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Lifethreatening tracheal obstruction in a child caused by nodular fasciitis: case presentation

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Abstract

Background: Nodular fasciitis (NF) is a rare non-neoplastic inflammatory tumor and usually presents as a painless, rapidly growing subcutaneous soft tissue mass. The head and neck are relatively common locations for nodular fasciitis, particularly in children. NF in the trachea is rare and may evolve to a fatal condition, especially due to its rapidly growing nature that can cause life-threatening tracheal obstruction.

Case presentation: We report the case of a 5-year-old child with NF in the trachea and subtotal tracheal obstruction. Bronchoscopy and biopsy proved the diagnosis, and bronchoscopic excision of the tumor was performed.

Conclusions: NF is a rare airway tumor, occurring mostly in adults, and may presenting with pneumonia-like symptoms. Early detection of the lesion is essential to avoid life-threatening airway obstruction.

Keywords: Nodular fasciitis, Tracheal obstruction, Subcutaneous pseudosarcomatous fibromatosis

Background

NF is characterized as a non-malignant, proliferative reactive lesion containing fibroblasts and myofibroblasts [1]. It can generally be found in the extremities and in the trunk. NF prevalence is 13–20% in the head and neck area. The precise causes of the lesion are unclear at the time [2]. The research clearly shows that prior trauma is a critical etiological element, but it is not often depicted in studies. It represents as a solid mass 1–5 ml in diameter. It may even spread through the tissue under the skin and muscles. Its occurrence rate is increasing, and is often misdiagnosed as malignant. NF needs to be diagnosed early [3].

Case presentation

We report the case of a 5-year-old boy who was referred to our emergency department with severe respiratory distress, and the patient was free of srtidor. Four months before presentation, the child had signs of a respiratory infection, interpreted as pneumonia. He had been admitted to the pediatric department three times. His mother confirmed that he had not inhaled a foreign body, and there is also no history of hemoptysis.

Nevertheless, a computed tomography (CT) scan was performed to exclude a radiolucent foreign body. It showed a space-occupying lesion in the trachea 3.2 cm above the bifurcation measuring 4 × 1.6 cm with a pneumonic consolidation in the right lower lobe (Fig. 1).

The patient was referred to our cardiothoracic surgery department and prepared for urgent rigid bronchoscopy performed under general anesthesia. It revealed a solitary soft tissue mass in the trachea with a subtotal obstruction. The mass appeared to be white, soft, circumscribed, vulnerable, and obstructed nearly the whole lumen of the trachea, leaving part of the lumen free. As a result of bleeding during the rigid bronchoscopy, the trachea was totally occluded.

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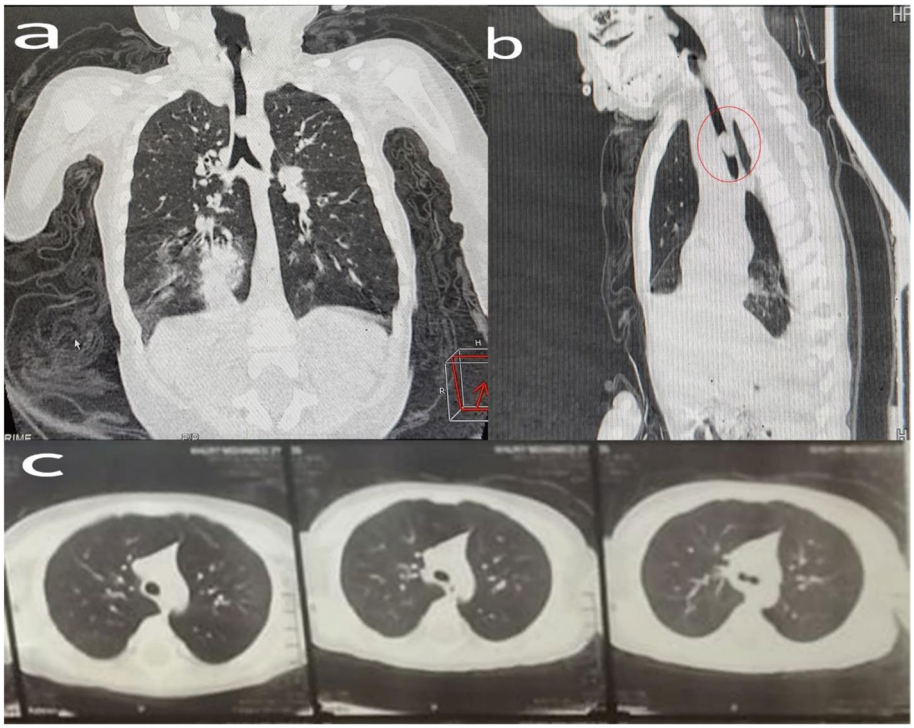

Fig. 1 CT scan showing mass in the trachea with a pneumonic patch in the RLL. **a** Coronal view **b** Sagittal view. **c** 6 months follow-up CT chest (cross-sectional view)

