An Unusual Presentation of Ossifying Fibromyxoid Tumor of the Mandible: A Case Report

Nur Mollaoglu* / Benay Tokman** / Sevil Kahraman*** / Sedat Cetiner**** / Sule Yucetas*****/ Omer Uluoglu******

A 13-year-old boy who complained rapid swelling on the left side of mandible is presented. Histopathological examination revealed ossifying fibromyxoid tumor (OFMT). OFMT is a rare soft-tissue neoplasm that occurs usually in the subcutaneous tissue of the extremities. Head and neck involvement is relatively rare. In this case, we present the diagnosis, surgical treatment and long-term follow-up of an OFMT due to its unusual site of occurrence. The precise clinical behavior of atypical and malignant types of OFMTs is still unclear. Thus, histopathology report is important, leading surgeon to decide how often and how long to follow-up patient with OFMT.

Key words: *Ossifying fibromyxoid tumor, mandible, enucleation*
J Clin Pediatr Dent 31(2):136-138, 2006

INTRODUCTION

Ossifying fibromyxoid tumor (OFMT) is a very rare soft-tissue neoplasm that was first described by Enzinger *et al.* in 1989.[1] It is an uncommon tumor localized in the subcutaneous tissue as a small, painless, well-circumscribed mass.[2,3] OFMT most often occur in the subcutaneous tissue of the extremities and usually attached to the deeper fascia, muscle, or tendon. Head and neck involvement is relatively rare around 23%. The typical age is adulthood.[3] The histological origin of OFMT is controversial. Most authors insist on Schwannian or neuronal cells origin. Although it is usually reported as a benign tumor, atypical or malignant OFMT have previously been reported.[4] We present the diagnosis, surgical treatment and long-term follow-up of an OFMT due to its unusual site of occurrence. The histogenesis of this lesion remains still uncertain.

CASE PRESENTATION

A 13-year-old boy was admitted to Gazi University, School of Dentistry, Department of Oral and Maxillofacial Surgery with a history of unspecified swelling which demonstrated rapid growing on the left side of the lower chin for the last four months in September 2004. Patient was a 13 year old boy. The medical history of the patient revealed mitral valve prolapse. Therefore, patient was referred to Gazi University, School of Medicine, Department of pediatric cardiology for consultation before performing an incisional biopsy. Antibiotic administration was suggested prior to surgical intervention by the pediatricians for prevention of bacterial endocarditis.

The swelling was located on the vestibular area of the left lower premolars. The size of the tumor was about 2 cm in diameter in MRI findings (Fig 1). Parents of the patient stressed that the swelling was about 1 cm in diameter when they first noticed. It was emphasized that the mass has shown a rapid growth within two months and reached the current size. On intraoral examination, there was a hard, warm, painless large mass on the vestibular side of the left lower premolar area with no ulceration or any color change on the mucosal surface. The panoramic radiograph revealed neither bony mass nor teeth migration in the area of swelling which was an evidence for existence of the soft tissue mass. However, there was a deminer-

