Rare disease

Intrapulmonary cystic teratoma mimicking malignant pulmonary neoplasm

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Benign cystic teratoma of lung is an extremely rare tumour, which was first described in literature by Mohr in 1839. Intrapulmonary teratoma is thought to be a derivative of the third endo-dermal pharyngeal pouch, which is an anlage of the thymus. The authors present a rare case of mature cystic teratoma in a young male involving the right upper lobe of the lung. Diagnosis is often missed and patients are treated for various infectious conditions. Treatment is complete resection for both benign and malignant teratomas and carries excellent prognosis. Benign cystic teratoma, if not excised may cause grave complications like life-threatening haemoptysis or malignant transformation with metastatic disease.

BACKGROUND

Benign cystic teratoma of the lung is a rare tumour. Diagnosis is often missed on imaging, bronchoscopy and tissue biopsy, and patients are treated for other causes (infectious or malignant) subjecting the patient to unnecessary antibiotics and even chemotherapy. If untreated, teratoma of the lung may cause significant and life-threatening complications like haemoptysis, compression of airways and rarely malignant transformation.

We write this case to increase awareness about this rare manifestation of teratoma, which often presents like commonly seen lung conditions, but needs accurate diagnosis and treatment by complete resection to avoid life-threatening complications.

Figure 1 Roentgenogram of the chest showing right upper lobe opacity.

CASE PRESENTATION

A 39-year-old Hispanic male immigrant ex-smoker residing in USA for last 5 years was referred from his primary care physician's office with a 2-year history of intermittent cough, recent haemoptysis and fullness in his chest. He had a weight loss of 10 pounds in the past 7 months. He recently had a Mantoux test done 2 weeks before presentation, which was negative. On examination, the patient had slightly decreased breath sounds in right upper chest, with egophony and dullness to percussion.

INVESTIGATIONS

Routine blood chemistries, cell count, urine analysis and quantiferon test were negative. Chest radiograph showed right upper lobe opacity (figure 1) and CT of the thorax showed large parenchymal opacity 8.5×8.9 cm and in right upper lobe extending into the anterior mediastinum (figure 2) abutting the aortic arch, superior vena cava and encasing the right upper lobe pulmonary artery, with central cavitation's and obstructing the right upper lobe bronchus. Also mediastinal and subcarinal lymph nodes were enlarged.

DIFFERENTIAL DIAGNOSIS

The patient was initially treated with clindamycin for suspicion of an abscess, but there was no response. CT guided biopsy of the mass was performed which showed dense fibrosis with patchy chronic inflammation, but no granuloma or malignancy. β human chorionic gonadotropin and α -fetoprotein levels were normal, and an ultrasound of scrotum did not reveal any testicular mass. CT scan of the abdomen and pelvis was unremarkable for any masses or metastasis, excluding any germ cell tumours.

Bronchoscopy revealed 70% narrowing of the right upper lobe bronchus with friable bulging mucosa and no frank intrabronchial lesion was seen (figure 3). Upon entering the anterior segment of the right upper lobe

there appeared to be necrotic mucosa with destruction of the normal airway anatomy. Bronchial lavage was negative for malignancy or acid-fast bacilli. Pulmonologist thereafter recommended a thoracotomy.

TREATMENT

On thoracotomy, a large mass was found in the right upper lobe surrounding the right upper lobe bronchus and extending into the mediastinum, which contained hair and particulate matter. On gross examination there was a mass of dull gray-tan rubbery tissue with attached hair. Microscopy revealed ectodermal components such as sweat, sebaceous glands and hair follicles along with multiple cystic areas consistent with a diagnosis of intrapulmonary cystic teratoma (figure 4). Multiple sections failed to reveal any dysplasia or metaplasia. There was evidence of organising pneumonia with dense fibrosis in the surrounding lung tissue (figure 5) and intra-alveolar hemorrhage secondary to disruption of the blood vessels around the alveoli with evidence of blood in the airways, which caused the patient to have episodes of haemoptysis.

