

# Mediastinal Teratoma with Pulmonary Parenchyma Fistula: A Rare Diagnostic Endeavour

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## ABSTRACT

Teratoma is the most frequent mediastinal germ cell tumour amongst young population. They are often detected due to pressure symptoms or as an incidental finding on chest X-ray done for any other reason. Mediastinal teratoma is usually benign and rarely possesses diagnostic or therapeutic challenge unless they rupture into surrounding viscera. In such scenario, they may lead to acute life threatening condition and need prompt diagnosis and surgical extirpation. Here, we are reporting our experience of managing a young lady, who presented with short duration breathing difficulty and cough, and was diagnosed with ruptured mediastinal teratoma on imaging, which was further confirmed intraoperatively and histopathologically.

**Keywords:** Extirpation, Mass, Pulmonary fistula, Ruptured

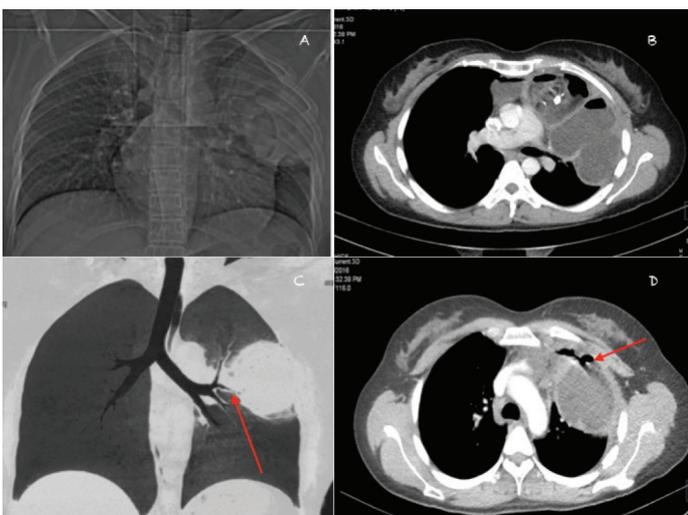
## CASE REPORT

A 27-year-old female presented to thoracic outpatient clinic with complains of breathing difficulty, cough and pain in back of 2-weeks duration. Her physical examination revealed reduced air entry in left upper lung field with no evidence of toxemia. Chest X-ray showed large left upper zone opacity. CT chest showed well circumscribed anterior mediastinal mass with internal fat fluid level and calcified component with evidence of localised rupture to anterior segment of left upper lobe of lung showing air fluid level. Lesion was inseparable from pericardium medially and chest wall laterally with fistulous tract between left upper lobe intra parenchymal component and anterior mediastinal mass, likely mature teratoma with pulmonary parenchymal fistula [Table/Fig-1]. Diagnostic bronchoscopy did not show any endobronchial abnormality. The provisional diagnosis of mediastinal teratoma with pulmonary parenchyma fistula was made.

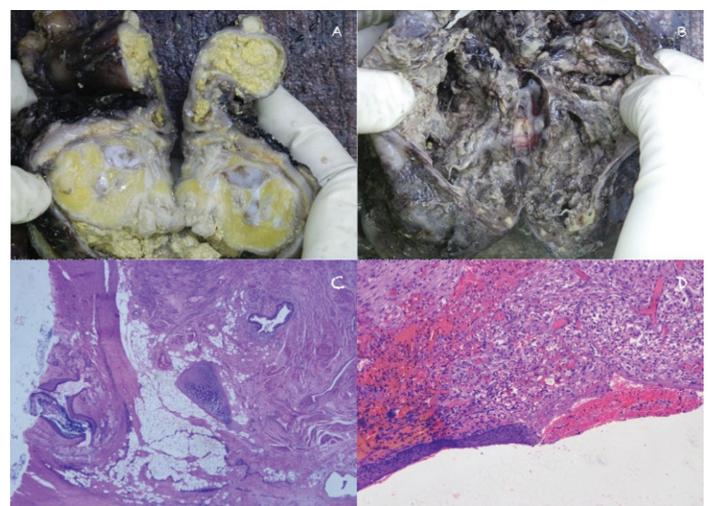
Tumour markers (AFP, hCG and LDH) were within normal limits. Abdominal USG was done to evaluate the retroperitoneum and

gonads, which showed bilateral ovarian simple cysts and was negative for retroperitoneal lymph nodes. Gynaecological opinion was taken and patient was planned for diagnostic laparoscopy during definitive surgery. On laparoscopy, benign looking left ovarian cyst was seen, for which laparoscopic drainage was done. Midline sternotomy was done for the mediastinal mass. There was a large multilobulated anterior mediastinal mass with left phrenic nerve invasion. The whole of left upper lobe was replaced by cystic mass leaving thin rim of lung parenchyma in the apico-posterior segment and lingua with focal infiltration. Non anatomical stapled resection of the left upper lobe was done to remove the mass en block. Perioperative period was uneventful.

