

Desmoid Fibroma: A Case Report and Literature Review

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Abstract

Desmoid fibroma is a rare benign tumor of fibrous tissue characterized by a high potential for locoregional invasion and recurrence. The mandibular location of this pathology is exceptional. In our work, we report a case of desmoid tumor observed in a 9-year-old boy. Clinically, he presented a large right cheek swelling that was immovable relative to the mandible. The diagnosis was based on clinical, radiological and pathological arguments. There is no therapeutic protocol for desmoid tumors, but surgery is the most commonly used treatment. Chemotherapy, radiotherapy and hormonotherapy are used in addition to surgery or in some cases of inoperable tumors.

Keywords: tumor, benign, chemotherapy, radiotherapy, mandibulectomy, histology.

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INTRODUCTION

Desmoid tumor or aggressive fibromatosis is a rare benign tumor of fibrous tissue that develops from supporting tissues and aponeuroses and characterized by a high potential for locoregional invasion and recurrence. It has the most commonly a cervico-facial location.

Its mandibular location is exceptional. It is predominant in young children. Its clinical and radiological presentation is not specific, but it poses a difficulty of differential diagnosis with malignant tumors because of its very infiltrative nature.

Histological diagnosis by tumor biopsy completed by an immunohistochemical study is essential before any therapeutic strategy.

Its treatment remains poorly codified. Indeed, several therapies are used; surgery being in the first line, chemotherapy, radiotherapy and hormonotherapy are used in addition in some inoperable cases.

The quality of the surgical treatment determines the occurrence of subsequent recurrences. In our work we present a case of a tumor in a 9-year-old boy and we discuss the clinical aspects and the diagnostic and therapeutic features.

OBSERVATION

We report the case of a 9-year-old male child without any medical history, who presented a voluminous right mandibular tumor evolving, progressively evolving during the last 4 months in a context of a good general status, associated with nocturnal snoring and odontalgia. The clinical exam found an eupneic child, in a good general condition, presenting a voluminous right jaw tumefaction with a shiny skin above and telangiectasias. The tumefaction was painless, movable relative to the superficial plan and immovable relative to the mandible (Figure 1), measuring 9 cm on its longest diameter with a preserved sensitivity of the lower part of the face. The endobuccal examination found a convenient dental occlusion with an absence of the 43 and a slight vestibular filling without neither linguoversion nor vestibuloversion of the teeth of the lower dental arch.



Fig-1: Large right jaw tumor

The orthopantomogram revealed a right lytic lesion extending from the parasympyseal region to the mandibular angle, heterogeneous, with irregularity of the mandibular base and periosteal reaction (Figure 2).



Fig-2: Heterogeneous lytic mandibular lesion with periosteal reaction and irregularity of the mandibular base extending from the parasympysis to the right angular region

The CT scan of the facial bones noted the presence of a right mandibular pathological process, centered on the horizontal branch, well limited with regular contours, hypodense, discreetly enhanced after injection of the contrast product, containing peripheral calcifications and responsible of a cortical lysis with periosteal reaction (Figure 3; Figure 4) coming into contact with the masseter muscle outside and the tongue inside without a separating fatty border. The process pushes back the sternocleidomastoid muscle and the laryngopharyngeal axis to the left with bilateral jugulocarotid and spinal lymphadenopathy.

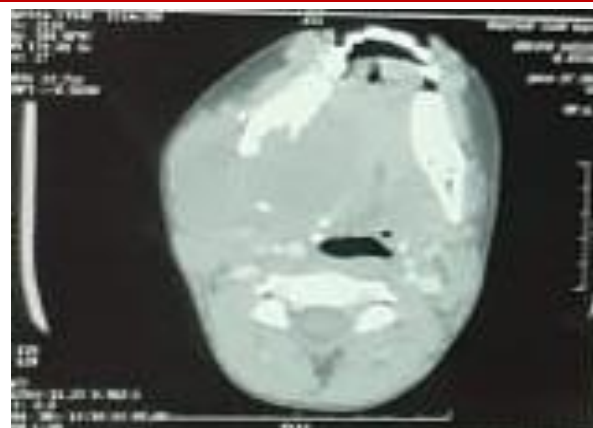


Fig-3: Axial CT scan showing a lytic tumoral process of the right hemimandible measuring 72x54x92mm

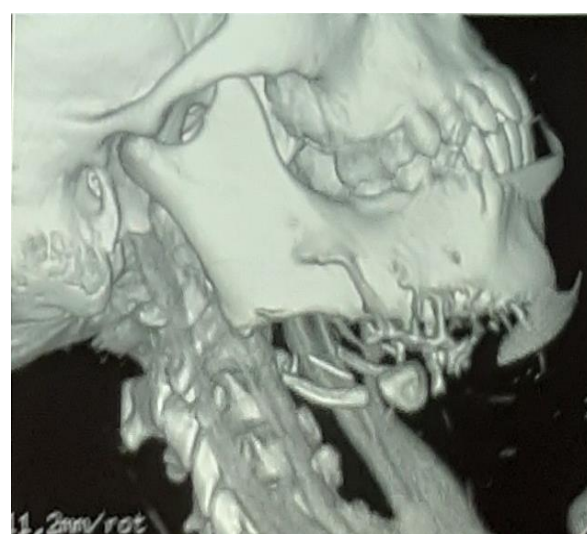


Fig-4: 3D CTscan showing a lytic process of the right hemimandible extending from the parasympyseal region to the angular region

An extraoral biopsy was performed, in which the anatomopathological study revealed a cell proliferation with an intermediate density, made of fusiform cells arranged in long bundles that have an ovoid nucleus and a moderately abundant eosinophilic cytoplasm with the absence of cytonuclear atypia (Figure 5, Figure 6).

