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CASE SERIES AND REPORTS

Transtracheal endoscopic-assisted resection of a rare inflammatory myofibroblastic tumour in adult trachea: a case report

Resezione transtracheale endoscopio-assistita di un raro tumore miofibroblastico infiammatorio della trachea di un paziente adulto: case report

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SUMMARY

Inflammatory myofibroblastic tumours (IMTs) are rare and clinically benign in childhood, and malignant in adults. The aetiology of IMTs is not clear, and recent studies report it as true neoplasm rather than a reactive or inflammatory lesion. IMTs can involve any part of the body, but are usually common in lungs. These are rarely seen in adults and tracheal involvement is also rare in both adults and children. We describe an 18-year-old woman who presented with respiratory difficulty to the emergency department. On clinical examination, the patient had complete absence of breath sounds on the right side of the chest. CT of the chest and virtual bronchoscopy revealed a polypoidal soft tissue mass lesion involving the carina with occlusion of right main bronchus. Endoscopic-assisted resection was performed under general anaesthesia and the final pathological diagnosis was tracheal IMT.

KEY WORDS: Strider • Inflammatory myofibroblastic tumour • Trachea • Bronchus • Spindle cell proliferation

RIASSUNTO

I tumori miofibroblastici infiammatori sono rari in età pediatrica, età nella quale sono clinicamente benigni; sono invece maligni in età adulta. L'eziologia non è chiara, recenti studi affermano che essi siano delle vere neoplasie piuttosto che delle lesioni reattive o infiammatorie. I tumori miofibroblastici infiammatori sono raramente riscontrati negli adulti e il coinvolgimento tracheale è raro sia nei bambini sia negli adulti. Noi descriviamo il caso di una paziente di sesso femminile di diciotto anni, che si è presentata al pronto soccorso per difficoltà respiratoria. All'esame clinico della paziente si evidenziava assenza dei suoni polmonari a destra, pertanto si eseguiva TC del torace e la broncoscopia virtuale rivelava una lesione polipoide soffice che coinvolgeva la carena occludendo completamente il bronco principale di destra. La resezione endoscopio-assistita è stata eseguita in anestesia generale e all'esame istopatologico definitivo la diagnosi è stata di tumore miofibroblastico infiammatorio.

PAROLE CHIAVE: *Stridor • Tumore miofibroblastico infiammatorio • Trachea • Bronco • Proliferazione a cellule fusate*

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Introduction

Inflammatory myofibroblastic tumours (IMT) are rare tumours, commonly seen in children less than 16 years of age and with frequency of 0.04-0.07% of all respiratory tract tumours [1-6]. The World Health Organization defines it as a lesion consisting of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes and eosinophils [1 2 5 6]. The aetiology of the disease is still not clear [2]. Tracheal IMTs are rarely reported in adults who are malignant and benign in children [1 2]. In 1939, the first case of IMT was reported in the lungs [4]. A wide variety of names has been applied to IMTs which are mentioned in Table I [1-4 6-8]. Because of its rarity, we report the case of 18-year-old woman with an IMT in the trachea.

Case report

An 18-year-old woman reported to the emergency depart-

Table I. Other names of inflammatory myofibroblastic tumour.

Inflammatory pseudotumour
Plasma cell granuloma (heart)
Inflammatory myofibrohistiocytic proliferation
Histiocytoma
Xanthoma
Fibroxanthoma
Xanthogranuloma
Fibrous xanthoma
Xanthomatous pseudotumour
Plasma cell–histiocytoma complex (lung)
Plasmocytoma
Solitary mast cell granuloma
Inflammatory fibrosarcoma (bladder)

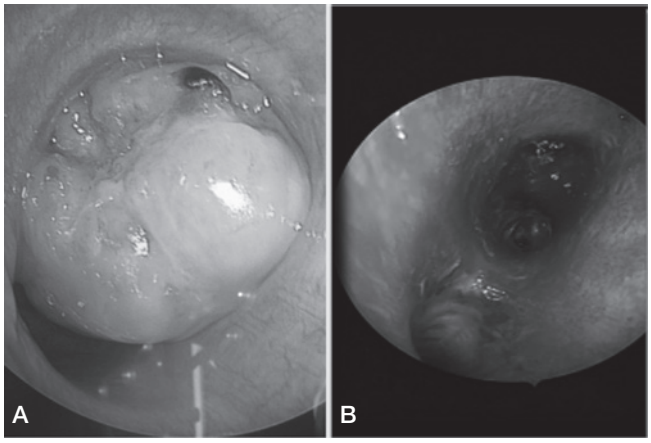

Fig. 2. (A) Endoscopic-assisted bronchoscopy showing the tracheal tumour occluding the right main bronchus. (B) After tumour removal.

