

Endotracheal inflammatory myofibroblastic tumour: A rare cause of central airway occlusion in adults

Albert Iruthiaraj Lourdesamy Anthony¹  | Taranjit Kaur Satnam Singh² |
Khai Lip Ng³  | Jamalul Azizi Abdul Rahaman⁴

¹Respiratory Unit, Hospital Taiping, Taiping, Malaysia

²Department of Radiology, Hospital Taiping, Taiping, Malaysia

³Department of Medicine, Hospital Melaka, Melaka, Malaysia

⁴Department of Pulmonology, Hospital Serdang, Kajang, Malaysia

Correspondence

Albert Iruthiaraj Lourdesamy Anthony,
Respiratory Unit, Hospital Taiping, Jalan Taming Sari, 34000 Taiping, Perak, Malaysia.
Email: albert5409@yahoo.com

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Abstract

Inflammatory myofibroblastic tumour (IMT) is an unusual myofibroblastic spindle cell neoplasm that is rarely discovered in the airway of adults. Previously, it was regarded as a reactive lesion and was infamously known as inflammatory pseudotumour before recent insights revealed that significant majority of cases harboured neoplastic genes. Diagnosis is difficult as clinical presentation and imaging findings are non-specific. Diagnosis and a favourable prognosis require the complete resection of the tumour. Detection of the anaplastic lymphoma kinase expression via immunohistochemistry expedites diagnosis. We report a young adult with an endotracheal mass occluding the central airway. The patient successfully underwent bronchoscopic resection using interventional techniques. IMT was diagnosed. No recurrence was seen after a year of surveillance. Due to the rarity of the disease, the recurrence rates for large airway disease is unknown. Recurrence rates for pulmonary lesions is reported to be lower compared to extrapulmonary IMTs and recurrence is unlikely if complete surgical excision is achieved.

KEYWORDS

anaplastic lymphoma kinase, central airway, endotracheal, inflammatory myofibroblastic tumour

INTRODUCTION

Inflammatory myofibroblastic tumour (IMT) is an uncommon mesenchymal neoplasm characterized by myofibroblastic spindle cell proliferation with a variable non-specific inflammatory cell infiltration mainly consisting of lymphocytes, plasma cells and eosinophils. Pulmonary IMT was first reported in 1939 and constitutes less than 1% of all adult respiratory tract neoplasms.^{1,2} Although it may commonly occur in the lungs of children and adolescents, pulmonary and endotracheal IMTs are extremely rare in adults.

In the past, IMT was regarded as a benign reactive lesion and a subset of inflammatory pseudotumour with interchangeable diverse nomenclatures which include plasma cell granuloma, xanthogranuloma and fibrous histiocytoma.^{3,4} Although the exact aetiology and pathophysiology of IMT remain unclear, recent insights revealed that 50%–70% of

IMTs harbour genes and chromosomal abnormalities which encode mitogen-activated protein kinase pathways.⁵

The diagnosis and prognosis are heavily reliant on the achievement of complete surgical excision. Histopathological diagnosis is often very challenging and requires excision of the tumour. Complete surgical resection when possible ensures good survival and low recurrence rates.⁶ Bronchoscopic techniques are minimally invasive and have the advantage to resect lesions localized in the airways without compromising lung function.⁷ We report a case of endotracheal IMT resected using diathermy snaring and debulking via rigid bronchoscopy in a young adult.

CASE REPORT

A 26-year-old man with no comorbidities presented with haemoptysis and dysphagia for 2 years. His haemoptysis was

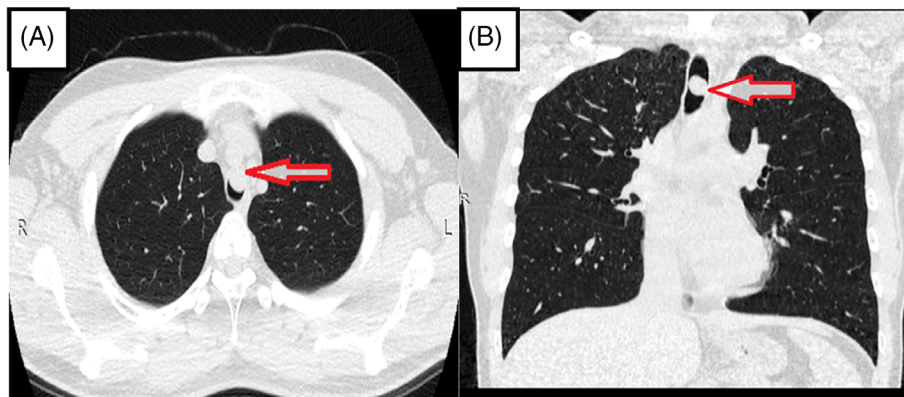


FIGURE 1 Computed tomography of the chest (A, axial view; B, coronal view) demonstrated a well-defined endotracheal round mass with smooth margins measuring 1.6 cm in length.

mild, not exceeding two tablespoons of fresh blood occurring once or twice in a month. He also experienced dysphagia to solid food which worsened 2 months prior to presentation. He had a history of smoking (1 pack per day for the past 10 years). Otherwise, physical examination, baseline laboratory investigations and chest radiograph findings were all unremarkable.

