Journal of Pediatric Surgery Case Reports 70 (2021) 101867

Contents lists available at ScienceDirect

Journal of Pediatric Surgery Case Reports

journal homepage: http://www.elsevier.com/locate/epsc

Resection of an aggressive nodular fasciitis of the mandible in an infant girl

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ARTICLE INFO

Keywords:
Nodular fascitis
Mandible
Myofibroblast
Sarcoma

ABSTRACT

Nodular fascitis is a benign pseudosarcomatous self-limited fibroblastic and myofibroblastic cell neoplasia that occasionally can be found in pediatric patients, in who head and neck is reported to be a common location. Although nodular fascitis could be suspected by imaging, histologic review is mandatory to certificate the diagnosis and rule out malignant conditions of similar behavior. We report the case of an infant girl with a big, solid, left submandibular mass with a rapidly progressive growth that erodes mandibular body. The diagnosis is proven by histopathology and immunohistochemistry. The lesion does not respond to triamcinolone intralesional infiltration and is finally surgically resected by a transcervical approach with good results and no recurrence at follow-up.

1. Introduction

Nodular fasciitis is a rare pseudosarcomatous condition characterized by benign proliferation of fibroblastic and myofibroblastic cells, which can appear as a nodule or subcutaneous firm mass. It can be painless or painful, and varies in size [1,2].

Although it often affects the soft parts of the upper limbs or trunk in adulthood, at pediatric age it appears to be more common in the head and neck [3].

Given the rapidly progressive growth, its erosive behavior in relation to adjacent tissues seen at diagnostic imaging, and the finding of fusional cells in proliferation and mitosis in histopathology, nodular fasciitis can be confused as a malignant lesion, its main differential diagnosis [1,2], along with the extensive chapter of benign fibromatosis.

The case of an infant affected by a particularly aggressive and voluminous nodular fasciitis located in relation to the mandibular body, treated with surgical resection, is presented.

2. Case report

A 1 year- 3 months old female came to the hospital with swelling on the left submandibular area, perceived as progressive in nature by her mother. The swelling was noted as hard in consistency and painless on palpation, without fluctuation and not mobile, without any inflammatory signs in the overlying skin, and no palpable lymph nodes in the head and neck or other regions. The study with skull x-rays on that day did not show any bone lesions.

A cervical ultrasound was performed, which describes a solid oval shaped tumor in the depth of the subcutaneous cellular tissue, heterogeneous in structure with linear ultrasound areas interspersed with other hypoechogenic ones, measures were 3.3 × 4 × 2.8 cm, with some vascular paths inside to the Doppler-color view, which surrounds the lower edge of the left mandible causing small bone erosion (Fig. 1). The ipsilateral submandibular gland is displaced and unaltered in structure.

Diagnostic imaging is complemented with craniocervical magnetic resonance which describes a neoplastic lesion located in the left submandibular space with medial extension to the parapharyngeal space, which contacts and displaces the submandibular gland posteriorly and the muscles of the mouth floor medially. The lesion is markedly heterogeneous and hyperintense in T2/STIR, with significant reinforcement after the gadolinium, measuring 3.7 × 4.8 × 4.2cm (estimated volume 39 cc) and surrounds the mandible showing some erosion of it (Fig. 2).

Surgical incisional biopsy of the lesion is performed to guide therapeutic behavior. The histopathological findings demonstrate a tumor composed of fibroblastic-looking fusiform cells arranged in short bands partly swirled, microcystic areas, erythrocyte extravasation and few lymphocytes, with no cellular atypia. The report concludes a nodular fasciitis.

