Radiology Forum

Congenital Cystic Adenomatoid Malformation of the Left Lung Presenting in an Adult

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Key words: Congenital abnormalities, Lung, Adult.

Clinical Summary

A 37-year-old male presented with complaints of cough and dyspnoea on exertion since childhood. His general physical examination was unremarkable. Respiratory examination showed ipsilateral volume loss with bronchial breathing in all the areas of left side. Other systems examination was normal.

Investigations

Haemogram and biochemical tests were normal. Chest radiograph (postero-anterior view) showed shift of the mediastinum to the left with multiple cavities on the same side and contra-lateral compensatory hyperinflation (Figure 1). Sputum smears were negative for acid-fast bacilli on two occasions. High resolution computed tomography (HRCT) (Figure 2) showed mediastinal shift to the left and elevation of the left hemidiaphragm suggestive of volume loss in the left hemithorax. Multiple thin-walled, air-filled cavities of varying sizes were seen almost completely replacing the parenchyma of both lobes of the left lung, some

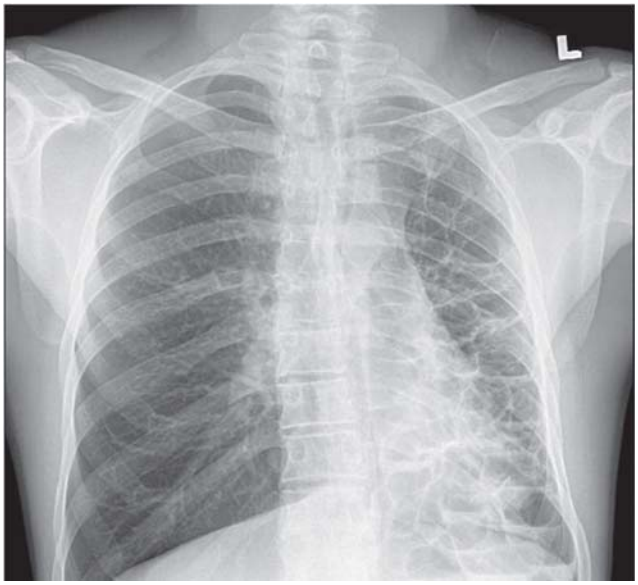
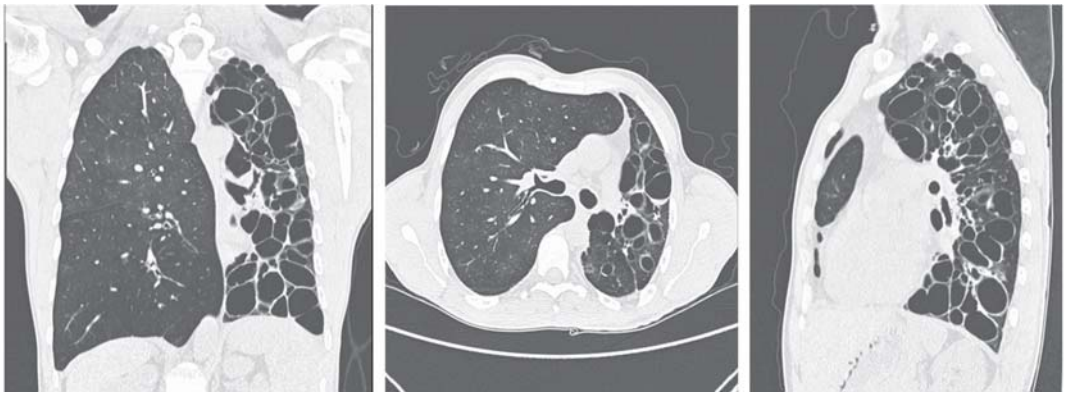


Figure 1. Chest radiograph (postero-anterior view) showing mediastinal shift to the left, multiple cavities on left side and hyperinflation on the right side.



A **B** **C**

Figure 2. Computed tomography of thorax (coronal section (A), axial section (B) and sagittal section (C) showing multiple cavities of varying sizes almost completely replacing the whole of left lung.

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communicating with the smaller bronchi. Most of the cysts were larger than 2cm, largest measuring 4.6cm × 4.2cm, among which few cysts showed air fluid levels. There was no significant central bronchiectasis or mediastinal lymphadenopathy. The patient later underwent left pneumonectomy. Histopathological examination of the resected lung specimen confirmed the diagnosis (Figure 3).

