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Case Report

Agressive pediatric myofibromatosis in a two-year-old child

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SUMMARY

Introduction: Aggressive paediatric myofibromatosis is an autosomal recessive disease characterized by fibroblastic proliferation from cells originated in muscle-aponeurotic tissue. Its etiology is unknown, and the average age of the reported cases is 7 years old. The tumor exhibits rapid painless growth and appears attached to muscle tissue and/or bone. The treatment of choice is conservative surgical excision despite of early relapses has been reported.
Observation: A 2-year-old patient, with no morbid history, presented with a large swelling in the left submandibular region, firm, neither defined limits nor inflammatory characteristics. Its size doubled 2 months after an incisional biopsy. CT images showed great compromise of the left mandibular body with expanded and thinned cortical bone. The MRI showed extension towards the pharynx. Histopathological findings were elongated fibroblastic and ovoid cells arranged in bundles and fascicles within fibromyxoid stroma, an image consistent with the diagnosis. The treatment consisted in a conservative exeresis of the tumor, preserving the jaw. Control 1 year after surgical removal shows no signs of relapse and the mandibular structure has been restored.
Discussion: The large size of the lesion and bone involvement at such an early age evidenced a very aggressive lesion, however, supported by a previous biopsy, we performed a conservative treatment, which only caused the loss of a dental germ, impossible to take off from the intraosseous tumor. The control of this type of lesions requires a longer follow-up.

1. Introduction

Aggressive paediatric fibromatosis is a recessive autosomal disease characterized by the spread of fibroblastic cells from muscle tissue, or aponeurosis [1]. Of unknown origin, its onset has been associated with traumatic, hormonal and genetic factors [2]. It is far more common in boys than in girls, by a factor of 18 to 1, and the average age of the reports is seven years [2].

Such tumours have been described in the neck, the parotid, submandibular region, lateral orbit, and the outer ear, among others. They grow quickly, are painless and normally adhere to muscle and/or bone tissue. The lesion can obstruct the airway if it grows toward the midline and reaches the oropharynx [3]. It is a relatively rare condition whose occurrence is 0.2–0.4 cases per 100,000, of which 12% are in the head and neck [4].

2. Observation

A two-year-old boy without a medical history was brought to us for consultation due to significant sub-mandibular swelling, painless and disfiguring, on the left side that had evolved over the last few months and which had doubled in volume over the last two months, since an incisional biopsy was carried out for diagnostic purposes. The histological diagnosis was fibromatosis (Fig. 1) The MRI images reveals a large tumour enveloping the left mandibular body, invading its left lingual side, passing along the back side of the ascending ramus to the vicinity of the inner lower pharynx, deforming the upper and lower hyoids on the left (Figs. 2 and 3) The lingual cortical plate of the mandibular bone structure was destroyed and the vestibular bone was worn and deformed, which led us to fear a fracture during surgery. The CT images of the chest, abdomen and pelvis did not reveal any other tumour sites.

We adapted an osteosynthesis plate using a 3D print of the mandible and small acrylic splints to fix it in the same position in the intraoperative period and to reinforce the structure during

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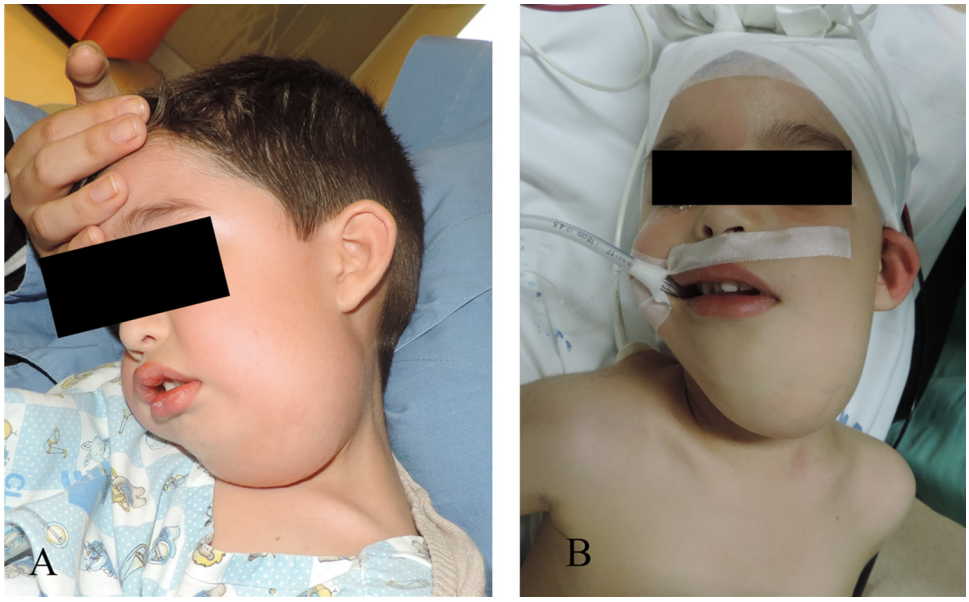

Fig. 1. A. Lateral view. B. Frontal view, two weeks later.

