Received: 5 May 2021 | Accepted: 27 July 2021

DOI: 10.1111/1759-7714.14104

CASE REPORT

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First report of an adult female patient with endobronchial inflammatory myofibroblastic tumor in Taiwan: A case report

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Abstract

An inflammatory myofibroblastic tumor (IMT) of the respiratory system is an uncommon disease. In Taiwan, there is a lack of previous studies on tracheobronchial IMT. The tumor is characterized by overexpression of anaplastic lymphoma receptor tyrosine kinase (ALK)-1. Surgical resection is the standard treatment of choice nowadays.

KEYWORDS

endobronchial tumor, inflammatory myofibroblastic tumor, lung collapse

INTRODUCTION

An inflammatory myofibroblastic tumor (IMT) of the respiratory system is a rare disease. It is difficult to confirm its prevalence because a definition is still evolving.[1] IMTs account for about 0.04% to 1% of all lung lesions.[2] It is also the most common primary lung lesion in children. Tracheobronchial IMTs are rare and have been reported in children and young adults.

About 30% of individuals with IMT of the respiratory system have cough, dyspnea, chest pain, or hemoptysis. Endobronchial IMT accounts for less than 5% of them. IMT was once thought to be a kind of inflammatory pseudotumor but it is now considered a neoplasm with myofibroblastic differentiation plus anaplastic lymphoma receptor tyrosine kinase (ALK)-1 overexpression.[3] This implies that IMT is a neoplasm mostly driven by kinase fusion.

Here, we report the first case in Taiwan of an endobronchial IMT in a middle-aged adult female patient who presented with breathlessness.

CASE REPORT

A 54-year-old woman presented with a history of dyspnea of two months duration. Auscultation confirmed bronchial breath sounds. Chest X-ray showed decreased left lung volume with mediastinal retraction. High-resolution computed tomography (HRCT) revealed a mass in the left main bronchus. Tumor markers were checked (CEA < 0.5 ng/ml and SCC = 0.70 ng/ml).

Flexible bronchoscopy revealed a left main bronchus tumor located 0.5–1 cm below the main carina which easily bled on manipulation (Figure 1). Rigid bronchoscopy was performed for tumor debulking the following day. Pathology indicated an inflammatory myofibroblastic tumor. The 18F-fluorodeoxyglucose positron emission tomography (FDG PET) disclosed neither nodal nor distant metastases.

The specimen showed a subepithelial nodule of spindle cells with minimally nuclear atypia, scattered lymphocytes, some mitoses, and focal myxoid stroma. Immunohistochemistry (IHC) study disclosed: the spindle cells are

