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Mandibular Resection in Pediatric Patient with Extensive Fibromyxoma: A Case Report

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ABSTRACT

Myxomas of the jaws are considered to arise from odontogenic ectomesenchyme. This pathology is most commonly found in young adults, but it may appear across a wide age group with an average age from 25 to 30 years with no sex predilection. We present a case of a 2-year-old male pediatric patient with the increased facial volume on the right side and six months of evolution without presenting symptoms, with the diagnosis of fibromyxoma, where a surgical resection through hemimandibulectomy with disarticulation and immediate costal graft reconstruction was performed. It was an uncommon case due to its extension and prolonged evolution time, which represented a challenge in our particular environment. Surgical resection may be required because myxomas are not encapsulated and, because of their consistency, they may infiltrate the surrounding bone. Long-term clinical and radiological follow-ups should be undertaken due to the risk of recurrence.

1. Introduction

The myxoma, or fibromyxoma, is a benign neoplasm with a non-defined histogenesis. It often behaves in a locally aggressive way, infiltrating tissues. Myxomas or fibromyxomas appear in the head and neck region deriving from the facial skeleton. Histopathologically, the lesion consists of rounded and angular cells placed in an abundant mucoid stroma, differentiating from myxoma by presenting a bigger quantity of fibrous cells.[1] Clinically, myxomas are regularly asymptomatic and present in the maxillofacial region as firm expansive masses; displacement or mobility of the associated dentition can also occur. There is no predilection for the mandible or the maxilla regarding its appearance. Their appearance age rate is wide, and they generally occur in individuals between the second and fourth decades of their lives. It is very unusual to find it in children and the elderly. It seems to have a very slight predilection for female patients.[2] The final diagnosis is defined by biopsy, and, in some cases, immunohistochemistry may also be helpful for a more accurate diagnosis. Complete ennuclation or radical excision is considered the best surgical treatment for fibromyxomas, depending on their size. Local recurrence is low when there is a complete tumor removal. In the literature, metastasis of this kind of tumor has not been reported.[3] The purpose of this study is to present an unusual clinical case of Fibromyxoma in a 2-year-old patient, as well as an updated review of the literature on this infrequent clinical presentation. According to the Helsinki declaration, the ethics committee approved this study of the University Hospital of Maracaibo, and the patient's mother signed informed consent.

2. Case Presentation

A 2-year-old male pediatric patient attended the Oral and Maxillofacial Surgery service of the University Hospital of Maracaibo, Venezuela, in January 2019, due to an asymptomatic increase in volume right side of his face with six months of evolution. The parent did not refer to any relevant medical history, and the patient was clinically stable. The large mass was specifically located at the right submandibular region. It was firm and painless to palpation (Fig. 1).

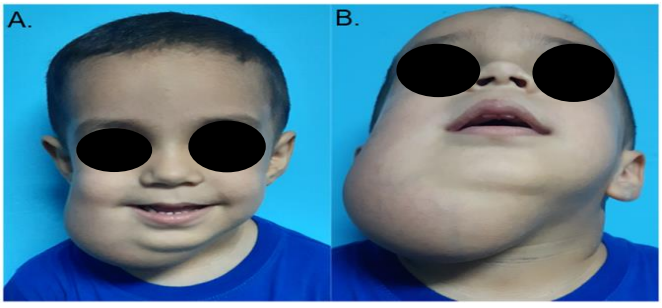

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Fig. 1. Clinical extraoral image of the patient.

Intraorally, there was evidence of preserved occlusion, and the teeth were in good condition. A CT scan was requested, and an isodense mass on the right side involving the mandible with soft tissue infiltration was observed. (Fig. 2).

