Case Report

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Juvenile ossifying fibroma in a 14-year-old female: A case report

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A B S T R A C T

Juvenile Ossifying Fibroma (JOF) is a rare benign bone neoplasm affecting age-group of 5-15 which tends to mimic malignant lesions. This is in opposition to its conventional variant which occurs mostly in middle aged cohort and is relatively docile. It's aetiology remains unknown and it is characterized by the substitution of normal bone by mineralized fibrous tissue. The rapid progression, aggressive nature and high recurrence potential are features that makes juvenile ossifying fibroma of vying attention. Additionally, its prognosis being greatly dependent on time and type of intervention employed is widely documented. We present as case of JOF in a 14 years old female. The affected bone was maxilla with dental and palatal involvement. Partial Maxillectomy and BIPP for reconstruction were applied as treatment of choice. Yearly follow up protocol for next 5 years was devised. No post op complications were observed.

Key Words: Juvenile ossifying fibroma. Pasammomatoid Ossifying Fibroma.

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Introduction

Ossifying fibroma is a rare, benign fibro-osseous neoplasm. It is marked by the replacement of normal bone by fibrous tissue containing varying amounts of mineralized substance. On clinic-pathological grounds, WHO has divided the lesion into two categories, conventional and Juvenile.[1] The former is a slow growing well demarcated lesion mostly affecting Maxilla and mandible and is most common in third to the fourth decade of life.[2] The latter in contrast presents between age 5 to 15, is clinically more aggressive and has a tendency to recur. These constitute only 2% of tumors in children. [1,2]

Both types are histologically similar and believed to arise from the multipotent mesenchymal cells contained in the periodontal ligament or the periosteum.[3] JOFs tend to mimic malignant tumors.[4] Mostly asymptomatic they can present as a painless, round, expansive mass which may deteriorate adjacent anatomical structures. Radiographically they vary in radiolucency.[5] The definitive pathology remains unknown and their diagnosis and treatment presents clinical challenges. We present as case of JOF in a 14 years old female with a history of seven months.

Case Report

A 14 year old female presented to the department of ENT of Pakistan Institute of Medical Sciences with complaint of unilateral expansile swelling of left cheek for seven months. She had no known premorbids and the swelling was noticed by patient as a small diffuse patch which progressively and rapidly increased in size and extent. The expansion was both intra-oral and extra-oral. It was painless and non-tender, associated with on and off gingival bleeding and caused loss of upper left molar teeth. There was no complaint of nasal obstruction, nasal discharge, anosmia, decreased vision, diplopia, ear ache or ear discharge. Systemic complaints were also absent.

Patient had not used any medication nor did she have history of any known allergies. She did not have any

significant past medical, surgical, personal or family history.

On extra-oral examination, the patient had a 6x8 cm firm to hard expansile swelling of left cheek extending from the left floor of orbit to the left maxilla and from lateral wall of nose to the zygomatic arch. It was extending to nasal cavity as a bulge on left lateral wall and also downward into oral cavity at left upper alveolus displacing the teeth. The overlying mucosa was intact. The swelling was well demarcated with regular margins, immobile, non-tender and adherent/fixed to overlying skin. The overlying skin was non-ulcerated and bore no anomaly. It was also non-fluctuant, non-reducible and non-compressible.

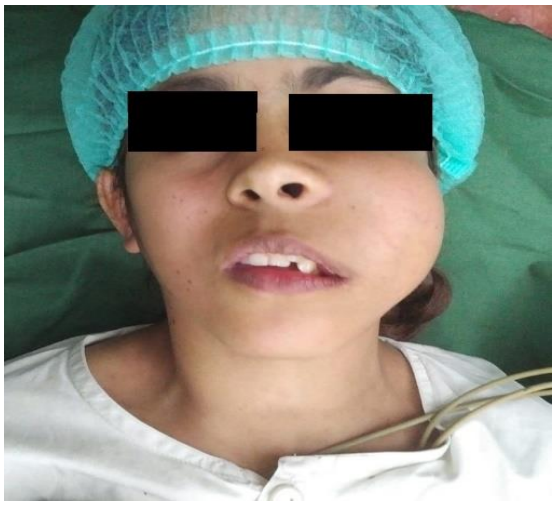

Figure 1- Unilateral swelling Left Cheek

On intra-oral examination, the swelling was partially edentulous, firm in consistency and noted in left posterior region of hard palate. It was causing obliteration of left infraorbital rim superiorly and was causing expansion of lower alveolus inferiorly. On the upper alveolar ridge (left) it was extending from incisor to molar teeth. It involved hard palate. Blood vessels were visible on the surface. The throat was clear and tongue movements non-affected. There was no associated lymphadenopathy. Systemic examination was normal and had no anomalies.