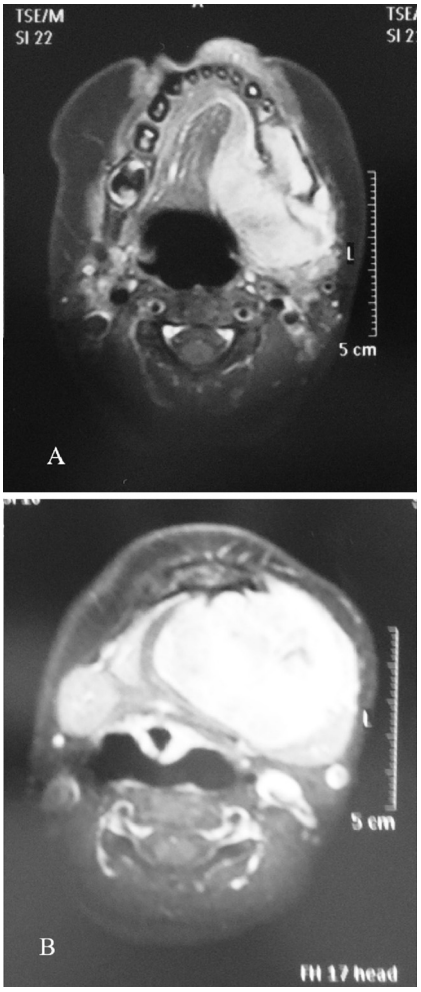

Fig. 2. A. NMR Picture, showing the tumor in the mandibular body. B. Lower image, passing by sub-mandibular space.

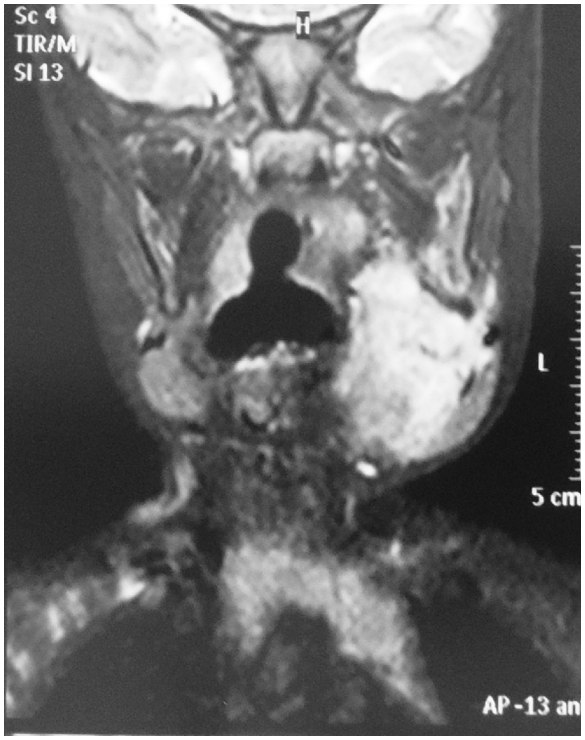

Fig. 3. Frontal view.

curettage (Fig. 4). The intervention was performed under general anaesthesia and we used a cervical approach to expose the most superficial part of the tumour, which presented a cleavage plane. We were forced to resect the superficial part of the tumour to reach the mandibular body (Fig. 5). We then exposed the mandible and resected the deep sub-mandibular part, which was well-encapsulated. Lastly, we placed the osteosynthesis plate and removed the tumour from the mandibular body, where a dental germ came out attached to the lesion (Fig. 6).

