Intralobar pulmonary sequestration presenting as a large multicystic mass

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ABSTRACT

A large multicystic mass lesion is not a common presentation of pulmonary sequestration. This presentation may result in failure to identify the supplying systemic artery due to multiplicity of the large cysts and, therefore, a wrong diagnosis. We discuss a case of intralobar pulmonary sequestration presented similarly. Chest x-ray and CT appearance gave an initial impression of hydatid disease which is endemic in Saudi Arabia. Fortunately, the supplying systemic artery was identified in the final CT review before surgery. Preoperative correct diagnosis of pulmonary sequestration is important to plan the appropriate surgery and to prevent possible intractable intraoperative hemorrhage. Awareness of this uncommon presentation of pulmonary sequestration helps to prevent such a fatal complication.

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A large multicystic mass lesion is an uncommon presentation of pulmonary sequestration. The supplying systemic artery can be overlooked due to multiplicity of the cysts and their large size. This may result in a wrong diagnosis leading to intraoperative hemorrhage which can be too severe to be controlled. We aim to increase the awareness of this uncommon presentation of pulmonary sequestration in order to prevent such a possible fatal hemorrhage.

Case Report. A 45-year-old woman presented with a recent history of chest tightness and one episode of hemoptysis. The patient gave a history of previous pneumonias on the right lung. The past medical history was otherwise, unremarkable. On examination, the right lower lobe was dull on percussion with decreased air entry. The chest x-ray showed multiple well-defined oval and rounded opacities in the right lung base **(Figure 1)**. A CT scan of the chest was performed and showed a large fluid filled multicystic mass in the right lower lobe **(Figure 2a)**. Initially, this was thought to be hydatid disease. Considering the symptoms of the patient and the size of the mass, excision of the lesion was contemplated. However, a feeding vessel from the descending aorta was seen during the final review **(Figure 2b)** and the diagnosis of sequestration was made changing the type of operation. The diagnosis was confirmed histopathologically. The post operative course was uneventful.

Discussion. Pulmonary sequestration represents an anomaly of tracheobronchial branching with retention of the embryonic systemic arterial supply.[1] The intralobar type accounts for 75% of pulmonary sequestrations and affects the lower lobes in 95% of cases.[2] The sequestered tissue usually derives its arterial blood supply from the descending thoracic aorta, although it can be from the upper abdominal aorta or one of its branches. Venous drainage in

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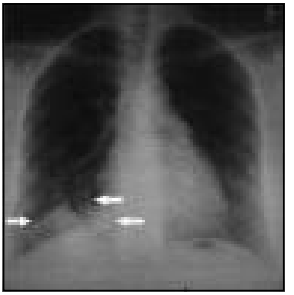

Figure 1 - Posteroanterior chest x-ray shows multiple oval and rounded lesions (arrows) in the right lung base.

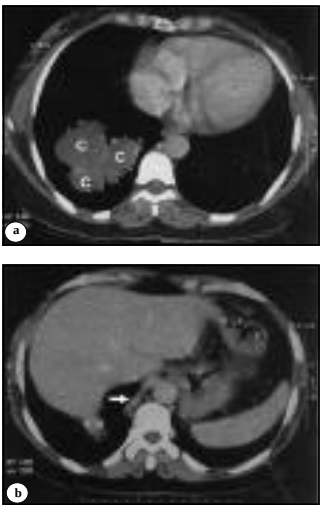

Figure 2 - A CT scan of the chest, **(a)** showing multiple fluid-filled cysts (C) in the right lung base, and **(b)** a lower cut shows the feeding systemic artery (arrow) originating from the distal thoracic aorta.

intralobar sequestration is usually to the pulmonary venous system, producing a left-to-left shunt; while extralobar sequestration commonly drains to the systemic veins.[3] The origin of intralobar sequestration, whether congenital or acquired is controversial.[1]

The major radiological patterns of intralobar sequestration depend on the degree of aeration and the presence of infection. The lesion may appear as a solid water-density mass or consolidation. Alternatively it may appear as an air-containing single or multicystic lesion.[2,3] On a chest radiograph, it presents most commonly either as a well-defined homogenous opacity or as a solitary lung nodule. It may mimic malignant lung tumor and, on rare occasions, may appear as an area of hyper-radiolucency. Pneumothorax may occur if the cysts rupture into the pleural cavity.[2] Pulmonary sequestration has a rare association with congenital cystic adenomatoid malformation (CCAM).[4] The cysts in CCAM usually contain air.[3,4] A completely fluid-filled large multicystic lesion is not a commonly described presentation of intrapulmonary sequestration in the literature. Our case is an example of such presentation. Unfamiliarity of this uncommon presentation explains the initial wrong impression of hydatid disease which is endemic in Saudi Arabia.[5] The feeding vessels can be overlooked radiologically leading to severe bleeding during surgery. These vessels are extremely retractile and friable and therefore must be carefully suture-ligated before excision.[4]

In a series of 24 cases,[6] the diagnosis of pulmonary sequestration has been achieved by imaging modalities in 21 of them (87.5%). The algorithm for diagnosis is controversial. With familiarity of the complex imaging findings, the diagnosis can be suggested by the findings on the chest x-ray and confirmed by showing the feeding systemic artery on ultrasound, CT, MRI or angiography.[2,6]

In conclusion, whenever there is a multicystic air-containing or fluid-filled lesion at the lung bases, careful search for a supplying systemic artery is recommended, as pulmonary sequestration may be the underlying pathology. Preoperative diagnosis of pulmonary sequestration helps to plan the appropriate surgery and to prevent a possible intractable intraoperative hemorrhage.

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