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Juvenile Psammomatoid Ossifying Fibroma of the Jaw: A Case Report

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Abstract: ***Introduction: Juvenile ossifying fibroma (JOF) is a rare benign fibro-osseous lesion in younger children. It has behaviour locally aggressive growth that differentiates from conventional ossifying fibroma (OF).Based on histopathology, JOF is divided into: juvenile psammomatoid ossifying fibroma (JPOF) and juvenile trabecular ossifying fibroma (JTOF). Case Report: A 6-years old girl patient complained of painless progressive swelling at the maxilla and mandible region since she was 4-years old. Excisional biopsy has been performed six times due to the recurrence since 5 years old. The patient is scheduled to periodically followed up to the age of 21 years until definitive therapy can be carried out. Discussion: Diagnosis of JPOF was made based on clinical feature, radiological characteristics, and pathological features. The site of the lesions, involvement of the surrounding vital tissue, patient age, growth rate and parental choice are the factors that must be considered by a surgeon when choosing the right therapy. Conclusion: JPOF lesions presented rapid growth and high rates of recurrence. Surgeon must educate families of JOF patient about tumor characteristics including relapse status, growth rate, treatment plan and the need for periodic follow up to improve the quality of life for JOF patients.***

Keywords:aggressive growth, juvenile ossifying fibroma**,** recurrence rate

1. Introduction

Ossifying fibroma (OF) is a benign fibro-osseous neoplasm of the jaw and craniofacial complex, which is still controversy in establishing the right diagnosis, classification and treatment.[1] Based on histopathology, ossifying fibroma is characterized by clear boundaries and cell-rich fibrosis, and contains varying amounts of calcified tissue resembling bone, cementum, or both. [2]This tumour can be divided into cemento-ossifying fibroma, and two distinct juvenile ossifying fibroma (JOF), called juvenile trabecular ossifying fibroma (JTOF) and juvenile psammomatoid ossifying fibroma (JPOF).[3]JOF has an early age of onset, a high recurrence rate, and a destructive effect over the cortical bone that causes severe morphological and functional changes. [4]Conventional OF presents as a slow growing mass that is usually without symptoms and rarely recurs, conversely, 38.5 % of JOF cases are associated with mandibular swelling and pain and short term rapid growth. However, there are no reports of cases of malignant transformation or metastasis. [3]

The management of JOF patients remains controversial. The surgeons should consider to clinical symptoms, radiological features, and pathological characteristics when diagnosing JOF. Therefore, proper diagnosis and appropriate treatment can improve the prognosis and quality of life of JOF patients.

2. Case Report

A 6–years old girl patient was referred to Oral and Maxillofacial Surgery Department of HasanSadikin Hospital Bandung with painless progressive swelling at the maxilla and mandible region since she was 4-years old. Initially there was a small swelling in the maxillary anterior which got bigger in about 8 months and began to spread to the maxillary posterior and mandible region. Extra oral examination, there was symmetrical face (**Figure 1**). Intraoral examination revealed gross expansion at maxilla and mandible region, painless and minimal tenderness on palpation (**Figure 2**).The preliminary case history revealed that the patient had undergone twice surgical excision when she was 5 years old at a private hospital, but is not well documented. On december 2016, when the patient came to our department, the lesion was of massive size at maxillary and mandible region.

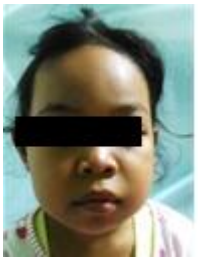

Figure 1: Extra Oral Examination

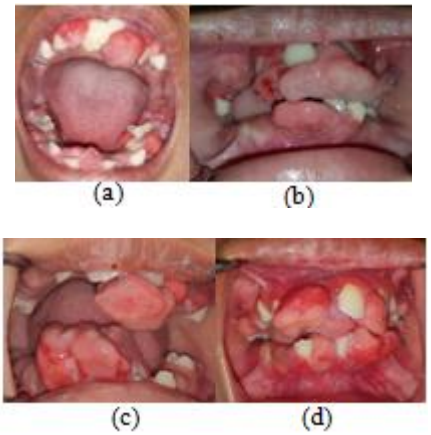

Figure 2: Intra Oral Examination . Gross expansion at maxilla and mandible region (a). December 2016 (b). September 2017 (c). September 2018 (d). July 2019.

Radiological feature showed mixed radiolucency and opacity according to the component and proportion of soft tissue and hard tissue in the internal structure (**Figure 3**).

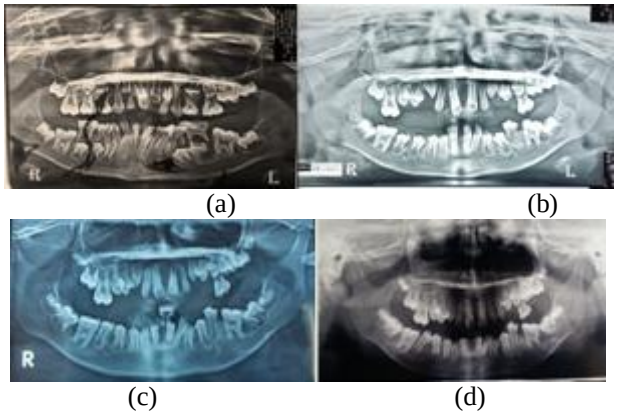

Figure 3: Panoramic Image (a). December 2016 (b). September 2017 (c). September 2018 (d). July 2019.

