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Case Report

A successful surgical management of pediatric ossifying fibroma in peripheral limited setting: a rare case report

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ABSTRACT

Ossifying fibroma (OF) is a rare non-malignant fibro-osseous lesion of the craniofacial area with the characteristic of slow-growing and locally aggressive. The incidence rate in paediatric patients is still not clear. The management is varied from observation and serial radiologic imaging to radical resection of the tumour. To provide knowledge and information about diagnosing and managing a rare case of craniofacial ossifying fibroma. A-5-year-old boy diagnosed with orbito-inferonasal region ossifying fibroma underwent extirpation of the mass with the lateral rhinotomy- Webber Ferguson approach, anterior orbitotomy by the ophthalmologist, and obturator placement by the prosthodontist. OF remains a rare disease, particularly in paediatric patients, which is locally aggressive although it's benign characteristic. Definitive excision is the treatment of choice for OF. Successful surgical outcome was due to a multidisciplinary team and surgical incision technique. We found that lateral rhinotomy Weber Ferguson incision showed excellent exposure. Vision and facial nerve function were preserved as well as good facial symmetry and aesthetics and recurrence prevention. Regular follow-up is necessary for young OF patients.

Keywords: Ossifying fibroma, Craniofacial, Surgery

INTRODUCTION

Ossifying fibroma (OF) is a rare non-malignant fibro-osseous lesion of the craniofacial area with slow-growing and locally aggressive characteristics. OF is made of fibrous cellular stroma with foci of mineralization or ossification, which replaces the normal bone. It is more common in the mandible followed by the maxilla and rarely in the paranasal sinuses, orbits and skull bones. The aetiology of OF is still unclear. However, it is known that developmental abnormalities of odontogenic or traumatic origin are predicted to be the underlying causes.[1] OF is classified based on histopathologic examination, including juvenile psammomatous, juvenile trabecular and cemento-ossifying fibromas.[2]

Patients with ossifying fibroma are often asymptomatic. Most cases are found incidentally when the patient is subjected to imaging examination for other indications. When a patient complains of mass effect manifestation, swelling of the face is the most common sign found.[1] Symptoms are often due to local tumour expansion. Visual complaints include proptosis, eyeball displacement, lagophthalmos, restricted eye movement, and decreased visual acuity. Lesion invasion to the sinus shows symptoms of recurrent sinusitis or nasal obstruction.[3]

Imaging examination is required to discover tumour location, extension, and involvement of nearby structures. Computed tomography (CT) scan is the preferred imaging, with OF manifesting as radiopaque uninoculated or multiloculated lesion with well-defined corticated borders,

without soft tissue involvement. We can find isointense calcification rim on T1-weighted images and hypointense on T2- weighted images on MRI examination. MRI with contrast will show bony shell enhancement, suggestive of tumour tissue rather than reactive hyperostosis.[3]

A multidisciplinary team is needed for managing the patient. Conservative management or watchful waiting is less favourable in young adults because the probability of a slow-growing tumour risks compressive symptoms.[2] Total excision of the lesion tends to be the definitive management of craniofacial ossifying fibroma. The surgical approach to tumour resection is determined by anatomical location and tumour size. Radiation therapy has not been recommended in the direction of ossifying fibroma.[1]

CASE REPORT

This was a case report of an unusual case of ossifying fibroma that presented to the outpatient clinic with advanced stage and early aged. The complexity of the medical, family history and clinical symptoms makes the therapy difficult, associating with multidisciplinary team and treatment. Here we study the clinical presentations, risk factor, and the diagnostic and multidisciplinary team challenges in the treatment of ossifying fibroma.

Case presentation

A 5-year-old boy was referred to our hospital in June 2021 with a lesion in the right eye, which caused incomplete right eye closure. The lesion was progressively enlarging in the past year.

