Case Report

Pulmonary sequestration cyst in a patient of cerebral palsy

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ABSTRACT

Pulmonary sequestration cyst is a rare entity in pediatric patients. Most of the time, it is diagnosed as an incidental finding. It is associated with other congenital anomalies, especially congenital diaphragmatic hernia. We report a patient of cerebral palsy presented with vomiting and recurrent chest infections. He was diagnosed to have hiatal hernia on computed tomography scan of chest. At operation, a pulmonary sequestration cyst along with hiatal hernia, malrotation, and meckel's diverticulum was encountered. The sequestration cyst was managed through transhiatal approach.

KEY WORDS: Bronchopulmonary sequestration, foregut malrotation, hiatal hernia, meckel's diverticulum

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INTRODUCTION

Pulmonary sequestration cyst is a very rare anomaly in pediatric age group. It is comprised of nonfunctioning primitive lung tissue, having no communication with the tracheobronchial tree, and contains a systemic arterial supply. The literature regarding its first-hand diagnosis is scarce. Pulmonary sequestration has been found to be associated with other congenital anomalies like congenital diaphragmatic hernia, congenital heart diseases, and gastrointestinal malformations. Its association with multiple anomalies is rarely reported.[1,2]

Vomiting and repeated chest infections in patients of cerebral palsy (CP) are usually attributed to the reflux and aspiration of the secretions in these patients. Rarely, other causes of vomiting and repeated chest infections are found in these patients.[3] We report a patient of CP who presented with vomiting and repeated chest infections, having causes other than CP for vomiting and repeated chest infections.

Access this article online

Quick Response Code:

Website: www.lungindia.com

DOI: 10.4103/0970-2113.83980

CASE REPORT

An 18-month-old male baby was presented to the medical emergency department of our institution with complaints of fever, cough, respiratory distress, and vomiting for 10 days. There was a history of repetition of such events. There was a history of multiple admissions in the local hospitals for such complaints. There was a positive history of birth asphyxia. The baby was a patient of CP and did not achieve developmental milestones with respect to the age. The baby could only hold his neck at this age. On examination, he was febrile with a temperature of 100°F; pulse, 110/min; respiratory rate, 40/min; and BP within normal limits. At inspection of the respiratory system, there were obvious nasal flaring, subcostal and intercostal retractions. On auscultation of the chest, there were course crepts on the left side along with reduced air entry on the same side. There was a bronchial breathing on the lower zone of the left half of the chest. Features of CP were present on CNS examination. A right-sided hydrocele and distal penile hypospadias were found on genital examination.

The baby was admitted in the medical emergency as a case of aspiration pneumonia and empirical antibiotics along with nebulization and chest physiotherapy instituted. A chest X-ray was requested, which revealed an impression of some cystic lesion in front of cardiac shadow, along with pneumonic patch, on left side of the chest [Figure 1]. His CBC report revealed mild anemia (Hb, 9 g/dl) and raised TLC indicating acute infection.

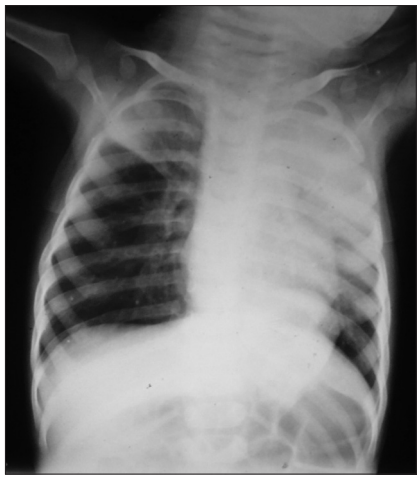

Figure 1: Chest X-ray showing left-sided bronchopneumonia and a shadow of cystic mass overlapping the cardiac location

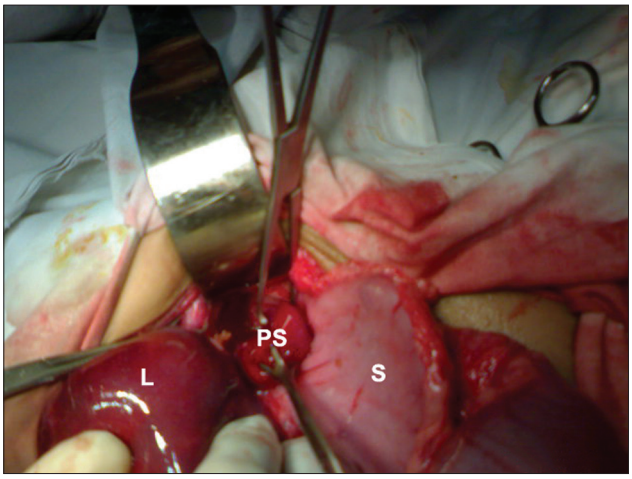

Figure 2: Operative view of pulmonary sequestration (PS). Note that the stomach (S) had been taken down from the hiatus and liver (L) was present on the opposite side

A computed tomography (CT) scan of chest was requested to delineate the cystic shadow in the chest. CT scan showed a widened hiatus and stomach was going up in the chest. A call was sent to the surgical unit for management of hiatus hernia. At the end of treatment for the bronchopneumonia, patient was shifted to our department and an exploratory laparotomy was planned to treat the hiatus hernia. After optimizing the patient for surgery, an exploratory laparotomy was performed by a midline vertical incision. Following were the operative findings:

1. On opening the peritoneal cavity, cecum and appendix (loaded with Fecalith) were present in the epigastrium with a band of Ladd running toward the duodenum.
2. There was widened hiatus, and stomach was present in the chest.
3. After delivering stomach back into the abdominal cavity, a cystic mass was present in the chest. That mass was not having any communication with the normal tracheobronchial tree and had its own pleural covering. The blood supply was identified to come from thoracic aorta [Figure 2].
4. A meckel's diverticulum with a thickened base was found as a coincidental anomaly.

