

## Case Report

# A huge nodular fasciitis in parapharyngeal space in a 7-year-old girl: a case report and review of literature

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**Abstract:** Nodular fasciitis (NF) is a benign and reactive fibroblastic growth extending from the superficial fascia into the subcutaneous tissue or muscle, with a morbidity of less than 20% in children. We report a case of a 7-year-old girl presented with a 3-month history of snore and mouth breathing. Image findings demonstrated a large soft-tissue mass in the right parapharyngeal space. The lesion was successfully eradicated by surgical removal. Pathological analysis established NF as the final diagnosis. Histopathological findings were notable for a reactive spindle-cell process composed of proliferative fibroblasts with extravasated red blood cells and interstitial edema. Immunohistochemical stains showed that the lesional cells were positive for smooth muscle actin (SMA), muscle-specific actin (HHF35), and epithelial membrane antigen (EMA), and negative for S100 protein. No clinical evidence of recurrence was noticed after 2 months of follow-up. Being the first report of NF in the parapharyngeal space of a child, this rare pediatric case points out the importance for otolaryngologists to keep NF in mind for differential diagnosis to avoid unnecessary wide resection.

**Keywords:** Nodular fasciitis, parapharyngeal space, children, solitary fibrous tumor

## Introduction

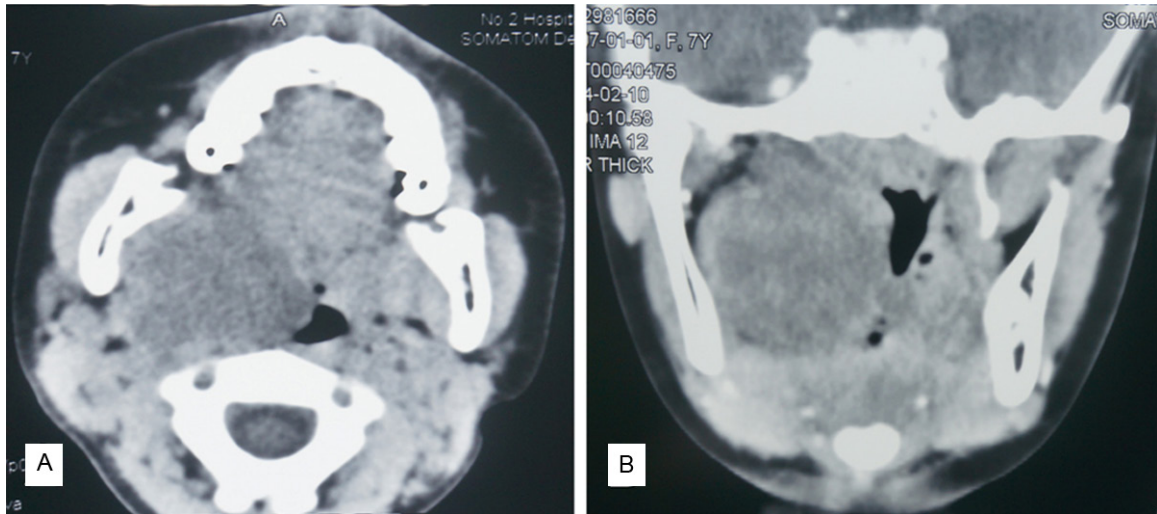
Defined by the World Health Organization as a benign and reactive fibroblastic growth extending from the superficial fascia into the subcutaneous tissue or muscle [1], nodular fasciitis (NF) was first described in 1955 by Konwaler et al. [2] who called the lesion subcutaneous pseudosarcomatous fibromatosis. The lesion commonly locates on the extremities, occasionally on the trunk, and infrequently on the head and neck in adults aging from 20-40 years, which is seldom larger than 3 cm [2, 3]. As far as we know, pediatric cases have been rarely reported in the literature. In this report, we present an unusual presentation of a NF in the parapharyngeal space on a 7-year-old girl.

## Materials and methods

A 7-year-old girl was referred to the Otolaryngology-head and Neck Surgery Clinic of the Second Xiangya Hospital, complaining of a 3-month history of snore and mouth breathing.

Physical examination disclosed a nodular mass with defined borders in the right pharynx. Measuring 4.0 × 3.5 cm, the mass was sessile, firm and immobile, and exceeded the median line. Tenderness existed, and the uvula was to the left. Enlarged bilateral lymphoglandulae submaxillares and left lymphoglandulae cervicales profundae superiores were presented with a diameter of 1-2 cm and no obvious tenderness. There was no history of trauma or inflammation in the region, and the medical history was noncontributory.

