CASE REPORT

A rare cause of recurrent chest infection in children-bronchopulmonary sequestration

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Abstract

Bronchopulmonary sequestration is one of the rare thoracic congenital anomalies. We report the case of a 6 year old boy with history of recurrent episodes of chest infection and breathing difficulty. This time admitted with fever and cough. Investigations revelaed neutrophilic leucocytosis, raised C-reactive protein and a retrocardiac opacity on chest radiographs. Contrast enhanced Computed Topography(CT scan) revealed a large, well defined, left lower lobe multi-loculated cystic mass with a vessel arising from the descending aorta supplying sequestrated lung portion. These CT scan findings were suggestive of diagnosis of an intralobar bronchopulmonary sequestration (IPS). Surgical resection of this opacity was done and histopathology report confirmed the diagnosis of bronchopulmonary sequestration.

Keywords: Bronchopulmonary sequestration, Children, Intralobar, Extralobar, Recurrent pneumonia.

Introduction

Pulmonary sequestration is a cystic or solid mass that comprises of nonfunctioning primitive lung mass, does not communicate with the tracheobronchial tree and has anomalous systemic blood supply.[1] Its divided into two types i.e. intrapulmonary sequestration (IPS) or extrapulmonary sequestration (EPS). In IPS(also known as intralobar pulmonary sequestration), the sequestrated portion is located within a normal lung lobe and lacks its own visceral pleura while in EPS (also known as extralobar pulmonary sequestration), the sequestrated portion is located outside the normal lung parenchyma and has its own visceral pleura. IPS is four times more common than EPS. IPS has an equal incidence in both males and females whereas EPS is present in males in >80% cases.[2]

Usual presentation of IPS is with recurrent chest infection and chronic cough whereas EPS may present with respiratory distress and cough. Surgical excision (usually lobectomy or segmental resection) is the curative treatment and is associated with minimal morbidity and mortality.[3] Moreover surgical treatment avoids fatal complications of malignant transformation and haemorrhage. Here, we report the case of a 6 year old boy who was having recurrent chest infections and was repeatedly treated as a case of pneumonia and reactive airway disease but later on advanced investigations revealed IPS. We are reporting this case because of its common clinical presentation but rare etiology and different management. Recurrent pneumonias and can be a presentation of a very rare disease. Parent's consent and approval of hospital ethical review committee were obtained for publication of this case.

Case Report

A 6 year old boy was admitted (April 2014) with history of fever and cough for last 05 days. Past history revealed episodes of similar illness and multiple admissions from age of 2 years and being treated as a case of reactive air way disease. His immunization status was up to date according to expanded programme on immunization (EPI) and there was no history of contact with tuberculosis patient or family history of allergic disorders.

On examination, he was febrile (100°F), tachypneoic (R/R

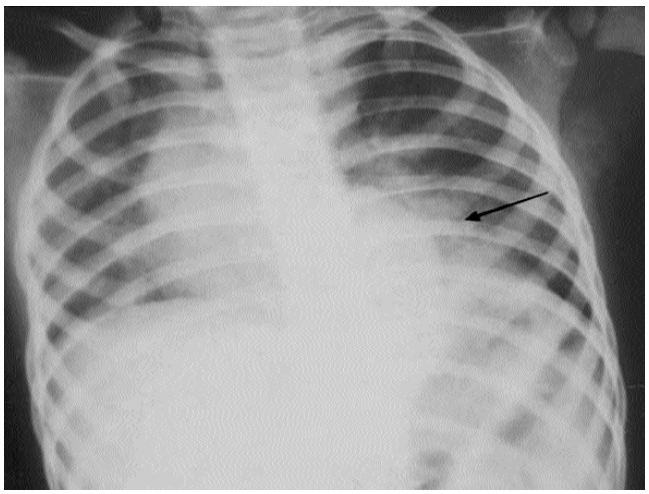

Figure-1: Chest X-ray PA view of patient showed a non homogenous retro cardiac opacity in left lower zone.

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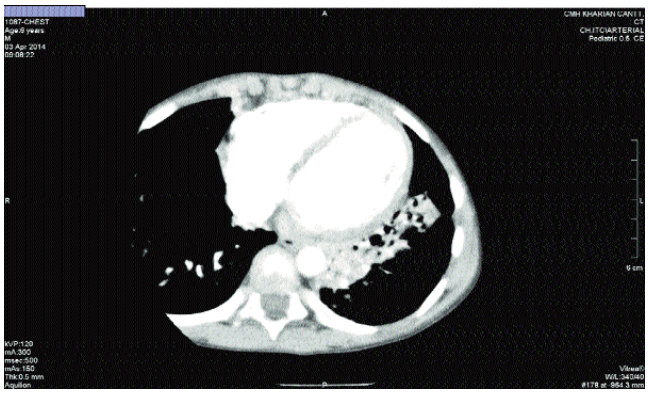

Figure-2: Plain CT scan of the child showing a classical patch of consolidation in left lower lobe with air bronchograms.