He also complained of a lesion on the right nose with nasal obstruction and a non-odorous clear secret. He had no complaint of vision changes and hearing impairment. History of nosebleed and decrease of appetite was denied. History of malignancy in the family was also denied. Biopsy was done in Sardjito General Hospital in October 2020. We found a fixed mass with hard consistency on the right nose anterior rhinoscopy from palpation

Computed tomography (CT) result showed a 5.1×5.5×5.0 cm iso-hypodensity lesion in right ethmoidal bone with regular, well-defined margin, ground-glass opacity expanding to left ethmoidal sinus, right maxillary sinus, bilateral sphenoidal sinus, right extraconal orbital, which caused proptosis of the right eye.

Histopathological examination revealed an immature bone fragment (woven bone) without osteoblastic rimming with fibroblast proliferation. The tumour cell was relatively monomorphic with a round, oval, spindle fine chromatin nucleus and no sign of malignancy. EMA, B catenin, and S100 were negative, Vimentin and Ki67 was positive.

Based on the light of the current clinical, radiological, and histopathological findings, the patient was diagnosed with orbito-inferonasal region tumour (ossifying fibroma). The patient management plan involved an extirpation of the mass with the lateral rhinotomy - Webber Ferguson approach, anterior orbitotomy by the ophthalmologist, and obturator placement by the prosthodontist.

The procedure was performed with lidocaine infiltration at the incision site. Lateral rhinotomy Weber Ferguson incision was done layer by layer until the anterior maxilla bone was completely visualized. The ophthalmologist did anterior orbitotomy to expand the operation area up to orbit. Anterior maxilla bone was drilled starting from the lateral border of piriformis aperture up to the lateral border of anterior maxilla sinus, inferior border of piriformis aperture as triangle form, until anterior maxilla bone wall can be removed. Palpation and complete extirpation of the mass were done. Bleeding control was done using tampons and cauterization. Hernial mesh was placed behind the right oculi for eye support to prevent enophthalmos and radiologic film to separate orbit space from nasal and maxilla spaces to ensure that the right and left eye positions are symmetrical. Then sutured was done layer by layer neatly. Histopathology examination was performed to determine the definitive diagnosis, and it was confirmed as an ossifying fibroma. Post-operative complications such as epistaxis, blocked nose, headache or aesthetic problems of the nose were not found.

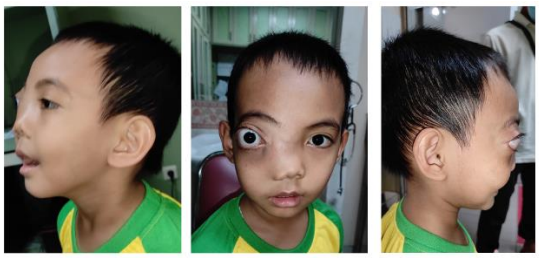

Figure 1: Clinical presentation of the patient.

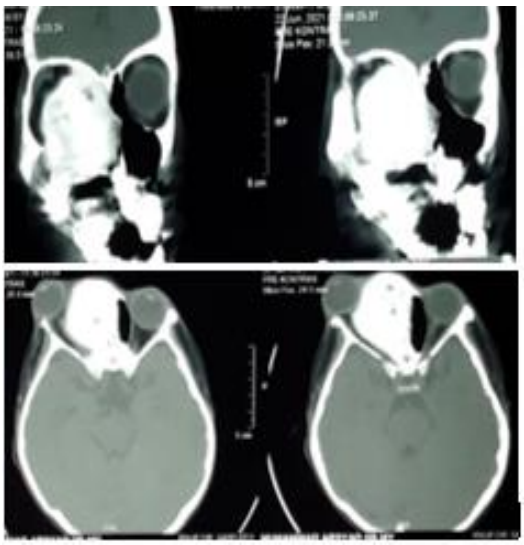

Figure 2: CT scan showed a 5.1×5.5×5.0 cm iso-hypodensity lesion.

