Ossifying Fibromyxoid Tumor; An Unusual Tumor in Oral Cavity: Case Report

Ossifiye Fibromiksoid Tümör: Oral Kavitede Sıradışı Bir Tümör

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ABSTRACT Ossifying fibromyxoid tumor (OFT) is a rare soft tissue neoplasm which arises mostly in the extremities. Only few cases of oral cavity are described with limited follow up time. A 23 year old pregnant woman in her second trimester presented with a mass in vestibular region of the mandible complaining of pain for the last six months. The mass was excised and histopathological examination revealed lobules of fibromyxoid tissue with proliferation of small, round to oval epitheliod and spindled cells. The tumor had a partial fibrous capsule surrounded by an incomplete shell of metaplastic bone trabeculae. The diagnosis was typical for OFT. No recurrences were observed after three years of follow up. Although most cases of OFT follow a benign clinical course, recurrence and metastasis are reported, therefore long term follow-up is necessary.

Key Words: Mouth neoplasms; mandible

ÖZET Ossifiye fibromiksoid tümör (OFT), ekstremitelerde görülen nadir bir yumuşak doku neoplazmasıdır. Oral kavitede tanımlanan az sayıda vaka ise sınırlı takip süresine sahiptir. Yirmi üç yaşında ikinci trimesterda olan kadın hasta, alt çene vestibül bölgesinde son altı aydır görülen ağrı ve şişlik şikayetiyle başvurdu. Kitle eksize edildi ve histopatolojik incelemede fibromiksoid doku lobüllerinde küçük yuvarlak-oval epitel ve iğsi hücreler saptandı. Tümör, metaplastik kemik trabekülleri içeren tamamlanmamış bir kabukla döşeli yarı fibröz bir kapsül ile çevriliydi. Tanı tipik OFT olarak konuldu. Üç yıllık takip sonrasında herhangi bir rekürens görülmedi. Çoğu OFT olgularında selim bir klinik tablo görülse de rekürens ve metastazlar da rapor edildiğinden uzun dönem takip gereklidir.

Anahtar Kelimeler: Ağız neoplazileri; mandibula

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Ossifying fibromyxoid tumor (OFT), first described by Enzinger et al. in 1989, is a rare tumor of uncertain differentiation which most commonly arises in the extremities. Tumor represents itself as a small, painless, well-defined, often lobulated subcutaneous mass usually attached to the deeper fascia, muscle, or tendon. In approximately 70% of cases, extremities are involved; less commonly involved sites include the trunk, head and neck, mediastinum and retroperitoneum.[1] OFT almost exclusively affects adults, with rare examples reported in children.[2] The presentation in the oral cavity is rare though a few cases are reported.[2-4] Tumor is mostly regarded as benign. Most often, the tumor is cured with complete

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excision.[5] The percentage of recurrence is about 27% of OFT cases, recurrence in head and neck region is reported as 21% and metastases are extremely rare.[1,6]

CASE REPORT

A 23- year- old woman was admitted to Istanbul University, Faculty of Dentistry, Department of Oral Surgery with swelling and pain complaints in right vestibular region of the mandible for the last 6 months. Prior to admittance, she mentioned that she had visited two other doctors complaining of toothache and minor swelling and she had received antibiotic treatment and a tooth extraction in the affected area.

Her medical history revealed that she was 4 months pregnant. Intraoral examination showed a mass of 4x3x3 cm located in the vestibulum of right mandibular premolar region. The firm mass was ulcerated on the occlusal area due to trauma (Figure 1). Lower right canine and first molar showed a slight migration and mobility, being non-vital in vitality test. The orthopantogram revealed an irregular radiolucent mass with an incomplete ring of peripheral calcification and several sclerotic foci (Figure 2). Considering the patient's pregnancy, advanced screening methods such as CT was not preferred. Along with the aggressive appearance of the lesion and pregnancy, not to risk a possible hematogenous spreading, total excision of the lesion was decided immediately instead of a biopsy. Also, giving the patient local anesthetics for both biopsy and the following surgery was not preferred. Under local anesthesia, excision of the mass with the extraction of right lower first molar, canine and lateral teeth was performed. During the operation, the tumor seemed to be infiltrated to the surrounding bone and vestibular bone showed depression. Also, it was found that the mental nerve was in close relation with the tumor. Mental nerve was not damaged and the patient did not complain of any paresthesia following the surgery.