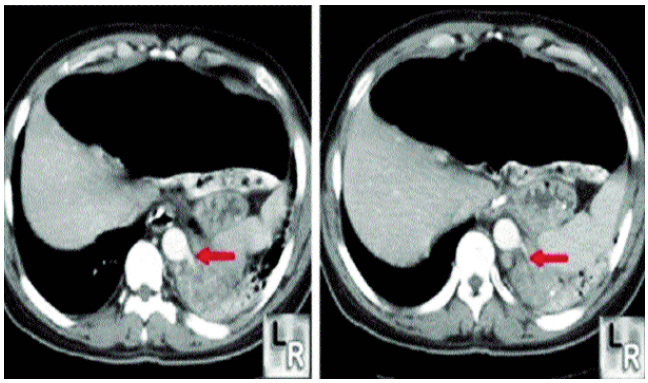

Figure-3: Contrast enhanced CT scan of patient. Red arrows are pointing towards a vessel which is arising from aorta and supplying sequestrated lung portion.

60/min) with no evidence of cyanosis, clubbing or lymphadenopathy. His chest examination revealed decreased air entry, decreased vocal resonance and crepitations in left lower chest and scattered bilateral ronchi. Rest of the systemic examination was unremarkable. He was managed as a case of pneumonia with antibiotics and supportive investigations were requested.

Laboratory investigations showed leucocytosis (16,200/mm^3) with neutrophilia (85%), erythrocyte sedimentation rate 20 mm/hour and C-reactive protein 12 mg/dl. Chest radiography showed a non homogenous retro cardiac opacity in left lower zone (Figure-1).

His symptoms improved markedly but chest findings persisted. Repeat chest X-ray revealed same findings. On basis of clinical suspicion of non resolving opacity, we requested CT scan (plain and contrast) of chest. Plain CT scan revealed a patch of consolidation in left lower lobe with air bronchograms (Figure-2). To further augment our diagnosis we did a contrast enhanced CT scan. It showed a large, well defined, left lower lobe multi-loculated cystic mass with multiple enhancing internal septae and a vessel arising from the descending aorta supplying sequestrated lung portion thus suggesting diagnosis of an intralobar bronchopulmonary sequestration with superadded infection (Figure-3). Echocardiography was normal.

Thoracic surgeon examined the patient and planned thoracotomy. At thoracotomy left lower lobe lobectomy was done, aberrant artery arising from thoracic aorta ligated and venous drainage into left pulmonary vein identified and transected. The gross histopathological examination revealed a 250 gram mass measuring 10 X 6 X 6 cm with multiple necrotic and haemorrhagic areas within the mass. Microscopy revealed evidence of chronic inflammation, fibrosis, microabscesses along with obliterative changes in blood vessels consistent with bronchopulmonary sequestration. Post operative course was uneventful and patient discharged with follow up advice. On follow up visits his chest X-ray was normal and had no subsequent chest infections for next four months.

Discussion

Pulmonary sequestration (PS) was first described by Rektorzik in 1861 and reported incidence is 0.15% - 6.4% of all congenital pulmonary malformations.[4] Embryologically, different theories have been proposed regarding its development. The accepted theory says that in utero, an accessory lung bud develops from the ventral aspect of the primitive foregut. This bud, a pluripotential tissue, migrates in a caudal direction with the normally developing lung. This lung bud receives its blood supply from an aberrant artery arising from the aorta and covers the primitive foregut. Accessory lung bud ultimately develops into sequestered tissue that is not attached to the pulmonary arterial blood supply. As a result, this sequestered tissue is not connected to the normal bronchial airway architecture and it fails to function and contribute to respiration. A left-right shunt is created at later stages where oxygenated blood from the systemic artery flows in a shortcut into sequestered tissue and returns to the heart (left atrium) via pulmonary veins thus increasing the work load of the heart. This sequestrated tissue may ultimately become cause of life threatening haemorrhage and recurrent infections as seen in our patient. Lower lobes of left lung are involved in more than 90% of the cases as observed in this patient and bilateral involvement is rare.[5]