Taking into account potential surgical morbidity after resection of the mass, the parents were consulted and it is decided to evaluate the

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https://doi.org/10.1016/j.epsc.2021.101867
Received 16 March 2021; Accepted 11 April 2021
Available online 22 April 2021

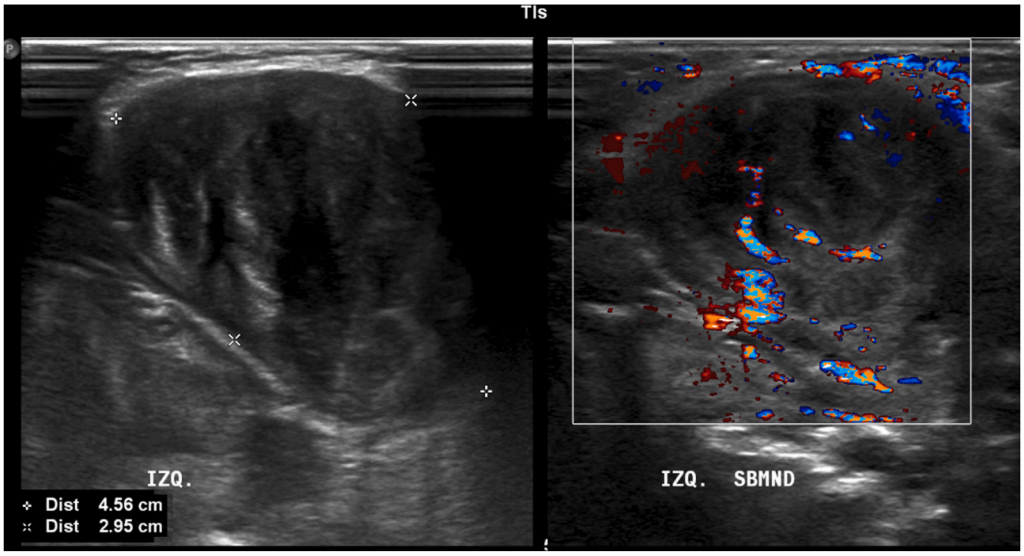

Fig. 1. Ultrasound features of the initial submandibular lesion.

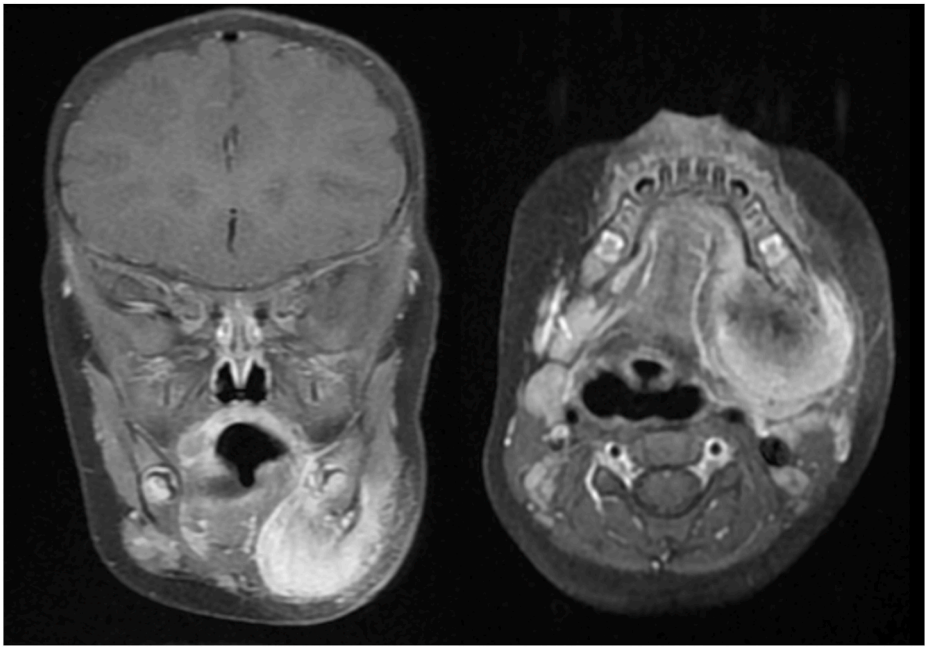

Fig. 2. MRI findings of submandibular lesion.

response to one attempt of local infiltration with glucocorticoids (solution of 3ml of triamcinolone in 7ml of saline), looking for a possible decrease or detention of growth of the mass. After 2 weeks, no objective response is seen and even a slight increase in size is noted. By that date, the patient does not show stridor or other ventilatory difficulties, the tumor remains painless and with similar clinical features, but some feeding difficulty is reported by the parents.

