Cervical nodular fasciitis in a 10-year-old girl: a case report of a rare condition

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Nodular fasciitis (NF) is a benign fibroblastic proliferation that usually involves superficial fascia in the subcutaneous tissue or muscle. The rapid growth, high cellularity and mitotic activity can simulate sarcoma. Thus, as clinical and morphologic characteristics are similar to those of malignant tumours, this entity is often misdiagnosed, with a few cases described in the literature. We present a case of nodular fasciitis involving the retropharynx and hypopharynx of a 10-year-old girl and discuss the importance of considering this rare diagnosis in different soft tissue masses. ***Ann Pediatr Surg*** **9:144–146 © 2013 Annals of Pediatric Surgery.**

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Introduction

Nodular fasciitis (NF) is defined by WHO as a benign and reactive fibroblastic growth. It was first described in 1955 by Konwaller, Keasbey and Kaplan, and was initially termed pseudosarcomatous fibromatosis. This entity is rare in children and more common in the second or fourth decades of life. Whereas in adults it appears in the upper extremities, in children, lesions typically involve the head and neck. We present a case of cervical NF, with rapid growth and consequent respiratory symptoms, initially interpreted as a malignant tumour, the histological evaluation only later confirming the diagnosis as NF.

Case report

The authors describe the case of a 10-year-old child who presented at the emergency department with right cervical swelling, showing rapid and progressive growth over 2 months, with associated increasingly persistent snoring in the last 5 days. There was no relevant personal or family history. Upon examination, the bulky cervical mass was hard, painless and mobile with swallowing. Ultrasound showed a right laterocervical lesion with defined contours and a longest axis of 43 mm, with calcifications. The MRI showed the mass in the posterior hypopharynx, restricting and pushing to the fore of the respiratory tract and epiglottis. It had a spherical configuration, well-defined apparent origin in the posterior retropharyngeal tissues, a 44-mm-long axis and showed an intermediate signal on T1 and a hyperintense signal on T2, which may correspond to a high cellularity, including lymphoma (Fig. 1). As the imaging findings pointed to an aggressive neoplastic process and given the clinical respiratory conditions, surgery was proposed. Excision was performed under general anaesthesia. The intramuscular mass engaging hypopharyngeal and retropharyngeal muscle was solid, hard and had well-defined contours, although in close proximity to the larynx. During block excision of the lesion, the larynx sustained a laceration of about 0.5 cm that resolved without any further complications. Subsequently, the histology showed a well-defined lesion, completely excised, moderately to intensely cellular expansive, consisting of spindle cells forming a loosely fascicular arrangement with few collagen fibres, without cytologic atypia, mitosis or intratumoural necrosis. The most probable diagnostic hypotheses suggested were an inflammatory myofibroblastic tumour and NF. Immunohistochemistry showed an intense and diffuse reaction of the cells to vimentin, CD68, smooth muscle-specific actin mostly in the cells and CD10 in 60% of cells. All the remaining antibodies were negative: anaplastic lymphoma kinase, CD34, desmin, S100 protein, chromogranin, Bcl2, AE1/AE3 and epithelial membrane antigen. Histology and immunohistochemistry enabled the diagnosis of NF (Fig. 2). Two years after resection, the child has no clinical evidence and/or imaging of lesion.

Discussion

NF is a benign condition that can simulate an aggressive oncologic entity. The incidence is unknown because the lesion is misdiagnosed frequently as sarcoma and other soft tissue tumours. Only 10% of lesions occur in children and it appears predominantly in boys [1]. Although the aetiology is unknown, a history of recent trauma, local or inflammatory process is present in 10–15% of cases [2,3]. The most common clinical presentation of NF is a solitary, rapidly growing mass, with variable consistency, located in the head and neck. This rapid growth in conjunction with mild tenderness, present in ~50% of cases, may arouse suspicion of a more aggressive process [4]. The size of the lesions can vary from 0.5 to 10 cm, most being less than 4 cm, as in our case. Radiologic investigations of choice include ultrasound and MRI, but in NF, there are no characteristic appearances. On MRI, characteristics are highly variable, reflecting the variable histologic make-up of lesions. NF shows a nonspecific signal intensity ranging from homogeneous to mildly inhomogeneous and from hypointense to a slightly hyperintense on T1-weighting images to an intermediate-to-high signal on T2 weighting [1,3]. In our case, in MRI, the lesion was solid and well circumscribed,