IPS is usually diagnosed later in childhood, adolescence or adulthood when the patient usually presents with

pulmonary infection as occurred in our case. A communication with other bronchi or lung parenchyma may co exist thus favouring pulmonary infection to occur. In EPS pulmonary infection is rare, the child may be asymptomatic, presents with an intrathoracic mass or may be diagnosed in the first six months of life with respiratory distress and feeding difficulties.[6] IPSs are not associated with other cardio-pulmonary anomalies but EPS may be found in association with cardiac or, more frequently, diaphragmatic anomalies in half of cases. IPS demonstrates no preference for either lung whilst EPS is found in the left lung in majority of cases. In IPS, the systemic blood supply can be from the descending aorta (72%), abdominal aorta and splenic artery (21%), intercostal artery (3.7%) or rarely the subclavian, internal mammary and pericardiophrenic arteries.[7] In our patient, the blood supply to the sequestrated portion was from an artery arising from descemding aorta.

Different radiological modalities are used to diagnose pulmonary sequestration with an aim to define the nature and extent of the lesion, identify the aberrant arterial supply and venous drainage.[8] Chest radiographs can give a reasonable diagnostic clue to pulmonary sequestration in combination with clinical correlation, but some are too small to be detected by conventional radiography. Contrast enhanced CT scan has > 90% accuracy in diagnosis of sequestration, although morphology of lesion varies. Aberrant blood supply and venous drainage may not be evident on CT scan, for which MRI is the investigation of choice. However in a few cases, conventional angiography can be done to identify the exact anatomic location of blood vessels.[9] In our case, diagnosis was established on contrast enhanced CT scan (Figure-3).

Surgical resection is the only recommended treatment for symptomatic pulmonary sequestrations and even in asymptomatic (especially IPS and subdiaphragmatic EPS) cases because of the risk of pulmonary infection, haemorrhage and malignant transformation.[10] Our patient also responded well to the surgical treatment. Resection of asymptomatic ELS is controversial.[11]

Sometimes extremely rare diseases can mimic common illnesses as in this case but a very high index of suspicion is required to diagnose such rare disorders like pulmonary sequestration.

A timely identification and treatment of this otherwise challenging diagnosis can effectively manage this disorder with minimal morbidity and mortality.

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Conflict of Interest: None.

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References

1. Aryal G, Pathak V. Bronchopulmonary sequestration presenting as recurrent pneumonia. WMJ 2011; 110: 240-2.
2. Bratu I, Flageole H, Chen MF, Di Lorenzo M, Yazbeck S, Laberge JM. The multiple facets of pulmonary sequestration. J Pediatr Surg 2001; 36: 784-90.
3. Savic B, Birtel FJ, Tholen W, Funke HD, Knoche R. Lung sequestration: report of seven cases and review of 540 published cases. Thorax 1979; 34: 96-101.
4. Nijagal A, Jelin E, Feldstein VA, Courtier J, Urisman A, Jones KD, et al. The diagnosis and management of intradiaphragmatic extralobar pulmonary sequestrations: a report of 4 cases. J Pediatr Surg 2012; 47: 1501-5
5. Litt D, Gandhi S, Bhinder S, Blitz M, McIntyre K. Incidental finding and management of intralobar sequestration of the lung in a 24-year-old man. Can Respir J 2013; 20: 403-5.
6. Samuel M, Burge DM. Management of antenatally diagnosed pulmonary sequestration associated with congenital cystic adenomatoid malformation. Thorax 1999; 54: 701-6.
7. Abbey P, Das CJ, Pangtey GS, Seith A, Dutta R, Kumar A. Imaging in bronchopulmonary sequestration. J Med Imaging Radiat Oncol 2009; 53: 22-31.
8. Gupta S, Kim S. Images in clinical medicine. Pulmonary sequestration. N Engl J Med 2013; 368: e6.
9. Mautone M, Naidoo P. A case of systemic arterial supply to the right lower lobe of the lung: imaging findings and review of the literature. J Radiol Case Rep 2014; 8: 9-15.
10. Kambayashi T, Suzuki T. Intralobar pulmonary sequestration treated by resection of the sequestrated segment. Kyobu Geka 2011; 64: 1082-5
11. Yue SW, Guo H, Zhang YG, Gao JB, Ma XX, Ding PX. The clinical value of computer tomographic angiography for the diagnosis and therapeutic planning of patients with pulmonary sequestration. Eur J Cardiothorac Surg 2013; 43: 946-51.