All base line blood work was in normal range. Aspiration of the lesion was negative. On CT Para-nasal sinuses the lesion presented as a large, well defined, rounded, unilocular, expansile avidly enhancing soft tissue attenuation area measuring 5.1 x 5 x 5.7cm. It had calcified walls and internal opacification arising from superior alveolar arch of left aspect of maxilla with erosion of bone and impacted molar tooth. Superiorly the lesion was completely opacifying the left maxillary sinus causing expansion of its walls and extending up till floor of left orbit. However, there was no intra-orbital extension. Medially it was causing mass effect on ipsilateral nasal cavity. Anteriorly it was causing bulge on overlying skin and subcutaneous tissue. Inferomedially it was bulging into oral cavity with erosion of hard palate. Remaining CT Paranasal sinuses appeared normal.

Tissue removed from affected Maxilla was taken for histopathology. On microscopic exam, the specimen was noted to be invaded by a tumor comprising of fascicles of fibroblasts like cells with interspersed bone formation and showing osteoblastic rimming at some foci and areas of calcification. No areas of mitoses and necrosis were identified. There were numerous large dilated vascular spaces lined at most areas by endothelial cells. Bone was seen adjacent to lesion. On gross appearance lesion consisted of round white tissue. The mass was homogenous white in appearance. All the margins of the specimen were involved except for the bone.

Surgical removal of lesion was carried out. The patient was put under general anesthesia and a nasogastric tube was placed. Weber Ferguson approach was used and an incision was made from vermillion border along the lip extending around base of nose and along the facial groove. It was then infraorbitally extended to the lateral canthus 3-4mm below the cilium.

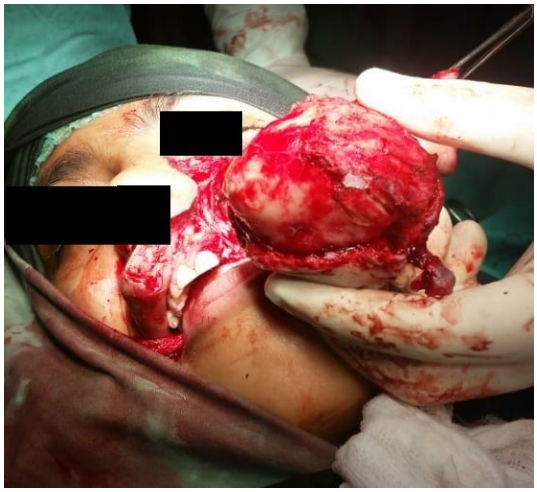

Figure 2- Excised lesion

After opening the incision and raising a flap, lesion was removed completely. Five osteotomies were done i.e. lateral nasal wall, inferior portion of floor of orbit sparing the rest, pterygoid plate, medial maxillary wall and hard palate. Incision was closed in two layers, Vicryl was used

for inner layer and Prolene for outer. Defect was filled with Bismuth Iodoform Paraffin Paste (BIPP) and booster was placed. Margins were sent for histopathology. BIPP and booster were removed on tenth post-operative day. Prosthesis was placed to fill the defect after clearance of margins on histopathology report. Oral trial was given after the fixation of obturator which was well tolerated by patient. Nasograstic tube was removed and patient was allowed to take orally.

