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Pulmonary Sequestration Infant with Unusual Presentation: A Case Report

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Abstract

Background: Pulmonary sequestration (PS) is characterized by a separate segment of the lung that receives anomalous vascular supply separate from the pulmonary arteries. Here we report the case of a 4-year-old girl with dyspnea and recurrent pneumonia who was finally diagnosed as a case of PS.

Case presentation: A 4-year-old girl was admitted to Imam-Reza hospital, Mashhad city, Iran, with a history of coughing and dyspnea from two years ago. On the CXR, consolidation can be seen in the left lower lobe. As there was a high likelihood of aspiration or pneumonia following her past medical history, the patient underwent bronchoscopy. More investigation with HRCT revealed vascular anomaly. CMRI (Cardiac Magnetic Resonance Imaging) showed no cardiac abnormality. More findings showed a large collateral artery originated from left lateral side of abdominal aorta. This collateral artery went upward to the LLLL (the Lower Lobe of Left Lung) and anastomosed directly with two large posterior segmental tributary of the left lung pulmonary veins. Hyper-vascular pattern of both lungs was also observed in the MRI.

Conclusion: To avoid misdiagnosis, PS should be considered in differential diagnosis in infants with chronic cough and dyspnea. These patients should be referred to a tertiary center to receive appropriate treatment.

Key Words: Chronic Cough, Diagnosis, Dyspnea, Infants, Pulmonary Sequestration, Recurrent Pneumonia.

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1- INTRODUCTION

Pulmonary sequestration (PS) is a rare condition with an unknown etiology (1, 2). It is characterized by a separate segment of the lung that receives anomalous vascular supply separate from the pulmonary arteries. PS is classified into two groups: Intralobar sequestration (ILS), and Extralobar sequestration (ELS). ILS is more common and accounts for more than half of the PS cases (3). Here we report the case of a 4-year-old girl with dyspnea and recurrent pneumonia who was finally diagnosed as a case of PS.

2- CASE PRESENTATION

A 4-year-old girl was admitted to Imam-Reza hospital, Mashhad city, Iran, with a history of coughing and dyspnea from two years ago. She was a late preterm infant; born at 34 weeks of gestation, having a successful natural delivery. She has had a normal growth and development (z- score: 0.45). At the time of hospitalization, the child weighed 15 kg, and had a height of 96 cm. Two years prior to this, she developed dyspnea and cough, and was treated with Seretide, Salbutamol, and Flixotide as a case of children asthma. During this two years, she had history of constant hospitalization due to recurring incidences of pneumonia. She underwent bronchoscopy to rule out aspiration or infection due to a foreign body; the report showed that the mucosa of both bronchi were erythematous and inflamed. Therapeutic aspiration with normal saline was performed in both bronchi and then the suction of secretion was done. No foreign body was observed. The chest X- ray (CXR) showed a circle shadow behind the heart. For further investigation, High-Resolution Computed Tomography (HRCT) was used. The report showed a vascular anomaly. Furthermore a Cardiac Magnetic Resonance Imaging (CMRI), and angiography were performed on the patient. CMRI findings revealed that the cardiac situs was normal. Also, the cavities, valves, and great vessels' connections were normal. Interatrial and interventricular septums were intact. The aorta and the pulmonary artery including their branches were normal. The pulmonary vein, and its branches had normal size and anatomy. The systemic veins were also normal. However, the main abnormal anatomy was a large collateral artery (as big as renal artery in diameter), originating from the left lateral side of abdominal aorta, superior to renal artery origin. This collateral artery went upward to the lower lobe of left lung (LLLL) and anastomosed directly to two large posterior segmental tributary of the left lung pulmonary veins. 3D Magnetic Resonance Angiography (MRA) showed hyper-vascular pattern of both lungs that could explain the reason of dyspnea (**Fig. 1**).

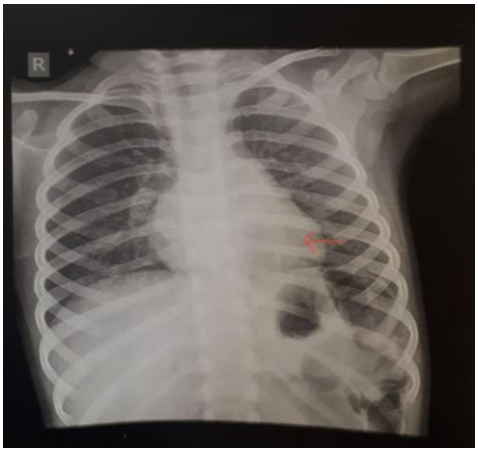

Fig. 1: X-chest showing an opacity in the left lower lobe of the lung

Angiography also confirmed the presence of a large collateral artery (**Fig. 2**).

