A Rare Case of Esophago-Pulmonary Fistula in a Patient with Carcinoma Oesophagus

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Abstract: A 39-year-old female presented with complaints of cough for 1 month, fever for 3 days, and dyspnoea for 1 week. The patient is also complaining of dysphagia for both liquids and solids. The patient was advised HRCT to evaluate the cause of dysphagia and dysphagia. On CT, there is carcinoma esophagus with middle mediastinal infiltration and a necrotizing pneumonic cavity in the right lung. There is fistulous communication between the esophagus and cavity at the level of the D7 vertebra. There is mediastinal and right hilar necrotic lymphadenopathy.

Keywords: Esophago-respiratory fistula, Esophago-pulmonary fistula, Carcinoma oesophagus, CT scan, Necrotic cavity

1. Introduction

Esophago-respiratory fistula is a rare disease defined as an abnormal tract between the esophagus and the respiratory tract. Types of fistulous connections include tracheoesophageal, broncho-esophageal or tracheo-bronchoesophageal, and esophago-pulmonary fistulas.

They are either congenital or acquired. Congenital fistulas are rare. They develop when the septum between the esophagus and tracheal is incomplete or perforated [1]. It is seen in congenital abnormalities such as vertebral atresia, esophageal atresia, anal atresia, and cardiac malformations as part of the VACTERL/VATER association [2]. This combination of defects may include vertebral defects, anal atresia, cardiac defects, trachea-esophageal fistula (TEF), renal anomalies, and limb abnormalities. Acquired esophageal respiratory fistulae of the oesophageal tracheal or oesophago-bronchial type are well-recognized complications of infections, traumatic lesions, malignant neoplasms, or surgery of the esophagus or tracheobronchial tree [3]. On the contrary, esophago-pulmonary fistulae are very uncommon life-threatening conditions.

Of all esophago-respiratory fistulas, the Esophago-tracheal subtype is the most common (52–57%), followed by the esophageal bronchial (37–40%) and esophago-pulmonary (3–11%) subtypes [4]. In patients with malignant ERF, 92% have esophageal cancer, 7% have bronchogenic carcinoma, and the rest are related to other mediastinal malignancies such as lymphoma, malignant mediastinal node disease, thyroid or laryngeal carcinomas [5,6]. Radiotherapy is described as a late cause of ERF[3]. Most occur spontaneously due to tumor invasion or as a complication of cancer therapies, including surgery, radiation, chemotherapy, laser treatment, instrumentation, or pressure necrosis caused by a previous stent [7]. The median time from the diagnosis of esophageal cancer is eight months but can be the initial presentation in 6% of cases. Most patients die from respiratory infections and poor nutrition if not treated [8].

2. Case Report

History: A 39-year-old female presented with complaints of fever, cough for 1 month, and dyspnoea for 1 week. The patient is also complaining of dysphagia for both liquids and solids. There was a significant weight loss since the past three months. The patient is not yet diagnosed with any malignancy and is not under radiotherapy at the time of presentation. The patient underwent CECT thorax for further evaluation.

Examination:

a) BP-140/90 mm of Hg.

b) Temperature: 38 degrees Celsius.

c) Respiratory rate: >24 breaths/ sec.

c) Anaemic.

Investigations: CECT (i.v) CHEST:

There is a diffuse circumferential wall thickening with irregular intra-luminal growth involving predominantly the middle third esophagus extending from the level of D5-D9 for a length of approx. 7.5-8cm. There is mediastinal infiltration in the form of obliteration of peri-esophageal fat planes, splaying of carina with obliteration of sub-carinal fat, thereby partial encasement of right and left main bronchi, abutment of right and left pulmonary arteries and pulmonary vein confluence region, by partial encasement of descending thoracic aorta of less than 180 degrees.

There is a large thick-walled cavity with internal frothy secretions noted in the posterior segment of the right upper lobe and superior segment of the right lower lobe. The cavity shows inner wall lobulations and few septa – Likely Necrotizing pneumonia cavity. Surrounding lung parenchyma shows diffuse ground-glass attenuation (?alveolar hemorrhage) and irregular consolidation changes. There is fistulous communication between the esophagus and cavity at the level of the D7 vertebra.

The right middle and bilateral lower lobes show mild bronchiectasis with bronchial wall thickening and extensive centrilobular infiltrates – Bronchitis. Right lower lobe dependent lung shows patchy consolidation changes. Few
enlarged and normal-sized necrotic superior mediastinal, right paratracheal, carinal, sub-carinal, and right hilar nodes noted, largest measuring approx—2.1x2.8 cm at the superior mediastinum, causing extrinsic compression over the esophagus. The trachea, cardia, and rest of the vessels appear normal. There is no evidence of pleural effusion bilaterally. Visualized bones appear normal.

A: Topogram showing a cavitatory lesion in right middle zone of the lung (arrow). B: There is a thick walled necrotizing pneumonic cavity with inner wall lobulations and a fistulous communication between oesophagus and cavity.

C: Necrotizing cavity with septa and frothy secretions.

D: Surrounding ground glass attenuation, irregular consolidation changes and few areas of bronchial thickenings.

E: Circumferential wall thickenings with irregular intraluminal growth in middle third of esophagus.
F: Heterogenous enhancement of intraluminal growth.

G: Esophago-pulmonary fistula and necrotizing lung cavity in right lung.

3. Discussion

The malignant esophago-pulmonary fistulas are rare and usually present in patients with locally advanced squamous cell carcinomas when invading lung parenchyma rather than tracheobronchial tree. This causes necrosis and fistula formation. In this patient the cause of fistula formation could be due to parenchymal invasion, but not radiotherapy as it is a newly diagnosed case of carcinoma oesophagus and did not underwent any treatment for such.

Patients may have symptoms such as cough, dyspnoea, dysphagia, fever, hemoptysis and weight loss. It is a fatal complication and can lead to death. Immediate intervention could be initiated after the diagnosis. Treatment includes endoscopic stents, esophageal bypass, wide resections/drainage depending on condition of the patient. In the absence of appropriate treatment, they can lead to pneumonia, lung abscess, and eventually sepsis or acute respiratory distress syndrome [3]. Early treatment is mandatory for malignant fistulae and relies on palliative treatment such as stenting, which provides immediate symptom relief.

The accurate investigation of fistulae should comprise CT scanning and videofluoroscopy, the latter being particularly useful to detect cases that have been misdiagnosed on the basis of CT scans [3]. While diagnosis can be suspected from medical history and symptoms, thin-cut computed tomography (CT) of the chest with oral and intravenous contrast or a contrast esophagogram are useful for diagnosis and for localization of fistula.

4. Conclusion

Esophago-pulmonary fistula is a rare presentation. They mostly present due to congenital or acquired causes such as infections, trauma or malignancy. It is a life threatening condition and early diagnosis can help in further intervention. CECT can help in diagnosing esophagopulmonary fistulas. Videofluoroscopy, bronchoscopy or esophagogram can help for confirmation for any type of esophago respiratory fistulas.

References


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