We identified the cystic mass as pulmonary sequestration cyst and sent for histopathology after surgical resection. Hiatus hernia was repaired and meckel's diverticulectomy performed in view of having heterotrophic mucosa in it. At the end, Ladd's procedure was performed to correct the malrotation.

The postoperative recovery was uneventful. Nasogastric tube was removed on fourth postoperative day. Patient was started orally on fifth postoperative day and discharged on eight postoperative day. The histopathology of the submitted tissue revealed a pulmonary sequestration cyst enclosed in a pleural covering. The patient was on regular follow up and showed a remarkable improvement in episodes of repeated chest infections.

DISCUSSION

Pulmonary sequestrations are rare anomalies of primitive lung tissue in pediatric patients. There are two types of pulmonary sequestrations, identified on the basis of its anatomical location, intrapulmonary sequestration, and extrapulmonary sequestration. Intrapulmonary sequestrations are surrounded by normal lung parenchyma without separate pleura, whereas extrapulmonary sequestration is enclosed completely in its own pleural sac.[1,2] In our case, the sequestration was lying outside the main pleural cavity and had its own pleural investment. It was an extrapulmonary sequestration.

There are two commonly accepted schools of thought regarding the etiology of the pulmonary sequestrations. According to one theory (the accessory lung bud theory), there developed an accessory lung bud from the ventral aspects of the primitive foregut. The accessory lung bud received its blood supply from vessels that connect to the aorta and that cover the primitive gut. If the event of accessory lung bud development occurred early in the course of embryogenesis, it would give intrapulmonary variant; on the contrary, later development of the accessory lung bud would result in the extrapulmonary type. According the other school of thought, the necrotizing pneumonia causes obliteration of lower zone bronchi that may lead to the development of sequestration.[4] We favor the first theory because the second theory cannot explain the systemic arterial supply of the sequestration, which can only be acquired during developmental phase of life.

Intrapulmonary sequestration is the most common variety and four times the extrapulmonary variety. Both of the varieties differ in location, venous supply, and age of presentation. The venous drainage of intrapulmonary variety is pulmonic, whereas that of extrapulmonary variety is systemic. Intrapulmonary sequestration presents in adulthood; on the other hand, most of the cases of

extrapulmonary sequestration present in the initial few years of life.[1,2-4]

The presentation of pulmonary sequestration is mostly as an incidental anomaly in patients operated for some other surgical disease. Sometimes, they may simulate other lesions of the chest like congenital lobar emphysema. The main presentation is that of repeated chest infections.[1-4] In our case, the patient was a diagnosed case of CP and had been admitted multiple times for repeated chest infections and vomiting. Vomiting and repeated chest infections in these patients are usually considered the sequel of the CP itself,[3] but in our case, vomiting and repeated chest infections were due to the hiatus hernia and pulmonary sequestration cyst.

CT scan can identify such lesions,[1] but as in our case, overlapping of stomach and sequestration might preclude the radiologist in delineating the sequestration cyst. We had the suspicion of some other associated anomaly in this patient due to a cystic shadow on the chest radiograph.

Extrapulmonary sequestration is found to be associated with congenital diaphragmatic hernia in about 30% cases. Other associated anomalies are congenital heart diseases and gastrointestinal malformations.[1] In our case, the extrapulmonary sequestration was present along with with hiatus hernia, malrotation of intestine, meckel's diverticulum, hypospadias, and hydrocele.

The management options are surgical resection or arterial embolization. The most common approach for surgical resection is left posterolateral thoracotomy; however, VATS (video-assisted thoracoscopy) has also been advocated by some authors. During the operation, phrenic nerve should be carefully identified and saved; otherwise, a diaphragmatic palsy might occur. Such a complication had been reported in literature. In that case, there developed a diaphragmatic eventration after excision of pulmonary sequestration; and patient had another operation for the iatrogenic eventration of diaphragm.[4-7] We excised the sequestration cyst successfully through the transhiatal route.

To conclude, the association of other anomalies like hiatus hernia and pulmonary sequestration as a cause of vomiting and recurrent chest infections in a patient of CP is very rare and a high index of suspicion is required to diagnose it. Vomiting and repeated chest infections are not always the characteristics of the CP itself, other causes like pulmonary sequestration should also be considered in these patients.

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How to cite this article: Mirza B, Saleem M, Ijaz L, Qureshi A, Sheikh A. Pulmonary sequestration cyst in a patient of cerebral palsy. Lung India 2011;28:209-11.
Source of Support: Nil, **Conflict of Interest:** None declared.