Surgical resection of the tumor is decided by the team, and an eventual mandibular reconstruction is considered. For surgical planning, the preoperative study is completed with maxillofacial computed tomography, which shows a clear increase in size compared to magnetic resonance imaging and measures, after of 2 months period interval. A solid mass is described, located in the left masticatory space, which surrounds the left mandibular body and angle, with clear erosion of the adjacent bone and periosteal reaction at its boundaries of non-infiltrative characteristics. CT measures are now 7 × 5.7 × 4 cm (estimated volume: 83.5 cc) (Fig. 3).

In the operating room, general anesthesia and orotracheal intubation is performed without any difficulty. The surgical approach is performed by a high left cervicotomy resecting the previous biopsy scar, located parallel and 2cm inferior to the mandibular margins, from the chin to the mandibular angle. Upper and lower flaps are raised, bringing the dissection to a deep plane to the superficial musculature, with mono- and bipolar electrocoagulation and blunt dissection. Once the tumor was surrounded externally, a direct relation with the mandibular body margin (in its inferior and medial area) was noted. This feature was previously noted in diagnostic imaging, causing periosteal reaction and cortical erosion, without extending beyond the inferior third.

At this deeper surgical level, blunt dissection, periosteal lifting and bone regularization with bone rongeur are uneventfully performed. Posterolateral dissection and resection of the tumor is completed, and the hypoglossal nerve and the submandibular gland are visible and intact. Hemostasis is checked carefully, repair and closure are performed

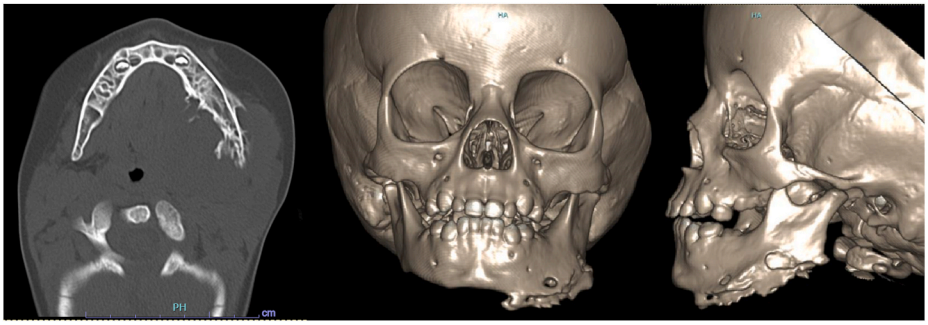

Fig. 3. MCT features and 3D reconstruction, where mandibular erosion and periosteal reaction are noted.

by planes, with no need of aspiration drainage, and the patient is extubated in the operating room (Fig. 4).

The patient is monitored in the PICU during the first postoperative day, showing no signs of ventilatory difficulty or any other surgical or anesthetics complications. Due to positive postsurgical performance the patient was transferred to a basic care unit and oral feeding was resumed. Mild postoperative edema and ecchymosis are observed with progressive regression after 3 days. A subtle smile asymmetry due to presumed marginal nerve apraxia is noted. She is discharged from hospital on the fifth postoperative day with analgesics.

Histopathological examination report describes the operative piece as a tumor composed of fusiform cells arranged in randomly interlaced bundles, with lax stroma areas, myxoid-microcystic and other denser degenerations, with erythrocyte extravasation foci, scarce mitosis and foci of infiltration into adjacent skeletal muscle tissue at the upper and medial border. This report is associated with immunohistochemical techniques that show negative skeletal muscle actin in neoplastic cells, positive in vessel walls; positive smooth muscle actin intense diffuse, cytoplasmic; weak positivity for Betacatenin, focal granular cytoplasmic (negative at nuclear level, ruling out fibromatosis); and negativity to the study with S100, desmin, myogenin, MyoD1, CD34 and CD117, with a Ki 67 of 1%. The findings are compatible with nodular fasciitis (Fig. 5).

At a 12 months postoperative follow up, a clear improvement of contour is noticed, with only subtle jaw irregularity that can be palpated but not seen, and a final adequate and cosmetic scar. The subtle smile asymmetry resolves spontaneously and completely by the third postoperative month. The 12 month ultrasound examination, no signs of local recurrence is found, and adjacent structures features such as the submandibular gland are uneventful (Fig. 6).