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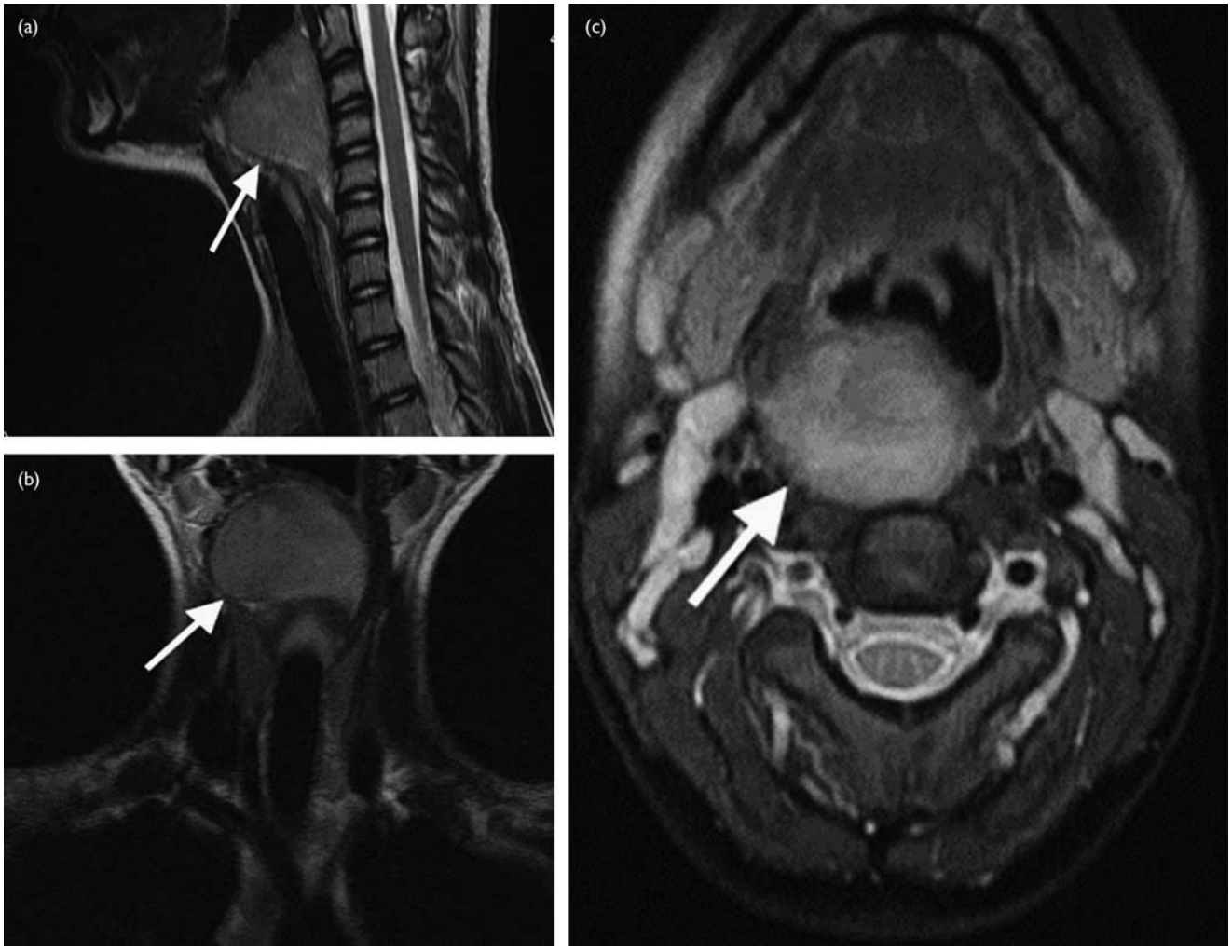
Fig. 1



Sagittal (a), coronal (b) and axial (c) MRI images show a well-circumscribed cervical mass. Arrows refers to a well circumscribed cervical mass.

with an intermediate signal on T1 and a moderate hyperintense signal on T2.