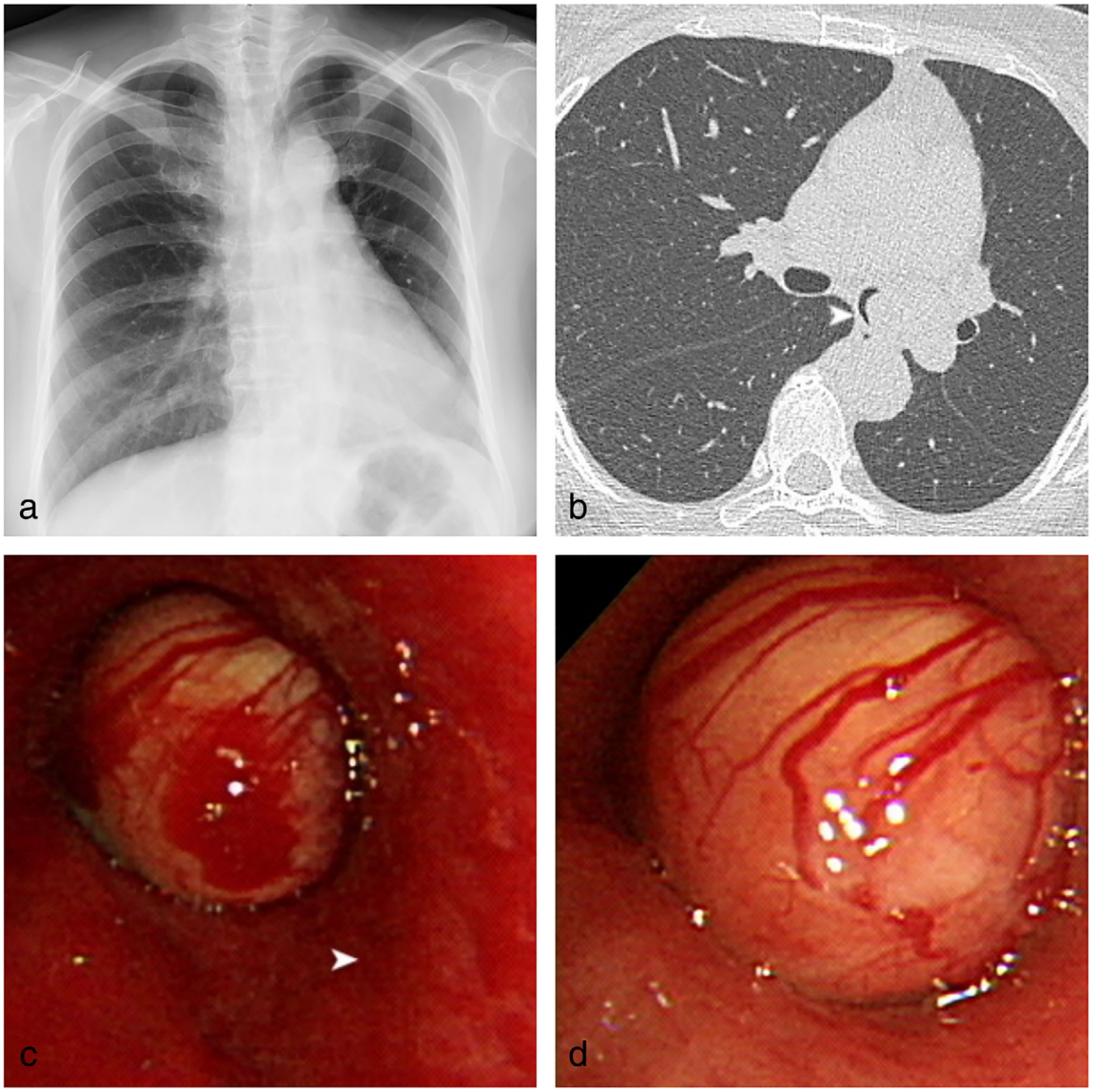

FIGURE 1 (a) Chest X-ray of the patient, (b) left endobronchial tumor (arrow) under HRCT and (c) endobronchial tumor under bronchoscopy. A left main bronchial tumor was located 0.5–1 cm below the main carina (arrow); (d) Close-up view of the endobronchial tumor

positive for ALK; focally positive for SMA, desmin (D33), and EMA(E29)(Figure 2(a)–(d)); negative for cytokeratin, CAM5.2, ER, PR, TTF-1, HMB 45, Melan-A, S-100, calponin, STAT6, and TLE1. The diagnosis was confirmed.

This patient recovered smoothly after tumor removal without complication. There was neither dyspnea nor tumor recurrence in a serial outpatient follow-up.

DISCUSSION

This is the first report of an adult female patient with an endobronchial inflammatory myofibroblastic tumor in Taiwan. IMT of the lung is usually presented as a solitary, well-defined mass on a chest X-ray. There is no specific radiographic feature on CT scan to differentiate IMT from other pulmonary lesions. FDG-PET scans may also show increased uptakes.[4]

IMTs are characterized by chromosomal translocations leading to activation of the ALK tyrosine kinase in approximately one-half of reported cases.[5] In a study using next-generation sequencing (NGS), 67% (6 of 9) of ALK-negative IMT tumors present fusions involving ROS-1 or PDGFRβ genes. This suggested that IMTs are mainly kinase fusion-driven tumors.[2]

The typical pathological finding of IMT is polymorphic mononuclear lymphocyte infiltration on a spindle cell proliferation background. These spindle cells are often in