3. Discussion

Fibroblastic and myofibroblastic tumors comprise a broad spectrum of mesenchymal neoplasms corresponding to 12% of soft tissue tumors at pediatric age. Their clinical manifestation is usually nonspecific, and some of their histopathological characteristics may overlap. They are classified as benign, intermediate (locally aggressive and rarely metastatic) and malignant (sarcomas). Benign ones can be subclassified into fibroids, fibromatosis and pseudosarcomas [3].

Nodular fasciitis (NF), a benign pseudosarcomatous entity that accounts for approximately 10% of benign soft tissue tumors, occurs in up to 20% of cases at the pediatric age, and is more common in adolescents. Previously attributed to the reparative process of local trauma, today NF is recognized as a neoplastic clonal proliferation attributed to MYH9-USP6 gene fusion [4].

At pediatric age, NF usually occurs in the head and neck (61%), followed by the limbs (22%) or trunk (17%) [3], as a small, solitary subcutaneous nodule that grows rapidly from weeks to months. In a series of 15 pediatric patients with head and neck NF, aged between 2 months and 18 years (average 9.3 years), lesions were reported in the maxillary or JAWS, forehead, scalp, suboccipital, post-auricular, chin and neck, and up to 4 cm in diameter (average 2.17cm) [5]. Another report of 18 patients with NF in head and neck, considering adult and children patients, includes the case of a 2 year old infant with a submandibular lesion of 4 cm [6].

The appearance of NF in images is usually nonspecific and variable depending on their location. They are usually found as an oval shaped, well circumscribed lesion, of approximately 2cm in diameter, with wide contact with the fascia that is usually projects a “fascial tail” [3]. On ultrasonography, it is portrayed as a solid hypoecogenic lesion with moderate increase in Doppler-color vascularization, without shadows or calcifications. On MRI, it is described as isointense to striated muscle at

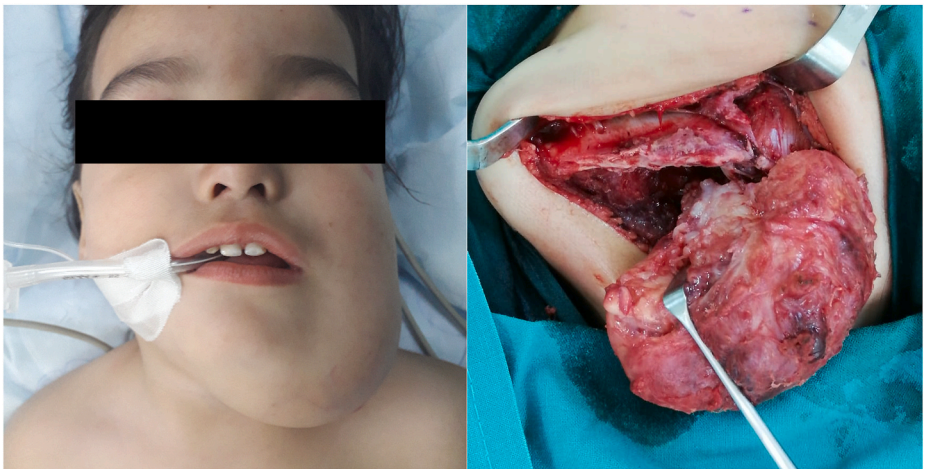

Fig. 4. Left: preoperative submandibular tumor aspect. Right: tumor dissected and mobilized from mandibular margin.