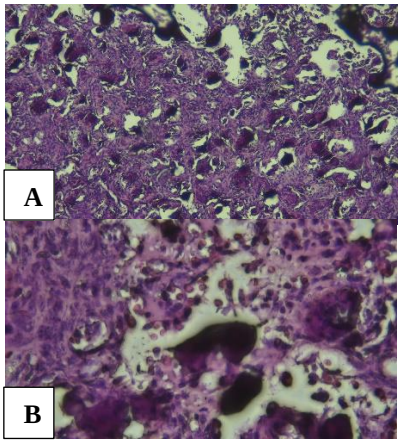

Figure 3: Histopathological examination result at (a) 100X magnification; and (b) 400X magnification.

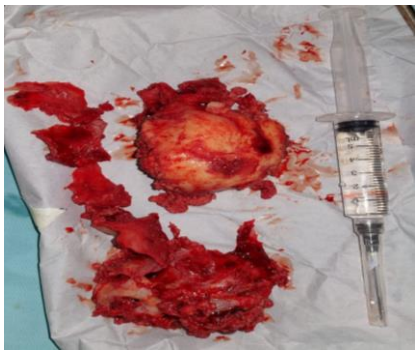

Figure 4: Complete surgical excision of the specimen.

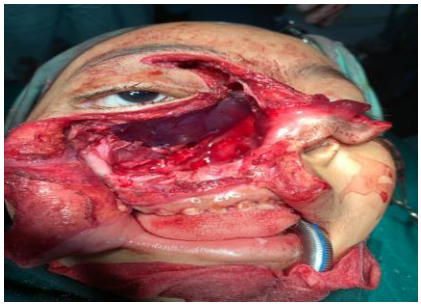

Figure 5: Hernial mesh and radiologic film were placed behind the right oculi space.

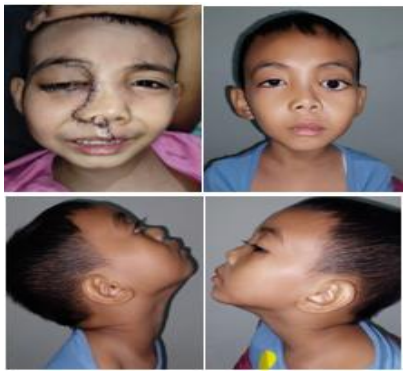

Figure 6: Post-operative clinical photograph.

DISCUSSION

OF is a rare non-malignant fibro-osseous lesion of the craniofacial area with a characteristic of slow-growing and locally aggressive.[1]

WHO (2017) classifies OF as odontogenic origin (cemento OF) and juvenile ossifying fibroma (JOF), which is further divided into Trabecular Juvenile Ossifying fibroma (TrJOF) and Psammomatoid Juvenile Ossifying fibroma (PsJOF). Based on several studies, the most common prevalence is cemento OF (71.4%), followed by JOF (25.4%). For the JOF subtype, the prevalence of TrJOF (10.3%) was higher than that of PsJOF (8.9%).[6]

Generally, OF affects patients in the second to third decades of age. The incidence rate in paediatric patients is still unclear; however, about 10% of cases are reported in patients under 18 years of age.[7] Several studies reported OF is more common in women than men, 55.4% vs 44.6%. The dominant age group is under 18 years (51.6%), and over 18 years is 40.8%.[6]

OF all craniofacial bones, OF is more common in the mandible, followed by the maxilla. It has been reported to rarely occur in the ethmoid sinus, orbit, and skull base.[1] Yet, when it affects paranasal sinus, OF is most common in the ethmoid sinus and nasal cavity, followed by sphenoid, maxillary, and frontal sinuses, as we found in the patient.[7]

OF tends to be asymptomatic. Symptoms are often due to local tumour expansion to surrounding tissues or organs because of the tumour locally aggressive behaviour, such as facial asymmetry, pain, and local destruction.[5,8] Visual complaints include proptosis, eyeball displacement, lagophthalmos, restricted eye movement, and decreased visual acuity.

