Dentistry
ISSN: 2161-1122
OPEN ACCESS Freely available online
Research Article

Juvenile Ossifying Fibroma of the Maxilla: A Case Report of a 15 year Old Girl

Syeda Rawnak Jahan[1]*, M. A. Awal[2]

[1]Family and Community Empowerment Support (FACES), Dhaka, Bangladesh; [2]Department of Oral and Maxillofacial Surgery, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh

ABSTRACT

Juvenile ossifying fibroma is a benign, but locally aggressive fibro-osseous tumor of craniofacial bones with high recurrence rate. It usually occurs in young children and arises in maxillofacial region. This pathology would be diagnosed and treated as early as possible because of its rapidly progressive and osteolytic nature which may complicate the surgical removal if left untreated over a long period. The present case is diagnosed as Juvenile ossifying fibroma involving the maxilla causing facial deformity, proptosis and nasal obstruction in a 15 year-old female child.

Keywords: Juvenile ossifying fibroma; Fibro-osseous lesion; Maxillary sinus; Paranasal sinus

INTRODUCTION

Juvenile Ossifying Fibromas (JOF) is classified as a group of progressive, benign fibro-osseous tumors of the craniofacial bones. It exclusively found in young children [1]. According to the new classification of World Health Organization (WHO), ossifying fibromas that appear as fast-growing mass between 5 years and 15 years of age, are radio logically well-bordered and consistent with ossifying fibroma histologically are referred as JOF [2]. JOF clinically may present or extend into the orbital bones, paranasal sinuses and mandible and may cause gross facial deformity. 85% of JOF lesions originate in the facial bones (Maxilla being more common than mandible), among which 90% arise within paranasal sinuses (ethm oid>frontal>maxillary>sphenoid sinus). 12% occur in the calvarial bones, and 4% are extra-cranial. Based on the bony involvement, symptoms can vary from proptosis, exophthalmos, nasal symptoms and sinusitis [3].

The present study describes a case of JOF of left maxilla involving left nasal cavity and protruding left orbit towards upward and laterally, mimicking malignancy in a 15 years old female child. It is a rare clinical entity, which is often misdiagnosed and mismanaged due to its benign but aggressive nature. JOF is treated by surgical excision and may recur if local resection is not complete. Long-term follow-up is necessarily owing to its locally aggressive nature and high recurrent potentials [4]. An early and correct diagnosis, supported by histological and radiological findings, can help to avoid large bone resection in young patients, which may decrease complication and comorbidity. The purpose of this case presentation is to emphasize correct and quick diagnosis to avoid rapid growth of this type of aggressive tumor.

CASE STUDY

A 15-year-old girl was presented with the complaint of a gradually progressive swelling left side of her face with upward and outward bulging of the left eye for last one and half year. Initially the patient noticed a small hard swelling between her left eye and nose, which was gradually increasing in size for first one year. However, in the last 6 months, the lesion enlarged rapidly and occupied almost upper left side of her face (Figure 1).

Figure 1: Frontal, right and left lateral view of the patient's face.

***Correspondence to:** Dr Syeda Rawnak Jahan, Family and Community Empowerment Support (FACES), Dhaka, Bangladesh, Tel: 8801716602741; E-mail: dr_srjahan@yahoo.com

Received: October 29, 2021, **Accepted:** November 12, 2021, **Published:** November 19, 2021

Citation: Jahan SR, Awal MA (2021) Juvenile Ossifying Fibroma of the Maxilla: A Case Report of a 15 Year Old Girl. Dentistry. 11:607

The large swelling pushed her left eye upward and laterally. Vision was intact in the left eye. Clinical examination found no signs of oculomotor paralysis and no loss of visual acuity or visual field abnormalities. There was history of occasional episodic epistaxis from left nostril, which was oozing type, red in colour, scanty in amount. Bleeding continued for 4 to 5 days and stopped spontaneously. There was a pus discharging sinus at the middle of the left check 6 months ago. From which discharging of pus occurred for 3 months, which cured and healed with medication. A round scar mark was present below the left zygomatic buttress, measuring 2 cm in diameter. Her nose was also deviated to right side and the left nostril was almost obliterated with polyp like soft tissue mass (Figure 2).