To close the large abdominal aortic collateral artery, the child underwent surgery. The patient was posited on right lateral decubitus and a thoracotomy was operated at the sixth intercostal space. Lung parenchyma was normal. A large abdominal aortic collateral artery connected to the lower lobe of left lung (LLLL) was found; upon seeing the bronchial artery (at the anatomical location no.1) a no. 20 catheter was placed posteriorly, with the skin being intact and the chest tube being ligated.

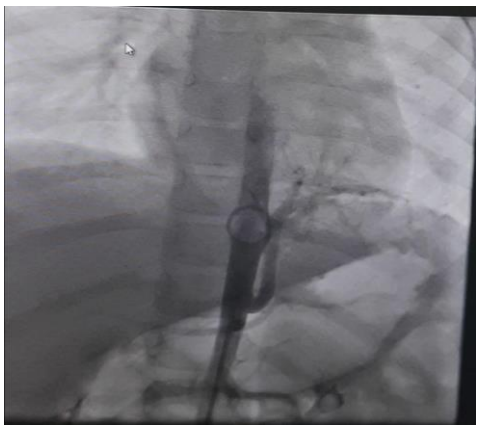

Fig. 2: Angiography showing collateral artery originated from abdominal aorta to the lower lobe of left lung

3- DISCUSSION

PS is classified into two groups: intralobar sequestration (ILS), and extralobar sequestration (ELS). ILS is defined as a sequestration segment within the common visceral pleura of the whole lung while ELS has its own visceral pleura. The other difference that separates these two types is the venous drainage; ILS venous drainage ends in the pulmonary vein, whereas ELS venous drainage is into systemic veins, frequently the azygos or hemiazygos veins. ILS accounts for 75% of pulmonary sequestration and is more often involved in the lower part of the left lung, while ELS constitutes 25% of the cases and is typically found in the posterior costophrenic sulcus (4, 5). ELS atrial supply is from systemic vessels including thoracic and abdominal aorta, celiac trunk, and splenic artery (3). Altogether it seems that our PS case was an ELS. Coexistence of ILS and ELS is extremely rare; however, some cases have been reported (6). PS should be suspected in any infant with recurrent respiratory distress, or in anyone with recurrent pneumonia (3); however, the only presentation of PS may be congestive heart failure. Choplin et al. (7) reported three cases of PS presenting congestive heart failure, these children were suspected to congenital heart disease. Shunting via sequestration may result in this presentation; as it was shown by solit et al. (8), PS could imbalance the cardiac output. This issue emphasized on excluding and ligation of abnormal vessels' connection in management of some PS patients. The other presentation of PS may be cyst on radiographic investigation. This may wrongly diagnose as pneumatocele, Cystic Adenomatoid Malformation (CCAM), bronchogenic cyst or infected pulmonary cyst, specially, if it coexists with fever or other presentations