ment with difficulty in breathing since four months with insidious onset. The patient had significant loss of weight and there was no history of wheezes, chest pain, haemoptysis, or fever. On examination, the patient was thin with stable vitals and complete absence of breath sounds on right side of chest. Patient was subjected to CT of chest and neck and virtual bronchoscopy revealed a polypoidal soft tissue mass lesion involving the carina with occlusion of right main bronchus causing collapse of the right lung with crowding of right sided ribs and ipsilateral mediastinal shift (Fig. 1). The senior author (SA) performed rigid bronchoscopic examination under general anaesthesia to confirm the above findings and simultaneously the woman was admitted for surgery (Fig. 2 a).

Under jet ventilation general anaesthesia the supine position, anterior tracheotomy was performed and the trachea was transposed anteriorly by using 1-0 prolene suture. Through the tracheotomy a 4 mm 0° rigid endoscope was passed and the tumour mass removed with insulated instruments (Fig. 2 b). The tracheotomy opening was closed with 1-0 vicryl suture and the incision was closed in layers. The mass sent for histopathological examination, which was suggestive of inflammatory myofibroblastic tumour of the trachea (Fig. 4).

The patients was given tapering doses of corticosteroid postoperatively for 10 days and to date we are following the case without recurrence.

Discussion

IMTs are rare tumours, commonly seen in children less than 16 years of age and with a frequency of 0.04-0.07% among all the respiratory tract tumours [1-6]. The World Health Organization defines it as a lesion consisting of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes and eosinophils [1 2 5 6]. Adult IMTs occurring in the trachea are malignant [1 2].

A variety of names are applied for IMTs as mentioned in

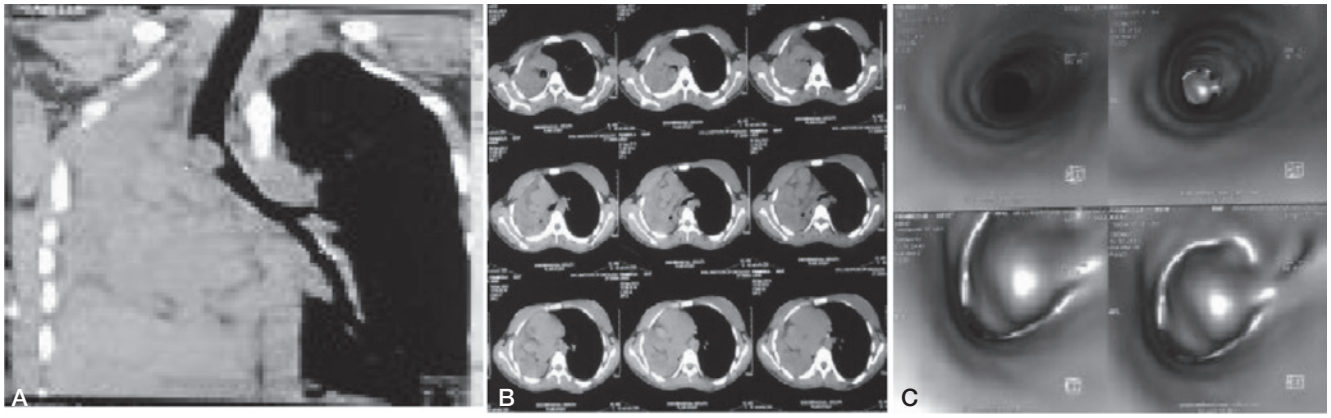

Fig. 1. (A) and (B) CT of chest images. (C) Virtual bronchoscopy image showing a polypoidal mass obscuring the right main bronchus.