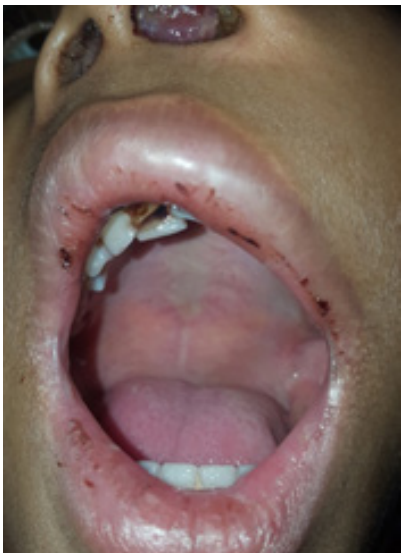

Figure 2: The lesion protruded through the nasal cavity and hard palate.

Patient had no history of pain or numbness in left side of her face. The color and the temperature of the overlying skin were normal and the overlying skin is free. Intra-orally, hard palate is swollen and pushed towards oral cavity, which is hard and non-tender. Upper Left alveolar process was expanded bilaterally. Teeth were not mobile or displaced. Mandibular movement is normal with adequate mouth opening. No cervical lymphadenopathy was present. Plain and contrast CT scan showed- heterogeneously contrast enhanced large mixed density mass lesion in the left maxillary sinus. The mass was involved and extended in nasal cavity, anterior and posterior ethmoidal sinus, sphenoidal sinus and frontal sinus with remodeling of nasal turbinates, nasal bone, bony walls of maxillary sinus, left orbital walls. In some extent the lesion involves anterior cranial cavity. The mass was deviated the left globe antero-leterally. The right maxillary sinus is also deviated to the right. Walls of right maxillary sinus are markedly thickened and osteo-meatal complex is completely blocked (Figures 3 and 4).

An incisional biopsy was performed intraorally. Histopathologic assessment showed a proliferation of fusiform cells with regular nuclei arranged in bundles and storiform pattern associated with multinucleated giant cells. Within these irregular strands, mineralized pieces in the form of trabeculae were found. The stratified squamous epithelium surface showed moderate acanthosis and hyperkeratosis. A diagnosis was confirmed through history, clinical examination, radiological and histopathological investigation as a definite case of JOF. Treatment plan was made by surgical excision of the lesion. But, after several attempts of counseling patient and her guardian were not agree to go for surgery.

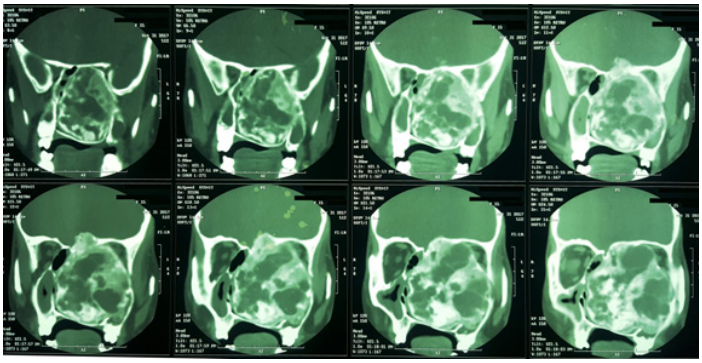

Figure 3: CT scan of coronal view showing the mass has involved and extended in maxillary sinuses, sphenoidal sinus, and frontal sinus and in some extent the lesion involves cranial cavity.

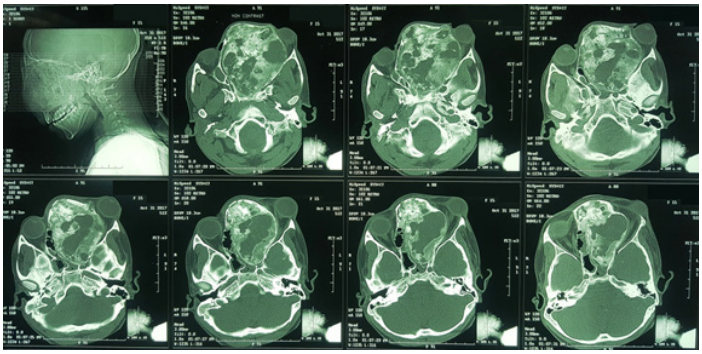

Figure 4: CT scan of axial view showing the mass has involved and extended in maxillary sinuses, anterior and posterior ethmoidal sinus, sphenoidal sinus.