Due to high rate of recurrence, this patient had been performed four times excisional biopsy in our department since December2016 until July 2019 (**Figure 4**).She is scheduled to periodically followed up to the age of 21 years until definitive therapy can be carried out.

Figure 4: Post Treatment (a). December 2016 (b). September 2017 (c). September 2018 (d). July 2019.

In December 2016, third excisional biopsy and multiple extraction of teeth was performed in our department, obtaining the diagnosis of ossifying fibroma. Histopathological feature showed subepithelialfibrocolagen connective tissue stroma appears with fibrocyte cells that are hyperplastic, the nucleus is within normal limits. Among them visible "woven bone' without osteoblastic rimming and also visible "psammoma bodies". There was also visible proliferation of capillary blood vessels. No sign of malignant tumor cells (**Figure 5a**).

In September 2017, fourth excisional biopsy was performed in our department, obtaining the diagnosis of peripheral ossifying fibroma. Histopathological feature showed subepithelialfibrocolagen connective tissue stroma appears with fibrocyte cells that are hyperplastic, the nucleus is within normal limits. Among them visible "woven bone' with osteoblastic rimming and also visible "psammoma bodies". On the other side, there is a stroma of fibrocolagen connective tissue mixed with massive inflammation of lymphocyte cells, plasma cells, histiocytes, PMN cells and multinucleated giant cells accompanied by dilatation of blood vessels. There is no sign of malignant tumor cells (**Figure 5b**).

In September 2018, fifth excisional biopsy was performed in our department, obtaining the diagnosis of juvenile psammomatoid ossifying fibroma. Histopathological feature showed subepithelialfibrocolagen connective tissue stroma appears with fibrocyte cells that are hyperplastic, hypersellular, compact, rounded nucleus, oval, fine chromatin, and found mitosis. Between them appear psammoma like bodies and osteoclast cells. It appears that the osteoid matrix partially obtained osteoblastic rimming. No sign of malignancy (**Figure 5c**).

In July 2019, sixth excisional biopsy was performed in our department, obtaining the diagnosis of juvenile psammomatoid ossifying fibroma .Histopathological feature showed subepithelialfibrocolagen connective tissue appears with lymphocytes inflammation cells, plasma cells, histiocytes, PMN cells accompanied by dilatation and blood vessel dams. Among them appear tumour masses consisting of fibrocyte cells that grow hyperplastic, compact, rounded nucleus, oval, mitosis found. Between them appear psammoma like bodies and osteoclast cells. No sign of malignancy (**Figure 5d**).

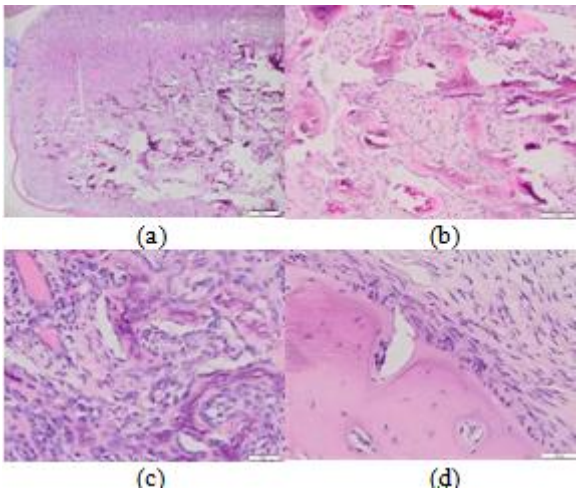

Figure 5: HistopathologicalFeature (a). Stroma appears with fibrocyte cells that are hyperplastic. Visible "woven bone' without osteoblastic rimming and also visible "psammoma bodies". (b). Stroma mixed with massive inflammation of lymphocyte cells, plasma cells, histiocytes, PMN cells and multinucleated giant cells (c). Stroma appears with fibrocyte cells that are hyperplastic, hypersellular, compact, rounded nucleus, oval, fine chromatin, and found mitosis. Appear psammoma like bodies and osteoclast cells and partially obtained osteoblastic rimming (d). Appeartumour masses consisting of fibrocyte cells that grow hyperplastic, compact, rounded nucleus, oval, mitosis found. Appear psammoma like bodies and osteoclast cells.