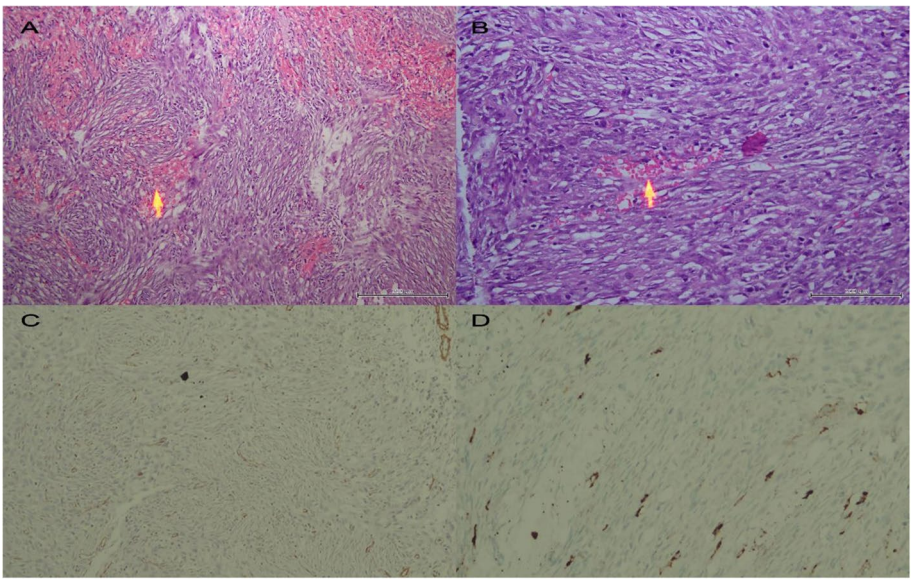

Fig. 2 **A**, **B** Cross section of soft tissue mass sample with different power of magnifications fibroblastic spindle cells arranged in a fascicular pattern. **C** SMA positive immunostaining of cells of nodular fasciitis. Magnification = × 100. **D** CD10 positive immunostaining of cells of nodular fasciitis. Magnification = ×200

This caused a hypoxic cardiac arrest for the duration of 1 min. The bleeding was controlled after applying a topical vasoconstrictor, and debulking of the tumor was performed, and a biopsy was sent for histopathological examination. The patient was transferred to the pediatric intensive care unit (PICU), and steroid therapy was administered. Histopathologic examination revealed fibroblastic spindle cells arranged in a fascicular pattern with scattered inflammatory cells, without malignant cells, SMA and CD_{10} stains were positive, confirming NF, while P_{53} was negative (Fig. 2).

The patient underwent rigid bronchoscopy under general anesthesia. The electrocautery was used to excise the tumor. The thermal effect degenerates proteins, thus inducing tissue necrosis. Thereafter, the necrotic tissue could be easily removed using a biopsy forceps. The postoperative period was uneventful and he stayed in the hospital for 3 days. The patient was followed six months after surgery with CT chest, which revealed no local recurrence of the tumor (Fig. 1).

Discussion

NF is a benign soft tissue inflammatory tumor composed of fibroblastic cells with myofibroblastic proliferation. It is also known as subcutaneous pseudosarcomatous fibromatosis [1]. The etiology is still unclear, but it might be caused by unusual myofibroblast proliferation preceded by an inflammatory proces, infection, or local injury [4]. Although NF may occur in patients of any age, it is most common in adults (20 to 40 years). Only 14% of the patients are < 10 or > 60 years old. The sex distribution is equal [5]. Although it may occur virtually anywhere on the body, most of the cases present with superficial soft tissue masses in the extremities or trunk.

Fabre et al. reported a high recurrence rate of pulmonary inflammatory pseudotumors that ranged from 18 to 40% and concluded that complete surgical resection had excellent long-term results [6].

The preferred treatment for NF is local excision. Recurrence is rare if totally excised as it has a good prognosis and metastases do not occur. Rare spontaneous regression and rapid resolution with intralesional corticosteroid injection in lesions that cannot be excised have been reported [7].

Conclusion

In conclusion, NF is a rare airway tumor, occurring mostly in adults, and may presenting with pneumonia-like symptoms. Early detection of the lesion is essential to avoid life-threatening airway obstruction. Also, the complete surgical resection with termino-terminal anastomosis is the gold standard of treatment if feasible.

Abbreviations

CT: Computed tomography; NF: Nodular fasciitis; PICU: Pediatric intensive care unit; SMA: Smooth muscle actin.

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Authors' contributions

YS: writing, editing. MK: reviewing. ME: writing. AM: writing, editing. All authors read and approved the final manuscript.

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Availability of data and materials

The datasets used or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

This study protocol conforms to the ethical standards of the Helsinki Declaration and approval was obtained from Qena University Hospital, Faculty of Medicine, South Valley University. Committee's reference number is not available. Informed written consent to participate is obtained.

Consent for publication

Appropriate written informed consent was obtained for publication of this case report and accompanying image from patient parent.

Competing interests

The authors declare that they have no competing interests.

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