of infection. This condition has been, previously, reported by choplin et al. (7). PS may be asymptomatic and be accidentally found in CXR. It was reported that a mass was accidentally found on CXR of a 14-year-old girl with scoliosis. At first, mesothelioma or carcinoma was suspected but more investigation confirmed PS (7). As it was discussed, CXR can provide informative presentation for diagnosis of PS; however, it is hard to differentiate between carcinoma, pneumatocele, Cystic Adenomatoid Malformation (CCAM), bronchogenic cyst, infected pulmonary cyst, mediastinal tumor, and diaphragmatic hernia by the information provided through CXR. Lin et al. showed that CT/MR angiography are valuable methods for obtaining a definitive diagnosis; they declared that CT angiography may be the gold standard for diagnosis (9). The complications of PS include hemoptysis, hemothorax, congestive heart failure and malignant transformation. Although mass lesion in CXR may propose PS or carcinoma, the diagnosis of PS cannot rule out carcinoma because localized carcinoma in the sequestrated lung tissue has also been reported as a rare complication of PS (1). Obstructive emphysema may also develop following compression of broncus (7). Edema is another complication caused by PS. In PS, increasing the overload of the left ventricular following significant venous return to left atrium can lead to pulmonary edema. This can explain the hyper-vascular pattern seen in the lung and frequent drug-resistant dyspnea (10). Resection of lesion is the treatment of choice for management of PS. To apply the right treatment for PS, all of the components should be considered, especially venous drainage; because anomalous venous drainage may involve the entire lung on the same side. Closing the collateral artery and resecting the PS without radical lobectomy has been proven to be helpful in management of PS and relieving the complications including pneumothorax, pneumonia, hemothorax, and malignancy (11-13). Rossi reported on a two month-old boy with PS, manifested with respiratory distress, and didn't respond positively to drug therapy; his physical examination was normal and his CXR showed consolidation in left lower lobe (similar to our case). A CT-angiography confirmed the consolidation in LLL and revealed a collateral artery originating from the left side of distal thoracic aorta ending at the site of consolidation. Also, a large vein was observed leaving the consolidation to the left lower pulmonary vein (14), although our patient had no significant cardiac complication. Rossi reported that their case suffered from dilation in left lower pulmonary vein, left atrium, and left ventricle. In addition, a Doppler echocardiography showed turbulent flow across the vessels at the site of consolidation. He applied PS excision on the patient and venous branches from PS were ligated for treatment. Rossi also investigated the pathologic specimen of the PS (11). The pathology section revealed prominent vascular lumina with the PS parenchyma containing a thickened artery with focal subendothelial fibrointima proliferation. There were also thickened abnormal airspace walls with poorly subdivided airspace containing numerous macrophage infiltration. This may explain the reason for inflamed mucosa (14). Wani et al. also reported a case of a 2-year-old boy with history of fever and cough. As CXR showed a consolidation in left lower lobe; CT angiography was performed. Their report showed that there was an anomalous artery originating from descending thoracic aorta. Wani et al. ligated the anomalous artery and then resected the sequestrated lobe (15).

4- CONCLUSION

Pulmonary sequestration (PS) is characterized by a separate segment of the lung that receives anomalous vascular supply separate from the pulmonary arteries. This condition should be suspected in any infant with recurrent respiratory distress, or in anyone with recurrent pneumonia. To avoid misdiagnosis, PS should be considered in differential diagnosis in infants with chronic cough and dyspnea. These patients should be referred to a tertiary center to receive appropriate treatment.

