Journal of the Postgraduate Institute of Medicine 2018; 5 (2): E73 1-4
http://doi.org/10.4038/jpgim.8187

Case Report

Successful embolization of aberrant pulmonary artery before elective lobectomy in a 13-year-old presenting with pulmonary sequestration complicated with bronchiectasis

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Keywords: Full Blood Count, Neutrophil/Lymphocyte Ratio, Cardiovascular Prognosis

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Submitted: July 2018, Accepted: August 2018, Published September 2018
Competing Interests: Authors have declared that no competing interests exist

Introduction

Pulmonary sequestration (PS) is a congenital malformation with localized, dysplastic lung tissue that lacks communication with the tracheobronchial tree but is fed by the systemic circulation [1].

Recent evidence shows that trans-catheter embolization is a minimally invasive, effective treatment option for symptomatic pulmonary sequestration [2,3]. We report a 13-year-old girl with bronchiectasis, presenting with an episode of massive haemoptysis who was successfully treated with embolization of the aberrant artery. This prevented the need for an emergency lobectomy and enabled a definite diagnosis of intrapulmonary sequestration.

Case

A 13-year-old girl diagnosed to have left lower lobe bronchiectasis at the age of 10 years, presented with massive haemoptysis of about a litre of blood over a 24-hour duration. She is the only child of a non-consanguineous marriage and has not had major respiratory concerns other than a productive chronic cough since the age of 3 years.

At the age of 10 years she had presented to the regional hospital with an episode of massive haemoptysis and received one blood transfusion. Chest X-ray was reported as left lower lobe pneumonia and she was treated for smear negative pulmonary tuberculosis based on the clinical history of a chronic cough, radiological features of the chest X-ray and the occurrence of haemoptysis. A high-resolution computer tomography (HRCT) was done during follow up which revealed left lower lobe bronchiectasis. However, follow up was defaulted due to social reasons.

She presented with two episodes of massive haemoptysis just prior to admission. She was pale but haemodynamically stable. Haemoglobin was 7.5 g/dl and the rest of the investigations were normal. She was given a blood transfusion and an HRCT was done which showed bronchiectasis confined to the left lower lobe. Although an elective lobectomy would have a better outcome, she was referred to a cardiothoracic surgeon for emergency lobectomy as she continued to have haemoptysis. However, as the emergency procedure would have been highly risky, the patient was referred to an interventional radiologist for selective bronchial artery embolization. The angiogram revealed an aberrant systemic artery originating from the descending thoracic aorta just adjacent to the sequestrated lung and embolization was performed.

An interval lobectomy was planned three months later, and she remained asymptomatic up to that point. Histology showed intra lobar sequestration in the removed 'lobe'. She is currently asymptomatic with improved lung function.

