



## CASE REPORT

### OSSIFYING FIBROMYXOID TUMOR- A CASE REPORT

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#### ABSTRACT

Ossifying fibromyxoid Tumor (OFMT) is a neoplasm of varying histopathology; it is rare and affects males of middle age. It has an intermediate malignancy potential (rarely metastasizing). The clinical picture of OFMT presents as a slowly expanding mass, which arises from subcutaneous tissue and is not fixed to underline bone. Presented here is a case of eleven years old female presented with a chief complaint of aesthetically displeasing, slowly enlarging painless mass on the right side cheek of the face.

#### INTRODUCTION

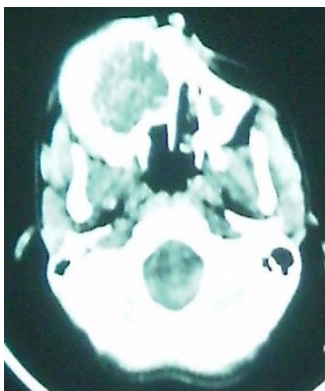
Ossifying fibromyxoid Tumor (OFMT) is a soft tissue tumor of a mostly benign nature infrequently encountered by health care professionals was first documented in 1989 by Enzinger et al. He described OFMT as a rare, soft tissue tumor that occurs mostly on the trunk, extremities, and maxillofacial region.<sup>1</sup> The clinical picture of OFMT presents as a slowly expanding mass, which arises from subcutaneous tissue and is not fixed to underline bone. The cause of tumor has been an enigma as histopathological studies suggested that it follows an uncertain line of differentiation;<sup>2</sup> however, a recent study has declared that OFMT is a translocation-based neoplasm with specific genetic material repeatedly arranging itself.<sup>3,4</sup> Prevalence and incidence of OFMT is rare<sup>1,2</sup> due to which it is often misdiagnosed as some other common soft tissue tumor. It has male predilection, and the age range is from 14–83 years.<sup>2</sup> OFMTs are considered as tumors having intermediate malignancy potential, meaning they rarely metastasize. Most of these tumors are benign in nature. However, metastasis to distant sites has also been reported in the literature. OFMTs also have the potential to recur locally.<sup>5</sup> Hardly any case of OFMT in the maxillofacial region is documented from Pakistan.

#### CASE REPORT

An 11 years old female patient presented with a history of slow-growing painless mass on the right side of her palate that was also causing right cheek swelling for the last one year. On extraoral clinical examination, there was firm to hard, non-tender, swelling approximately 6x8cm in size present on the right side cheek region of the face. It was extending mediolaterally from the right lateral side of the nose, obliterating the right nasolabial fold, stretching the right corner of the upper lip, and laterally till 3 cm away from the tragus. In the superioinferior plane, the swelling was extending 1 cm away from the right orbital margin until 3 cm above the mandible's inferior border. The bulk of swelling was bulging inferiorly. Overlying skin temperature and the color were normal as compared to the adjacent skin. Vision was intact. There was no nasal blockage. All cranial nerves were intact.

Intraorally, the swelling was 5x5 cm in size, extending anteriorly from the upper right lateral incisor extending posteriorly till the right maxillary second molar. It was causing the obliteration of the right maxillary buccal vestibule. Maxillary dentition was embedded in the mass, and it was causing the displacement of teeth. Overlying mucosa of the mass was normal with underlying

hard mass (Figure 2). The right Stenson duct was not appreciable due to mass. Mass had indentations of lower occluding teeth with it. Neck examination was unremarkable with no lymphadenopathy. Computer tomography was carried out that showed it to be the mass on the right side of the maxilla and palate bulging downward on the mandibular teeth and causing destruction of the right side of the maxilla (Figure 1).



**Figure 1: Axial view showing the extent of the lesion**



**Figure 2: Intraoral view showing mass**



**Figure 3: Intraoral defect after tumor removal**

Informed consent was taken. Alginate impressions were taken and model surgery was performed on the models. The acrylic palatal plate was made to use after resection in post-operative healing time. General anesthesia was given with left nasotracheal intubation. Tumor margins were identified, and peri-lesional incisions were made (Figure 3). Bristow elevator was used for tumor retrieval (Figure 4,5). Pressure packs with sponges were applied for bleeding control. There was not significant bleed, and it stopped after five minutes for sponge pressure. Bone ronger was used to trim the sharp edges of the bones that were further smoothed by the flame bur with saline irrigation. The remnant of sinus linings was removed by curettage, and copious irrigation was done to remove all bone remnants. Polyfex pack was placed in the right defect and an acrylic plate was applied for comfortable healing of the wound. Post-operative antibiotics and analgesic were administered, and she was discharged from the ward on third post-op day. Pack was changed after 48 hours and she was recalled after five days. The tumor was subjected to histopathology, which showed it to be the ossifying fibromyxoid tumor of the maxilla. She was followed up for three years. Her defect was epithelized and there was bone formation. Later dental rehabilitation with removable denture was done.



**Figure 4: Superior view of the resected tumor**



**Figure 5: Occlusal view of the resected tumor**

## DISCUSSION

Ossifying fibromyxoid Tumor (OFMT) was first documented almost 30 years ago in 1989 by Enzinger et al. based on his study of 59 cases for 25 years. He described OFMT as a rare, soft tissue tumor arising from subcutis, which occurs mostly on the trunk, extremities, and maxillofacial region.<sup>1</sup> In order of frequency of lesion according to the site, extremities are most commonly affected, followed by trunk and then head and neck region.<sup>1</sup> Median age is 50 years ranging from 14-83 years; the

male population is more affected, with male to female ratio of almost 1.5:1.<sup>2</sup> Ossifying fibromyxoid tumor is a slowly growing mass, typically small in size, which is usually painless.<sup>3</sup>

The tumor name implies the histology of the tumor. A thick fibrous band or capsule makes the outer lining of the tumor mass, fibrous bands projecting inside the tumor mass, giving it a lobulated appearance divided by fibrous septa.<sup>2</sup> It also has osseous content, mostly present as a layer of bone underneath the fibrous capsule, but in some cases, the osseous content represents focal areas of calcification. Small spindle cells with round or oval nuclei are distributed in fibromyxoid stroma. The tumor has a rich vasculature.<sup>6</sup>

Radiologically, on CT evaluation, it is noted that the tumor is bounded peripherally by an incompletely formed bony ring in about 60-70 percent of cases.<sup>1,2,5</sup> The tumor has a significant intra-lesional component of calcification.<sup>2</sup> Overall, a moderate enhancement of lesion is seen on contrast-enhanced CT scan. Peripheral bony shell or metaplastic bone appears on T1, and T2-weighted magnetic resonance imaging (MRI) as low signal intensity structures, whereas the fibromyxoid matrix of the

lesion appears isointense to adjacent muscle on T1 weighted MRI and intermediate to high signal intensity is seen T2 weighted MRI.<sup>2</sup>

Treatment of choice is the surgical removal of the tumor. Complete surgical excision must be carried out in order to prevent the local recurrence. Recurrence is not uncommon, with 17-27% of recurrence documented in literature after complete surgical removal of the tumor mass,<sup>1,7</sup> with 22% of all recurrence cases reported after more than ten years of surgical excision.<sup>5</sup> Since these lesions have intermediate malignancy potential,<sup>8</sup> a 5% metastatic recurrence rate is reported.<sup>5</sup>

## CONCLUSION

Although OFMTs are rarely encountered by health care professionals, it is recommended that it should be considered in the differential diagnosis of soft tissue lesion, particularly those which are firm to hard and on histopathological evaluation have significant ossification.

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