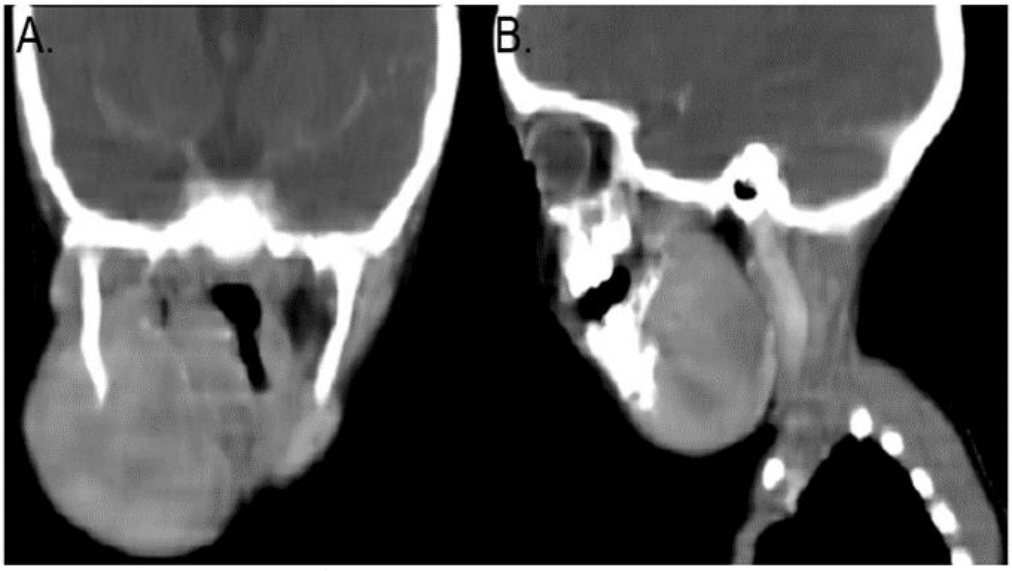

Fig. 2. CT scan of the head showed an isodense mass on the right mandible.

The first clinical impression was lymphoma or rhabdomyosarcoma. An incisional biopsy was performed, and a diagnosis of Fibromyxoma was obtained (Fig. 3), for which the pathologist requested immunohistochemistry to discard any malignant pathology and confirm the diagnosis. The results from the latter study were consistent with odontogenic fibromyxoma. Due to these findings, an excisional biopsy was performed under general anesthesia, involving a hemimandibulectomy with disarticulation and immediate placement of a costal graft with plate osteosynthesis for the reconstruction (Fig. 4).

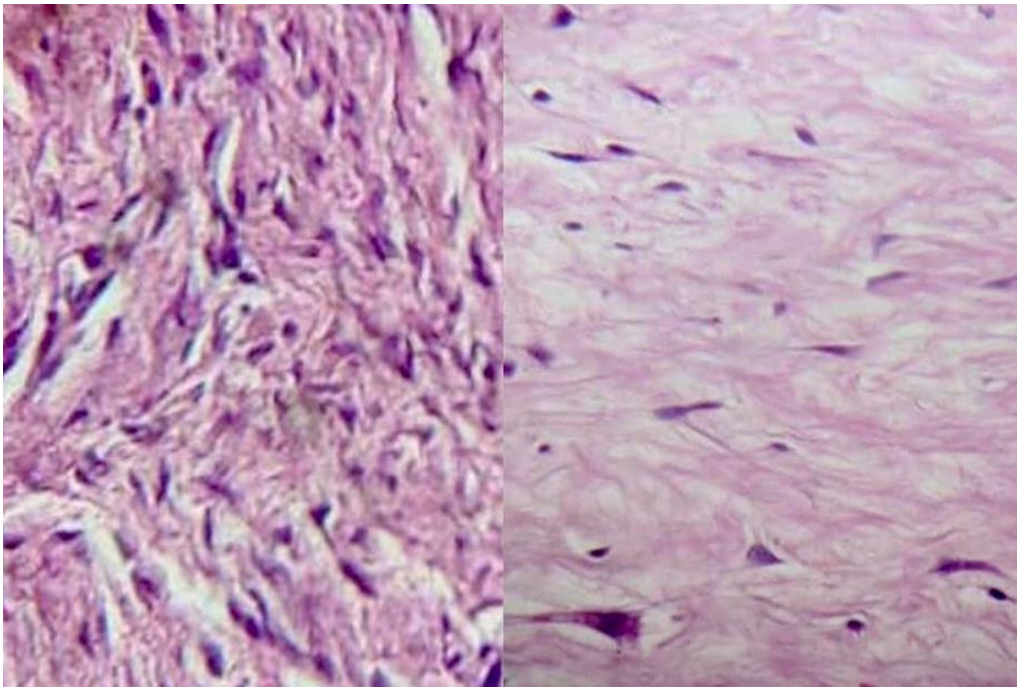

Fig. 3. Left, dense fibrous connective tissue stroma with round and spindle-shaped fibroblasts (200X). Right, loose myxoid fibrous connective tissue stroma. Round, stellate and spindle-shaped fibroblasts (400X).

