DESMOPLASTIC FIBROMA OF THE MANDIBLE: A CASE REPORT

A. Pınar Sumer, DDS PhD  
Professor, Department of Dentomaxillofacial Radiology,  
Faculty of Dentistry, University of Ondokuz Mayıs,  
Samsun, Turkey

Mahmut Sumer, DDS PhD  
Professor, Department of Oral and Maxillofacial Surgery,  
Faculty of Dentistry, University of Ondokuz Mayıs,  
Samsun, Turkey

Armağan Çalışkan DDS  
Research Assistant, Department of Dentomaxillofacial Radiology,  
Faculty of Dentistry, University of Ondokuz Mayıs,  
Samsun, Turkey

Ömer Günhan, MD, PhD  
Professor, Department of Pathology,  
Gülhane Military Medicine Academy,  
Ankara, Turkey

ABSTRACT
Desmoplastic fibroma of bone is an uncommon myofibroblastic tumor characterized by local aggressive behavior that is associated with a high-recurrence rate. The lesion most frequently occurs in the mandibular posterior area in children or young adults. Radiographically, it appears as a multilocular or occasionally unilocular radiolucency with well-defined or ill-defined margins. The treatment of patients with fibromatosis consists of wide surgical excision. Radiation therapy sometimes has been used. This report describes a case of desmoplastic fibroma in a 20 year old female presenting as a firm, painless mass in the labial side of the left mandibular lateral incisor with a radiolucency of adjacent bone on radiographs.

Keywords: Desmoid Tumor, Fibromatosis, Mandible


Submitted for Publication: 01.23.2015
Accepted for Publication: 04.06.2015
MANDİBULADA GÖRÜLEN DEZMOPLASTİK FİBROMA:
VAKA RAPORU

ÖZ

Anahtar Kelimeler: Dezmoid Tumor, Fibromatosis, Mandible
INTRODUCTION

The fibromatoses also called desmoids tumors are rare neoplasms that develop from the tissue of the musculoaponeurotic system. In the soft tissues of the head and neck, these lesions are frequently called aggressive fibromatoses or extraabdominal desmoids. Similar lesions within the bone have been called desmoplastic fibromas. The lesion is characterized by local aggressive behavior but it has no metastatic potential. It most frequently occurs in children and young adults however it can affect any age group. The age range of reported gnathic examples is from 10 months to 59 years, with a mean of approximately 16 years. Although some studies have found a male preponderance, a female preponderance was found whereas other studies suggest no sex preponderance. The etiology remains unclear. However, a genetic predisposition, trauma, hormonal factors, an association with familiar adenomatous polyposis and Gardner’s syndrome have been implicated. Aslan et al reported a case of fibromatosis that occurred after a trauma. Kindar described a case that occurred at the incision scar which is extending to the superior peducular screw insertion point after thoracal fracture stabilization. Most of desmoplastic fibromas have occurred in the mandible, especially in the molar angle-ascending ramus area. Patients usually complain of facial swelling, rarely pain and dysfunction may be observed. Said-Al-Naief reported a case of desmoplastic fibroma of the mandible in an 8-year-old boy who initially presented with a 2-month history of a rapidly expanding, painless mass along the right inferior border of his mandible. Radiographically, the lesion appears as a multilocular or unilocular destructive radiolucency with well-defined or ill-defined margins. Because of lesion’s locally aggressive nature, the preferred treatment is wide surgical excision with a clear margin. Adjuvant treatments such as radiotherapy, chemotherapy, hormonal therapy, and non-steroidal anti-inflammatory drugs have also been used. In this article a case of desmoplastic fibroma in a 20 year old female is presented.

CASE REPORT

A 20 year old female was referred with a one month history of an expanding, firm, painless swelling of the left lower jaw. The patient’s medical and family history was unremarkable. Intraoral examination revealed a 0.5x0.5cm in size, hard, non-tender mass on the labial side of the left mandibular lateral incisor. No mucosal surface ulceration was present. Radiographically, a radiolucent destruction area was observed between the mandibular left lateral incisor and canine teeth in panoramic and periapical radiographs (Figure 1). As the lesion was smaller in size, conventional radiographs were sufficient. Computed tomography (CT), and magnetic resonance imaging (MRI) were not required. The lesion was removed surgically via an intraoral approach under local anesthesia and the surgical specimen was sent for histopathologic examination. In histopathologic examination, tumoral tissue parts were observed infiltrated irregularly into the striated muscle. The tumor was composed of ill-defined spindle fibroblastic cells and variable amount of collagen. No atypical mitosis was seen. The diagnosis of the lesion was fibromatosis (Figure 2). The patient was followed up for four years. Figure 3 shows the clinical appearance and radiological imaging of the lesion in four years time without any recurrence.