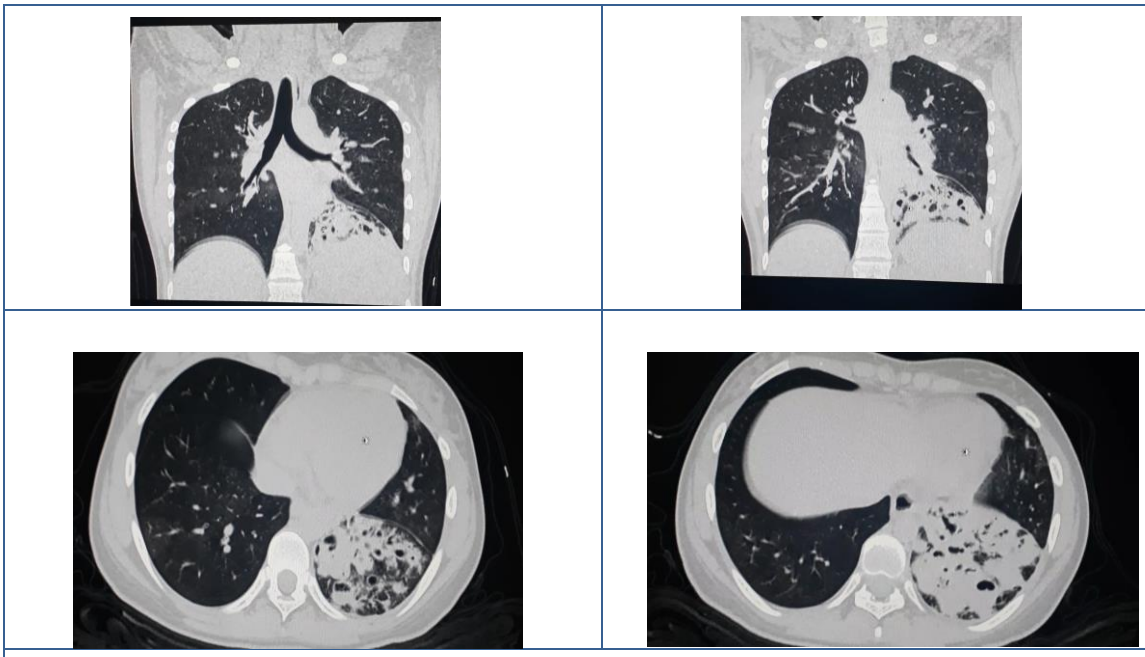

Figure 1: Sections of high-resolution computed tomography (HRCT) taken during current admission showing left lower lobe lesion

Discussion

Pulmonary sequestration (PS) is a rare congenital broncho-pulmonary malformation that represents 0.1–6% of all structural lung diseases and developmental malformations. Associated complications include infections, such as fungal infection or tuberculosis, fatal haemoptysis, haemothorax and malignancy [4].

Finding an abnormal feeding artery from the systemic circulation is the key to confirm PS. Classification into intra lobar sequestration (ILS) and extra lobar sequestration (ELS) is based on the location of the dysplastic lung tissue in relation to the parietal pleura,

presence or absence of its own visceral pleura and pulmonary versus systemic venous drainage [1,5].

The commonest form of ILS is contained within otherwise normal lung tissue, lacks its own visceral pleura and the veins normally drain into the pulmonary system. ELS, which is less common, is located outside the normal lung, has its own visceral pleura and the venous drainage, although variable, is commonly through the azygos system [6,7].

Our patient presented with recurrent episodes of productive cough from the age of 3 years with two episodes of haemoptysis. In a large case series of 2625 cases of pulmonary sequestration, documented in the Chinese National Knowledge Infrastructure from 1998 to 2008, Yong *et al* describe that the majority presented with cough, sputum production, fever, haemoptysis or chest pain and only 13% were discovered as incidental findings on imaging, without any respiratory symptoms [8].

Preoperative diagnosis of pulmonary sequestration is not possible in most cases, mainly due to the rarity of intrapulmonary sequestration in the paediatric age group [9]. Even after repeat HRCT, the diagnosis of our patient did not extend beyond bronchiectasis until the angiogram was done at the time of embolization. A surgical intervention was warranted as a life saving measure and selective bronchial artery embolization was offered as an elective lobectomy (after pre-operative assessment of lung functions) would carry a better prognosis [10].

In the past, all sequestrations were surgically resected to prevent recurrent pulmonary infections [11,12] but this has now become controversial based on current evidence. Embolization is preferred as it is associated with less complications compared to surgical resection. Spontaneous regression of the sequestration, with fibrosis, has been noted after embolization of the aberrant artery. In addition, embolization of the artery is thought to result in infarction of the dysplastic lung, leading to progressive regression that reduces intraoperative bleeding during the subsequent lobectomy [13]. The uncomplicated post-operative period seen in our patient after the lobectomy could be explained as a consequence of the preoperative embolization.

Conclusion

Bronchiectasis presenting with massive haemoptysis is rare in children and an underlying structural malformation needs to be suspected. The diagnosis of pulmonary sequestration could be challenging due to the variable presentations. Accurate identification of the vascular supply by a computer tomographic angiogram is important, especially prior to resection /lobectomy. Selective bronchial artery or aberrant artery embolization is life saving and carries multiple benefits.

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