ORIGINAL RESEARCH PAPER

Radiodiagnosis

A RARE PRESENTATION OF INTRALOBAR PULMONARY SEQUESTRATION RECEIVING ITS BLOOD SUPPLY FROM THE CELIAC ARTERY

KEY WORDS: Pulmonary sequestration, intralobar sequestration, celiac artery

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ABSTRACT

Pulmonary sequestration is a rare congenital malformation characterized by accessory lung tissue with no direct connection to the tracheobronchial tree and is supplied by an aberrant systemic artery. The correct diagnosis may be suspected on clinical grounds and should be confirmed by identifying the lesion and aberrant artery on imaging studies. Identifying the aberrant artery is a difficult problem when resecting a pulmonary sequestration. Abnormal blood supply is most commonly from thoracic and abdominal aorta. However, arterial supply from the celiac artery is quite rare. We present a rare case of 16 yr old female presenting with hemoptysis and diagnosed with intralobar pulmonary sequestration receiving blood supply originating from the celiac artery.

INTRODUCTION:

Pulmonary sequestration is a rare congenital malformation, an estimated incidence of 0.15% to 1.8% of the general population (4). There are two types of sequestrations: intralobar and extralobar, depending on whether they share their pleural with the rest of the lung or not [2,4]. Sixty percent of these lesions are diagnosed within the first decade of life and are more common in males by a 3:1 ratio [3]. Symptoms may vary and typically are related to chronic respiratory infection although sequestrations may be discovered incidentally on radiographic studies.

CASE HISTORY: -

A sixteen year old female with history of occasional cough, was admitted to the hospital complaining of hemoptysis and productive cough with no history of fever since three days.The patient was well built, vital signs were normal, physical exam was positive for decreased breath sounds and dullness on percussion over the right lower zone. Laboratory values were normal. Sputum AFB smear was negative.Her chest radiograph **(Figure 1)** showed a large thin walled cavity with air fluid level in the right lower zone. Computed tomography (CT) **(Figure 2)**of the chest with iv and oral contrast showed well defined heterogenous solid tissue with air lucenies measuring 6.3x4.7x6.5cm with in the medial & posterior basal segment of the right lower lobe, pulmonary veins seen traversing the tissue and anterior air component with arterial supply noted arising from the celiac artery **(Figure 3)** and extending upwards along posterior surface of liver traversing the diaphragam, artery divided and supplying superior and inferior margin of lesion, suspicious communication seen with lower lobe bronchus and no evidence of communication with oesophagus. Given with the history, imaging features characteristic of intralobar sequestration, and ongoing symptoms, the patient was referred to right lower-lobe resection. Right lower lobectomy was done containing thin walled Cavity contain thick mucus mixed with blood, one small bronchial communication with identification of aberrant vessel from celiac trunk directly perforating the underlying diaphragam. Pathology of the resected (Figure 4) specimen (10 x 8 cm) showed inflammatory lung parenchyma, with multiple cystic spaces with thin septa filled with purulent secretions with abdundant neutrophils. The pathological examination revealed an intralobar sequestration.

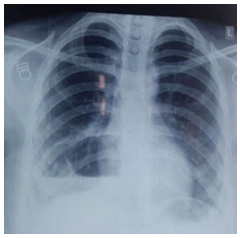

FIGURE 1 – CHEST X RAY -showing thin walled cavity in right lower zone.

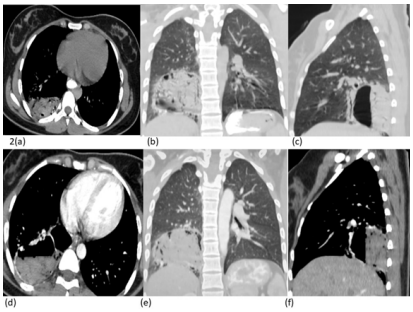

FIGURE 2 - CT of chest plain with oral and IV contrast AXIAL (a,d), CORONAL (b,e), SAGITTAL (c,f) showing intralobar pulmonary sequestration in right medial posterior basal segment .

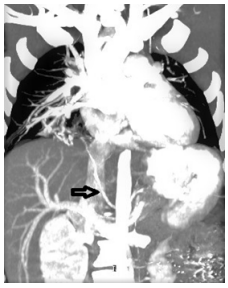

FIGURE 3 - Reformatted Maximal intensity projection (MIP) image showing the aberrant arterial supply from celiacartery(arrow), branch of abdominal aorta.

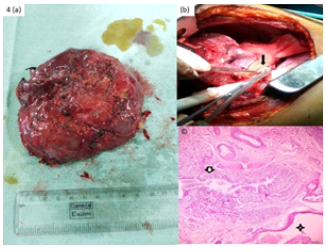

FIGURE 4 - (a) Resected gross specimen of sequestrated lung tissue (b) intraoperative image showing the aberrant arterial supply(arrow) arising from coeliac trunk (c) histopathology image showing bronchiole lined with epithelium(arrow), cartilage and surrounding blood vessels (star).