During the hospital stays, relevant examinations were performed on this patient. An electrocardiogram and chest X-ray were unremarkable. Complete blood count, coagulant function and serum electrolytes serology were normal. A computed tomography (CT) scan showed an oval soft-tissue mass measuring 4.8 cm in the right paranasopharynx, and projected into the nasopharyngeal cavity obviously with a poorly defined boundary. Bone destruction was not discovered (**Figure 1**). Magnetic resonance



**Figure 1.** A. Axial CT scan: Axial CT scans showed an oval soft-tissue mass measuring 4.8 cm in the right paranasopharynx, and projected into the nasopharyngeal cavity obviously with a poorly defined boundary. B. Coronal CT scan: Coronal CT scans demonstrated an oval soft-tissue mass measuring 4.8 cm in the right paranasopharynx, and projected into the nasopharyngeal cavity obviously with a poorly defined boundary.

imaging (MRI) demonstrated an irregular-shaped soft-tissue mass in the right parapharyngeal space with a relative defined boundary. The mass was isointense on T1-weighted images, hyperintense on T2-weighted images, and was significantly enhanced with a size of  $4.7 \times 2.6 \times 5$  cm. Enlarged lymph nodes were observed in the right parapharynx and bilateral cervical regions (**Figure 2**). This case report was approved by the ethics committee of The Second Xiangya Hospital of Central South University. Before the survey, the patient's guardian was asked to sign an informed consent to identify their willingness to take part in this study and to ensure their rights of voluntary participation, knowing, as well as privacy.

## Results

The child was scheduled for resection of the lesion because of its continued proliferation. Under endoscopy, the lesion was resected by direct trans-soft palate approach through the oropharynx. The mass was smooth, hard, and fixed, and projected into the nasopharyngeal cavity. The huge mass measured  $4.5 \times 5.0$  cm, and the upper bound of it reached pharynx nasalis, the lower bound reached epiglottis, and it extended to parapharynx and internal carotid artery bilaterally.

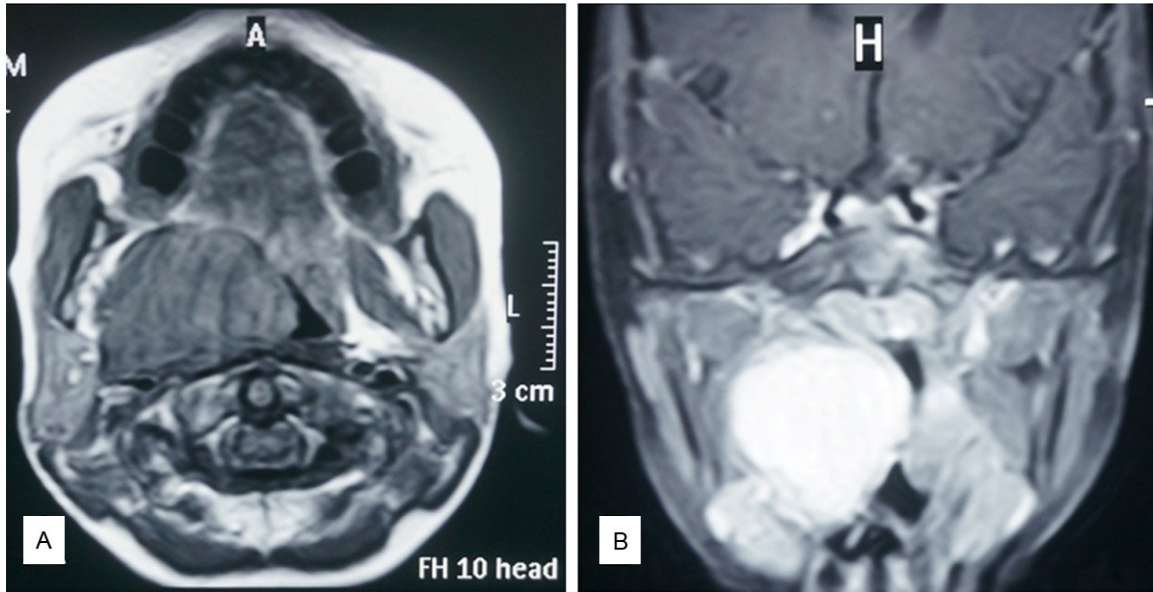
Histopathological findings were notable for a reactive spindle-cell process composed of pro-

liferative fibroblasts with extravasated red blood cells and interstitial edema (**Figure 3**). Immunohistochemical reactions were performed, showing that the proliferative cells were negative for desmin, bcl-2, CD34, CK, and S100, and positive for smooth muscle actin (SMA), muscle-specific actin (HHF35), and vimentin, and slightly positive for Ki-67 (**Figure 4**). These findings were consistent with the diagnosis of NF. After 2 months of follow-up, the patient showed no clinical evidence of recurrence.