OUTCOME AND FOLLOW-UP

Right upper and middle lobectomies were performed along with mediastinal lymph node dissection, which

was complicated by bleeding due to significant inflammatory reaction around the surrounding tissue. The patient recovered well postoperatively and was discharged home.

DISCUSSION

Teratomas are most commonly found in the gonads and the most common extra pulmonary site is the mediastinum. Teratoma of the lung is a rare tumour, and only few cases have been reported in literature since its first description by Mohr in 1839.¹ Some cases have cited presence of thymic tissue which provides clues towards the origin of intrapulmonary teratoma as a derivative of the third ento-dermal pharyngeal pouch which is an anlage of the thymus.² Although teratomas originally arise at an early stage of embryonic development they are only discovered later in life due to their slow growth.³ They can be found in female's in age group 16 to 60 years, and more commonly seen in the left upper lobe of the lung.⁴ Our patient had right upper lung involvement, also cited by Trivedi *et al* and Ali *et al*.^{5 6} Most patients present with chest pain, cough and commonly haemoptysis, and rarely with expectoration of hair called 'Trichoptysis' which is very specific to intrapulmonary teratoma and signifies communication with the bronchus.⁷ Based on the pathology, benign teratomas may consist of only one germ cell

Figure 2 CT scan of the chest (axial and coronal views) showing the large parenchymal mass measuring 8.5×8.9 cm abutting the aortic arch and SVC with surrounding area of pulmonary consolidation.

Figure 3 Bronchoscopy revealing significant narrowing of the right upper lobe (RUL) bronchus with friable bulging mucosa (right), and RUL anterior segment necrotic mucosa with destruction of the normal airway (left).

Figure 4 Microscopy showing well-formed hair follicles along with prominent hair shaft (left), sweat glands and sebaceous glands (right).

Figure 5 Microscopy showing keratin debris in the bronchioles (left) with surrounding organising pneumonia (right).

layer (ectoderm) and are called dermoid cyst, or may contain entodermal components like pancreatic acinar tissue but sometimes contain all three germ cell layers.⁸ Those tumours with pancreatic acinar tissue cause bronchiectasis secondary to exocrine secretions from the tumour.

Diagnosis is often not established until thoracotomy. Resection is curative and recommended due to complications associated with growth of the tumour and invasion into bronchioles, causing life-threatening haemoptysis, lung collapse or postobstructive pneumonia. Secretions from a teratoma may cause a myriad of problems as seen in our case where the ectodermal germ cell layer components produced keratin debris and sebaceous secretions creating areas of organising pneumonia. Teratomas with malignant transformation (TMT) have often been reported, including transformation to sarcomas which can be difficult to diagnose because malignancy might be restricted to localised area and may affect only one of the tissue components of an otherwise benign looking tumour.⁴ Surgical resection is the primary modality of treatment because TMTs are considered to be resistant to radiation and systemic chemotherapy. TMTs limited to the testis have a good prognosis. TMTs with metastatic disease have a poor prognosis and are usually treated with chemotherapy. No standard chemotherapy exists and recommendations are based on anecdotal reports/case reports. Regimens containing doxorubicin and cyclophosphamide can be used in sarcomatous TMTs. Cisplatin based regimens can be used for carcinomatous TMTs.^{9 10}

Learning points

- ▶ We present an extremely rare case of mature cystic teratoma in a young male involving the right upper lobe of the lung.
- ▶ Intrapulmonary teratoma is thought to be a derivative of the third ento-dermal pharyngeal pouch, which is an anlage of the thymus.
- ▶ Treatment is complete resection of the tumour and carries excellent prognosis, but if not excised may carry grave risk of complications like life-threatening haemoptysis, or malignant transformation.
- ▶ Malignant transformation is difficult to diagnose and considered to be resistant to radiation and systemic chemotherapy.

Competing interests None.

Patient consent Obtained.

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