



Case report

Management of juvenile trabecular ossifying fibroma of bone of the maxilla in a child: A case report at a tertiary hospital in Northern Tanzania

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ARTICLE INFO

Keywords:

Juvenile ossifying trabecular fibroma

Diagnosis

Management

Case report

Tanzania

ABSTRACT

Introduction and importance: Juvenile trabecular ossifying fibroma (JTOF) is a rare variant of ossifying fibroma. Though it is benign, it has aggressive clinical behavior. JTOF may pose diagnostic and therapeutic difficulties due to their clinical, radiological and histological variability. Herein, we describe our experience in encountering this unusual disease entity in terms diagnostics as well as surgical procedure and the differential diagnoses to be considered.

Case presentation: An 8-year old female child presented to our facility because of a rapidly growing right maxillary swelling. Clinical examination revealed facial asymmetry resulted from the swelling on the right side of her face extra-orally, especially in her posterior upper jaw which was about 12 cm × 7 cm in dimension. Intra-oral examination revealed solitary mass, well-defined, firm in consistency and non-tender. There were no palpable lymph nodes. Infra-orbital nerves were bilaterally intact. Based on the clinical findings and history, the differential diagnoses of ossifying fibroma, ameloblastoma and fibrous dysplasia were given. Clinico-pathological and radiological correlation confirmed the diagnosis of JTOF. Hemimaxillectomy and reconstruction of maxilla with a rib were performed. Afterwards, the child reported with normal facial appearance, acceptable aesthetics and better chewing function.

Clinical discussion: The clinical presentation of JTOF, and its rapid growth, can cause alarm of other pathologies such as osteosarcoma. The radiological features should reassure the practitioner and a histological examination confirmed the diagnosis.

Conclusion: JTOF is a benign tumor and it should be operated at an early stage because of its rapid growth.

1. Introduction

Fibro-osseous lesions of the jaw represent a rare, benign group of lesions that share similar clinical, radiological, and histopathological features and are characterized by progressive, variable replacement of healthy bone tissue by fibrous connective tissue [1]. Ossifying fibromas (OF) are benign fibro-osseous lesions that are principally encountered within the jawbones and most common neoplastic component of the fibro osseous lesions [2]. OF can be present in two forms: Conventional and Juvenile. The first usually occurs in the mandibles of 30–40-year-old female. On the other hand, the active juvenile form is very rare, may grow rapidly, affects children or young adults, and is more common in

the maxilla and it occurs before age 15 years in 80 % cases [3]. Juvenile Trabecular ossifying fibromas (JTOF) appear as fast growing mass usually between 5 and 15 years of age, radiologically well bordered, and consistent with ossifying fibroma histologically [3]. Herein, we report the case of JTOF of maxilla in a female child who was successfully treated at Kilimanjaro Christian Medical Centre (KCMC); the zonal referral and academic center in Northern Tanzania. This case may provide an insightful guide to consider in terms of diagnosis and treatment when encountering similar cases in future. A brief literature review is also highlighted. This work has been reported in line with the SCARE 2021 criteria [4].

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<https://doi.org/10.1016/j.ijscr.2022.107746>

Received 15 September 2022; Received in revised form 8 October 2022; Accepted 8 October 2022

Available online 13 October 2022

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2. Presentation of the case

An 8-year old female child was brought to dental clinic of the KCMC because of a chief complaint of a swelling in the upper posterior maxilla. The mother of the child was conscious of the swelling since three years. It was slow in onset, enlarging over the years to the present size and was not associated with the history of any toothache or trauma in that region of the jaw. Initially the swelling was asymptomatic but a history of associated pain since one year was noted and had become painful on closure of the jaw because of the contact of the lower teeth with the lesion. The patient had no significant past medical or surgical history and all vital signs were normal.