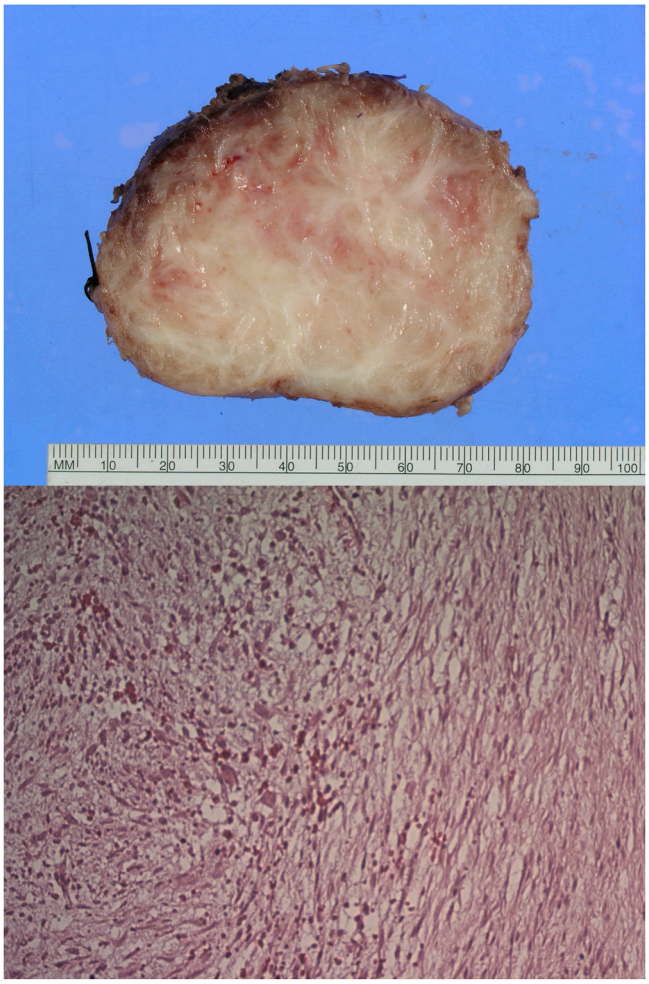

Fig. 5. Macroscopic and histopathological examination of ressected lesion.

T1, heterogeneous signal of moderate to high intensity at T2, and heterogeneous contrast attenuation. Limits are usually well defined, often associated with a margin of perilesional edema by the associated fascial plane. They can be seen locally aggressive, with transcompartmental extension and bone involvement [3].

Although NF may be suspected on imaging, the diagnosis is established with histopathology. Typically, it is a non-encapsulated, circumscribed tumor, smaller than 3 cm. Histopathological characteristics [12] vary according to the stage of growth. In early lesions, fusiform and stellate myofibroblasts surrounded by a myxoid matrix confer a fibroblast cultured appearance, also extravasated erythrocytes and lymphocyte and plasmacyte infiltrates are common. In the classical well-developed phase, cellularity is higher with fusiform cells arranged in short fascicles mixed with collagen bundles. There may also be zonation with myxoid, hyaline and more peripheral fusiform areas, osteoclastic type cells and swollen ganglion cell type myofibroblasts may be present in variable amounts. Over time, the myofibroblast bundles become wider and in a suture-like arrangement, the myxoid matrix decreases and is replaced by thick bands of collagen and microcystic spaces with mucoid material are observed inside. Mitosis is more numerous in the early phase [7]. Immunohistochemistry shows strong reaction with smooth muscle actin and vimentin, can also express specific muscle actin, desmin and calponin, with in the absence of h-caldesm and beta-catenin.

Although spontaneous regression or regression after incisional biopsy has been reported, the diagnosis of NF should always be tested by histopathology to exclude malignant tumors, and should usually receive active treatment because of its rapid and unpredictable behavior. Resective surgery usually has a curative and characteristically recurrence-free result [1–3], even in partial resections [5]. In fact, the recurrence after resection in these cases makes the revision of the histopathological diagnosis mandatory [3].

More conservative therapeutic alternatives have been described, particularly in dermatological literature. A clinical response has been demonstrated to intralesional injection of glucocorticoids (triamcinolone), with or without association with CO2 point laser [8,9], susceptible to subsequent resection in the event of lack of response. In case report, infiltration was attempted with a higher volume of triamcinolone than that used by Graham and collaborators for a flattened lesion of 5 cm in diameter [9], in multiple punctures seeking to dispense the drug throughout the tumor, without achieving an objective response within 14 days. Considering the progressive growth of the lesion and the potential risk of airway obstruction, we proceeded to complete resection.

