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Maxillofacial Surgery and Stomatology

Desmoid Fibroma of the Jaw in Pediatric Patients: How should it be Treated?

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Abstract **Case Report**

Desmoid fibroma of the jaw in pediatric patients presents unique challenges in treatment due to its local aggressiveness and tendency for recurrence. The management approach typically involves the following strategies: surgery, observation or adjunctive therapies. We report a case of a 15 years old children with a desmoid fibroma of the jaw operate bye a hemimandibulectomy and reconstruction. The management of desmoid tumours is often surgical due to the risk of extension and recurrence and in children, the impact on growth and facial morphology must be taken into consideration.

Keywords: Desmoid Fibroma, Jaw, Child.

INTRODUCTION

Desmoid fibroma is a rare bone tumor that primarily affects adolescents and young individuals under 30 years of age, most commonly affecting the mandible especially at the angle and ramus. It is characterized by local aggressiveness and tendency for multiple recurrences following surgery, leading to repeated operations. Definitive diagnosis is histological, which can sometimes be challenging, as desmoid fibroma shares similarities with its tissue homologs.

In this article, we will explore the management challenges associated with desmoid fibroma in children, using a clinical case and a review of the relevant literature.

OBSERVATION

A 15-year-old child without pathological antecedents consults for a mouth opening limitation and a right mandibular swelling evolving for 3 months and associated with an uncounted weight loss (figure 1).

The examination of the facial massif finds a mandibular swelling in front of the ramus on the right side, which is hard and no mobile and is part of the bone.

The mouth examination notes a vestibular filling with a healthy mucous membrane, the sensibility and the motricity are preserved. In front of this clinical aspect, a check-up made of standard biological sample which came back normal and a radiological check-up made of an orthopantomogram which shows multilocular radioclar lesions which intrude the angle and the ramus of the mandible on the right side which swell the cortical in places without any ricealysis (figure 2).

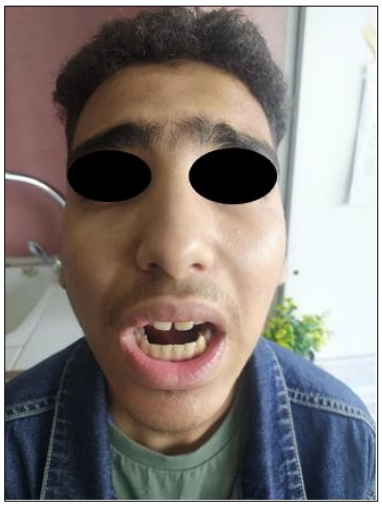

Figure 1: limitation of mouth opening

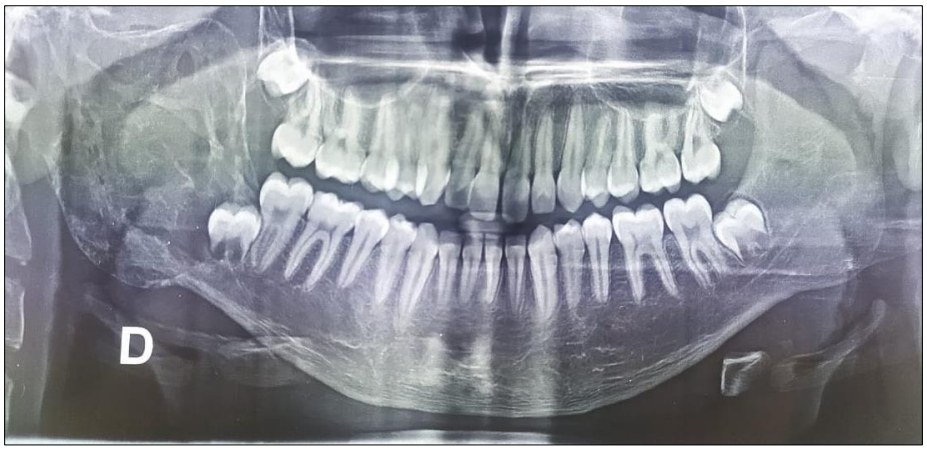

Figure 2:othopanthomogram showing the lesion on the right hemimandibula

A complementary scan was performed which also showed a multicompartmental bone mass extended to the mandibular ramus and the condyle (figure 3).