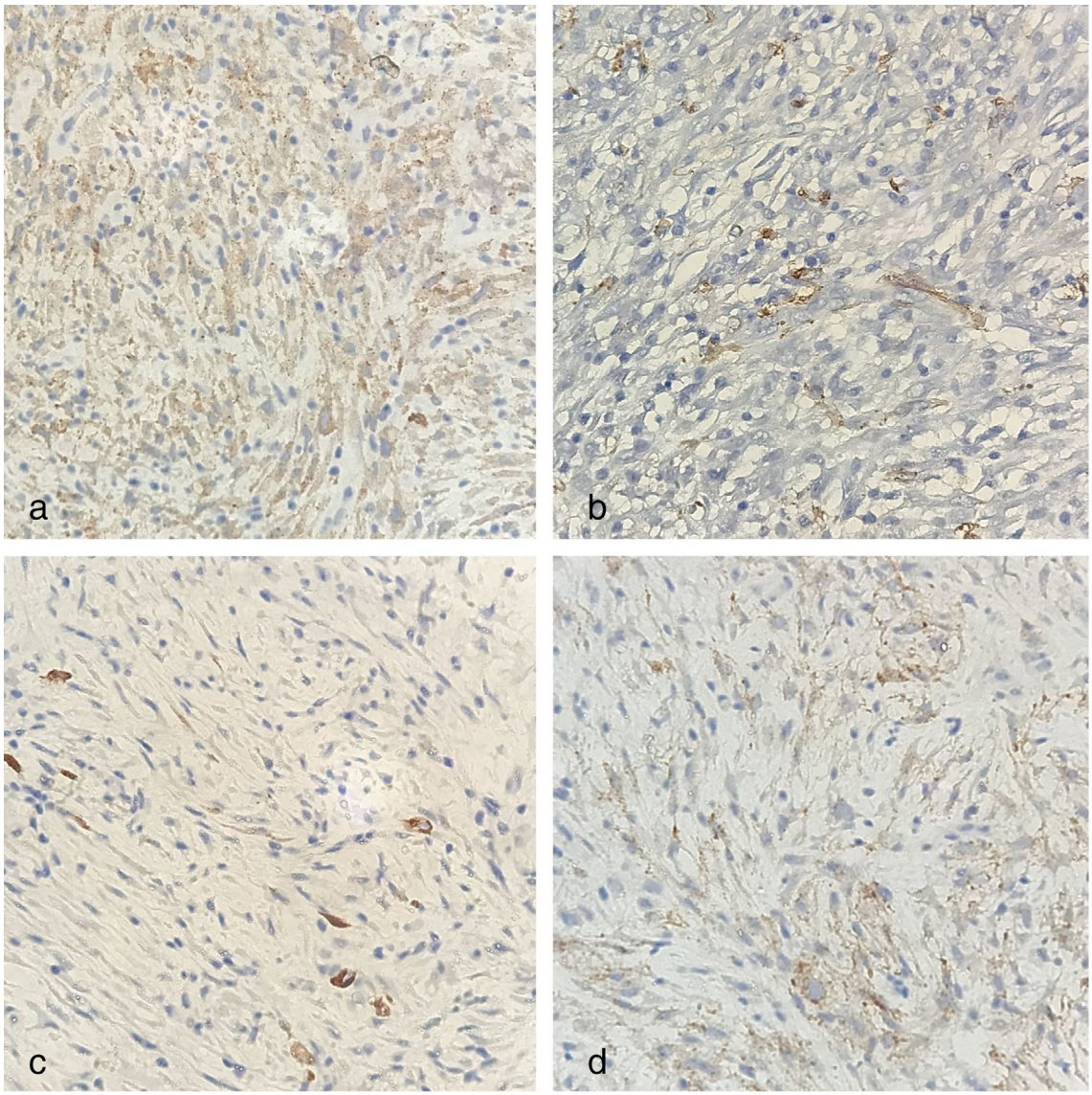

FIGURE 2 (a) Positive for ALK; (b)–(d) focally positive for SMA, desmin, and EMA

short fascicles with a storiform (cartwheel-like) architecture. Immunohistochemical studies of these spindle cells present fibroblasts as well as myofibroblasts in different proportions.

Surgical resection is the optimal choice in the treatment of IMTs.[3,6] The operation may be performed by open thoracotomy, video-assisted thoracoscopy surgery, or bronchoscopy. A frozen section or biopsy will help decide the range of excision, which is usually minimized to preserve pulmonary function. Chemotherapy and radiation are not considered primary treatments.[7] A 5-year survival rate of up to 91% has been reported in patients with IMT who have undergone complete resection.[8] Although recurrence of IMT is rare in those who have undergone complete tumor resection, it is up to 60% in those who have experienced incomplete resection.[8]

CONFLICT OF INTEREST

None of the authors have any conflict of interest to declare.

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How to cite this article: Lin Y-T, Huang S-H, Chang C-H, Hsu P-C, Wang C-W, Lee C-S. First report of an adult female patient with endobronchial inflammatory myofibroblastic tumor in Taiwan: A case report. Thorac Cancer. 2021;12:2807–10. https://doi.org/10.1111/1759-7714.14104