CASE REPORT

Cervical nodular fasciitis in a 17-month-old child

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SUMMARY

Nodular fasciitis (NF) is an uncommon fibroblastic proliferation that usually arises in the extremities or trunk, less frequently in the head and neck region. It is frequently mistaken for a malignant tumour owing to its rapid growth, but it is ultimately a benign condition with spontaneous regression. We describe a case of a 17-month-old African girl with cervical NF in which spontaneous and complete regression of the lesion occurred following the diagnosis by fine needle aspiration cytology. We emphasise the importance of making an accurate and timely diagnosis, and the imperative role of fine needle aspiration cytology in this process, potentially avoiding further unnecessary surgical procedures.

BACKGROUND

We believe this is an interesting case because it is about a rare condition that presents as a common physical finding with worrisome characteristics that may represent a diagnostic challenge for the physician. In addition, once the diagnosis of nodular fasciitis (NF) is made, even though known as benign, the most frequent treatment for this condition is still surgery. What we intend to show is that fine needle aspiration cytology (FNAC) is a minimally invasive technique that can make the diagnosis and offer a watchful waiting strategy.

CASE PRESENTATION

A 17-month-old African girl presented to the emergency department with dry cough, fever and a left cervical mass of sudden onset—noticed by the child's mother the day before. She had no history of previous trauma or any other symptoms. Family history was irrelevant. The child had a medical history of preterm birth (at 26 weeks and 4 days), extremely low birth weight (490 g), hyaline membrane disease, *Staphylococcus* coagulase-negative sepsis, distal ischaemia of the left hand with amputation of fourth and fifth right fingers, and bronchopulmonary dysplasia during neonatal admission in the intensive care unit. She had an adequate psychomotor development.

On admission, she presented feverish (38.7°C—axillary temperature); the cervical mass was 4×3 cm in size, firm, with poorly defined limits and was fixed but not attached to the skin; there was some tenderness to palpation, but no other inflammatory signs on the overlying skin. She had no enlarged lymph nodes in the neck nor enlarged liver or spleen. Oropharynx had no signs of infection and the remaining physical examination was irrelevant.

INVESTIGATIONS

The chest X-ray was normal. Laboratory tests revealed leucocytosis (21 930/μL) with 41%

neutrophils and 44% lymphocytes, C reactive protein 2.3 mg/dL, sedimentation rate 110 mm/h, thrombocytosis 737 000/μL, aspartate aminotransferase 83 U/L, alanine aminotransferase 66 U/L and lactate dehydrogenase 551 U/L (normal range: 100–190 U/L). There were no atypical cells or any other morphological or quantitative alterations on peripheral blood smear.

Initial evaluation of the mass by ultrasonography identified an inflammatory process of the left submandibular gland (Doppler study was not performed). With the initial diagnostic hypothesis of cervical adenitis, the girl was admitted and started on empiric intravenous antibiotics (amoxicillin and clavulanic acid), with no clinical response. Cervical CT scan revealed an anterior cervical mass of the subcutaneous tissue with no contrast captation and compressing the ipsilateral jugular vein and carotid artery. Tuberculin skin test, as well as bacilloscopy and culture of gastric aspirate for *Mycobacterium tuberculosis* were negative. Serologies for HIV, cytomegalovirus, Epstein-Barr virus and toxoplasmosis were also negative. *Bartonella* serologies were not performed because there was no suggestive epidemiology.

On the fourth day of disease, after 72 h of intravenous antibiotics, the child remained febrile and the diameter of the mass kept increasing, maintaining the other characteristics. She was then submitted to FNAC under local anaesthesia. Cytopathological examination of the lesion demonstrated moderately cellular smears, with inflammatory infiltrate and scarce necrosis, isolated skeletal muscle cells, loose collagen stroma, spindle-shaped fibroblasts and cells with enlarged nuclei, sometimes binucleated, with prominent nucleolus and vast azurophilic cytoplasm, suggesting myofibroblastic origin; there were no malignant cells. These findings suggested NF (figures 1 and 2).

Figure 1 Cells with enlarged nuclei, prominent nucleolus and vast azurophilic cytoplasm.

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Figure 2 Inflammatory infiltrate, loose collagen stroma, spindle shaped fibroblasts.

TREATMENT

The child completed 10 days of intravenous antibiotic treatment.

OUTCOME AND FOLLOW-UP

The child became afebrile on the seventh day and after the 12th day the mass started to become progressively smaller (1×1 cm at discharge), softer and with no tenderness. After 6 weeks she was clinically well, with total regression of the mass and showed no evidence of recurrence in the following 3 years.