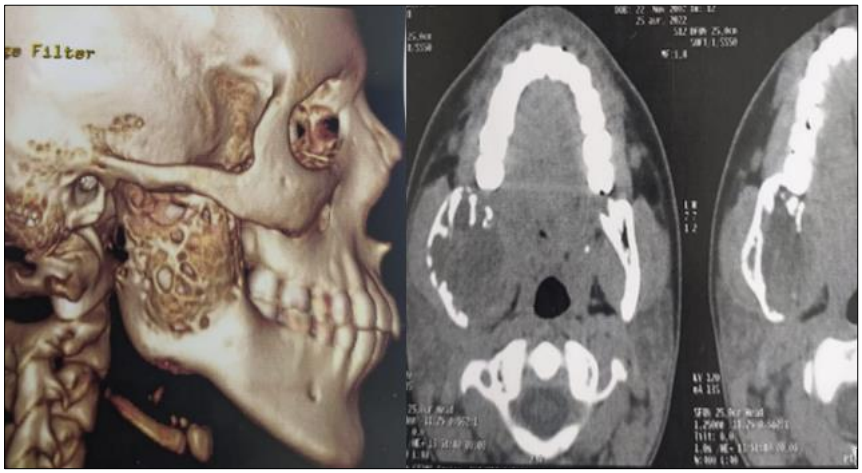

Figure 3: CT image of mandibular lesion

In front of these radioclinical data a surgical biopsy was performed under general anesthesia and which came back in favor of a desmoid fibroma given the local aggressiveness of the tumor, the risk of recurrence a hemimandibulectomy was performed, and the reconstruction was done by surgical cement given the age of the patient and a reconstruction by a free fibula transfer is planned after the end of the growth phase of the patient.

The evolution was marked by a clear improvement of the mouth opening under an early rehabilitation (figure 4).

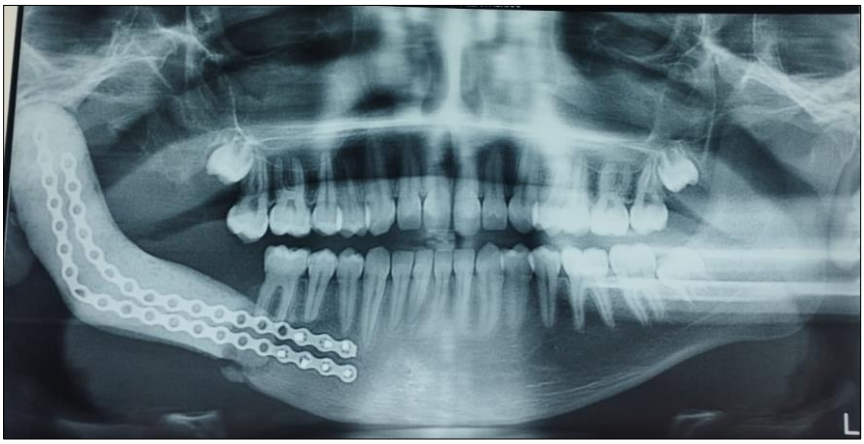

Figure 4: orthopanthomogram at one-year postoperative follow-up

DISCUSSION

Desmoid fibroma or desmoplastic fibroma is a rare, non-odontogenic, fibrous bone tumor that represents about 0.06% of all bone tumors and 0.3% of benign tumors [1].

The mandibular location is the most frequent: 86% of cases, all ages are concerned but with a peak during the second decade and an average age of 25 years, most reviews report an equal distribution of sexes. The occurrence of desmoid fibomas is often sporadic but also observed in the GARDNER syndrome which associates familial adenomatous polposis with bone tumors and other manifestations [2].