The final histopathology study reported a stroma of dense fibrous connective tissue in some areas, myxoid in others, composed of fusiform cells, some of them with a slightly pleomorphic and hyperchromic nucleus, others with a wavy and palisade pattern, organized in fascicles and presence of collagen fibers (Fig. 3). The same diagnosis was confirmed by immunohistochemistry, which reported positive for Vimentin, which favors mesenchymal origin, and negative for S-100, discarding neurofibroma as one of the differential diagnoses (Fig. 5). Seven days after the surgery, the patient was discharged, and the postoperative period was uneventful. (Fig. 6).

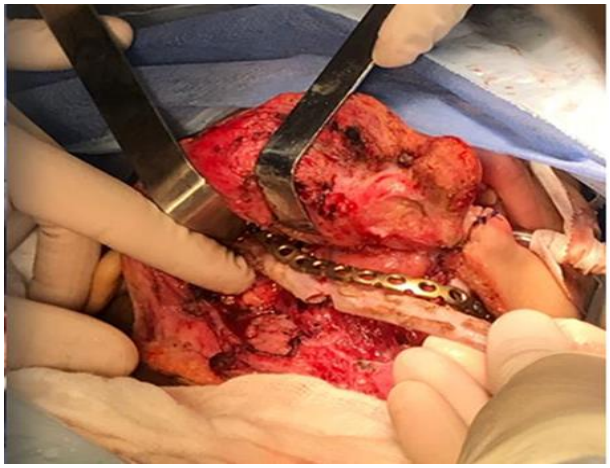

Fig. 4. Close view of the surgery, hemimandibulectomy with disarticulation, and costal graft with plate osteosynthesis.

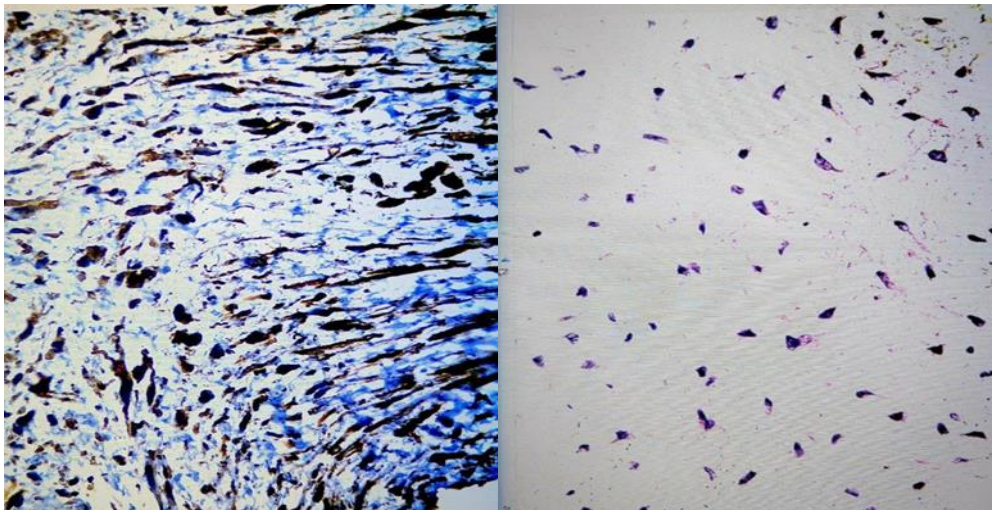

Fig. 5. Immunohistochemically reported tumor cells negative for S-100 discarding neurofibroma and positive for Vimentin. Left, Vimentin (400X). Right, S-100 (400X).