Contrast computed tomography (CT) of the chest was abnormal and demonstrated a well-defined endotracheal round mass with smooth margins measuring 1.6 cm in length. The mass was 4.5 cm from the main carina and the tracheal lumen was narrowed more than 50% with the narrowest diameter of 0.5 cm. There were no other significant lung masses or nodules, pleural effusion or lymphadenopathy reported (Figure 1).

Urgent rigid bronchoscopy revealed a polypoidal mass occluding 90% of the tracheal lumen (Figure 2A). The tumour was completely resected using a diathermy snare and the tumour base was debulked and treated with diathermy. Haemostasis was secured with endotracheal instillation of adrenaline (Figure 2B).

On gross pathology, a 2-cm polypoidal mass with greyish surface was retrieved through bronchoscopic resection (Figure 2D). Subsequent histological evaluation revealed spindle cell tumour cells with prominent fusiform or oval nuclei and eosinophilic cytoplasm arranged in fascicles and vague storiform pattern. The stroma had an intense inflammatory cell infiltration especially by plasma cells and lymphocytes (Figure 2E). On immunohistochemistry, tumour cells stained positively for smooth muscle actin with a tram-track pattern and a cytoplasmic pattern for anaplastic lymphoma kinase (ALK) (Figure 2F) as well as negatively for cytokeratin stains, desmin for smooth muscle cells, CD 34 and S-100 ruling out a granular tumour. IMT was diagnosed.

Surveillance flexible bronchoscopy after 6 months showed a well-healed tumour base. Serial bronchoscopy (Figure 2C) and CT scans showed no recurrence in 12 months.

DISCUSSION

IMT is a rare soft tissue neoplasm of mesenchymal origin that may arise in a wide variety of sites such as the abdomen, mesentery, lung, head, neck, extremities, genitourinary tract and orbit. However, it commonly occurs in the lungs of children and adolescents. Previously, IMT was perceived as a benign reactive lesion and was largely known as inflammatory pseudotumour. Airway IMTs are extremely rare in adults with a prevalence of not more than 12% among pulmonary IMTs.⁸ The data on the epidemiology of airway IMTs are limited due to the low prevalence of the disease in the adult population. To the best of our knowledge, only 11 cases of adult tracheal IMTs have been reported, although the prevalence is speculated to be higher due to potential unreported cases (Table 1).^{7,9–18} The disease is mostly seen before the age of 50, but has also been described as late as in the eight decades of life. There was also no gender predominance or ethnic and geographic predispositions for the development of the disease.¹⁹

The clinical and radiological diagnoses of IMTs are often difficult due to the non-specific presentation. Although the majority of patients have asymptomatic disease that are found incidentally on imaging, patients with tracheobronchial IMTs are often recognized earlier as they are likely to present with symptoms of airway obstruction, respiratory distress and lung collapse such as dyspnoea, cough and haemoptysis.²⁰ On imaging, pulmonary IMTs may be detected as peripheral lung nodules and masses as well as hilar and mediastinal lymphadenopathies.²¹ Our patient presented 2 years from onset of symptoms due to the lack of obstructive respiratory symptoms. The dysphagia which was experienced by our patient was an atypical symptom and was attributed to the compression of the oesophagus as it resolved after the surgical excision of the endotracheal mass.

The ALK gene locus on 2p23 encodes a classical receptor tyrosine kinase that was detected in anaplastic large-cell lymphoma, diffuse large-cell lymphoma and non-small cell lung cancer, and was also discovered in 50% of IMTs.^{22,23}

