosis of the soft tissue, including positive b-catenin (nuclear-positive) and vimentin expression, whereas the expression of Desmin, S-100, CD34, and MDM2 was negative (5). However, a recent study revealed that there were no mutations in exon 3 of CTNNB1, encoding for B-catenin, thus genetically distinguishing desmoplastic fibromas of bone from desmoid-type fibromatosis (18). In the present case, we used some immunohistochemical markers such as S-100, vimentin, desmin and proliferation marker that confirmed the diagnosis.

Due to the locally aggressive behavior of DF and its high recurrence rate, choosing the appropriate treatment is important (12). The recurrence rate has been reported in various articles, ranging from 17% to 55% following resection or curettage (12). Other studies presented significant rates of recurrence (up to 67%) that have been associated with DF, depending on the completeness of surgical removal (10).

Different treatment methods have been suggested for DF that are the source of controversy. Enucleation, curettage, resection, radiotherapy and chemotherapy are usual implemented treatment modalities. Radiotherapy due to its mutagenic effect is not recommended in most pieces of literature. This method is likely to cause post-radiation sarcoma (9). Most clinicians seem to favor surgical resection of the lesion with a wide margin, in order to avoid recurrence [Hopkins *et al*., 1996; Iwai *et al*., 1996; Templeton *et al*., 1997; Said-Al-Naief *et al*., 2006] (21).

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