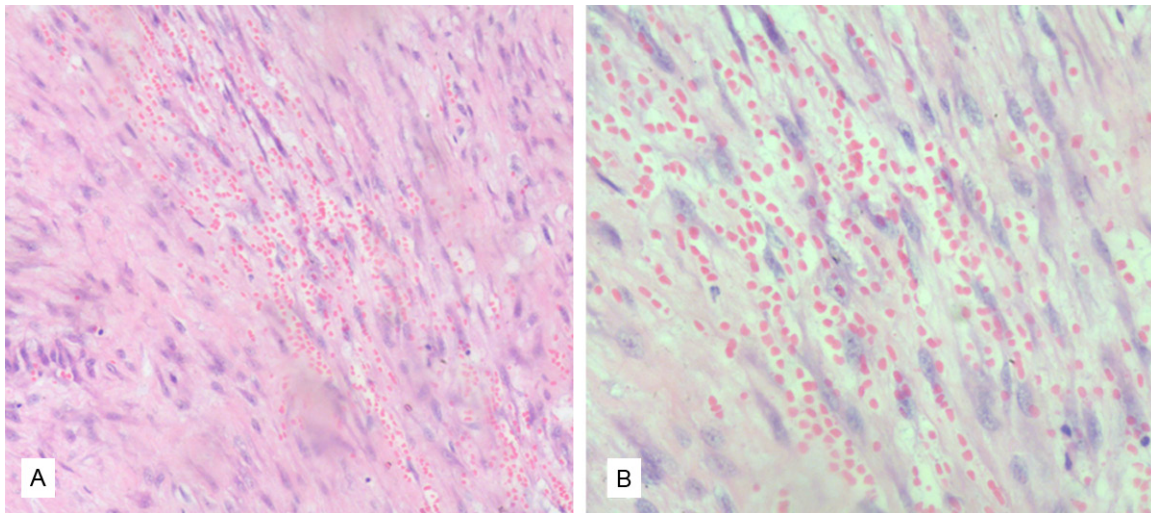
## Discussion

NF is believed to occur as a benign fibroblastic proliferative and reactive process of the soft tissues related to fascia, mainly triggered by local injuries or inflammatory processes. NF can be divided into subcutaneous, intramuscular, and fascial types depending on its relationship to anatomic location [4]. Lesions in NF could be separated into three types based on a range of histological features: myxoid, cellular, and fibrous, which are roughly correlated with the duration of the nodule [5].

NF is extremely uncommon in children. It accounts for 0.025% of all pathologic diagnoses, with less than 4% of those cases occurring in children aged 0-9 years old [6]. NF is equally distributed between genders and most commonly presents in the third to fifth decades of



**Figure 2.** A. Axial T1-weighted MRI: Axial T1-weighted MRI showed an isointense soft-tissue mass in the right parapharyngeal space with a relative defined boundary. B. Coronal T2-weighted MRI: Coronal T2-weighted MRI demonstrated a hyperintense soft-tissue mass in the right parapharyngeal space. Enlarged lymph nodes were observed in the right parapharynx and bilateral cervical regions.



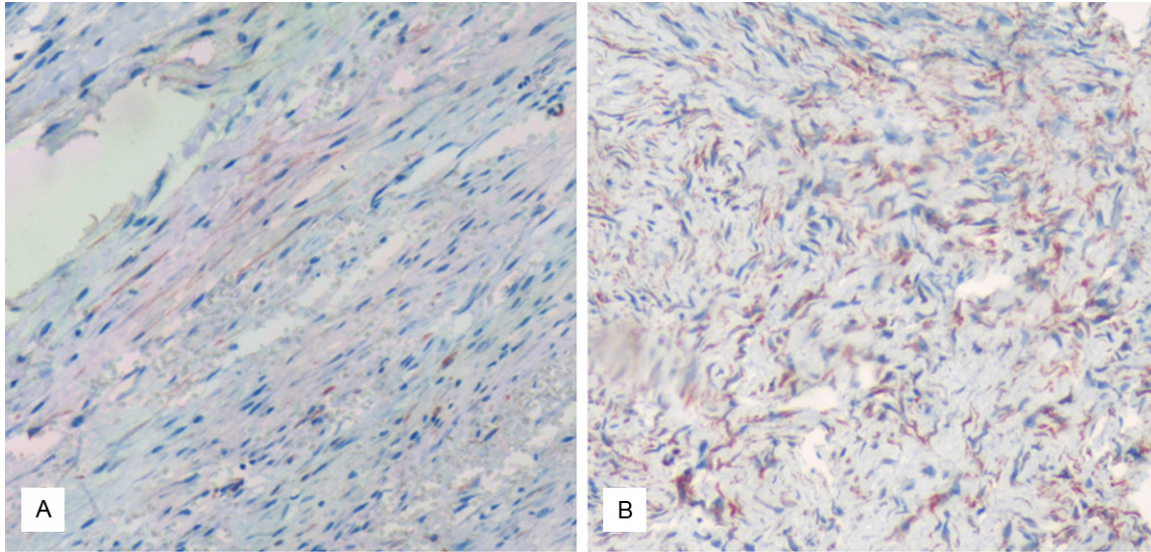
**Figure 3.** Histopathology: Proliferative fibroblasts with extravasated red blood cells and interstitial edema. Original magnification: A  $\times 100$ , B  $\times 200$ .