DISCUSSION

Amongst the odontogenic fibro-osseous lesions, Juvenile Ossifying Fibroma (JOF) is uncommon, yet important entity. JOF is a benign neoplasm of bone with a predisposition of early age of onset, locally aggressive behaviour, typical bone pattern and high tendency to recur [5]. JOF is thought to arise from mesenchymal cell differentiation of the periodontal ligament-which is a precursor to cementum, fibrous tissue and osteoid [6]. Other names of JOF include juvenile active ossifying fibroma, aggressive ossifying fibroma or young ossifying fibroma. Most common age group for its occurrence ranges from 8 to 12 years [7]. Clinically JOF demonstrates rapid growth, causing local bone destruction and facial asymmetry. Paranasal sinus or orbital bone involvement is very common. It may also cause nasal obstruction, proptosis, exophthalmia and visual disturbance. JOF tend to be massive, but painless expanding swelling if left untreated. The recurrence rate for JOF is reported to range from 30% to 56%, with few cases described to recur for more than once [8].

Radio graphically, JOF may show multilocular expansile lesion with scalloped borders and destruction and displacement of adjacent structures. Depending upon the extent of the presence of mineralized material, the lesion may appear completely radiolucent to mix radiolucent-radiopaque in nature [9]. JOF must be distinguished from other ossifying fibroma lesions that are characterized by the replacement of normal bone tissue with more or less cellular fibrous tissue including ossified structures. In fibrous dysplasia, which develops slowly, can cause bone pain

or deformities. Radio logically; fibrous dysplasia has a somewhat homogenous, frosted glass appearance and continuity with the normal bone unlike JOF. A sarcomatous transformation is reported in 0.5% of cases [10]. In cemento-osseous dysplasia, a reactional or dysplastic lesion surrounding the alveolodental ligament, may occur close to dental apices. It is asymptomatic and generally does not exceed 2 cm. These lesions must not be biopsied because of the risk of a secondary healing disorder [11]. And in osteosarcoma, especially in its trabecular form that can mimic JOF. Clinically, the symptomatology is generally painful, unlike JOF [12].

The present study describes a case of JOF of left maxilla involving left nasal cavity and protruding left orbit towards upward and laterally. Despite such large lesion patient had no history of pain or numbness in left side of her face. Radiology showed a large expansile lesion with heterogenously contrast enhanced mixed density mass in the left maxillary sinus. The mass was involved and extended into anterior and posterior ethmoidal sinuses, sphenoidal sinus and frontal sinus with remodeling of nasal turbinates, nasal bone, bony walls of maxillary sinus, left orbital walls. In some extent the lesion involves middle cranial cavity. The right maxillary sinus is also deviated to the right. Microscopic examination of JOF may show proliferation of fusiform cells with regular nuclei arranged in bundles and storiform pattern associated with multinucleated giant cells. Within these irregular strands, mineralized pieces in the form of trabeculae may found [13]. It has two histologic subtypes- Juvenile Psammomatoid Ossifying Fibroma (JPOF) and Juvenile Trabecular Ossifying Fibroma (JTOF). JTOF mainly occur in the maxilla, mandible, and fronto-ethmoid complex, and JPOF occurred in the paranasal sinus, calvarium, maxilla and mandible [14]. Juvenile trabecular ossifying fibroma showed a mixture of cellular osteoids without osteoblastic rimming, and there were trabeculae of immature bone with osteoblastic rimming. Ossifying fibroma also showed irregular trabeculae, similar to those of JTOF, but the trabeculae in JTOF were longer and slenderer, with an anastomosing pattern resembling paintbrush strokes [15]. In this case diagnosis was confirmed after assessing radiological and histopathological features and comparing with other fibro-ossious type of lesions.

