Intratracheal Nodular Fasciitis: a Rare Lesion Case Report

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Research

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Abstract

Nodular fasciitis is a benign proliferation of myofibroblasts that usually arises in subcutaneous tissues of the trunk, neck, head, and upper extremities of young to middle-aged adults. It is not reported to arise in the intratracheal. We present a patient with chest tightness and asthma for three months. Chest computed tomography showed no displayed of left bronchus. The trachea of the left main bronchus was blocked by organisms under bronchoscope. The organisms were extracted and pathologically diagnosed as a rare, benign, intratracheal nodular fasciitis. One month later, the patient relapsed again and underwent resection.

Introduction

Nodular fasciitis (NF) is a benign myofibroblastic proliferation that usually found in the subcutaneous tissue of young to middle-aged adults [1]. NF commonly appears rapid growth. The most common sites are the upper limbs[2], trunk[3], head[4] and neck[5]. Sometimes, such as salivary glands[6], breast[7], mesentery[8], and vulva[9] may be involved where were unusual locations. There have been no reports of it in the trachea. We report a case in which nodular fasciitis grew fast in the patient's left main bronchus and recurred in the same site about one month after surgical resection.

Case Report

A 52-year-old man presented with a history of chest tightness and asthma for three months. Physical examination showed left lung breathing sound disappeared without other positive signs. There was nothing special in past history of the patient.

Radiology findings

The patient received chest computed tomography(CT) at outpatient department which revealed the trachea of the left main bronchus was blocked and atelectasis was found in the left lung. (Fig. 1A.B) Three-dimensional reconstruction of trachea also showed the left main bronchus disappeared from the main bronchus divided into about 1 cm behind the left main bronchus. (Fig. 1C)

Bronchoscope findings

The patient underwent fiberoptic bronchoscopy after hospitalization under general anesthesia. At the opening of the left main bronchus, fish like new organisms can be seen to block the lumen to complete occlusion. The new organism had abundant blood supply and was prone to bleeding and was derived from distal lateral wall of left main bronchus. (Fig. 2) The new organisms were removed by electric snare cutting and cryotherapy and was submitted for pathological examination. The operation was finished after high-frequency electric knife and electrocoagulation hemostasis treatment at the lesion. The patient received chest CT again after operation and image showed the left bronchus was open. (Fig. 3A-C)

Pathology findings

On microscopic examination proliferative spindle cells with infiltration of myxoid stroma and inflammatory cells were seen. Immunohistochemistry demonstrated that the cells were positive in patches for SMA, and negative for Desmin, CD34 in lesion cells. Ki-67 stained about 10% of cells. (Fig. 4A-F) Fluorescence in situ hybridization (FISH) showed USP6(17p13) translocation and no translocation in ALK. Nodular fasciitis was considered in combination with immunohistochemical molecular pathological results.

Follow-up

The symptoms of chest tightness and asthma were relieved after operation. Re-examination of chest CT showed that the left lung recovered ventilation. The patient had chest tightness and asthma again 1 month after operation and received treatment under tracheoscopy. The results of pathological examination after new biological resection showed that nodular fasciitis. The patient went to the clinic for local radiotherapy and the radiation dose was 30Gy. No new organism was found again in the patient's left main bronchus until the paper was written.

Discussion

Nodular fasciitis is a benign self-limited process, which was firstly reported by Konwaller et al in 1955[10]. It was previously thought that the inducement had a history of trauma while many were not. The definite etiology of NF is still uncertain. A large part of NF arises in the subcutaneous tissue and underlying fascia and grows rapidly. There is no relevant report about NF occurring in intratracheal at present. In 2011, Erickson-Johnson et al found high expression levels of USP6 mRNA in 2 examples of nodular fasciitis. USP6 belongs to a subfamily of deubiquitinating enzymes that may serve as oncogenes. A recurrent MYH9-USP6 fusion gene was discovered by molecular cytogenetic and indicated that NF may be a clonal neoplastic disorder[11]. USP6 translocation can be found in our cases.