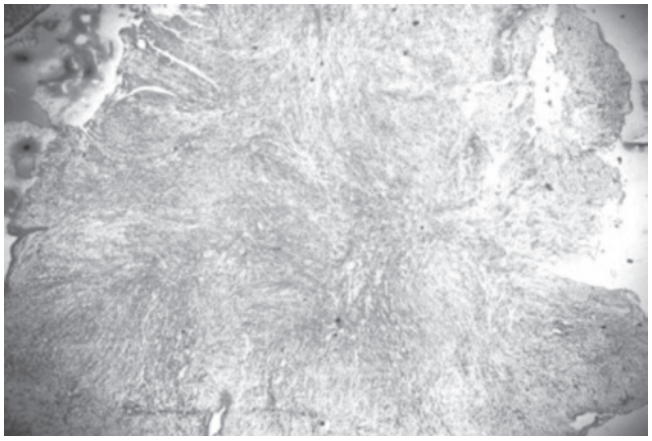

Fig. 3. Histopathological examination suggested inflammatory myofibroblastic tumour.

Table III. Basic histologic patterns of inflammatory myofibroblastic tumour (Coffin et al.)

1. Myxoid/ Vascular pattern: Fasciitis-like appearance and loosely arranged plump spindle cells in an oedematous myxoid stroma with prominent vasculature. It has inflammatory infiltrate of more neutrophils, eosinophils and few plasma cells than the other two patterns.
2. Compact spindle cell pattern: Cellular proliferation of spindle cells with fascicular or storiform architecture in a collagenous stroma and typically show numerous plasma cells and lymphocytes mixed with spindle cells, but discrete lymphoid follicle and aggregates of plasma cells are common.
3. Fibromatosis-like pattern: Relatively hypocellular with elongated spindle cells in a densely collagenous background containing scattered lymphocytes, plasma cells and eosinophils.

Table I [1-4 6-8]. The aetiology of the disease is still not clear, but is probably the cause is an inflammatory reaction secondary to trauma, immune reaction, or infection [2 4 6]. IMTs are most commonly seen in lungs, but can develop in any part of the body (Table II) [1-6 8-11]. The symptoms of IMTs are usually nonspecific and depend on its location. Most respiratory tract IMTs are presented with dyspnoea, stridor, chronic cough, haemoptysis and pleuritic chest pain.

Radiological evaluation with PA and left lateral chest radiograms, CT imaging and endoscopy (bronchoscopic) examination are the diagnostic methods for evaluation. The radiological evaluation gives the information about the tracheal lumen [1 3 8]. IMTs are in the differential diagnosis with other tracheal tumours and tissue biopsy is needed for definitive diagnosis. Immunohistochemical study of IMTs is positive for vimentin, muscle-specific actin, SMA and cytokeratin, which are characteristic for myofibroblats [1 3 4 13].

Table II. Common sites for inflammatory myofibroblastic tumour.

Airway	**Chest**
Lung	Oesophagus
Nasal cavity	Heart
Nasopharynx	Breast
Larynx	**GI**
Trachea	Stomach
Head & Neck	Liver
Orbit	Spleen
Oesophagus	Pancreas
Thyroid	Kidney
Tonsil	Adrenal gland
Maxillary sinus	Retroperitoneum
Fourth ventricle	Diaphragm
Spinal cord meninges	Mesentery
Central nervous system	**Genitourinary system**
	Testis
	Bladder
	Uterus

Simple surgical excision of the tumour with a normal rim of tissue is the treatment of choice. However, endoscopic-assisted resection may be a choice of approach to remove the endoluminal tumours [1-5 7 11 12 14]. CO2 laser or electrocautery is also documented in the literature. Radiotherapy and chemotherapy are reserved for cases of recurrence [1-4 11 12 14].

Histologically, IMTs have variable cellular spindle cell proliferation in a myxoid to collagenous stroma with a prominent inflammatory infiltrate of plasma cells and lymphocytes with some eosinophils and neutrophils. Coffin et al. described three histologic patterns which are tabulated below (Table III) [9]. Salvatore et al. described three types depending on the predominant cell types [7]:

1. Organising pneumonia type with predominant fibroblast-like spindle cells;
2. Fibrous histiocytoma type;
3. Lymphoplasmacytic typeDeath can occur by local recurrence and in cases with infiltration to mediastinal organs or rarely due to distant metastasis [8].

Conclusions

IMT of the trachea are rare and pose a diagnostic dilemma. Most cases present with nonspecific symptoms. Radiological and endoscopic assessment are useful, but tissue biopsy is needed for definitive diagnosis. Simple surgical resection is the treatment of choice; RT and CT are reserved for unresectable cases. Death can occur in local recurrence and infiltration to mediastinal organs, and rarely to distant metastasis. The prognosis of patients who undergo radical resection is excellent.

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