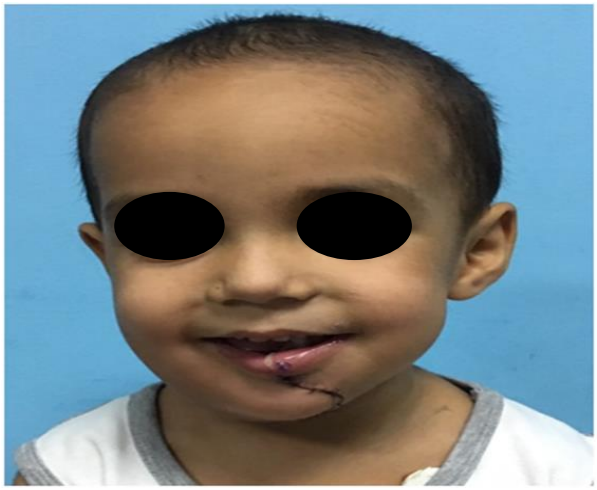

Fig. 6. Postoperative evolution of the patient after seven days.

3. Discussion

The Myxoma is considered a tumor present as a slow-growing, persistent, locally aggressive, destructive but histologically benign, non-encapsulated lesion. When this kind of tumor appears in the jaw, they are considered odontogenic myxomas. Adults in their third decade of life are the most prone patients, and it is rarely seen in pediatric patients.[4, 5] In our case, the tumor grew rapidly in 6 months, and the patient was only two years old. These two facts have been rarely reported in the literature. When rapid growth and expansion are seen, it is usually associated with a mucoid ground substance's production by stellate tumor cells. The lesion is benign, but bone perforation, root resorption, and tooth displacement and mobility can occur, making it a locally aggressive tumor.[6] In this reported case, in which the jaw has been compromised, all these characteristics were observed, and thus, a more aggressive approach was decided.

The Fibromyxoma presents macroscopically as a soft, glistening, gelatinous, and non-encapsulated lesion. Microscopically, it consists of haphazardly arranged stellate, spindle-shaped, and round cells in an abundant, loose myxoid stroma that contains only a few collagen fibrils.[7] Sometimes, the histopathological study can not define if the lesion is a myxoma or a fibromyxoma. Immunohistochemistry must be requested to confirm the diagnosis and discard other lesions such as lymphoma rhabdomyosarcoma, especially in fast-growing tumors and pediatric patients. A myxoma, such as the rare chondromyxoid fibroma or the myxoid neurofibroma, can be microscopically confused with other myxoid jaw neoplasms.[8, 9] At first instance, the presented case was diagnosed by an incisional biopsy, which was confirmed by immunohistochemistry, presenting the reported characteristics. Tumor cells stained negative for S-100 and CD34 and positive for Vimentin, ensuring its mesenchymal origin.

Conservative surgery, such as curettage and enucleation, has been reported to be performed when tumors are less than 3 cm in diameter. Surgical resection with safety bony margins of 1.0 mm to 1.5 mm may also be performed when tumors are large and aggressive.[9, 10] The present case was highly aggressive, and thus, it was treated by surgical hemimandibulectomy with disarticulation and immediate costal graft reconstruction. Depending on the tumor's location, focusing on pediatric patients may be considered an important factor in determining the resection extension. The percentage of recurrence for this lesion must be taken into account as well.[7] The tumor may reappear during the first two years. In patients with this pathology, long-term follow-up for at least five years is ideal, so regular clinical and radiological examinations are necessary for successful management.[10]

4. Conclusion

It was a case of a massive fibromyxoma in a 2-year-old pediatric patient. The prolonged evolution time, the rapid growth of the tumor aggravated the challenges faced in its management in an environment with a shortage of resources such as the one in this case. More studies are needed to assess the prognostic importance of growth rate and age in presenting this tumor type. Oral and maxillofacial surgeons should know about this tumor's clinical characteristics to perform a correct diagnosis and management, considering immunohistochemistry as a helpful resource, especially when the histopathological study does not generate a clear diagnosis.

Conflict of Interest

The authors declared that there is no conflict of interest.

Acknowledgements

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