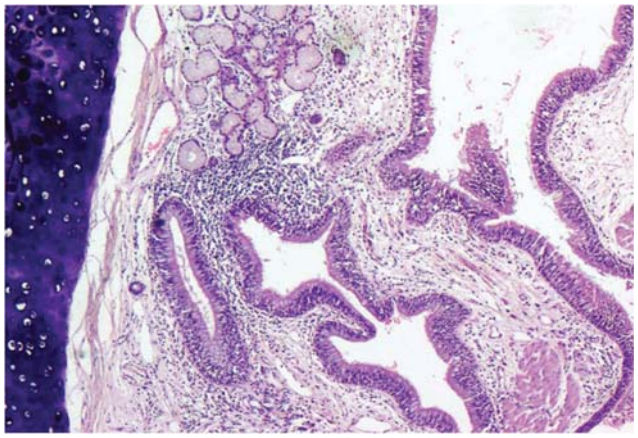

Figure 3. Photomicrograph of resected lung showing cystically dilated and cleft-like spaces lined by pseudo-stratified ciliated columnar epithelium with fibromuscular stroma of the cyst wall showing cartilage, mucous glands and lymphocytic infiltrate (Haematoxylin and Eosin × 40).

Diagnosis: *Type 1 congenital cystic adenomatoid malformation (CCAM) of left lung.*

Discussion

Congenital cystic adenomatoid malformation of the lung is a rare developmental anomaly involving the terminal respiratory structures.[1] It may involve either a part of or a whole lobe or rarely an entire lung.[2] The reported incidence is about 1 in 25000-35000 pregnancies and it forms about 25% of all congenital lung lesions.[1,3] However, these malformations cause significant morbidity and usually most of these cases are detected in neonatal period or during infancy. About 90% of them are reported within first two years of life and they most commonly present with respiratory distress or pneumonia.[4] It is rare that this disease goes unrecognised till adulthood;[5] CCAM in adults is often unilateral.

It occurs due to failure in the fusion of proximal bronchial system which arises from laryngo-tracheal bud with the alveolar tissue derived from the mesoderm. Thus, the affected lung consists of disorganised pulmonary tissue with excess of air passages that resemble terminal bronchioles along with mucus cyst of varying sizes.[2]

Congenital cystic adenomatoid malformation has been classified into 3 types by Stocker and colleagues.[2] Type I is characterised by single or multiple large cysts (>1cm), while Type II consists of multiple small cysts (<1cm) and Type III includes solid mass of airless tissue. Type I is the most common while Type III carries the worst prognosis.[2]

About 25% of the cases are still-born. Many of the cases die within few hours of birth. Recurrent infection and respiratory distress are common presenting complaints in those who survive into infancy and childhood. The survival and diagnosis in adulthood is rare and may be due to associated complications which include recurrent pneumonia, abscess formation, pneumothorax and haemoptysis.[6] Surgical resection of the affected parts of the lung is the treatment of choice.

This radiological image is being presented here because this case highlights the importance of imaging modalities beyond chest radiograph in diagnosing disorders affecting the lung architecture. Cases of CCAM of the lung have been reported in adults, but involvement of whole of the lung is rare. Because of this rarity and radiographic similarity, these cases may be mis-diagnosed as other common disorders like pulmonary tuberculosis or bronchiectasis. Hence, a high degree of suspicion is warranted.

References

1. MacSweeney F, Papagiannopoulos K, Goldstraw P, Sheppard MN, Corrin B, Nicholson G. An assessment of the expanded classification of congenital cystic adenomatoid malformations and their relationship to malignant transformation. *Am J Surg Pathol* 2003;27:1139-46.
2. Seaton A, Seaton D. Developmental disorders of the lungs. In: Seaton A, Seaton D, Leitch G, editors *Crofton and Douglas's Respiratory Diseases*; 5th edition, Vol 2. Delhi: Wiley India; 2002:p.1313.
3. Aichaouia C, Farah S, Dabboussi S, Moatamri Z, M'hamdi S, Bougrine F, *et al*. Multiple bilateral pulmonary nodules revealing a congenital cystic adenomatoid malformation. *Rev Pneumol Clin* 2012;68:261-5.
4. Patz EF Jr, Muller NL, Swensen SJ, Dodd LG. Congenital cystic adenomatoid malformations in adults: CT findings. *J Comput Assist Tomogr* 1995;19:361-4.
5. Vicidomini G, Santini M, Baldi A, Cesarano T, Di Marino MP, Baldi F, *et al*. Cystic adenomatoid malformation of the lung in an adult. *Minerva Chir* 1997;52:469-73.
6. Morelli L, Piscioli I, Licci S, Donato S, Catalucci A, Nonno FD. Pulmonary congenital cystic adenomatoid malformation, type I, presenting as a single cyst of the middle lobe in an adult: case report. *Diagn Pathol* 2007;2:17.

