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Thoracoscopic resection of an unusually hypervascular extra-lobar pulmonary sequestration that resembled an arteriovenous malformation in a 2-year-old boy

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ABSTRACT

There is a risk for high-output congestive heart failure to be associated with an extra-lobar pulmonary sequestration (ELS) when there is left-to-left shunting caused by a large systemic arterial supply to a sequestration and venous drainage via the pulmonary veins into the left atrium. We present a 2-year-old boy who underwent thoracoscopic resection of an unusually hypervascular right ELS with a high output left-to-left shunt between the aorta and the left atrium via a pulmonary vein. This case is of particular interest because computed tomographic angiography identified a hypervascular nidus with indistinct borders between arterial and venous vessels suggestive of an arteriovenous malformation.

1. Introduction

Pulmonary sequestration may be extralobar (ELS) or intralobar (ILS). ELS has its own visceral pleura and comprises some 25% of all sequestrations. There is a male preponderance. Venous drainage is almost always into the pulmonary veins in ILS, but in ELS is only 21% [1]. Here, we present a case of ELS that appeared to be associated with a high output left-to-left shunt resembling an arteriovenous malformation (AVM) on computed tomographic angiography (CTA) that was excised thoracoscopically to prevent potential congestive cardiac failure (CHF).

2. Case report

Our case was first identified as a potential problem pregnancy when fetal magnetic resonance imaging (MRI) performed elsewhere confirmed the presence of a solid mass in the right mediastinum at 30 weeks gestation (Fig. 1). The pregnancy was otherwise unremarkable and a boy was born by spontaneous vaginal delivery at 38 weeks gestation. He was discharged well from hospital soon after birth. CTA performed elsewhere when our case was 2 months old was suggestive of a right pulmonary sequestration that was initially considered to be an ILS because it appeared to drain into the inferior right basal pulmonary vein. The boy was asymptomatic; CTR was 46%. Growth and development progressed unremarkably without any major health issues and repeat CTA performed when our case was 2 years old identified enough hypervascularity to suggest that an AVM with a feeding artery arising from the thoracic aorta may be present (Fig. 2a and b). CTR was 53%. At this point, the boy was referred to our institute for further management of an ELS complicated by a high output left-to-left shunt suggestive of an AVM (Fig. 3a and b). Surgical removal was indicated for the potential for an AVM-like lesion to precipitate CHF and parental anxiety.

Thoracoscopic resection was performed when the patient was 2 years old under general anesthesia using single lung ventilation in a lateral position. The operating surgeon and scopist stood on the left side of the patient with the monitor

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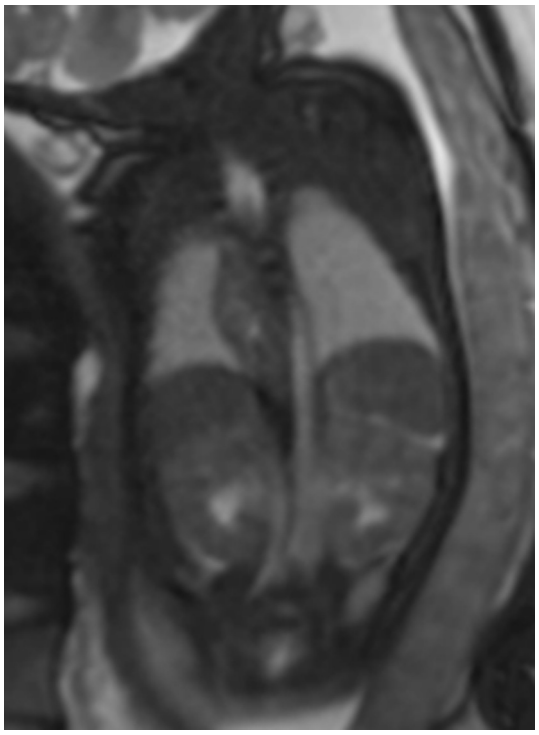

Fig. 1. T2-weighted fetal MRI images showing a solid lesion in the right mediastinum with partial high signal intensity.