DISCUSSION

Fibromatosis is a rare benign tumor accounting for about 0.03% of all neoplasms and less than 3% of all soft tissue tumors. Head and neck desmoids tumors are estimated to constitute only 12% of all desmoids tumors throughout the body. The most common locations are the mandible, femur, pelvic bones, radius and tibia. In Sharma et al’s retrospective review, a total of 10 cases of fibromatosis of the head and neck were identified from a total of 340 cases of fibromatosis of the whole body. Pena et al evaluated 97 cases of fibromatosis in the head and neck in the pediatric population and found that the majority of tumors were of the mandible (38%), and other locations included the submandibular area, neck, tongue, sinuses, upper lip and hard palate. In a literature review, 84% cases of desmoplastic fibromas involved the mandible and 70% of these cases were located posteriorly. Fibromatoses show an aggressive and infiltrative behavior. The tumor does not have a capsule and its margins are often poorly defined so it tends to interdigitate with the underlying bone or soft tissue and invades adjacent neurovascular structures. A soft tissue mass will be occurred when the desmoplastic fibroma erodes through the cortex that may cause difficulty to determine whether the lesion is a desmoplastic fibroma of bone with soft tissue extension or a soft tissue fibromatosis with an extension into the bone. Seper et al evaluated 37 published cases of fibromatosis of the mandible and reported that most patients (89%) felt...
Figure 1. Intraoral photograph shows 0.5x0.5cm in size mass on the buccal side of the left mandibular lateral incisor (A). Panoramic (B) and periapical (C) radiographs show the destruction between the mandibular left lateral incisor and canine teeth.

Figure 2. Intraoperative photograph showing the lesion being removed (A), Macroscopic appearance of the lesion (B), Histopathologic examination shows the desmoids tumor composed of ill-defined spindle fibroblastic cells infiltrated into the striated muscle (HEX100) (C)

Figure 3. Postoperative intraoral photograph (A), panoramic (B) and periapical (C) radiographs.
no pain, and the signs such as tooth mobility, limitations of mouth opening were infrequent. A painless swelling of the affected area is the most common initial complaint. Tuncer et al. reported a case of fibromatosis in an 11 year old girl noticed by an extraoral swelling. Fibromatosis should be considered in young patients submitting with a mass in head and neck region in early childhood. In the present case the lesion was on the buccal side of the left mandibular lateral incisor and caused no pain or tooth mobility. X-ray examination, ultrasonography (US), CT, and MRI might be helpful for tumor management, preoperative planning and for follow-up. The radiographic features of desmoplastic fibromas are nonspecific. These include a unilocular or multicellular, well-defined or ill-defined radiolucency with variably expressed marginal sclerosis. Smaller lesions appear to be unilocular and when the lesion is large the internal aspect may be multilocular. Desmoplastic fibromas of bone can expand bone and often perforate the outer cortex. In Said-Al-Naief literature review about desmoplastic fibromas of the jaw, a 30% of cases had cortical perforation and 10% of cases had cortical expansion. MRI used for the assessment of the nature and size of the soft tissue lesion and involvement of surrounding structures. T1-weighted MRI, the internal structure has a low signal, which helps in determining intraosseous extent because of the contrast with the high signal from the bone marrow. CT is valuable in determining the tumor’s relation to bone and US will reveal a heterogenous hypoechoigenicity and increased surrounding vascularity. Usually CT scan and MRI are required to determine the exact soft tissue extent of the lesion. Especially in children, US or MRI was preferred to CT for the follow-up to prevent unnecessary radiation unless bone was involved. In the present case, panoramic and periapical radiographs showed a radiolucent destruction area of adjacent bone and because the lesion was small, there was no need for further radiologic examination. Differential diagnosis should be made with fibrosarcoma, fibroma, giant cell lesions and fibrous dysplasia. Radiographically, the periphery of the lesion most often is ill defined and has an invasive characteristic usually seen in malignant lesions. Distinguishing this neoplasm from a ill-defined fibrosarcoma may be difficult so all desmoplastic fibromas of bone should be considered potentially malignant.

The management of fibromatosis is based on an accurate diagnosis by histologic and clinical examination. Because of its lack of capsule and infiltrative growth, a wide excision of the tumor is advocated. Surgical resection was recommended for most of the cases similar to the present case and sometimes chemotherapy and radiotherapy was given. Wang et al. reported that a conservative resection with preservation of form and function should be given greater priority in all age groups for the treatment of aggressive fibromatosis. Additionally, postoperative adjuvant therapy was vital for patients to obtain recurrence-free survival.

A 23% recurrence rate has been reported for oral and paroral fibromatosis but higher recurrence rate has been noted for other head and neck sites. Kruse et al. reported 30.07% recurrence of a total of 143 published cases of fibromatosis of the head and neck and found no correlation between recurrence and localization of the lesion. Küçük et al. reported a 29% recurrence rate for 31 cases diagnosed as aggressive fibromatoses and suggested adjuvant radiotherapy with surgical treatment for satisfactory clinical results. Close follow-up at least two but preferably more years because of the late relapses was necessary. Recurrences are treated essentially in the same way as primary tumors. The present case is different from other cases in the literature as it was smaller in size so surgical excision was used as treatment and no recurrence was observed for four years. Appropriate radiographic imaging and timing are important for the follow-up for satisfactory clinical results.

ACKNOWLEDGEMENT

The study was presented as a poster presentation in the 17th International Congress on Oral Pathology and Medicine at May 25-30, 2014 held in Istanbul, Turkey

REFERENCES