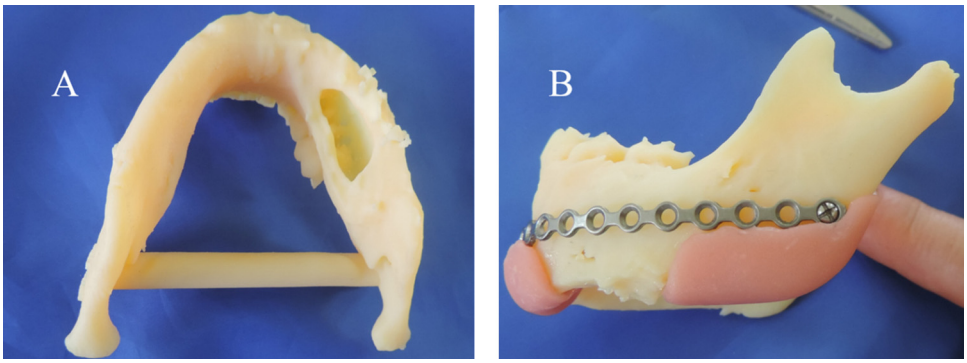

Fig. 4. A. 3d image of the mandible, showing the bone destruction. B. The osteosynthesis plate and the acrylic splints for assuring the same position.

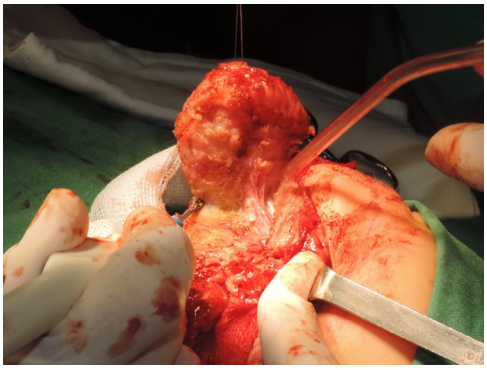

Fig. 5. The hemi section of the tumor.

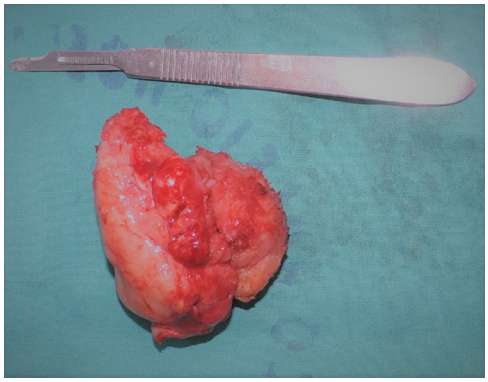

Fig. 7. The surgical specimen.

The surgical specimen as a whole was the size of a tennis ball, and the consistency was more solid on the periphery than in the centre (Fig. 7). Histologically, it was described as a fibroblastic tumour with a low mitotic index, without necrosis or inflammatory infiltration. H&E staining revealed spindle-shaped and ovoid fibroblastic cells arranged in bundles in a fibromyxoid environment. The histochemical study was Vimentin (+); S100 (−); CD34 (−), Desmin and Ki67 (15%), and smooth muscle actin in the spindle cells was identified, confirming the diagnosis of fibromatosis (Fig. 8).

Postoperative evolution has been highly satisfactory. Follow-up images a year after surgery, upon removing the osteosynthesis plate, do not show any sign of relapse in the soft tissue, the jawbone structure has been rebuilt and is showing normal growth (Fig. 9).

3. Discussion

Aggressive paediatric neurofibromatosis can occur as a single lesion or multiple lesions that can affect several sites, with or without visceral compromise [6], meaning that when it is diagnosed its occurrence in other locations must be ruled out with CT scans of the chest, abdomen and pelvic areas.

The majority of cases occur in male children and evolve as solid masses with poorly defined edges that show swift and progressive growth, and without inflammation [6] they can grow to a large size, making their removal difficult and favouring relapses [7,8].

With regard to relapse rates, different numbers have been reported: Peña [5] reports 16% in a series of 97 cases, while Tostevin calculates 50% in a series of just 6 cases. Local recurrence is partly due to a certain potential for infiltration on the part of the

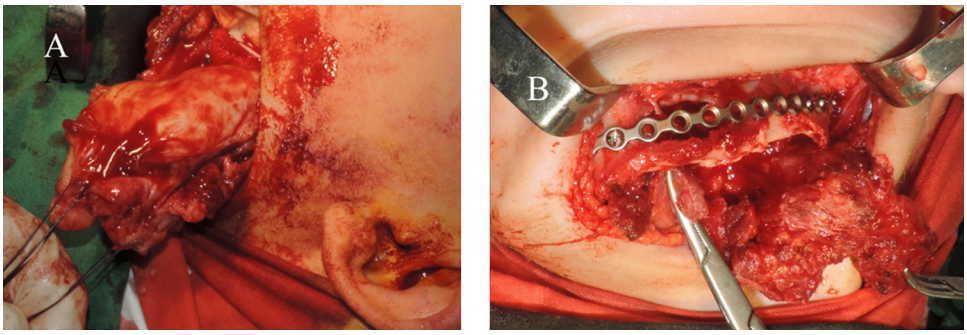

Fig. 6. A. Excision of the deeper part of the tumor, with a capsule. B. Mandibular aspect after bone curetage.