Radiology Forum

Spontaneous Oesophago-Pleural Fistula

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Case Summary

A 54-year-old male presented with acute onset breathlessness and left-sided chest pain of three days duration. He also had slight difficulty in deglutition for the last two days. These symptoms appeared following an episode of sudden onset of nausea and projectile vomiting after heavy meal and had progressively increased over the next two days. There was no history of fever, cough, palpitation, syncope, trauma or any history of undergoing diagnostic procedures. The past medical history was negative for any medical illness or hospitalisation. He was non-smoker and non-alcoholic.

On examination, pulse was 110/min, blood pressure was 130/80mmHg and respiratory rate was 24/min. Mild surgical emphysema was evident in the chest wall and neck. Rest of the general physical examination was unremarkable. On respiratory system examination, reduced thoracic movements, reduced vocal fremitus, dull percussion note, reduced intensity of breath sounds were evident on both sides but were more prominent on the left side. Further, crepitations and rhonchi were also present on the left side. Rest of the systemic examination was unremarkable.

Investigations

His complete blood count, haemogram and biochemical tests were within normal limits. Electrocardiogram (ECG) revealed sinus tachycardia; no ST-T changes were seen. Chest radiograph (Figure 1) showed hydropneumothorax on the left side with pleural effusion on the right side. Pleural fluid analysis was as follows: protein content 1.24 g/dL, glucose 74.5 gm%, chloride 84.3 mEq/L, cell count 1500/mm^3 (polymorphs 50%, lymphocytes 50%) and adenosine deaminase (ADA) 46 IU/L.

Broad-spectrum antibiotics were initiated and closed tube thoracostomy was performed on the left side. Subsequent chest radiograph showed expansion of left lung with drainage of effusion but an increase in pleural effusion on the right side. Radiograph barium swallow showed abnormal extravasation of the barium and passage of contrast into the pleural space

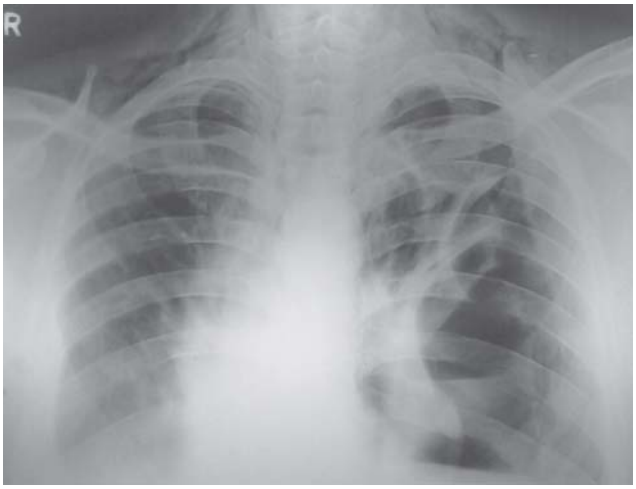

Figure 1. Chest radiograph (postero-anterior view) showing hydropneumothorax on the left side, pleural effusion on the right side and subcutaneous emphysema.

(Figure 2). It was also observed that food particles and swallowed liquids (e.g., milk) were partly draining through the chest tube. The patient was immediately referred to thoracic surgical unit at higher centre for further management. He was lost to follow-up thereafter.

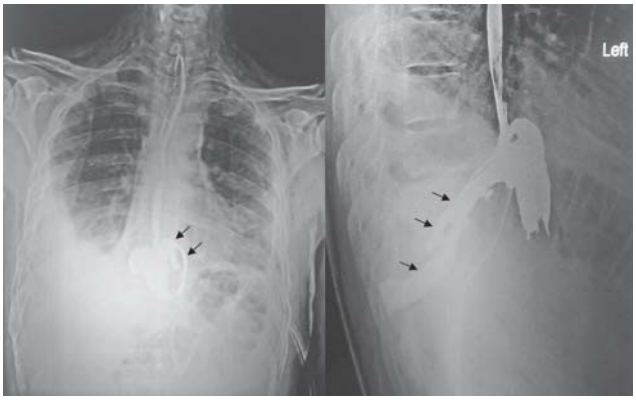

Figure 2. Radiograph barium swallow (PA and lateral views) showing abnormal extravasations of the contrast into pleural space (arrows).