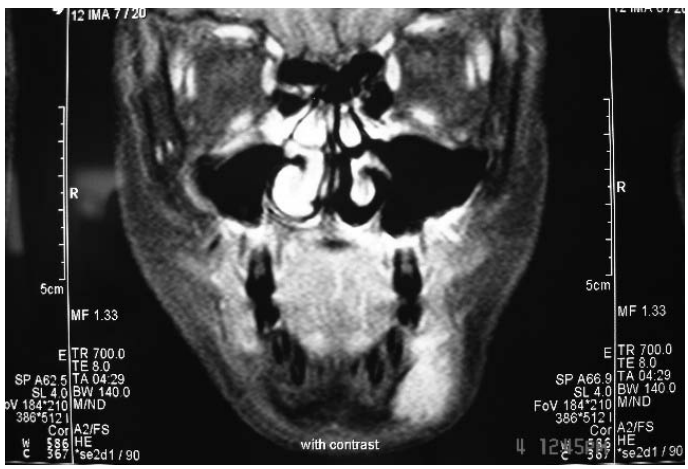

Figure 1: Image of the tumor in MRI

*Nur Mollaoglu. AssociateProfessor., Gazi University, School of Dentistry, Department of Oral & Maxillofacial Surgery
**Benay Tokman. Assistant.Professor, Gazi University, School of Dentistry, Department of Oral Pathology
***Dr.Sevil Kahraman, Gazi University, School of Dentistry, Department of Oral and Maxillofacial Surgery.
**** Sedat Cetiner. Associate.Professor., Gazi University, School of Dentistry, Department of Oral & Maxillofacial Surgery
*****Sule Yucetas. Professor, Gazi University, School of Dentistry, Department of Oral & Maxillofacial Surgery
******Omer Uluoglu. Professor, Gazi University, School of Medicine, Department of Pathology

Send all correspondence to: Prof. Nur Mollaoglu, Bestekar sok. No:61/8, Orta giris, Kavaklidere, 06680, Ankara-Turkey.

Fax: ++ (90)(312) 223 9226

Email: nurmolla@gazi.edu.tr

alization around the apices of the left premolar and molar teeth on the radiograph. Furthermore, the bilateral carotid angiography revealed that the border of the soft tissue mass was not related to border of internal and external carotid arteries which indicating that the lesion was not vascular tissue originated (Fig 2).

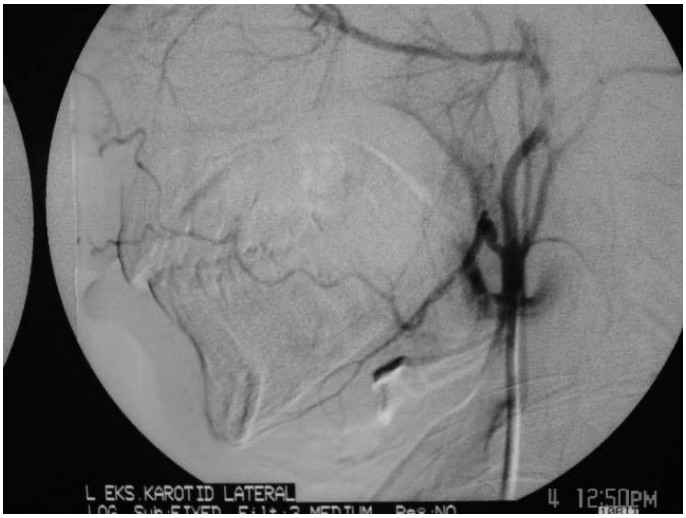

Figure 2: Carotid angiography of the tumor site

An incisional biopsy was performed prior to total excision of the tumor under local anesthesia. The lesion was reported as a benign mesenchymal tumor and in respect of this histopathological report total enucleation of the tumor was planned and patient was operated on October 2004. During the operation, it was noticed that the tumor was located in the area between the lower 2nd incisor and the 1st molar tooth, seated on the surface of the vestibular bone of mandible and made a depression on the bony surface. The mental nerve was in close relation to the tumor. No damage was given to the mental nerve and no paresthesia occurred after the operation (Fig 3). In addition, the tumor was found to be infiltrated to surrounding subcutaneous tissue and muscle in the anterior end. The tumor was enucleated with a wide dissection.