Lesion invasion to the sinus shows symptoms of recurrent sinusitis or nasal obstruction.[3] In this case, the patient complained of a lesion in the right eye, which caused uncomplete right eye closure. The lesion was progressively enlarging in the past year. He also complained of a lesion on the right nose with nasal obstruction and a non-odorous clear secret. He had no vision changes, hearing impairment, or history of nosebleeds.

CT scan is the preferred imaging, presented as a unilocular or multilocular radiolucency, radiolucent-radiopaque, or radiopaque lesion.[9] The pathognomonic sign of OF is a well-defined margin (62.9%), followed by an ill-defined margin (12.2%) and a moderate-defined margin (0.9%).[6]

CT scan result in this patient showed a 5.1×5.5×5.0 cm iso-hypodensity lesion in the right ethmoidal bone with regular, well-defined margin, ground-glass opacity, which expanding to left ethmoidal sinus, right maxillary sinus,

bilateral sphenoidal sinus, right extraconal orbital, which causes proptosis of the right eye.

Based on OF aggressiveness, the management is varied from observation and serial radiologic imaging, which is preferable for asymptomatic patients, to radical resection of the tumour.[10]

Some studies reported that enucleation or curettage is the initial treatment of choice for OF. However, many studies showed that radical surgery, including local, marginal, and total resection, is superior to conservative surgery.[8] It is supported by data that the recurrence rate after surgery was 14.4%, with most recurrence occurring in conservative surgery. The recurrence rate after conservative surgery (19.7%) was higher compared to radical surgery (10.6%).[6] It is hypothesized that the infiltrative nature of the tumour leads to incomplete excision and cause a recurrence.[4] Therefore, total excision of the lesion is considered the definitive management of craniofacial ossifying fibroma.[4,10]

In our case, the patient management involved an extirpation of the mass with the lateral rhinotomy - Webber Ferguson approach, anterior orbitotomy by the ophthalmologist, and obturator placement by the prosthodontist. Hernial mesh was placed behind the right oculi for eye support to prevent enophthalmos and radiologic film to separate orbit space from nasal and maxilla spaces to ensure that the right and left eyes' positions are symmetrical. Postoperative complications such as epistaxis, blocked nose, headache or aesthetic problems of the nose were not found.

The surgical approach to tumour resection is determined by anatomical location and tumour size.[10] Surgical management with lateral rhinotomy, bilateral craniotomy, trans-facial, craniofacial or trans-maxillary approaches has been applied to paranasal sinuses and skull base OF resection. However, this approach has drawbacks, namely the morbidity in the form of facial scars.[10] Management of blood loss during surgery is very important in paediatric patients because massive bleeding can cause several perioperative complications. Preoperative embolization to reduce bleeding is an effective way for haemorrhagic neoplasms, including hemangiomas and angiofibroma.[7]

We conclude that multidisciplinary teamwork is crucial for the resection of these difficult tumours. Complete surgical excision is beneficial, accompanied by long-term follow-up to monitor function and craniofacial skeleton development.[5]

There is a possibility of tumour growth in young patients during the follow-up period. Growth hormone plays a role in tumour growth. Excess growth hormone becomes a risk factor for the occurrence of compressive optic neuropathy. Therefore, careful and regular follow ups are necessary in young OF patients. The surgeon must be aware of tumour regrowth, complications, or malignancy transformation in teenage patients.[11]

CONCLUSION

OF remains a rare disease, particularly in paediatric patients, which is locally aggressive although its benign characteristic. Definitive excision is the treatment of choice for OF. Successful surgical outcome was due to a multidisciplinary team and surgical incision technique. We found that lateral rhinotomy Weber Ferguson incision showed excellent exposure. Vision and facial nerve function were preserved as well as good facial symmetry and aesthetics and recurrence prevention. Regular follow-up is necessary for young OF patients.

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