DISCUSSION: -

Pulmonary sequestration (PS) is a rare congenital malformation that is believed to develop from an accessory lung bud [1-16]. It was

firstly described by Rektorzik in 1861, classified it in two groups: intralobular (ILS) and extra lobular sequestration (ELS [1-16]. The resulting mass of dysplastic lung tissue has no direct connection to the tracheobronchial tree and is supplied by an aberrant systemic artery [1,4,6]. Multiple theories of the pathogenesis of pulmonary sequestration have been proposed, but all remain controversial. The most widely accepted hypothesis is that it results from formation of an accessory lung bud inferior to the normal lung buds during development and gets its blood supply from foregut vascular [5,10]. If the accessory lung bud develops before formation of the pleura, both the normal and sequestered lung tissue are covered by the same pleura, resulting in ILS; If the accessory lung bud develops after formation of the pleura, the sequestered lung tissue forms its own pleural covering, resulting in ELS [10].. The original connection with the foregut involutes in most cases, but it may sometimes persist, giving rise to a communication with the gastrointestinal tract. In such cases a tubular structure lined by columnar or squamous epithelium and containing intramural cartilage connects the esophagus or stomach to anomalous pulmonary tissue supplied by a systemic artery. These have been referred to as esophageal lung or gastric lung [17].

Localization of pulmonary sequestration

Almost 97% of PS cases were located in the lower lobe, and PS in the left lower lobe is two to three times more common than that in the right lower lobe [2-10] . As in our patient, the sequestration was in the right lower lobe (Figure 2)

Aberrant arterial supply of pulmonary sequestration

In most cases, PS has a single feeding artery,occasionally there are multiple systemic arteries. Arterial blood circulation is generally through systemic arteries such as thoracic aorta (73%), abdominal aorta (21%), intercostal artery (4%), the other feeding aorta include intercostal artery, diaphragmatic artery, aortic arch, subclavian artery, pulmonary artery, left gastric artery, coronary artery, celiac trunk and renal artery [2-4, 6]. In our patient, the enhanced CT scan demonstrated that the feeding vessel coursing from the celiac trunk directly into the cystic solid mass (Figure2).

Symptoms of pulmonary sequestration

Symptoms include recurrent respiratory infections, dysphagia, growth retardation, dyspnea and other respiratory problems. Symptoms are often nonspecific, the most common symptom of PS was cough or expectoration, fever, hemoptysis and chest pain [2-6].. Although the aetiology of the hemoptysis is uncertain, it is thought to be due to high-pressure blood flow in the sequestered lung from the anomalous systemic arteries. As in our patient, the remarkable symptoms were cough and hemoptysis.

Imaging of PS

Ultrasound plays an important role in the prenatal diagnosis, findings are nonspecific and has limited value in the antenatal, neonatal periods and in adults.as differential diagnosis is wide, visualization of a systemic feeding artery arising from the thoracic or abdominal aorta is a useful finding that distinguishes a sequestration from other masses such as a congenital cystic adenomatoid malformation or bronchial atresia [10].

On MRI sequestration typically appears as a well-defined mass in the chest that has a T2 signal intensity that is higher than that of the normal lung. MRI can demonstrate signal voids of the aberrant feeding vessel on T2-weighted imaging, and magnetic resonance angiography can demonstrate the systemic blood supply to the sequestered lung[17].

ELS most commonly manifests as a well-defined pyramidal, oval, or round mass in the pleural space near the posteromedial aspect of the ipsilateral hemidiaphragm and can be seen on fetal sonography as early as 16 weeks gestation. Because of separate pleural investment, ELS never contains air. On CT or MRI, ELS appears as a well-defined mass of uniform soft-tissue attenuation, there is often a single anomalous artery arising from the thoracic or abdominal aorta [10]

ILS has diverse imaging features. Plain chest radiograph is usually nonspecific, showing an ill-defined consolidation that mimics pneumonia,or shows a solitary soft tissue mass or nodule,or a cystic or multicystic lesion [4, 5]. On CT, ILS seen as a mass lesion, cystic lesion that may be filled with fluid, air, or both and cavitary lesion [3-5]. CT scan with intravenous contrast and preferably CT angiography (CTA) is the method of choice for identifying the arterial supply[7]

CURRENT TREATMENT

Definitive treatment involves resection of the affected lung segment. There are several key elements that should be considered: (I) a preoperative course of antibiotics in the setting of a pneumonia exacerbation can be beneficial by limiting the inflammation found at the time of surgery, (II) accurate preoperative identification of the arterial blood supply is crucial since inadvertent injury of these systemic vessels and (III) great care should be given to securing the systemic arterial branches at the time of surgery. Lobectomy is appropriate in scenarios when it is difficult to distinguish sequestered tissue from functioning parenchyma [1,8,10]. Asymptomatic ILS is often treated by surgery to avoid the risk of death in adulthood due to massive hemoptysis [1, 5]. Surgery usually involves lobar resection via standard thoracotomy or video assisted thoracic surgery (VATS). Since Wan et al. first described VATS lobectomy for treating pulmonary sequestration in 2002[10], the use of VATS for pulmonary sequestration resection has been used widely. Percutaneous endovascular embolization of the feeding systemic vessel has been performed for definitive treatment. However, the major concerns of the embolization include possible incomplete occlusion of vascular supply, subsequent evolution of the sequestered tissue and possible recurrence of symptoms [11].

CONCLUSION

Pulmonary sequestration refers to lung tissue that receives aberrant arterial supply from the systemic vessels, mainly the aorta and it`s major branches. In this case, the sequestered lung tissue receiving blood supply from celiac trunk,branch of the abdominal aorta, perforating through diaphragam.

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