life [2, 3]. When seen in children, it may be confused with other more common soft tissue tumors such as fibroma, lipoma, desmoids tumor, sarcoma, chondroma, myxoma, malignant fibrous histiocytoma, schwannoma, atypical fibroxanthoma, or parotid tumor [1, 7-10]. Meanwhile, lesions are usually small, and are commonly located on extremities [2, 3]. Khuu et al. [11] reported an average diameter of 2.6 cm ranging from 1.8-3.5 cm in six pathologi-

cally confirmed cases of NF. In our case, the huge mass measured 4.5  $\times$  5.0 cm. To our knowledge, this is the first reported case of NF occurs in children's parapharyngeal space with an extremely large size.

The pathogenesis of NF remains unknown. However, it seems to be reactive or inflammatory, involving fibroblastic or myofibroblastic proliferation, rather than a true neoplasm [4].





**Figure 4.** A. Immunohistochemical reactions were positive for SMA. B. Immunohistochemical reactions were positive for vimentin.

Some investigators have suggested that local trauma could trigger myofibroblast proliferation [12]. Another hypothesis, based on the observation of some cases of pregnant and lactating women presenting with NF, proposes that stimulation of estrogen receptors in myofibroblasts could be a contributing factor to the proliferation of those cells [12, 13].

Image is helpful to guide diagnosis of NF. On Ultrasonography, the mass is often hypoechoic [11]. While on MRI, the mass is isointense to muscle on T1-weighted sequences, hyperintense to muscle on fluid-sensitive sequences, and enhanced avidly but heterogeneously [11]. In addition, on  $^{18}\text{F}$ -fluorodeoxyglucose positron emission tomography ( $^{18}\text{F}$ -FDG PET), the lesion is visible on CT while having only low uptake on the PET images, rendering malignancy less probable [14].

The diagnosis is based on the typical histopathological findings as it is difficult to arrive at a correct diagnosis clinically due to lack of clear cut clinical features. The nodule consists of numerous whorls of pleomorphic fibroblasts growing haphazardly in the stroma that is often highly vascular and contains varying amounts of mucoid substance, reticulum and collagen fibers [9]. Usually the lesion is well delimited, but non-encapsulated, and abundant mitotic activity can be seen with no cellular atypia [8-13]. Besides, some lesions display multinucleated giant cells [13]. A scattered chronic

inflammatory infiltrate and erythrocytes are frequently evident within the background [10].

The immunohistochemical profile of NF is consistent. Lesions reliably stain for HHF35, SMA, and vimentin, indicating myofibroblast or fibroblast proliferation; while they do not stain for S100, desmin, and keratin [7-13]. Immunostaining of CD34, CD56, CD99, and bcl-2 aids in excluding solitary tumors. Moreover, CD68 can be applied to confirm the presence of giant cells [13]. Ki-67 has been suggested to assess the proliferative activity of NF [1, 12]. Both high and low proliferative activities have been reported by Ki-67 staining [1, 12]. In present case, slight staining was marked by the Ki-67, showing a low amount of cell proliferation. The findings of the present case corroborate the general pathologic characteristics reported previously, suggesting a diagnosis of NF.

Given the benign nature of NF, local resection is usually curative. Besides, response to intralesional steroid injections has also been reported [15]. Meanwhile, prognosis upon resection is satisfactory with a rare recurrence rate quoting as 0.4-1.0% [3, 5]. In fact, the development of a recurrence likely indicates an incorrect preliminary diagnosis rather than a true recurrence [5].

In conclusion, one should bear a suspicion to diagnose with NF when a rapidly growing nodule occurs. Histopathologic and immunohisto-

chemical examinations play an essential role in differentiation NF from other soft tissue masses. Once the diagnosis of NF is confirmed, the only treatment option is complete surgical excision of the lesion.

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## Disclosure of conflict of interest

None.

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