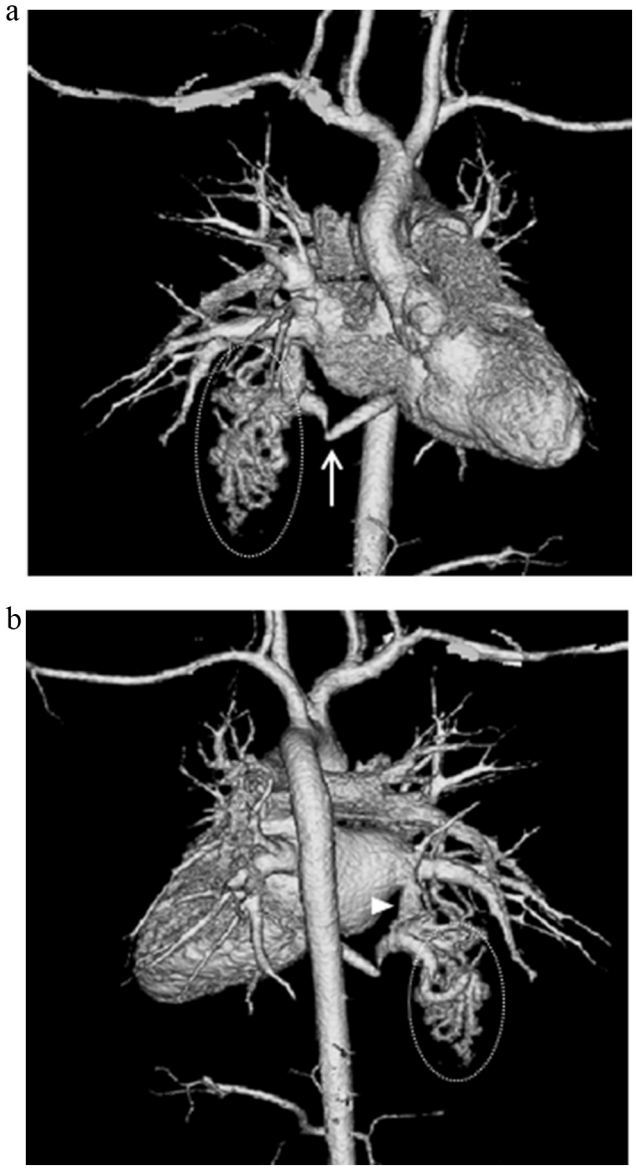

Fig. 2. a, b CTA delineates a hypervascular right pulmonary sequestration (dotted circle) with a feeding artery arising from the thoracic aorta (arrow) and draining into the inferior right pulmonary vein (arrowhead). There is a nidus of extra blood vessels with indistinct borders between arterial and venous vessels.

opposite them on the other side of the operating table. The first port was placed in the posterior axillary line in the fourth intercostal space. Additional ports were placed in the anterior axillary line in the third, fourth, and sixth intercostal spaces, and a final port was placed in the posterior axillary line in the eighth intercostal space.

The sequestration was confirmed to be an ELS, located in the right inferior mediastinum between the right diaphragm and the inferior lobe and was found to drain into the inferior right basal pulmonary vein, as shown on preoperative CTA. The feeding artery from the thoracic aorta and the draining vein were very close but with great caution, were successfully separated, hemo-clipped and divided (Fig. 4a, b, c, d). The ELS was extracted through one of the trocar sites. No chest tube was inserted. The postoperative course was unremarkable and discharge from hospital was possible on postoperative day 2. CTR had improved from 53% preoperatively to 47% at the time of discharge. Histopathology of the excised sequestration confirmed the presence of a proper visceral pleural membrane typical of ELS (Fig. 5a) with alveolar epithelium, bronchus with cartilage, and increased abnormal thick walled arteries and veins. Vessel walls were fragile and tortuous, but there was no evidence of bleeding or thrombus. AVM was excluded because there were no direct connections between arteries and veins observed (Fig. 5b).

3. Discussion

When an ELS drains via pulmonary veins, a left-to-left shunt can develop and be the cause of high output CHF. Such a shunt is so uncommon that extreme caution is warranted during preoperative investigations to assess the anatomy of the suspected anomaly accurately and may also exacerbate symptoms in patients with incidental intracardiac anomalies [2–4]. Spinella et al. [2] reported that high-output congestive heart failure (CHF) may be associated with an ELS when there is left-to-left shunting caused by a large systemic arterial supply to a sequestration and venous drainage via the pulmonary veins into the left atrium. Thus, an ELS could function as an arteriovenous malformation (AVM) because it shunts large amounts of systemic arterial blood back to the heart to cause high output CHF.

Conventionally, angiography is used to assess vascular flow once a diagnosis of pulmonary sequestration is made but it is an invasive procedure, particularly in children. Here, we used CTA because it is less invasive than angiography and also has the advantages of