The clinical symptomatology is atypical and consists of a mass syndrome evolving progressively over time, which delays the diagnosis. in 65% of cases it is a non-painful swelling that increases in volume progressively [3]. The appearance of jaw constriction is a sign in favor of an invasion of the masticatory muscles and/or the temporomandibular joint [3]. Sensitivity disorders are rare in the mandible and in some cases pathological fractures can be observed [4]. dental displacement and mobility are found in 18% of cases [4].

In general, DF mimics other common or unusual pathologies of the jaws, which poses many differential diagnoses [5]. Radiologically, the images are not specific and include well-limited or irregular uni or multilocular radiolucent images with thinning or swelling of the cortices; this destruction is found in 74% of cases. Intramedullary extension and soft tissue invasion are best assessed by MRI.

Histologically, the desmoid fibroma is characterized by sparse spindle-shaped fibroblasts within a striated collagen stroma. The nuclei are elongated or ovoid without mitotic activity. The differential diagnosis is a well-differentiated fibrosarcoma. The absence of mitotic activity, hyperchromatism or nuclear atypia helps to make the distinction. On immunochemical analysis, the tumor cells react to anti-vimentin and anti-smooth muscle actin antibodies. They do not react to S-100 protein and MIB-1 (Ki67), an excellent marker of proliferative activity activity of sarcomas. Malignant transformation has been reported but never established and probably these tumors should have been initially classified as low grade fibrosarcoma which poses a real histological differential diagnosis [6].

The management is surgical, and wide resection with safety margins is the reference treatment for this locally aggressive and recurrent lesion. Conservative approaches such as enucleation are reserved for small lesions but with disappointing results due to frequent recurrences which are between 40% and 70% depending on the technique. Interruptive resections leave substance losses that require immediate or delayed reconstruction. In the growing child, mandibular reconstruction is a source of debate. Delayed reconstruction will cause latero-deviation and jugal depression responsible for an aesthetic prejudice that can be a source of psychological suffering for the child.

Thus, given the age of the child, the reconstruction must be as simple as possible with less morbid postoperative effects. Therefore, microsurgical reconstruction is ideally left to the end of the growth phase.

In our patient, given the extent of the resection, we performed a surgical cement reconstruction that will serve as a space maintainer and a free transfer of the fibula will be performed after the end of the growth phase to allow the patient a dental rehabilitation.

CONCLUSION

In conclusion, the treatment of desmoid fibroma in pediatric patients should be individualized, balancing the need for effective tumor management with the potential impact on the child's growth and development

BIBLIOGRAPHY

1. Kahraman, D., Karakoyunlu, B., Karagece, U., Ertas, U., & Gunhan, O. (2021). Desmoplastic fibroma of the jaw bones: a series of twenty-two cases. *Journal of Bone Oncology*, *26*, 100333.
2. Fricain, J. C. (2014). “Thanks Jacky!,” Med. Buccale Chir. *Buccale*, *20*(2), 73–74, 2014, doi: 10.1051/mbcb/2009037.
3. Ben Slama, L., Zoghbani, A., Hidaya, S., & Ruhin, B. (2023). “¨de Fibrome desmoı,” 327–328.
4. Elmonofy, O., ElMinshawi, A., Abdelsalam, S., & Mubarak, F. A. (2021). Desmoplastic fibroma of the jaws: case report. *Advances in Oral and Maxillofacial Surgery*, *4*, 100162.
5. Skinner, H. R., Vargas, A., Solar, A., Foncea, C., & Astorga, P. (2017). Desmoplastic fibroma of the mandible in a pediatric patient: a case report of resection and reconstruction with a six-year follow-up. *Journal of Oral and Maxillofacial Surgery*, *75*(7), 1568-e1.
6. Schneider, M., Zimmermann, A. C., Depprich, R. A., Kübler, N. R., Engers, R., Naujoks, C. D., & Handschel, J. (2009). Desmoplastic fibroma of the mandible-review of the literature and presentation of a rare case. *Head & face medicine*, *5*, 1-5.