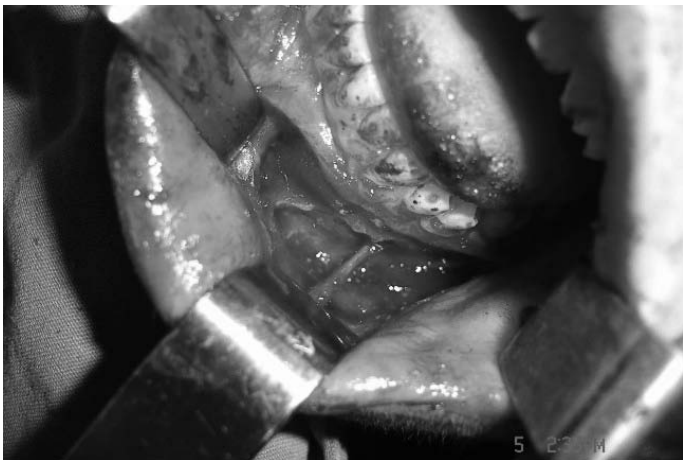

Figure 3: Intra oral image of the tumor cavity following enucleation

Histopathological examination of the lesion showed lobules of fibromyxoid tissue with varying cellularity and metaplastic osteoid formation (Fig 4). The tumor cells were uniform with round-oval vesicular nuclei and ill defined eosinophilic cytoplasm. The matrix

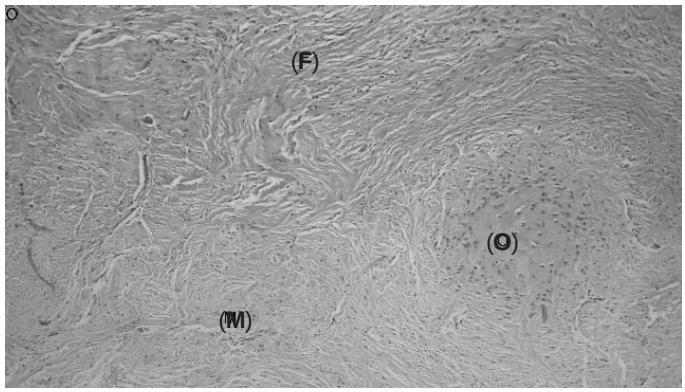

Figure 4: Lobular myxoid matrix (M) and fibrous areas (F) with osteoid (O) formation in OFMT (X10 H&E)

ranged from myxoid to hyalin and richly collagenous. Prominent thin walled capillaries, extravased erytrocytes and scattered mononuclear cells were also present. There was no mitotic activity, pleomorphism or necrosis. Immunohistochemically, cells were intense positive with vimentin and focally positive with smooth muscle actin and glial fibrillary acidic protein. However, cells were negative with S-100, epithelial membrane antigen, pankeratin, desmin, Leu-7 and CD 34. The histomorphology along with the immunohistochemical results was consistent by OFMT.

Patient was followed up once a month for the initial two months following the operation, thereafter called once every six months. No tumor recurrence was evidenced during the follow up visits of the patient.

DISCUSSION

OFMT is usually a slow-growing, well-circumscribed, painless mass that occurs in the subcutaneous tissue and sometimes within muscle.[1,5] It occurs in the 5th to 7th decade of life with a male predominance.[6] It most commonly involves the extremities, although the head and neck and the trunk can also be involved.[7] The mean greatest dimension of OFMTs is 4 cm. The range of 1.5 cm to 17 cm. large size of OFMTs is an indicator of poor prognosis.[8] In our case, the localization of the lesion and the age of the patient were extremely unusual. OFMT was located on the left vestibular mandibular area. In addition, patient was a teenager.

Most OFMTs are composed of small, evenly sized, round, tumor cells separated by variable amounts of osteoid, and, in 80% of the cases, the periphery consists of an incomplete shell of bone.[9] They are usually surrounded by a thick capsule.[2] In this present case, the tumor was about 2 cm in diameter and elliptic shaped. The outer surface of the tumor was surrounded by a thick capsule which was infiltrated to buccal tissues around mentum which was hard to separate from healthy soft tissue.

Since the histogenesis of OFMT is controversial, its diagnosis raises difficulties. The main histological and immunophenotypic differential diagnosis of this tumor should be made with a schwannoma, myxoid chondrosarcoma, low grade fibromyxoid sarcoma, smooth muscle tumors and benign fibro-osseous tumors.[1] In this case histomorphologic features along with the lack of S-100 and Leu-7 expression ruled out schwannoma and myxoid chondrosarcoma. In opposition to the tumor cells of this case, smooth muscle cells are stained strongly by desmin immunostain. In addition, benign fibro-osseous tumor is characterized with cellular fibrous stroma

and many foci of newly formed trabecule of immature bone or osteoid than OFMT. Other malignant mesencymal neoplasms such as fibrosarcoma or mesenchymal chondrosarcoma show hypercellularity and pleomorphism with mitotic activity, which are not consistent by OFMT. Low grade fibromyxoid sarcoma is one of the closest differential diagnose which raises a great difficulty. The distinction from low grade fibromyxoid sarcoma is almost impossible on both conceptual and practical ground. Therefore, total excision and a long follow up period is proposed for OFMT.