Histopathological examination of the lesion showed lobules of fibromyxoid tissue with proliferation of small, round to oval epitheliod and spindled cells (Figure 3a). The tumor had a partial fibrous capsule surrounded by an incomplete shell of metaplastic bone trabeculae. The stroma ranged myoxid to hyaline with prominent thin walled vessels. Extravased erytocytes, scattered mononuclear cells, metaplastic bone formation and calcifications were present in some areas of the tumor (Figure 3b). Mitotic activity, necrosis or pleomorphism were not observed. Immunohistochemical staining was performed and the tumor cells were negative for smooth muscle actin and S100 protein.

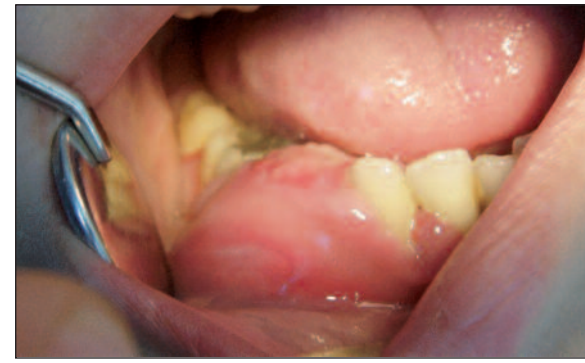

FIGURE 1: Pinkish firm mass with ulcerations on the occlusal surface.

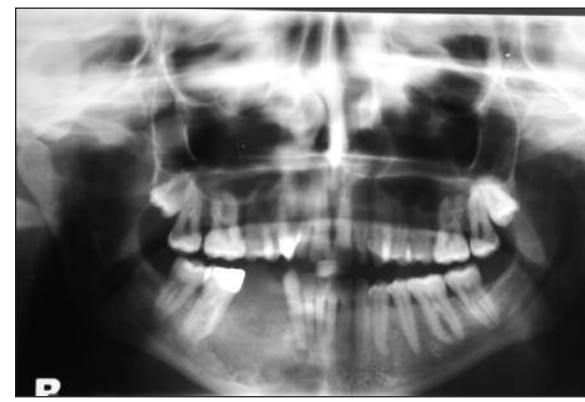

FIGURE 2: Panoramic radiography showing an irregular radiolucent mass with an incomplete ring of peripheral calcification involving adjacent teeth.

Patient was followed up once a month until her delivery. She had no complications post operatively and also regarding her pregnancy. She was followed twice in the first year and once in the following years. Three years of follow up did not show any signs of recurrence (Figure 4).

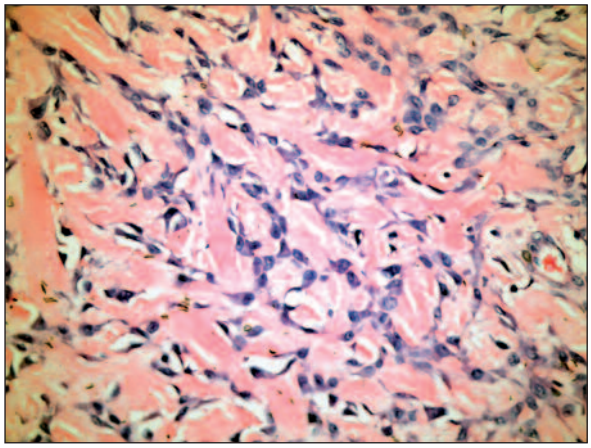

FIGURE 3a: Lobules of fibromyxoid tissue with proliferation of small, round to oval epitheliod cells (H&E x400).

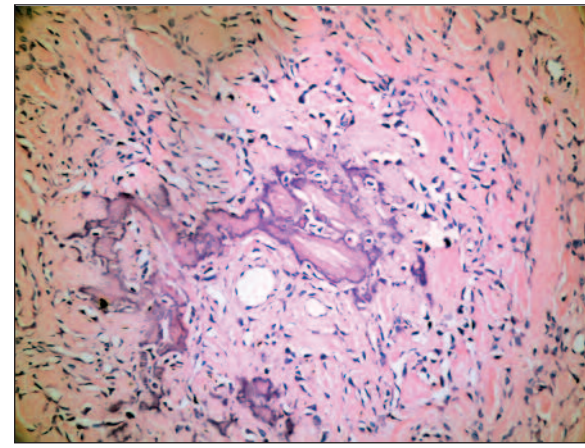

FIGURE 3b: Calcified areas are seen within the fibromyxoid tumor (H&E x200).