On gross pathological examination, a solid cystic mass containing pultaceous material with hair was noted. On microscopic analysis multiple components derived from all three germ cell layers in variable proportion was noted, suggesting mature cystic teratoma with a ruptured and infiltrative interface with lung, measuring 6x2 cm area with cystic areas [Table/Fig-2]. Patient was kept on regular follow up and was well six months after the surgery.



**[Table/Fig-1]:** a) Topogram revealing encapsulated left paramediastinal opacity with internal air and calcific foci associated with left upper zone consolidation; b) Axial post contrast CT image revealing encapsulated anterior mediastinal lesion with internal fat fluid level and calcified component with evidence of localised rupture to anterior segment of left upper lobe of lung showing air fluid level; c) Coronal lung window image with minimum intensity projection showing left upper lobe lesion and relation of involved lung segment with anterior segmental bronchus of left upper lobe (arrow); d) Axial post contrast CT image showing fistulous tract between left upper lobe intra parenchymal component and anterior mediastinal mass (arrow).



**[Table/Fig-2]:** a) Cut section of mediastinal mass with areas of pultaceous material; b) Cystic lesion into lung with ragged margins; c) Haphazardly arranged skin with adnexal structures, mature hyaline cartilage, skeletal muscle and adipose tissue fragments (H&E, 2X); d) Ulcerated cyst wall lining, partially lined by stratified squamous epithelium with subepithelial congestion and foreign body reaction (H&E, 10X).

Factor	Unruptured mediastinal teratoma	Ruptured/(PPTF)	Intra Pulmonary Teratoma (IPT)	Teratoma with lung abscess
Clinically	Young adult Asymptomatic Incidental	Young adult Almost always symptomatic	Children Often symptomatic Extremely rare	Young adult Always symptomatic Sign of toxemia
Imaging	Small masses	Large masses	Small masses	Variable size
	Pushing and compressing adjacent structures	Infiltration to surrounding structure	Multi lobulated well contained mass within lung parenchyma	No specific findings
	Homogeneity of their internal contents of each compartment	Inhomogeneous contents.	-	-
	Air Fluid Level (AFL) uncommon	AFL found often	AFL level common	AFL common
	Pleural effusion Uncommon	Pleural effusion > 57% cases	Pleural effusion uncommon	-
Bronchoscopy	Normal	Content/ defect may be visible	Content/defect may be visible	Non specific

**[Table/Fig-3]:** Differentiating features of mediastinal teratoma, ruptured/Pulmonary Parenchymal Teratoma Fistula (PPTF), Intra Pulmonary Teratoma (IPT) and teratoma with lung abscess.

## DISCUSSION

Mature teratoma is one of the most common causes of anterior mediastinal mass in young females. More than half of them are asymptomatic and the usual presentation is incidentally detected mass on chest radiography [1,2]. Rarely, they may also present with acute onset symptoms due to rupture into surrounding organs like lung, chest wall, and pericardium or rarely skin [1-3].

Pulmonary parenchymal or airway involvement of mediastinal teratoma is rare occurrence and variably labelled as pulmonary rupture or Pulmonary Parenchymal Teratoma Fistula (PPTF). The cause of these events is largely elusive and speculative including autolysis, ischaemia, infection and inflammation [2,4,5]. They pose diagnostic and therapeutic challenges.

The patient often presents with sudden onset of breathing difficulty. The differential diagnosis includes teratoma with secondary lung abscess, chemical pneumonitis and Intra Pulmonary Teratoma (IPT) [5-8]. The definitive differentiation is not always possible preoperatively particularly from large IPT. It is difficult to differentiate the two when a large mediastinal teratoma extends into the lungs [5,6].

The diagnostic clue includes acute onset of symptoms like breathlessness and haemoptysis, mass infiltrating into lung parenchyma, mass infiltrating airways, air fluid levels in imaging, and documentation of intraluminal growth/defect with hair bronchoscopically. Teratoma with lung abscess is often associated with sign of toxemia and more constitutional symptoms than other two conditions.

The ultimate diagnosis is often possible intraoperatively [2,4-8]. We were lucky to document PPTF on preoperative imaging, which is not always possible. Clinical history and examination, imaging with astute judgment help in the management of such cases. The basic principle is timely judgement for surgical intervention to prevent life threatening consequences. The clinical, bronchoscopic and imaging differentials are depicted in [Table/Fig-3].

The surgical extirpation is not straight forward in these cases. The surgical challenges include pleural symphysis, dense adhesions to vital mediastinal structures such as the phrenic nerve, vena cava, pulmonary, aortic vessels, pericardium and lung due to inflammation and infiltration of ruptured teratoma [1,3]. In our case dense adhesions were noted intra operatively, which required careful mobilisation and partial excision of the pericardium. Phrenic nerve was sacrificed as it was engulfed in the tumour along with non anatomical wedge resection of the left upper lobe.

## CONCLUSION

Ruptured mediastinal teratoma is a very rare event and poses a diagnostic challenge to the treating physician. As definitive diagnosis is not always possible preoperatively, the prompt decision making and desired surgical intervention, is the key to manage this challenging scenario.

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