Although OFMTs usually behaves in a benign fashion, recurrences and metastasis have been previously reported.[10] Presence of centrally placed osteoid, hypercellularity, high nuclear grade and increased mitotic activity are the signs of aggressiveness for OFMT. The malignant types of OFMT's are called as the "atypical" or "malignant" type.[7,10] Kilpatrick *et al.* suggested naming OFMT as "malignant" for those tumors that had evidence of metastasis and "atypical" for the aggressive masses without metastasis.[10] The present case was an atypical OFMT. Thus, patient's follow up visits are arranged as once a year following the first year.

In conclusion, OFMT is a rare soft-tissue neoplasm. In addition, precise clinical behavior of atypical and malignant types of OFMT is still unclear.[10,11] It may be associated by the growing group of translocation-associated sarcomas, such as extra-skeletal myxoid chondrosarcomas, that are characterized by histological features that diverge minutely from primary tumor to recurrence to metastasis.[7] Thus, histopathology report is off important leading surgeon to decide how often and how long to follow-up patient with OFMT.

REFERENCES

1. Enzinger FM, Weiss SW, Liang CY. Ossifying fibromyxoid tumor of soft parts. A clinicopathological analysis of 59 cases. Am J Surg Pathol;13:817–27,1989
2. Harish S, Polson A, Morris P, Malata C, Griffiths M, Bearcroft PW. Giant atypical ossifying fibromyxoid tumour of the calf. Skeletal Radiol.;24:1-6, 2005
3. Al-Mazrou KA, Mansoor A, Payne M, Richardson MA. Ossifying fibromyxoid tumor of the ethmoid sinus in a newborn: report of a case and literature review. Int J Pediatr Otorhinolaryngol.;68:225-30,2004
4. Ijiri R, Tanaka Y, Misugi K, Sekido K, Yokohama TN. Ossifying fibromyxoid tumor of soft parts in a child: a case report, J. Pediatr. Surg.;34:1294—1296, 1999
5. Ogose A, Otsuka H, Morita T, Kobayashi H, Hirata Y. Ossifying fibromyxoid tumor resembling parosteal osteosarcoma. Skelet Radiol;27:578–80, 1998
6. Schofield JB, Krausz T, Stamp GW, Fletcher CD, Fisher C, Azzopardi JG. Ossifying fibromyxoid tumour of soft parts: immunohistochemical and ultrastructural analysis. Histopathology;22:101–12, 1993
7. Folpe AL, Weiss SW. Ossifying fibromyxoid tumor of soft parts: a clinicopathologic study of 70 cases with emphasis on atypical and malignant variants. Am J Surg Pathol;27:421–3, 2003
8. Holck S, Pedersen JG, Ackermann T, Daugaard S. Ossifying fibromyxoid tumour of soft parts, with focus on unusual clinicopathological features. Histopathology;42:599–604, 2003
9. Nishio J, Iwasaki H, Ohjimi Y, Ishiguro M, Isayama T, Naito M, *et al.* Ossifying fibromyxoid tumor of soft parts. Cytogenetic findings. Cancer Genet Cytogenet.;133:124-8. 2002
10. Kilpatrick SE, Ward WG, Mozes M, Miettinen M, Fukunaga M, Fletcher CD. Atypical and malignant variants of ossifying fibromyxoid tumor. Clinicopathologic analysis of six cases. Am J Surg Pathol;19:1039–46, 1995
11. Goodlad JR, Fletcher CD. Recent developments in soft tissue tumors. Histopathology;27:103–20,1995