CONCLUSION

JOF is a benign but aggressive neoplasm occurred most commonly in children. A careful evaluation of the clinical, radiological, and histological components of this lesion is needed to establish the confirm diagnosis. Early diagnosis and complete surgical excision followed by long-term follow-up of the patient is very important. There are diagnostic challenges as its clinical features overlap with many commonly occurring fibro-ossious type of lesions. Histologic criteria for accurate classification is often controversial, Thus for accurate and reliable diagnosis of JOF requires a multidisciplinary approach with close attention to pertinent clinical history, radiographic impression, and correlation with histo-morphologic findings. The management of these lesions is directed at primarily surgical intervention and must consist of a complete surgical resection.

REFERENCES

1. Rallis G, Schoinohoriti O, Krasadakis C, Eftychiadis C, Nikitakis N, Constantinos Mourouzis. Trabecular Juvenile ossifying fibroma: Updated review of the literature and report of an interesting case. J Otolaryng Head Neck Surg. 2017;3(1): 1-5.
2. Barnes L, Eveson JW, Reichart P, Sidransky D. Pathology & Genetics Head and Neck Tumours. WHO, Press 2005: p- 430.
3. Vamanshankar H, Karunakaran V, Parida PK, Hegde JS, Ganesan S, Alexander A. Giant trabecular type juvenile ossifying fibroma of the maxilla: A case report. Rev Esp Cir. Oral Maxilofac. 2017;39(4): 234-238.
4. Kundu PR, Beniwal K, Singh B, Kaur S. Juvenile ossifying fibroma: An unusual case report and review of the literature. Saudi Surg J. 2014;2(2): 57-59.
5. Vamanshankar H, Karunakaran V, Parida PK, Hegde JS, Ganesan S, Alexander A. Giant trabecular type juvenile ossifying fibroma of the maxilla- A case report. Revista Española de Cirugía Oral y Maxilofacial 2017; 39(4), 234-238.
6. Cawson RA, Odell EW. Cawson's essentials of oral pathology and oral medicine. (8th edition), Churchill Livingstone Elsevier, Press 2008;p: 146-83.
7. Sun G, Chen X, Tang E, Li Z, Li J. Juvenile ossifying fibroma of the maxilla. Int J Oral Maxillofac Surg. 2007;36: 82-85.
8. Sarode SC, Sarode GS, Waknis P, Patil A, Jashika M. Juvenile psammomatoid ossifying fibroma: A review. Oral Oncol 2011;47(12): 1110-1116.
9. Tamgadge S, Avinasha T, Bhalerao S, Rajhans S. Juvenile psammomatoid ossifying fibroma with aneurysmal bone cyst in the posterior mandible. Ecancermedicalscience. 2014;8(10): 471.
10. Lemoine S, Cassagnau E, Bertin H, Poisson M, Corre P, Guiol J. Juvenile ossifying fibroma: Case report and literature review. J Oral Med Oral Surg 2018;24(2): 67-71.
11. Khonsari RH, Corre P, Bouguila J, Gayet-Delacroix M, Piot B. Multiple target-shaped periapical lesions. Rev Stomatol Chir Maxillofac. 2008;109(6): 403-404.
12. Odin G, Benchetrit M, Raybaud H, Balaguer T, Soler C, Michiels JF. A maxillary tumor not to be ignored; the juvenile ossifying fibroma. Ann Pathol. 2012;32: 65-67.
13. Aboujaoude S, Aoun G. Juvenile trabecular ossifying fibroma of the maxilla: A case report. Med Arch. 2016;70(6): 470-472.
14. Osunde OD, Lyogun CA, Adebola RA. Juvenile aggressive ossifying fibroma of the maxilla: A case report and review of literature. Ann Med Health Sci Res. 2013;3(2): 288-290.
15. Sohyung P, Bong JL, Jeong HL, Kyung-JC. Juvenile ossifying fibroma: A clinico-pathologic study of 8 cases and comparison with craniofacial fibro-osseous lesions. Korean J Pathol. 2007;41(6): 373-379.