Clinical examination revealed a giant swelling on the right side of her face extra-orally, with displacement of the nasal base at the right side, protrusion and cranial displacement of the right eye, lateral and ventral displacement of the zygoma leading to gross facial asymmetry (Fig. 1). The patient had a normal mouth opening. Intra-oral examination revealed the swelling was solitary, well-defined, not ulcerated, firm in consistency, non-tender and extending from behind the right tuberoses to the central incisor on the contra-lateral side and superiorly to infra-orbital rims with a slight palatal expansion. Because of huge size lesion, the patient was suffering from paraesthesia of infra-orbital nerve, difficulty in chewing and speaking. Marked alveolar asymmetry was noted. However, the associated teeth were firm, non-mobile and without any associated root resorption. There were no palpable lymph nodes and infra-orbital nerves were bilaterally intact. Radiological examination revealed maxillary bone unilocular radiolucency with well-defined borders and central opacification, (Fig. 2A). Based on the clinical findings and history, the differential diagnoses of ossifying fibroma, ameloblastoma and fibrous dysplasia were given.

Histopathology of biopsy form the lesion revealed the lesion to be composed of connective tissue stroma that was moderate to highly cellular with abundant collagen fibers and fibroblasts. Additionally, the presence of trabeculae of bone with plump fibroblasts and globules of calcification were associated giving an idea of JTOF, (Fig. 2B). Correlation of the clinical presentation, histopathological and radiological findings ruled out the differential diagnosis of fibrous dysplasia and thus, concluded it to be JTOF.

The patient was counseled for surgery, a written informed consent from the legal guardian was obtained. Treatment was scheduled where an en-block maxillectomy was undertaken and could be managed through an intra-oral approach. The dorsal, medial and lateral wall of the maxillary sinus, the pterygoid plates, part of the orbital rim and nearly the complete orbital floor turned out to be destroyed by the tumor (Fig. 2A). To provide support for the soft tissues of nasal base and cheek a free costal graft was inserted between the nasal spine and the lateral

part of the infra-orbital rim (Fig. 3A-D). As the palatal mucosa could be preserved, a primary closure intra-orally could be performed and no obturator was necessary. The lateral displacement of the zygomatic complex was accepted in the expectation that after removal of the pressure of the expanding tumor, in time the zygomatic complex would return automatically to its normal position. Direct post-operative the position of the right eye was normalized; the malposition of the zygomatic complex is not changed as calculated. The support of the soft tissues was adequate, (Fig. 3E). The surgery was performed by a team of experienced experts in the field of oral and maxillo-facial surgery.

One year after surgery, the child reported with normal facial appearance, good aesthetics and better chewing function. The position of the zygomatic complex has normalized as expected. No recurrence has been observed in 3-year following up now with CT-scan (Fig. 3F). Future plan is the replacement of lost teeth with fixed prosthesis.

3. Discussion

JTOF is an uncommon fibro-osseous lesion that occurs in the facial bones. It is also called aggressive ossifying fibroma due to its aggressiveness and the high tendency to recur, unlike other fibro-osseous lesions, such as cemento-ossifying fibroma, which may resemble radiographically [1–3]. As it was the case in our patient, clinically, JTOF presents as a painless, slowly growing mass in the jaw where displacement of teeth may be the only early clinical feature. This lesion is therefore frequently ignored by the patient until the growth produces a noticeable swelling and facial deformity [5,6].

JTOF is typically a lesion of the jaws characterized by the early age of onset under 15 years of age and are more common located more in posterior maxilla. In our case the lesion was located on the posterior maxilla and the age of the patient was 8 years old. Due to its distinct histological features, JTOF has been recognized as a separate histopathological entity among the fibro-osseous group of lesions [3]. Radiologically, the tumor has four radiographic patterns namely cystic radiolucency, ground glass appearance, sclerotic change and mixed type. The radiographic borders appear relatively smooth, well defined, and mostly corticated and the contour is regular [6,7]. As it was the case in our patient, (Fig. 2B); histopathologically, JTOF lesion typically displays cellular stroma consisting of spindled to stellate shaped fibroblasts with bland of steoid without osteoblastic rimming together with immature bony trabeculae surrounded by plump osteoblasts [5,6].