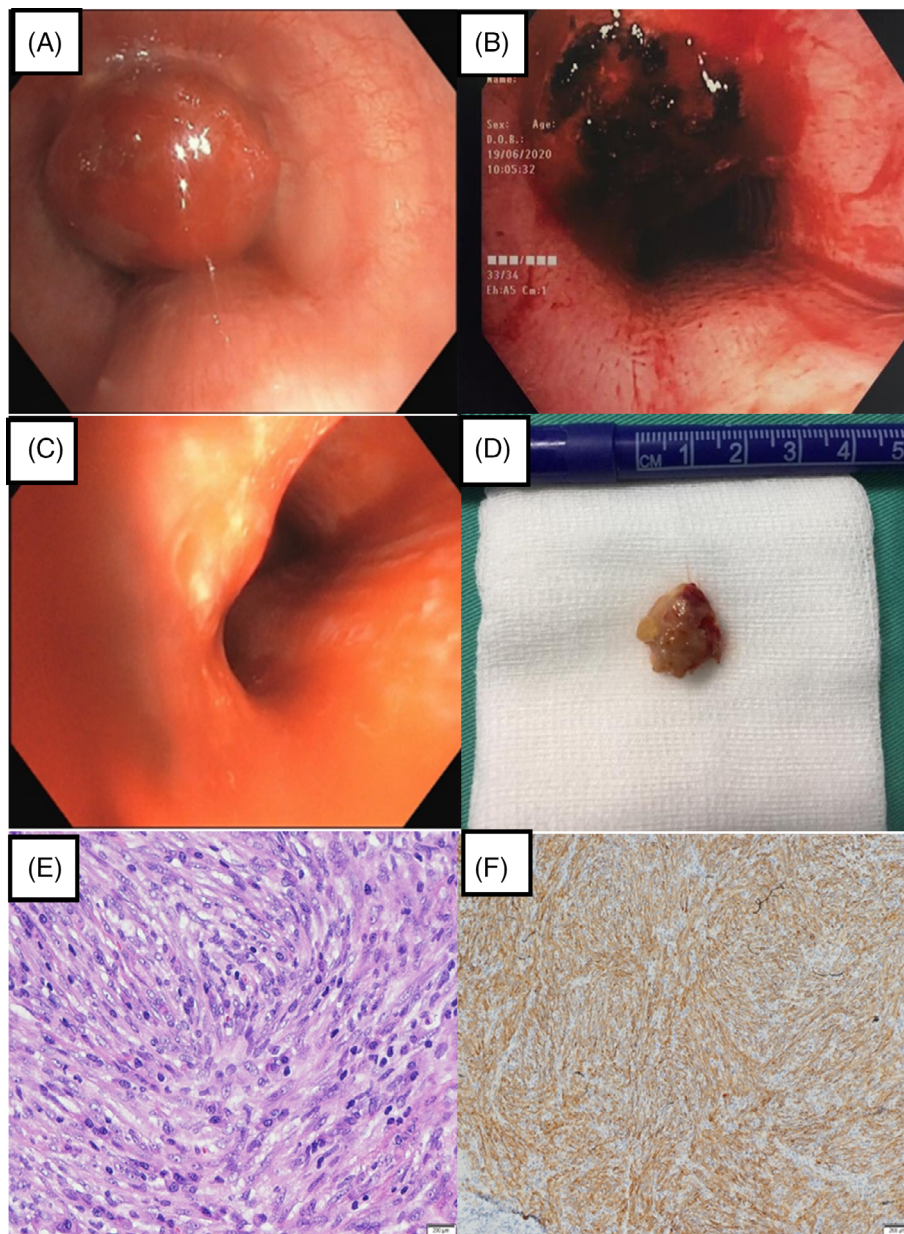


FIGURE 2 Bronchoscopic view of the endotracheal mass (A) prior to intervention, (B) post diathermy snaring and debulking and (C) surveillance 1 year later. Pathology findings: (D) gross appearance of the resected mass. Histopathology findings: (E) presence of spindle cell tumour cells with prominent fusiform nuclei and eosinophilic cytoplasm arranged in fascicles (haematoxylin and eosin stain, original magnification 40 \times). Immunohistochemistry staining: (F) anaplastic lymphoma kinase positive with cytoplasmic pattern (original magnification 100 \times)

This revelation gives us the understanding that IMTs are a true neoplastic process and they were classified by the World Health Organization as tumours of intermediate biological potential due to a tendency for local recurrence and the small risk for distant metastasis.²⁴ ALK expression can be detected by immunohistochemistry as well as fluorescence in situ hybridization, and is found to be more common in younger patients.²⁵ Despite ALK-positive IMT has a tendency for recurrence, it carries a favourable prognosis as ALK negativity is associated with metastasis.²⁶ Our patient manifested ALK expression which was detected by immunohistochemistry.