3. Discussion

A combination of age factor, growth pattern, imaging features and pathological characteristics play an important role in establishing the right diagnosis. The surgeons has noticed a high rate of recurrence in the OF cases of

children's jaw.[5] Therefore, this case must be treated properly for children's mental and psychological health. [6]

JOF is distinguished from other fibro-osseous lesions primarily by its age of onset, clinical symptoms, and potential behavior. [7]In the 2005 WHO definition, the age at onset of JOF is ≤ 15 years.[3]JPOF mainly affect children and young adults (mean age 20 years old) but conventional ossifying fibromas occur between the second and fourth decade (mean age 35 years old). [8]In this case, the first complaint of swelling in the maxilla and mandible occurs when the patient is 4 years old.

Locally aggressive growth is an important clinical charactheristics of JOF. However, there are no reports of cases of malignant transformation or metastasis.When the tumour is located in the upper jaw and becomes large, it can involve the surrounding tissues such as the eyes and nasal cavity, thereby resulting in exophthalmos, nasal congestion and other symptoms.[3] JOF has been reported to occasionally occur in areas of missing teeth. [9]

JOF is hypothesized to originate from the overproduction of the myxo-fibrous cellular stroma which is otherwise involved in the physiological growth of the septae in the paranasal sinuses as they enlarge and pneumatize. The hyaline material produced by the stromal cells ossifies and the cystic changes are initiated by the connective tissue mucin.Clinically the lesions tend to be massive, painless, expanding swelling with an aggressive growth pattern and high recurrence potential (especially cases dealt with inadequate surgical interventions).[10]In this case, intraoral examination revealed gross expansion at maxilla and mandible region, painless and minimal tenderness on palpation. Recurrence has occurred approximately six times during 2015 - 2019 with complaints of progressive swelling in the same region, which was rapidly increasing in size. During the time, the recurrence occured between 6 months until 1 year after treatment by excisional biopsy (**Figure 2**).

In recent study, immunohistochemistry examination showed the simultaneous of MDM2 (E3 ubiquitin-protein ligase gene) and RASAL1 (RAS protein activator like 1 gene) was significantly higher in JOF compared with non-JOF patients. This is not only as a potential molecular diagnostic marker, but may also be indicative of aggressive forms of the disease with a higher risk of recurrence, and therefore may indicate a more radical treatment plans.[3] However, in this case the immunohistochemistry examination was not carried out because the results of the histopathology examination were clear and sufficient to establish the diagnosis.

The imaging characteristics tumour can present as radiolucent, mixed, opaque, and ground grass-like.[3] In this case, panoramic images presented mixed radiopaque and radiolucent (**Figure 3**). The features of radiolucency, mixed radiolucency and opacity and complete radiopacity depending on the degree of mineralisation.[4]Pathologies that need to be distinguished from JOF using imaging studies include fibrous dysplasia, osseous dysplasia, odontoma and ameloblastoma.[3]

Histological appearance comprises of trabeculae of woven bone, without osteoblastic rimming and bands of cellular osteoid, within a cell-rich fibrous tissue.JPOF was characterized by fibroblastic stroma and basophils containing sand- like structure that resembled cementum. There is no sign of malignant tumor cells from the histological result of this case (**Figure5**). Fibrous dysplasia, aneurysmal bone cystosteosarcoma, osteoblastoma and cement-osseous dysplasia are the differential diagnosis of JOF.[9]

Surgery is the therapy of choice JOF cases by enucleation or curettage for smaller lesions and radical excision for larger ones.[9]Conservative therapy with curettage, enucleation and partial resection in young patients aims to limit the occurrence of facial deformity, growth and development dysfunction and preserve a normal appearance, chewing and nerve functions. The site of the lesions, involvement of the surrounding vital tissue, patient age, growth rate and parental choice are the factors that must be considered by a surgeon when choosing the right therapy in JOF patients.[3]

Since JPOF has an aggressive behaviour, incomplete excision has been associated with a high local recurrence rate.[7] Kaban et al. suggested a combination therapy of curettage or enucleation with adjuvant interferon therapy for one year. Long term follow up is necessary as recurrence rate vary from 30% to 58% and usually occurs between 6 months and 7 years.[10]However, the patient should undergo complete resection when there is a change in tumour growth pattern which usually characterized by rapid growth, cortical thinning or perforation, and the displacement of teeth or root resorption.

Based on the clinical feature, radiological feature, recurrence rate, patient age and parental choice, surgical excision is the therapy of choice in this case.Surgeon should aim to maintain or restore the patient's appearance during treatment to reduce the impact on their growth and also mental and psychological health the younger patient. She had been performed four times excisional biopsy in our department (**Figure 4**) and scheduled to periodically followed up to the age of 21 years until definitive therapy can be carried out. Since malignancies have not been reported, these neoplasms can safely be left when they are in stable state. [3]

4. Conclusion

Juvenile ossifying fibroma (JOF) has locally aggressive growth characteristics and a high recurrence rate. Our case presented high rates of recurrence after treatment by excisional biopsy only. Long term follow up and vigilance is required, because it'sreccurence rate is high. Surgeon must educate families of JOF patient about tumour characteristics including relapse status, growth rate, treatment plan and the need for periodic follow up to improve the quality of life for JOF patients.

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