Among the cases reported in the literature, the one published by Anehosur and cols stands out [2], as the most similar and useful for the management of our patient. Their report describes a 10-year-old male boy who consulted for a firm and minimally sensitive swelling on the left mandibular body, rapidly progressive and displacing the midline and cervical vessels. It is studied with fine needle aspiration puncture for cytology, which is complemented with incisional biopsy confirming NF. An elective tracheostomy in anticipation of post-operative airway edema after resection is preformed, and the definitive treatment involves extended left submandibular approach, resection of a $7 \times 5 \times 5$cm tumor surrounding the left mandibular body, angle and rami, and mandibular fixation with stainless steel mini-plate due to extensive mandibular cortical erosion. Our patient, although with a similar located and dimensions lesion, airway protection with tracheostomy

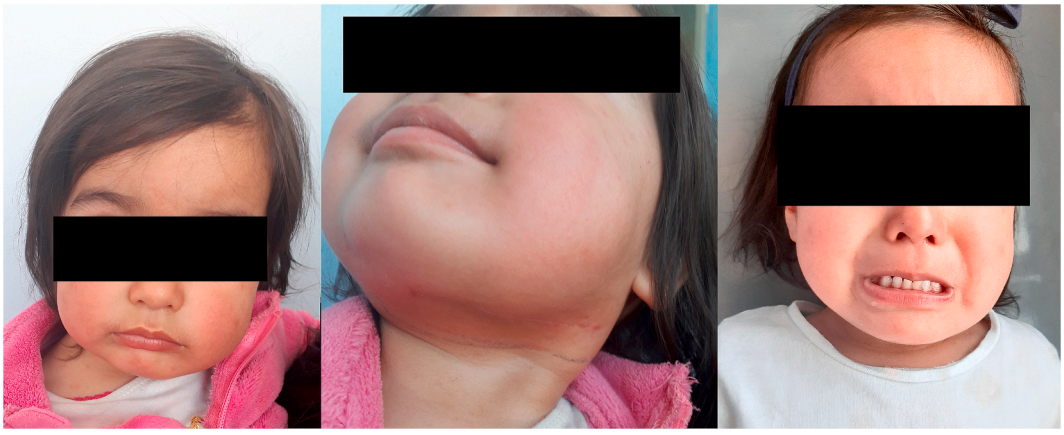

Fig. 6. Postoperative results.

was not a preventively preformed due to lack of airway obstruction risk, and even after resection it was possible to immediately extubate the patient, without any signs of ventilatory difficulties despite local edema. We believe, however, that it is undebatable to consider a preventive tracheostomy in cases deemed important for perioperative airway safety. In our case report, after tumor removal the mandibular bone structure showed no evidence of risk fracture, consequently no need for fixation plates and satisfactory local bone remodeling of the mandibular margin, which was confirmed in the monthly and 1 year follow up.

Other noteworthy reports include a 7-year-old girl case referred for 3 month snoring and mouth breathing, secondary to a parapharyngeal NF that reached 5 cm in diameter. This case was managed with transoral resection without evidence of recurrence at follow-up [10]. Another group reported the case of a 82 year old woman with 1 month progressive dyspnea that finally required emergency tracheostomy due to rapid growth of a cervical histological demonstrated NF that caused extrinsic compression of her trachea [11].

4. Conclusion

NF is a rare pathology at pediatric age, that should be included in the differential diagnosis of solid tumors of the head and neck in children. Despite its benign behavior, it can simulate sarcomatous lesions in imaging study because of its locally aggressive features, requiring histopathological exclusion of malignant conditions of similar course. When located in the neck, potential airway obstruction should be considered due to its rapidly progressive behavior.

The most accepted and definitive treatment for pediatric head and neck NF is surgical resection, which has demonstrated good results in the few reports available with minimal recurrence rate.

Funding

No funding was received for this work.

Intellectual property

We confirm that we have given due consideration to the protection of intellectual property associated with this work and that there are no impediments to publication, including the timing of publication, with respect to intellectual property. In so doing we confirm that we have followed the regulations of our institutions concerning intellectual property.

Research ethics

We further confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript.

Consent

Consent to publish the case report was obtained and signed by the child's parents.

Authorship statement

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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