The clinical management of ossifying fibroma is not so clear. Various approaches such as conservative surgery, enucleation, curettage, resection, local surgical excision, radical resection have been reported [2,8]. In our case, conservative surgery was not feasible due to the massive tumor size. Also, to reduce the possibility of recurrence, for such

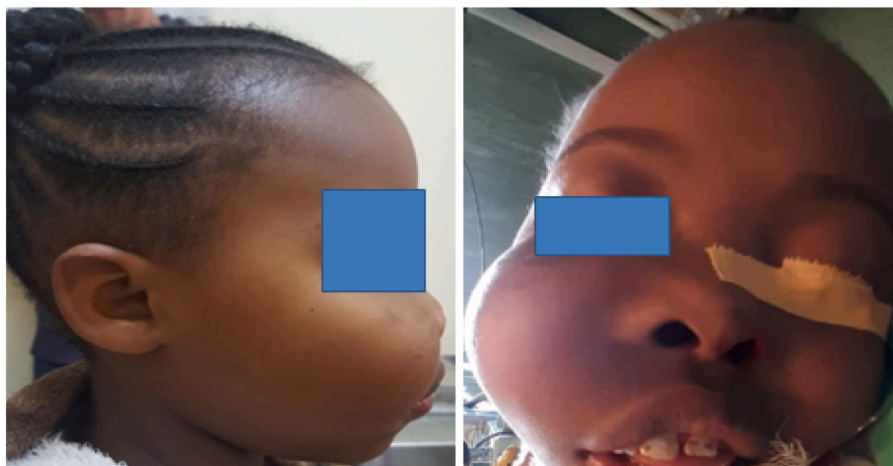


Fig. 1. Photograph of the patient displaying facial asymmetry due to right maxillary tumor.

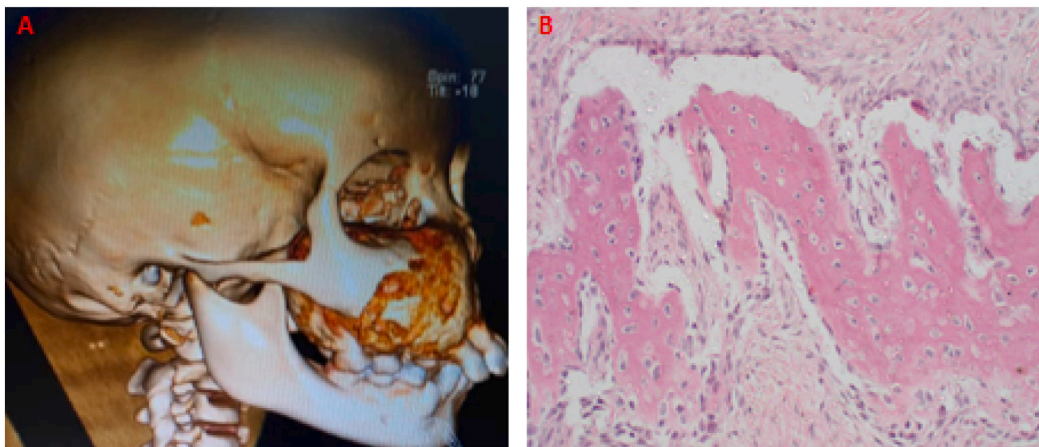


Fig. 2. 3D CT-SCAN of the head highlighting a large mixed density well circumscribed bone lesion with a ground glass appearance involving right maxilla and hard palate with mass effect to the maxillary sinus (A); histopathology of the lesion demonstrating connective tissue stroma that is cellular with abundant collagen fibers and fibroblasts along with trabeculae of bone with plump fibroblasts and globules of calcification were associated; H & E stained sections, 200× original magnification (2B).