DISCUSSION

NF is defined by the WHO as a benign and probably reactive nodular fibroblastic growth, extending from the superficial fascia into the subcutaneous tissue or muscle.^{1–3}

It was first described in 1955 by Konwaler *et al*,⁴ under the name of subcutaneous pseudosarcomatous fibromatosis or fasciitis and has also been called nodular fibrositis, subcutaneous fibromatosis and infiltrative fasciitis.^{1 5 6}

Although NF is a benign idiopathic condition, it is frequently mistaken for a sarcomatous tumour owing to its rapid growth, rich cellularity and high mitotic activity.^{5–7}

Its precise incidence is unknown,⁶ but it is a rarely diagnosed entity^{8 9} especially in children—fewer than 20% of cases occur under the age of 20-years-old.^{1 3 10} It is more common in young adults between 20 and 40 years old,^{1–3 5 6} without gender preference.^{2 5 11 12} Lesions have been described in almost every sites, although the most common locations are the upper extremities (48%), especially the flexor surface of the forearm, the trunk (20%) and less frequently (10–20%) the head and neck.^{5 10 11} Remarkably, about 80–90% of all head and neck lesions occur in children.⁵

The aetiology of NF remains uncertain. Some believe that the unusual proliferation of myofibroblasts is triggered by local injury or inflammatory process.^{5 6 8 9 12} However, only a minority of cases (10–15%) report local trauma^{2 5 6 11} and it remains unclear why there are not more lesions in the inferior limbs, more prone to trauma in children.^{6 11}

NF's clinical presentation is rather unspecific; the most common presentation is a solitary, rapidly growing solid mass, with frequently associated pain and tenderness. Lesions can vary in size, from 0.5 cm to 10 cm, but most are <2–4 cm. Larger or multiple lesions are rare.^{3 10 11}

Imaging features can help characterise the lesion and the surrounding anatomy but are also unspecific and, most of the times, insufficient to make the correct diagnosis or differentiate it from malignant lesions.^{9 10 13} Ultrasound generally reveals a well circumscribed, isoechoic or hypoechoic heterogeneous mass, with substantial colour Doppler arterial flow. The CT scan shows a hyperdense mass, sometimes with invasion of the surrounding tissues. MRI findings vary according to the nature of the lesion (if myxoid or cellular).^{5 13} Since neither CT scan nor MRI are able to reach firm conclusion, only cytological or histological findings confirm the diagnosis of NF. FNAC, a minimally invasive procedure, can correctly identify NF (its reliability in distinguishing benign vs malignant mesenchymal lesions is 90–95%).¹⁴

NF accounts for 0.025% of all pathological diagnosis, with less than 4% of those occurring in children aged 0–9 years.^{1 2} Its cytomorphology is rather characteristic, making it possible to recognise these lesions in fine needle aspirates by an experienced pathologist.^{8 9} Histologically it is characterised by a myxoid stroma with high cellularity of immature appearing fibroblasts. These spindle shaped cells are arranged haphazardly and occasional normal mitosis and giant ganglion-like cells can be seen. There is abundant ground substance, capillaries, intermixed lymphoid cells (chronic infiltrate is often present at the periphery of the nodule) and lipid-laden macrophages.^{6 12} The immunohistochemical profile of NF can further help the differential diagnosis: cells are positive for vimentin, smooth muscle actin and KP1 (histiocyte marker)—consistent with dual myofibroblastic and fibrohistiocytic differentiation—and are negative for cytokeratin, S-100 protein and desmin.^{1 6 11 12}

Several reports support that the lesion is self-limited, with spontaneous regression in a few months (longest known duration is 26 months).^{8 12 15 16} However, because of the difficulty in differentiating NF from aggressive tumours, complete local surgical excision remains the standard treatment.^{1–3 5 8} Nonetheless, since FNAC can accurately differentiate NF from malignancy and considering the self-limited evolution of the disease, in cases where there is no compression or invasion of the surrounding structures, it may be considered a watchful waiting strategy after a cytological diagnosis, preventing further complementary examination or unnecessary treatments such as surgical excision.^{7 8 14 15} The prognosis is excellent and recurrence very rare.^{8 11 12 17}

In our case, the rapidly growing mass in the presence of fever and upper respiratory symptoms prompted reactive or infectious adenitis as our first diagnostic hypothesis, which was supported by the ultrasound image. However, the negative serologies for the most probable infectious causes and the unresponsiveness to the antibiotics excluded these diagnostics. Despite the fact that immunohistochemical analysis was not performed, our pathologist recognised the cytological pattern of NF, excluding the need to search for other aetiologies such as atypical mycobacteria and allowing for a conservative treatment. Spontaneous resolution provided objective evidence that the FNAC diagnosis was correct, thus avoiding unnecessary surgery.