Thus, the diagnosis of NF is histologic, presents with hypercellularity and active mitotic figures without atypia and needs to be differentiated from other fibroblastic tumours such as fibromatosis, neurofibroma, fibrous histiocytoma and others [5]. NF is comprised of fibroblasts and myofibroblastic cells and can be divided into different subtypes, such as subcutaneous, intramuscular and fascial (intermuscular) according to the anatomic location. The subcutaneous type occurs 3–10 times more commonly than the other subtypes and presents as a subcutaneous nodule. The intramuscular type is most likely to mimic a soft tissue malignancy because it is typically larger and deeper. There is a specific subtype that presents exclusively in children younger than 2 years of age, in whom the lesion deeply engages skull fascia–cranial fasciitis. NF can still be classified into myxoid, cellular and fibrous subtypes on the basis of the amount and type of extracellular matrix. A relationship has been proposed between these subtypes and the age of the lesion: lesions with more cellular components correspond to younger lesions, whereas older ones are more fibrous. In our case, the lesion had moderate to elevated cellularity and thus appears to be recent. Moreover, the different proportion of myxoid, fibrous and cellular components allows a different relation with clinical and imagiologic features, but as yet, no classification has been developed. Some authors consider that histologic aspects limit the predictability of a prototype of NF in MRI [3,6].

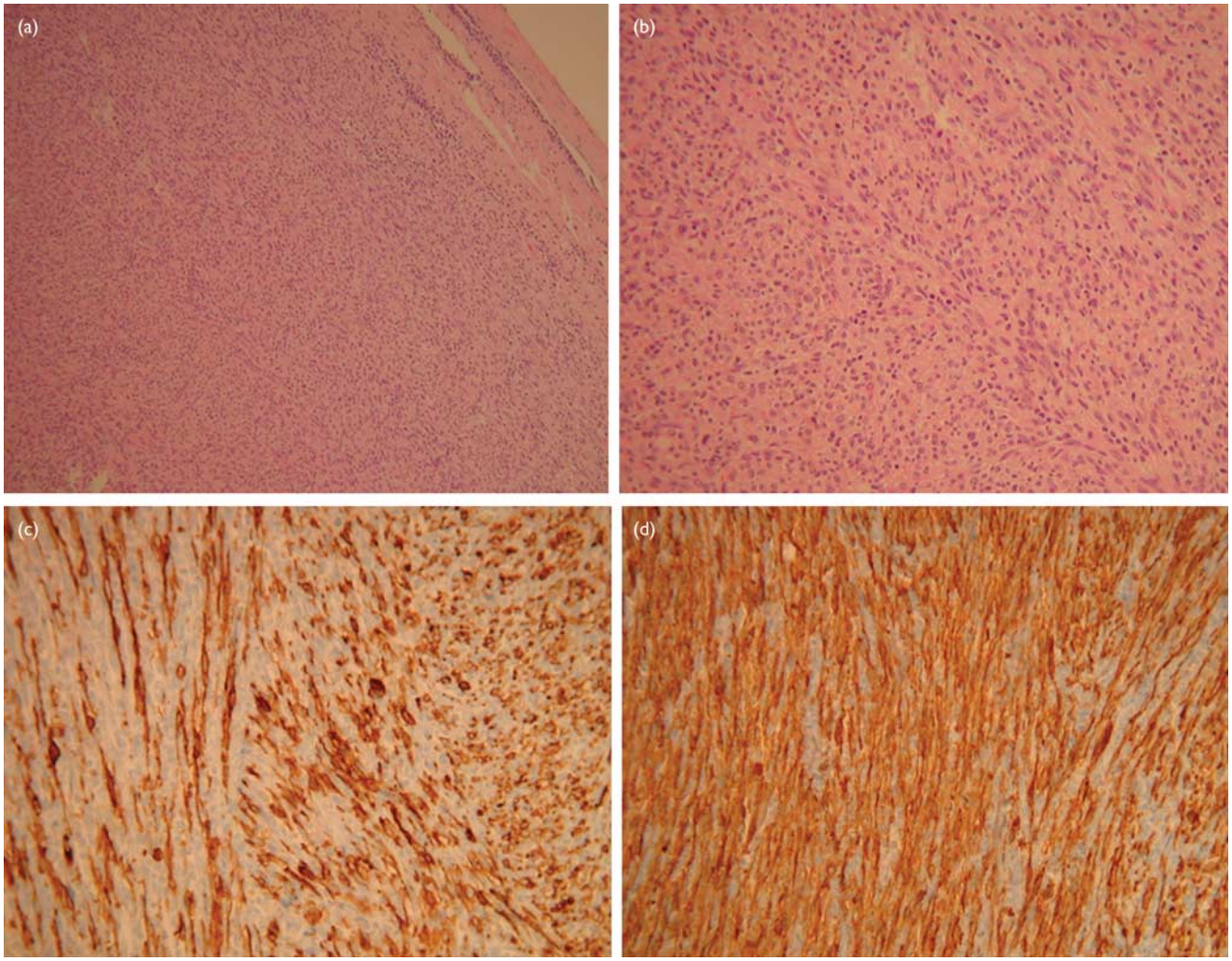
Despite histologic appearances, immunohistochemical characteristics confirm diagnosis: NF expresses actin and vimentin but not desmin or S100 protein, as in this case [4].