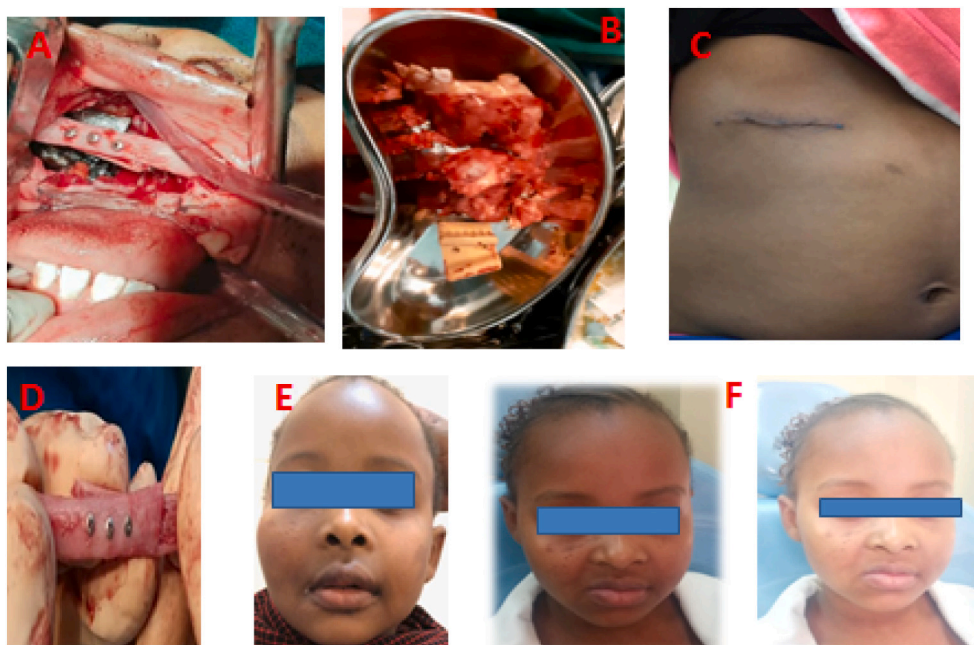


Fig. 3. Hemimaxillectomy surgical procedure (A); resected specimen (B); reconstruction of maxilla with a rib flap (C-D); appearance of the patient after one week (E) and three years (F) of surgery respectively.

large lesion, an enbloc resection of the jaw was ideal with an immediate reconstruction of defect by rib's bone flap. Successful reconstruction of jawbone defects by using autogenously rib flaps following tumor resection in children has been reported [9–12].

When the surgical resection is wide, added restoration using implants and bone grafts can be essential because of functional and esthetic problems, particularly when teeth are removed. In the case explained here, as the lesion ruptured all the cortical bones of maxilla thus radical excision with all involved teeth was performed. Since the region that could be subject to strong muscle area was widespread, it was decided to place a titanium reconstruction plate in order to retain the maxillary outline. After one year of surgery, the defect was reconstructed with the prosthetic obturator to fulfill the functions of chewing, speaking, swallowing and acceptable esthetic (Fig. 3E-F).

Due to high recurrence rate, immediate cosmetic reconstruction should be performed cautiously with close follow up. Secondary

reconstruction can be done sooner for slow-growing disease and delayed for fast-growing lesions. In the present case, cosmetic surgical approach was undertaken immediately considering the slow growth nature of the lesion, the child's age, aesthetics, the functional, growth as well as the fact that the lesions may cease to grow at adolescence [9–13]. In our patient, the tumor measured 12 × 7 cm in diameter but did not lose its encapsulation and was lobulated. This property of tumor made surgical excision easier as a single mass. The post-surgical wound was thoroughly inspected to confirm any involvement towards medial (nasal), roof (floor of the orbit) and posterior wall of the maxillary sinus.

4. Conclusion

JTOF is a rare, benign but locally aggressive lesion with a high potential for recurrence. It may pose diagnostic and therapeutic difficulties because of wide variability clinically, radiologically and pathologically.

Thus, diagnosis should base on combination of clinico-pathological and radiological correlation. Excision must be as complete as possible while ensuring the safety of the neighboring anatomical structures, and treatment should include regular patient follow-ups.

Consent

Written informed consent was obtained from the patient's legal guardian for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of competing interest

All authors have declared that no competing interests exist.

Acknowledgements

The authors would like to thank Petra Alexandra Meerwijk for supporting this study as well as the parent and the legal guardian of the patient for allowing us to use the patient's medical information for academic purposes.

Ethical approval

A waiver for ethical approval was obtained from Kilimanjaro Christian Medical University College Ethics Review Committee.

Funding

The work did not receive fund from any source.

Author contribution

All authors made substantial contributions to the paper. KAN conceived the study. KAN, DSR, SMB, SFM, ARM and AM reviewed the patients' medical records, planned and executed management. KAN and ARM were the lead surgeons. KAN made the initial manuscript draft. AM performed histopathological analysis and critically reviewed the manuscript. All authors read and approved the final manuscript.

Guarantor

Dr. Alex Mremi is the Guarantor of this work.

Research registration number

Not applicable.

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