Recently, Pateletal used an anchored multiplex polymerase chain reaction (PCR) assay to demonstrate that in nodular fasciitis, USP6 not only fuses to MYH9, but also a myriad of other partners (RRBP1, CALU, CTNNB1, MIR22HG, SPARC, THBS2, and COL6A2), all of which are promoter swaps that increase USP6 expression[12]. USP6 rearrangements are not specific for nodular fasciitis. It can also be found in cellular fibromas of tendon sheath[13] and aneurysmal bone cyst[14]. While the classic histology of each is unique and should consider morphologic distinction. The diagnostic should combine the clinical performance with the pathology. NF happened in such as subcutaneous tissue, muscles and intraarticular had different imaging findings.

The endobronchial tumors are divided into primary and secondary according to the source[15]. The pathological manifestations of bronchial tumors originating from different parts are also different. Endobronchial tumors are seldom seen in clinic. It is difficult to diagnose endobronchial tumors because they often has no characteristic symptoms and radiographic findings are also no-specific. CT

examination provides limited differential diagnosis. The awareness of NF that occurs in the bronchial is of great importance because this disease resembles endobronchial tumors in terms of its clinical features and imaging features despite its benign nature.

Pathological features of lesions are consisted by proliferation of spindle cells that should be included in differential diagnosis of NF. Especially, spindle cell carcinoma and sarcoma among malignant lesions, and fibromatosis and myofibroblastoma among benign lesions, morphologically resemble NF and need to be differentiated[16]. The clinical features and imaging results of the patient in this case were helpless to the diagnosis. NF has wide range of clinical manifestations and imaging results due to its' locations. It is very important to combine clinical effect, imaging features and pathological results when diagnosis needs to be made.

Conclusion

We reported a rare case of nodular fasciitis of the intratracheal which not yet seen before. The new organism grew fast during 1 month and blocked bronchus again after surgery. Local radiotherapy prevented the growth of the NF. The detection of the USP6 fusion gene supported a diagnosis of nodular fasciitis.

Declarations

Ethics approval and consent to participate

The data of the study population were obtained from the databases in our institution and the ethics has been approved by the Medical Ethics Committee of Nanjing Drum Tower Hospital, Medical School of Nanjing University (2019-011-11).

Consent for publication

We have asked for the patient's consent and signed the consent form.

Competing interests

The authors declare that they have no competing interests.

Availability of Data and materials

Please contact author for data requests.

Disclosure

All authors have and declare that: (i) no support, financial or otherwise, has been received from any organization that may have an interest in the submitted work ; and (ii) there are no other relationships or activities that could appear to have influenced the submitted work.

Authors' contribution

ZY collected data and wrote the manuscript. CMS and LXQ collected relevant references. MFQ analysed the results of pathology. ZX was responsible for the pictures in the page. CLL modified the manuscript. All authors read and approved the final manuscript.

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Figures

Fig. 1 Chest computed tomography(CT) at outpatient department. a. Coronal position indicated left atelectasis. b. Horizontal position indicates left atelectasis. c. Tracheal reconstruction suggested left main bronchial obstruction.

Figure 1

See image above for figure legend.

Fig. 2 New organisms removed by tracheoscopy.

Figure 2

See image above for figure legend.

Fig. 3 Chest computed tomography(CT) after operation. a. Coronal position indicated left lung recovered ventilation. b. Horizontal position indicates left lung recovered ventilation. c. Tracheal reconstruction suggested left main bronchial obstruction disappeared.

Figure 3

See image above for figure legend.

Fig. 4 The pathological findings of the intratracheal lesion. a. Low-magnification (hematoxylin and eosin, original magnification $\times 40$) image. b. Low-magnification (hematoxylin and eosin, original magnification $\times 200$) image. c. Negative for CD34 (original magnification $\times 200$). d. Negative for DESMIN (original magnification $\times 200$). e. Ki-67 was 10% (original magnification $\times 200$). f. positive in patches for SMA (original magnification $\times 200$).

Figure 4

See image above for figure legend.