Diagnosis of IMT is very difficult and often only possible after complete surgical excision of the lesion. Sampling from bronchoscopy or needle biopsy may yield small specimens that are insufficient for diagnosis and it was found that only 6.3% of IMT cases are diagnosed based on preoperative sampling methods.²⁰ These methods may be convenient but are unreliable as the pathologist may need to evaluate the complete lesion to make a confident diagnosis of IMT. In the past, patients with IMT commonly underwent multiple biopsy procedures to establish a diagnosis. Classical histological features of IMT include the presence of cellular spindle cell proliferation in a myxoid to collagenous stroma with a prominent

TABLE 1 Reported adult endotracheal inflammatory myofibroblastic tumour

Year	First author	Age	Presenting symptom	Histological diagnosis	Treatment	Outcome
1991	Satomi ¹⁰	55	Stridor	Plasma cell granuloma	Bronchoscopic resection with laser	Not disclosed
1993	Ishii ¹¹	61	Dyspnoea and wheezing	IMT	Surgery	Not disclosed
2002	Amir ¹²	21	Dyspnoea and stridor	IMT	Surgery	Not disclosed
2003	Restrepo ¹³	20	Dyspnoea and cough	IMT	Surgery	Not disclosed
2004	Nikanne ¹⁴	21	Dyspnoea and cough	IMT	Bronchoscopic resection	Not disclosed
2006	Belák ¹⁵	45	Dyspnoea and stridor	IMT	Surgery	Not disclosed
2006	Ono ¹⁶	45	Dyspnoea	IMT	Bronchoscopic resection with laser	Not disclosed
2009	He ¹⁷	28	Dyspnoea	IMT	Surgery	Not disclosed
2009	Fabre ⁹	19	Not available	IMT	Surgery	Not disclosed
2010	Andrade ⁷	31	Cough and dyspnoea	IMT	Bronchoscopic resection and lobectomy	No recurrence after 6 years
2011	Koch ¹⁸	57	Cough, haemoptysis	IMT	Bronchoscopic resection	No recurrence after 6 months

Abbreviation: IMT, inflammatory myofibroblastic tumour.

inflammatory infiltrate composed primarily of plasma cells and lymphocytes, with occasional eosinophils and neutrophils. Due to the predominance of inflammatory cells, immunohistochemistry is necessary to distinguish IMT from other lesions with similar histological features. Comprehensive histopathological and immunohistochemical evaluation are required to avoid misdiagnosis. Complete surgical excision of the endotracheal mass and the presence of classical histological features seen in our patient expedited diagnosis.

Radical bronchoscopic resection via rigid bronchoscopy has been the mainstay of treatment of airway IMTs to prevent recurrence.⁷ Incomplete resections of pulmonary masses may pose a 60% recurrence rate as described in recent studies.²⁷ Recurrences have been reported among patients with tumours in the larynx where complete surgical resection is challenging.²⁸ Complete surgical excision of a solitary lesion is necessary to ensure a favourable prognosis and improved survival. Evidence for efficacious treatment alternative to surgery remains limited. We performed diathermy snaring and debulking of the endotracheal mass via rigid bronchoscopy.

The recurrence rates specifically for tracheal IMTs are unknown, but for pulmonary IMTs it was reported to be low around 2%. Although it is remarkably lower than the recurrence rates for extrapulmonary IMTs, reported as 25% in previous studies, regular imaging and bronchoscopy to detect recurrence are recommended within the first 2 years after diagnosis.²⁹ Distant metastases is also rare, occurring in 5%–11% of cases especially in children and extrapulmonary IMTs. Common sites include the lung, brain, liver and bone.³⁰ Due to the rarity of the condition, predictors of disease behaviour remain understudied. Overall prognosis is excellent with a 10-year survival of 89% and is dependent on the size of primary lesion and whether complete surgical resection is feasible.⁹ Our patient remained asymptomatic following 1 year of surveillance without any evidence of recurrence.

In conclusion, tracheal IMT is a rare cause of central airway occlusion in adults. Prompt recognition will require early imaging and bronchoscopy. Endoluminal lesions should be

resected completely using bronchoscopic techniques to expedite diagnosis. In most cases, surgical resection alone is sufficient to deter recurrence and ensure a favourable prognosis. Surveillance imaging and bronchoscopy are recommended for the subsequent 2 years to anticipate recurrence.

AUTHOR CONTRIBUTION

Albert Iruthiaraj Lourdesamy Anthony drafted the manuscript. All authors were involved in the care of the patient. All authors critically revised the draft manuscript and approved the final manuscript.

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CONFLICT OF INTEREST

None declared.


DATA AVAILABILITY STATEMENT

Data are available on request from the authors.

ETHICS STATEMENT

The authors declare that appropriate written informed consent was obtained for the publication of this manuscript and accompanying images.

ORCID

Albert Iruthiaraj Lourdesamy Anthony  <https://orcid.org/0000-0001-6725-4572>

Khai Lip Ng  <https://orcid.org/0000-0002-9868-1398>

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