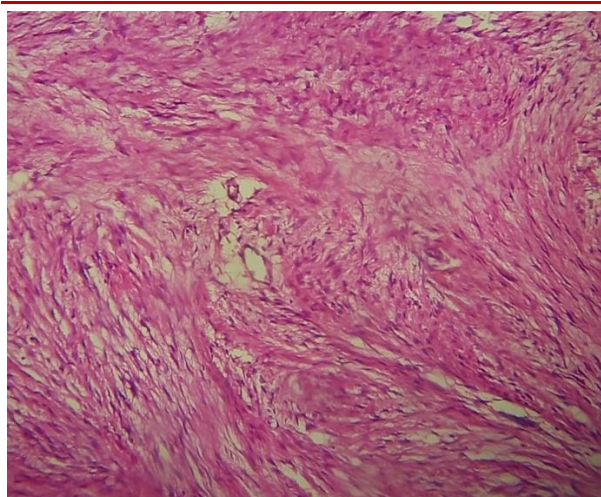


Fig-5: Macrophotography showing tumor proliferation of fusiform cells with low cell density arranged in bundles and dotted with vessels surrounded by a clear halo. HE GX10

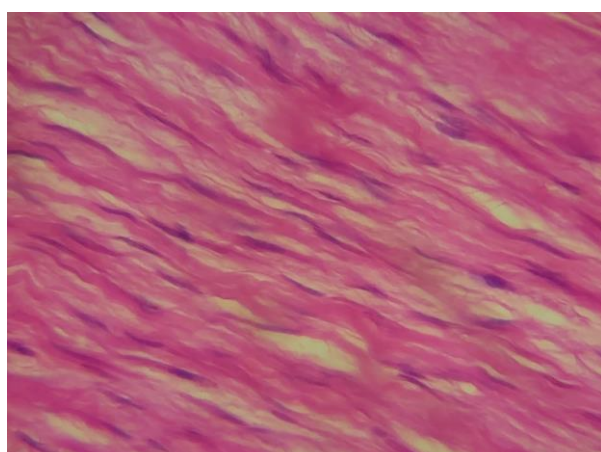


Fig-6: Tumor cells are endowed with elongated nuclei without atypia or sign of mitosis HE GX40

The immunohistochemical study showed positivity of tumor cells for anti-EMA antibodies and an estimated 5-10% labeling of tumor cells for anti-K67 antibodies indicating a desmoid fibroma.

The patient was admitted to the operating room. Under general anesthesia, we adopted a cervical approach of about 10 cm followed by a careful dissection from close to close preserving the mental nerve and allowing a complete resection of the tumor with regularization of the mandibular base (Figure 7). The patient did not present any postoperative complications and the clinical and radiological follow-up after 6 months was satisfactory.

The immunohistochemical study of the resected specimen (Figure 8) showed tumor cells clearly expressing betacatenin with diffuse nuclear labelling confirming the diagnosis of desmoid fibroma (Figure 9).

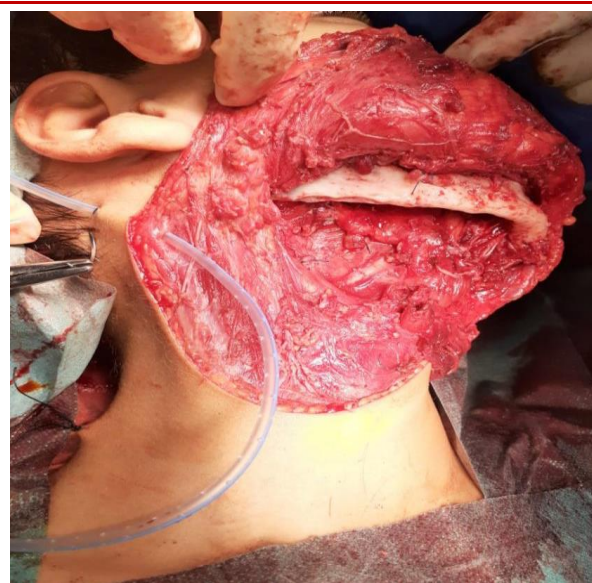


Fig-7: Intraoperative image after tumor resection and regularization of the mandibular base