Discussion

Ossifying fibroma was subtyped into the Juvenile variant in 1927 by Montgomery.[6] It is characterized by its earlier age of onset, recurrence potential and aggressive nature.[1,7] The mean age of onset has been reported as 11.8 years and 79% of cases occur before age 15.[7]

JOF has further been subdivided into Psammomatoid Juvenile Ossifying Fibroma (pJOF) and Trabecular Juvenile Ossifying Fibroma. (tJOF) on the basis of histopathology.[8] Samir et al reported their relative ratio of occurrence at 4:1.[9] Clinical course and outcome is unaffected by the distinction. In contrast to the COF, Juvenile variant carries no gender predilection although a male predominance has been noted in case of the Psamommatoid type.[10]

These lesions demand clinical interest due to their aggressive growth occurring with notable rapidity. [8-10] They are locally destructive invading into adjacent anatomical structures. Most of these involve facial bones.[6,7] Literature reports conflicting views as to which bone is most commonly affected with uncertainly lying between Maxilla and the Mandible.[6,7,10] Where mandible is affected, congenitally missing teeth due to defective socket development is observed.[7] Most cases involve sino-nasal areas and jaws. (90%) [8] In contrast to JOF, OF mostly originates from jawbones and the skull.Our patient supported the former view with her Maxilla and Maxillary sinus being affected moreover she presented with a short history of only seven months highlighting the rapid progression.

Initially asymptomatic.[6] The spectrum of presentation varies highly depending upon site of origin and adjacent involvement. Patients can present later on from facial asymmetry, sinus and nasal complaints to orbital and dental complaints in the form of proptosis and root involvement.[8] Facial disfigurement in contrast to the conventional form is a pertinent feature. Our patient concurred these findings as she presented with a painless swelling that involved her dental roots causing displacement and loss. Infection and ulceration can complicate the lesion and result in pain.[7]

Radiographically, these lesions present as unilocular or multilocular, osteolytic lesions having a variable radiolucency and can also exhibit cystic change.[10] These are regular and well demarcated. [8] The radiolucency depends on amount of calcification.[8] On CT these appear as hypodense and expansive bearing a shell of bone as circumscription. However CT and MRI are not diagnostic nor specific as many other fibro-osseous lesions present similarly.

Histological examination remains the most reliable method of diagnosis. Runwandi et al in a case series observed that all lesions were un-encapsulated which is in contrast to conventional ossifying fibromas.[10] Psamommatoid type bears Psammoma bodies as the pathognomic feature while trabecular type exhibits immature woven bone osteoid.[10] overall JOF has a heterogeneous morphology. Fibroelastic stroma with varying cellularity, myxomatous regions, internal calcification, giant cells, ossicles and trabeculae are often present.[8,] The histopatholgy variant has no effect on clinical course.[8]

The definitive etiology remains unknown. Genetic, traumatic and developmental origins have been suggested.[6] Differentiation of the multipotential precursor cells of the periodontal ligament to form aberrant tissue is most noteworthy theory.[3,8]

JOF due to its resemblance to other fibro-osseous lesions poses a diagnostic challenge and other differentials must be ruled out. The main cases being extra-cranial-meningioma, osteosarcoma, COF, and focal cemento-osseous dysplasia.[10] Clinical correlation in conjunct with the diagnostic modalities enumerated are vital to accurate diagnosis. This point is further emphasized given the aggressive nature and rapid growth of the lesion and the effect on outcome due to timely and appropriate intervention.

The treatment of choice is determined by behavior and size of the lesion.[6] For smaller lesions, curettage and local excision can be employed and in the case of para-nasal lesions, endoscopic route can also be taken. In bigger and aggressive lesions causing root and cortical involvement block resection and radical measures remain most efficient.[1] Our patient was treated with a partial Maxillectomy. Wide resection with clear margins on frozen sections is recommended.[8]

JOF has a reported recurrence rate of 30-58% following 6 months to 19 years.[6,8] This makes follow-up instrumental to complete cure. This also has to be kept in mind while

operating as immediate reconstruction is not advised. This tendency is age related with higher occurrence in the younger age group.[7] There is no standardized follow-up protocol in literature, however long-term follow up of at least 5 years is recommended.[8]

Despite the aggressive nature and recurrence potential, the over-all prognosis of JOF is good and no cases of malignant transformation have yet been reported [7] Radiotherapy however is reported to cause malignant transformation and is strictly contraindicated.

Conclusion

In younger age group, cure is incomplete and indefinite without close and careful follow-up. JOF is a potentially debilitating condition but which is amenable to therapeutic interventions given a timely diagnosis.

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