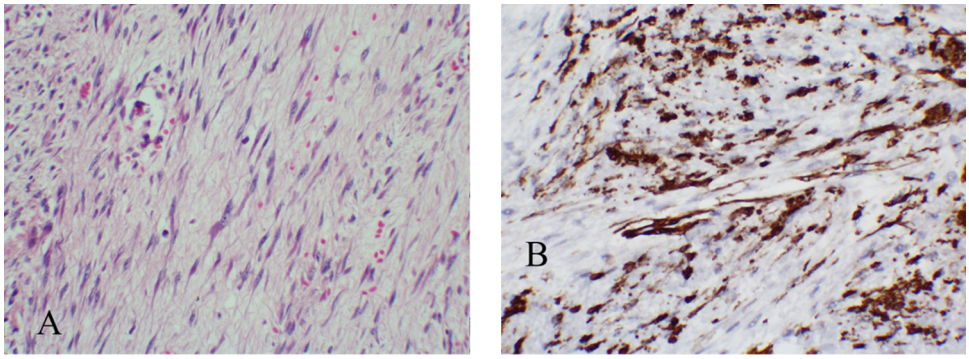

Fig. 8. A. Stained with HE. B. Immunoreactivity, showing the expression of actine ml.

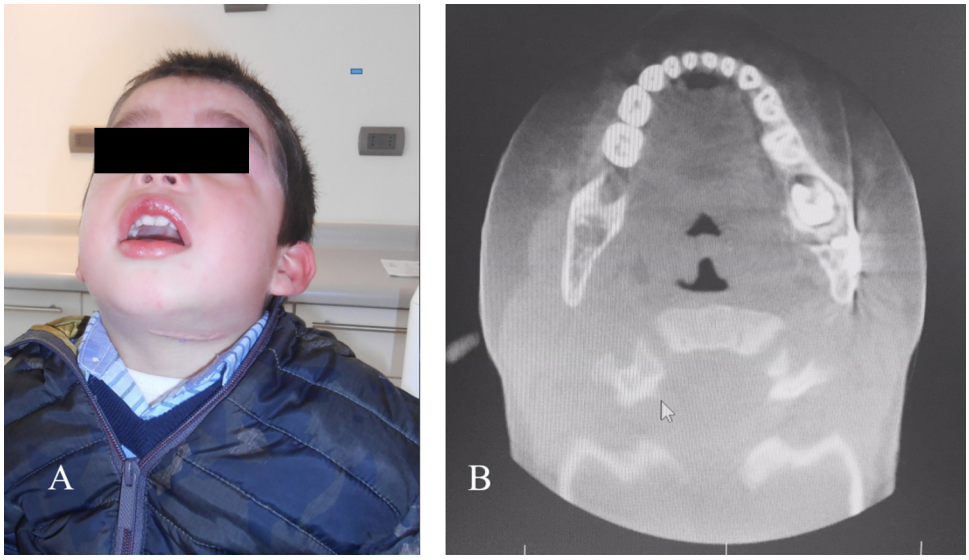

Fig. 9. A. Clinical aspect one year after surgery. B. CT image, without signs of recurrence, one year post-surgery.

tumours, given their weak or incomplete encapsulation, in addition to their normally large size in small patients, which often places the tumours in relatively close proximity to vital structures, further complicating their total removal.

These features have led these lesions to be considered locally aggressive, and wherever possible they are to be removed with a safety margin of 1 cm to ensure negative resection limits [8]. Postoperative follow-up must be rigorous and in case of relapse an early resection must be performed again.

Radiotherapy and chemotherapy have been used only in some cases where vital structures were compromised, given the toxicity and the growth problems associated with use of such procedures in children [1,4,7,8].

The treatment of choice is complete surgical resection, with a safety margin whenever possible. This is the procedure we followed in this case, where we performed a conservative surgery, and so far, 18 months after the operation, there have been no signs of relapse.

Disclosure of interest

The authors declare that they have no competing interest.

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