This case report shows that when a rapidly growing mass occurs in children, differential diagnoses from malignancies need to be considered. It also highlights the importance of FNAC, a minimally invasive technique, which allows a fast diagnosis thus avoiding overtreatment with surgical resection.

Learning points

- ▶ Nodular fasciitis is a benign condition but its rapid growth makes the differential diagnosis with malignancy sometimes difficult.
- ▶ Laboratory and imaging exams may not be helpful.
- ▶ Histological or cytological findings make the diagnosis.
- ▶ Fine needle aspiration cytology is a minimally invasive procedure that allows cytological analysis.
- ▶ Conservative treatment should be considered after histological diagnosis.

Contributors SMA, HV and AB were responsible for the child during admittance to hospital and actively participated in the writing of this article. MHO was the pathologist responsible for the histological analysis and also actively contributed on the writing of the present article.

Competing interests None.

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REFERENCES

- 1 Lenyoun EH, Wu JK, Ebert B, *et al.* Rapidly growing nodular fasciitis in the cheek of an infant: case report of a rare presentation. *J Plast Surg* 2008;8:e30.
- 2 Cohen JL. Nodular fasciitis in the parotid region of a child. *Head Neck* 1998;645–8.
- 3 DiNardo LJ, Wetmore RF, Potts WP. Nodular fasciitis of the head and neck in children. A deceptive lesion. *Arch Otolaryngol Head Neck Surg* 1991;117:1001–2.
- 4 Konwaler BE, Keasberry L, Kaplan L. Subcutaneous pseudosarcomatous fibromatosis (fasciitis). *Am J Clin Pathol* 1955;25:241–52.
- 5 Roberti A, Roberti MRF, Carneiro SS, *et al.* Fasciíte Nodular em Região Cervical - Relato de caso. *Rev Para Med* 2007;21:41–4.
- 6 Zuber TJ, Finley JL. Nodular fasciitis. *South Med J* 1994;87:842–4.
- 7 Satish S, Shivalingaiah SC, Ravishankar S, *et al.* Fine needle aspiration cytology of pseudosarcomatous reactive lesions of soft tissues: a report of two cases. *J Cytol* 2012;29:264–6.
- 8 Marki K, Sharma J, Kaur H. Nodular fasciitis of the hand—a potential diagnostic pitfall in fine needle aspiration cytology. *J Cytol* 2007;24:197–8.
- 9 Fernando AF, Chua AH, Sia-Vargas LL. Nodular fasciitis in a one-year-old male: a diagnostic dilemma. *Philipp J Otorolaryngol-Head Neck Surg* 2010;25:27–31.
- 10 Kim ST, Kim JH, Baek JH, *et al.* Nodular fasciitis in the head and neck: CT and MR imaging findings. *Am J Neuroradiol* 2005;26:2617–23.
- 11 Haddad AJ, Avon SL, Clokie CML, *et al.* Nodular fasciitis in the oral cavity. *J Can Dent Assoc* 2001;67:664–7.
- 12 Cyriac MJ, Celine MI, Kurien G, *et al.* Nodular fasciitis. *Indian J Dermatol Venereol Leprol* 2004;70:239–41.
- 13 Nikolaidis P, Gabriel HA, Lamba AR, *et al.* Sonographic appearance of nodular fasciitis. *J Ultrasound Med* 2006;25:281–5.
- 14 Aydin O, Oztuna V, Polat A. Three cases of nodular fasciitis: primary diagnosis by fine needle aspiration cytology. Blackwell Science Ltd. *Cytopathology* 2001;12:345–7.
- 15 Kong CS, Cha I. Nodular fasciitis diagnosed by fine needle aspiration biopsy. *Acta Cytologica* 2004;48:473–7.
- 16 Wong NL, Di F. Pseudosarcomatous fasciitis and myositis—diagnosis by fine needle aspiration cytology. *Am J Clin Pathol* 2009;132:857–65.
- 17 Shimizu S, Hashimoto A, Enjoji M. Nodular fasciitis: an analysis of 250 patients. *Pathology* 1984;16:161–6.

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