Diagnosis: *Spontaneous oesophago-pleural fistula following forceful vomiting (Boerhaave's syndrome).*

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Discussion

Boerhaave's syndrome is an uncommon cause of oesophageal rupture characterised by spontaneous full-thickness oesophageal perforation. It usually results from a sudden rise in the intra-oesophageal pressure secondary to contraction of cricopharyngeus muscle and closing of pyloric sphincter that occurs after forceful vomiting. Patients with Boerhaave's syndrome classically present with vomiting, subcutaneous emphysema and lower thoracic pain (Mackler triad).[1] This syndrome is uncommon and accounts for approximately 15% of traumatic oesophageal perforations.[2,3]

The symptoms and clinical signs of oesophago-pleural fistula are non-specific. Chest radiograph may show pleural effusion, pneumothorax, hydropneumothorax, subcutaneous emphysema or pneumomediastinum depending upon the site, duration, severity of perforation and integrity of the pleura. The contrast study of oesophagus using barium is very useful in lower oesophageal perforation and passage of oral contrast into the pleural space confirms the diagnosis of oesophago-pleural fistula. Other investigations that help in the diagnosis include thoracic ultrasonography, computed tomography, magnetic resonance imaging, upper gastrointestinal endoscopy among others.[4]

Management of this condition depends upon the site, size, duration of injury and the extent of mediastinal involvement. Conservative therapy includes drainage of empyema, local irrigation, tube feeding, gastrostomy or jejunostomy. Primary repair is the treatment of choice and should preferably be done within 24 hours. Other measures include cervical oesophagostomy, T-tube placement and plastic covered self expandible metallic stents, etc.[5]

The Boerhaave's syndrome should be differentiated from Mallory-Weiss syndrome that is characterised by longitudinal mucosal lacerations caused by forceful or long-term retching, vomiting or coughing. These patients present with gastrointestinal bleeding; the oesophageal mucosal tears heal spontaneously. This condition is commonly associated with hiatus hernia and alcoholism.[6]

Oesophago-pleural fistula may occur secondary to the iatrogenic trauma (endoscopy and allied procedures), barotraumas, following empyema thoracis, oesophageal malignancy, tuberculosis, corrosive oesophagitis, oesophageal ulcers, surgical procedures (especially, post-pneumonectomy) and radiation therapy.[7,8]

The clinical presentation may mimic aspiration pneumonia, lung abscess, pneumothorax, pulmonary embolism, myocardial infarction, aortic dissection, pericarditis, acute gastritis, acute pancreatitis and acute cholecystitis. Boerhaave's syndrome is an uncommon and a dangerous entity. Rapid diagnosis and early institution of therapy provides the best chance of survival.

References

1. Yagnik VD. Boerhaave's syndrome: spontaneous full thickness oesophageal perforation. *Lung India* 2012;29:197.
2. Jagminas L, Silverman RA. Boerhaave's syndrome presenting with abdominal pain and right hydropneumothorax. *Am J Emerg Med* 1996;14:53-6.
3. Gime´nez A, Franquet T, Erasmus JJ, Martý´nez S, Estrada P. Thoracic complications of esophageal disorders. *Radio Graphics* 2002;22:S247-58.
4. Wechsler RJ. CT of esophageal-pleural fistulae. AJR *Am J Roentgenol* 1986;147:907-9.
5. de Schipper JP, Pull ter Gunne AF, Oostvogel HJ, van Laarhoven CJ. Spontaneous rupture of the oesophagus: Boerhaave's syndrome in 2008. Literature review and treatment algorithm. *Diag Surg* 2009;26:1-6.
6. Weiss S, Mallory GK. Lesions of the cardiac orifice of the stomach produced by vomiting. *J Am Med Assoc* 1932;98: 1353-5.
7. Singla R, Bhagi RP, Singh P, Goel RN, Das PK. Oesophagopleural fistula secondary to empyema thoracis. *Indian J Chest Dis Allied Sci* 1988;30:225-9.
8. Lauwers P, Van Schil P, Schroyens W, Fierens H, Vanmaele R, Eyskens E. Oesophagopleural fistula: a rare sequel of pneumonectomy. *J Thorac Cardiovasc Surg* 1996;44:266-70.