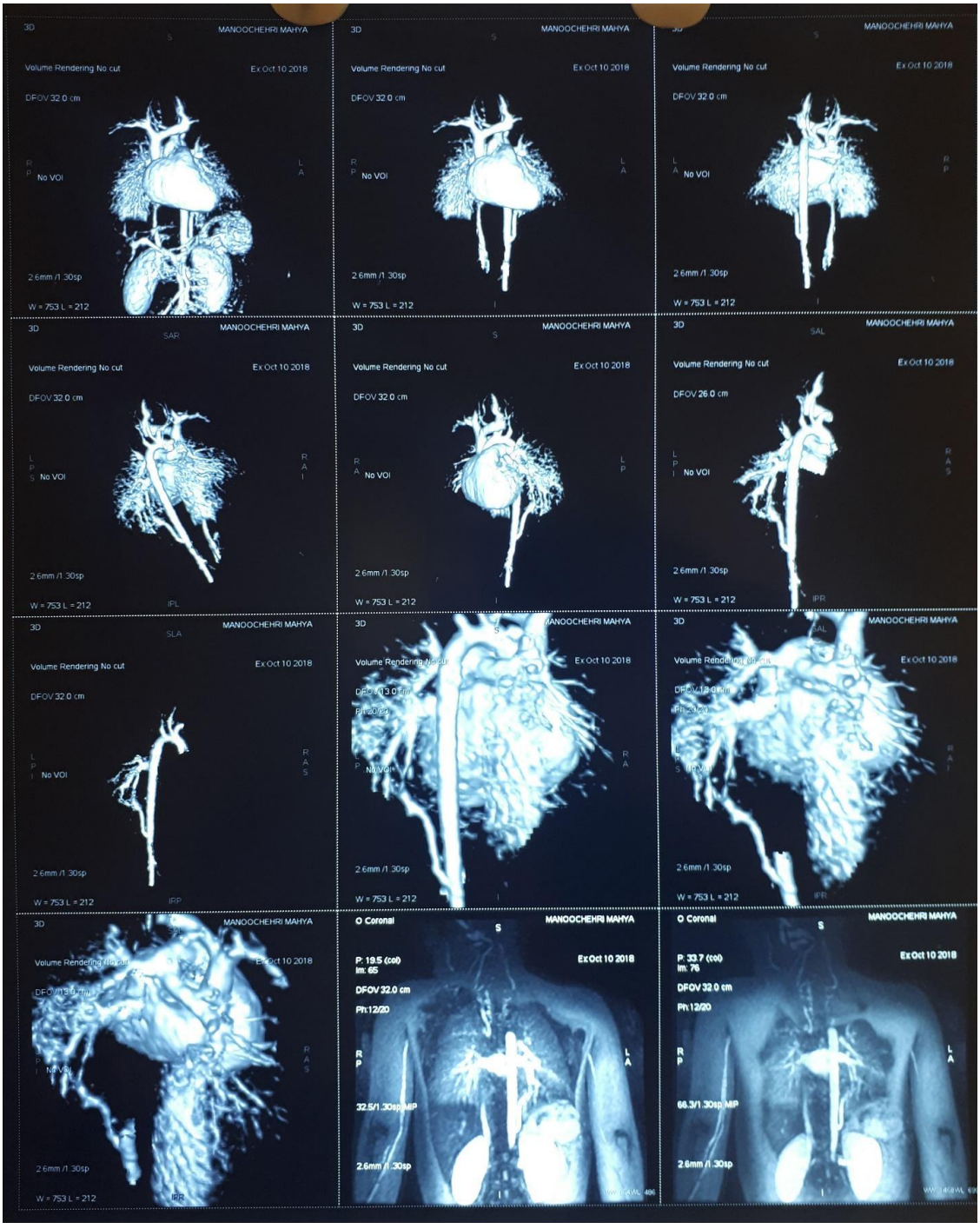

Fig. 3: 3D MRA showing collateral artery originated from abdominal aorta to the lower lobe of left lung and hyper vascular pattern of both lungs

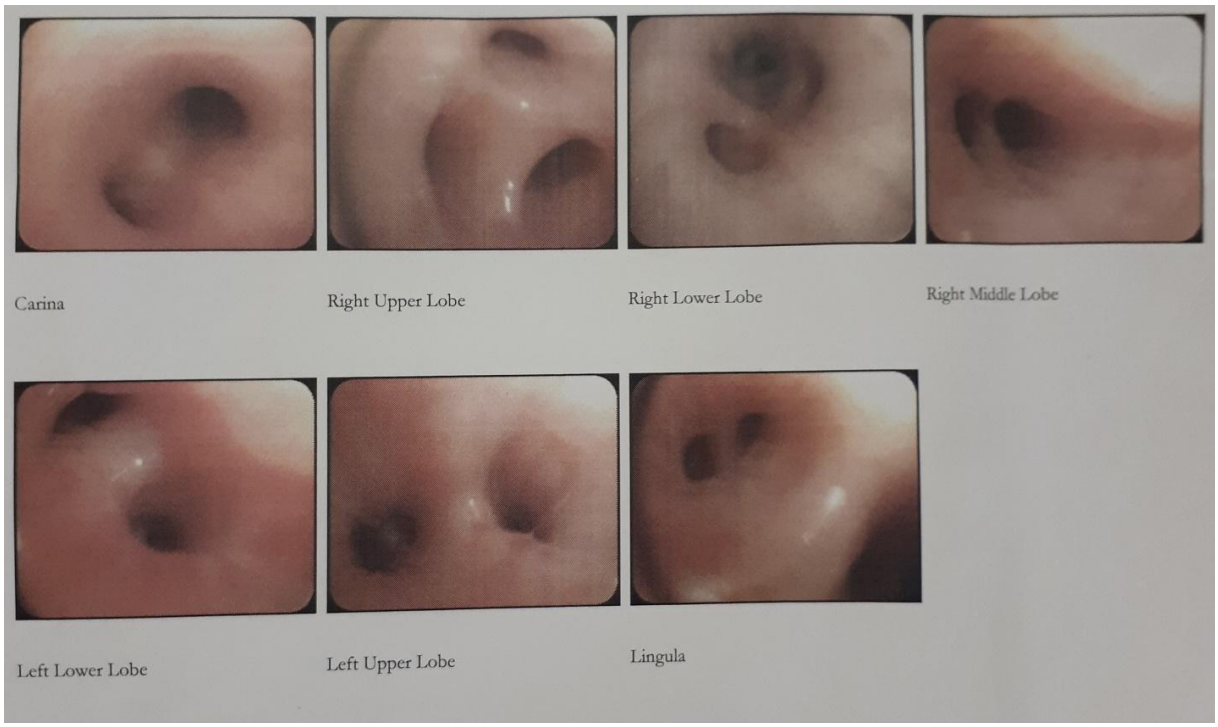

Fig. 4: Bronchoscopy showing erythematous and inflamed mucosa of both bronchi

5- ACKNOWLEDGMENT

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6- CONFLICT OF INTEREST

None.

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