Most lesions are treated by local excision and the prognosis of NF is favourable: it does not develop metastases and rarely recurs. If it does recur, a disease other than NF should be suspected. In fact, in some cases, recurrence has been associated with incomplete excision of the lesion, although contrary to this, some authors have reported that even incompletely resected lesions do not recur [7]. In this case, no recurrence was observed 2 years after the excisional biopsy.

Conclusion

NF is an unusual benign reactive process of soft tissue often misdiagnosed as sarcoma. Early diagnosis and nonaggressive treatment depends on acknowledging this entity, allowing us

Fig. 2



Histopathological examination: (a) the mass is well circumscribed, with a heterogeneous appearance, and (b) the cells are spindled and showed a reaction to (c) smooth muscle-specific actin and (d) CD10.

to consider this pseudosarcomatous lesion in the differential diagnosis of paediatric soft tissue masses.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

References

1 Nikolaidis P, Gabriel HA, Lamba AR, Chan NG. Sonographic appearance of nodular fasciitis. *J Ultrasound Med* 2006; **25**:281–285.
2 Zuber TJ, Finley JL. Nodular fasciitis. *South Med J* 1994; **87**:842–844.
3 Di Serafino M, Maurea S, Vallone G. Nodular fasciitis of the chest: case report of a rare presentation. *Musculoskelet Surg* 2011; **95**:251–253.
4 Mazura JC, Matrai C, Spigland N, Brill PW, Kovanlikaya A. Intramuscular nodular fasciitis of the rectus abdominis muscle in an 11-year-old girl. *Skeletal Radiol* 2013; **42**:147–150.
5 Tomita S, Thompson K, Carver T, Vazquez WD. Nodular fasciitis: a sarcomatous impersonator. *J Pediatr Surg* 2009; **44**:e17–e19.
6 Wang XL, De Schepper AM, Vanhoenacker F, De Raeve H, Gielen J, Aparisi F, *et al*. Nodular fasciitis: correlation of MRI findings and histopathology. *Skeletal Radiol* 2002; **31**:155–161.
7 Mallina S, Rosalind S, Philip R, Harvinder S, Gurdeep S, Sabaria MN. Nodular fasciitis: a diagnostic dilemma. *Med J Malaysia* 2007; **62**:420–421.