DISCUSSION

OFT usually occurs in adults and mostly located in the subcutaneous tissues of extremities.[2] In our case, the patient was a 23 year old woman with the lesion presenting in oral region. There are few reports of OFT in children and adolescents the youngest being 3 weeks old infant.[4] Only 7 cases are reported in the literature in which the lesions were located in oral region including buccal mucosa, mandibular gingiva, inner cheek and lower lip.[2,3,4,6,7] Most often the lesion is found in deep tissues, subcutis and muscles.[1] In the presented case, the lesion was found in the mandibular bone resulting in severe bone resorption which is not often seen with OFT.

Immunohistochemistry can be used as an adjunct method to confirm the histopathological diagnosis of OFT. Histological examination showed typical appearance of an OFT and immunohistochemistry did not generate diagnostic findings for this tumor. OFT is often positive for vimentin, S100 protein and CD10 and can show expression of α smooth muscle actin, Leu 7, desmin and neuron specific enolase.[7] In the case presented, α smooth muscle actin and S100 protein was negative. Studies indicate positivity rates ranging from 60% to 94% for S100 protein.[5,7] It is suggested that loss of S100 protein may be associated with atypia and malign transformation.[5] However, histological findings in the present case, the lack of necrosis, pleomorfism and mitotic activity does not agree with a possible malign transformation or atypia.

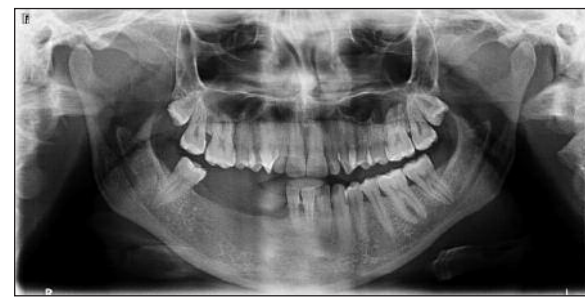

FIGURE 4: Panoramic radiograph of the patient 3 years after the surgery.

Differential diagnosis of OFTs includes myxoid chondrosarcoma, low-grade fibromyxoid sarcoma and schwannoma. Low-grade sarcoma is one of the most common specific sarcoma types to be misinterpreted as a malignant OFT with histological features such as atypical spindle cells with myxoid and collagenous matrix, whorled growth patterns, tumor cells with epithelioid morphology and immunohistological characteristics as lack of S100 protein. But features such as the presence of occasional hyaline rosettes can help in differentiating the lesions.[7]

Median preoperative duration of OFT is 4 years with a range up to 30 years due to the slow

and painless progression of the lesion.[7] The presented case had a relatively faster progression of the lesion with only 6 months of swelling and pain complaint. Oral cavity is important in mastication, phonation and aesthetics. Since most of OFTs occur in deep soft tissues, recognition and growth of the lesion can be overlooked by patients. Prior to the patient's admission to our clinic, trauma caused by the interventions of other dentists such as performing extractions in the tumor's location may have contributed to the progression of the lesion and increase in pain.

None of the cases in the literature show OFT in a young pregnant woman. She was in her second trimester during the surgery. Hormonal and immunological changes during pregnancy are known to effect oral tissues. Gingival enlargements and pregnancy tumors are among common oral manifestations.[8] Head and neck tumors arising during pregnancy are present in the literature, including melanomas, lymphomas, thyroid carcinomas, laryngeal carcinomas and squamous cell carcinoma.[9] Although there is no evidence to indicate direct correlation of pregnancy with these lesions, pregnancy is a state of increased metabolic and hormonal activity with relative immunosupression which may affect the progression and outcome of treatments as well as diseases.

The majority of OFT is histologically benign and pursue a benign clinical prognosis. However; typical, atypical and malignant forms of OFT were reported in literature.[5,10,11] Typical OFTs are composed of a low to moderate cellular proliferation of uniform, round-to-slightly ovoid cells embedded in a myxoid, fibromyxoid, or hyalinized stroma without necrosis and vascular space invasion. In a clinical study with a median follow up duration of 55 months show no metastases or local recurrences with typical OFT.[10] However, another study indicates the overall recurrence and metastatic rates of typical OFT as 17% and 5%, suggesting this tumor to be considered a lesion of intermediate malignancy.[5] With malignant and atypical OFT local recurrences and distant metastases are more common although this form of OFT is rare.[5] According to Folpe and Weiss' classification of OFTs, histological examination identified the lesion as a typical OFT in this case.[5] In accordance with the literature, follow up of 3 years showed no recurrence. However, recurrences are observed late, mostly after 10 years after treatment.[7] Therefore, long term follow up is necessary.

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