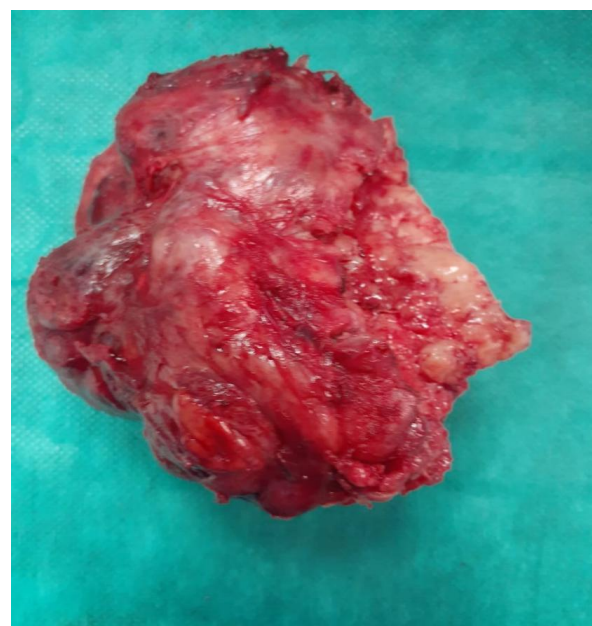


Fig-8: Tumor after an en bloc resection

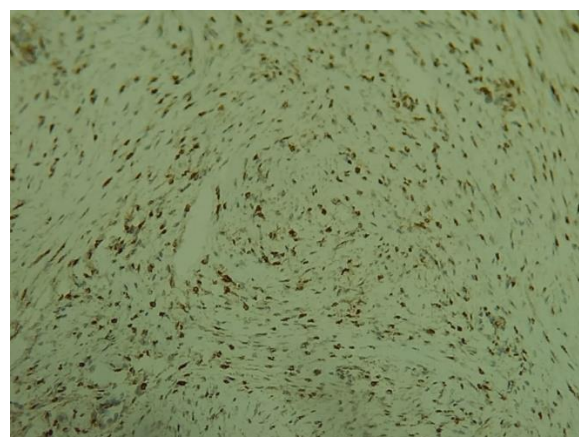


Fig-9: In immunohistochemical study tumor cells clearly expressing Betacatenin with nuclear labeling

DISCUSSION

Desmoid fibroma is a rare myofibroblastic tumor representing 0.06% of all bone tumors and 0.3% of benign bone tumors [1]. It is a tumor that can pose diagnostic difficulties [2]. Mandibular location is exceptional, representing less than 1% of mandibular tumors [3]. In the mandibular location, the angle and the ramus are the most frequently affected [4].

The exact causes are still unknown, but the desmoid tumor may be related to traumatic or hormonal factors [5]. Most cases are non-genetic, mainly related to the mutation of a gene in the tumor, which encodes the β -catenin protein, involved in the regulation of cell growth genes. Mandibular desmoid tumors affect mainly male children with a sex ratio of 5/3. The average age is nine [6].

The clinical symptomatology is polymorphic and nonspecific, but it is most often manifested by a firm and painless mandibular swelling [5] without neither facial sensitivity disorder nor associated tooth mobility, as is the case in our patient.

The uni- or multilocular osteolytic radiological images with or without condensation border often associated with dental displacements and rhizalyses are not specific [7]. On CT scan, a periosteal reaction and a cortical rupture with extension to the soft tissues can be found as well [8].

The diagnosis is histological and it is important to differentiate this tumor from malignant tumors, mainly fibrosarcoma with low-grade of malignancy whose treatment is different [6]. The anatomopathological study shows a proliferation of myofibroblastic fusiform cells, in a highly rich collagenous stroma. The proliferation is clearly invasive, and extending into adjacent tissues [9]. The absence of bone or osteoid substance makes it possible to exclude ossifying fibroma and fibrous dysplasia [10].

There is no therapeutic protocol for desmoid tumors, but surgery is the most common treatment. Radiotherapy is used in case of incomplete tumor resection and to prevent recurrence or in case of inoperable tumors. Chemotherapy can also be used alone or preoperatively to decrease the tumor size [11], but surgery remains the gold standard despite a high recurrence rate of up to 70% [9].

Several authors recommend radical treatment [6], in fact interruptive mandibulectomy allows a good tumor control at the cost of a considerable aesthetic and functional damage [9]. When the resection is interruptive, the mandible is repaired most often at the same time to avoid tissue retractions. Other authors prefer conservative treatment with a complete removal of the tumor in order to limit the impact on facial

growth. The high risk of recurrence requires a regular and prolonged follow-up.

CONCLUSION

Desmoid fibroma is a rare myofibroblastic tumor, its mandibular location remains exceptional. The diagnosis is histological. Although it is a benign tumor with no metastatic potential, its treatment remains difficult due to its aggressive nature.

SUMMARY

Desmoid fibroma is a rare benign tumor of fibrous tissue characterized by a high potential for locoregional invasion and recurrence. The mandibular location of this pathology is exceptional. In our work, we report a case of desmoid tumor observed in a 9-year-old boy. Clinically, he presented a large right cheek swelling that was immovable relative to the mandible. The diagnosis was based on clinical, radiological and pathological arguments. There is no therapeutic protocol for desmoid tumors, but surgery is the most commonly used treatment. Chemotherapy, radiotherapy and hormonotherapy are used in addition to surgery or in some cases of inoperable tumors.

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