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Case Report

Extra-lobar Pulmonary Sequestration with Associated Asymptomatic Congenital Diaphragmatic Hernia

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ABSTRACT

Background

Pulmonary sequestration is a rare aberrant formation of non-functional pulmonary tissue that does not communicate with the tracheobronchial tree or pulmonary blood supply. It can be associated with congenital diaphragmatic hernias (CDH) which typically present in the newborn as respiratory distress and feeding difficulties.

Case Presentation

A full term infant presents with an asymptotic left sided CDH with associated supra-diaphragmatic extra-lobar pulmonary sequestration intermittently sliding through the diaphragm. Prenatal imaging raised suspicion for CDH; however, postnatal imaging in an asymptomatic infant confirmed suspected sequestration and suggested intact diaphragm. The infant was taken to the operating room for elective resection of the suspected supra-diaphragmatic sequestration. Intraoperative findings demonstrated a supra-diaphragmatic extra-lobar pulmonary sequestration obscuring the undiagnosed CDH.

Conclusions

This rare presentation of a supra diaphragmatic pulmonary sequestration intermittently herniating through an unrecognized asymptomatic CDH emphasizes the potential for unexpected intra-operative findings of bronchopulmonary foregut malformation with associated CDH despite prior radiographic imaging results.

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Background

Pulmonary sequestration is the aberrant formation of non-functional pulmonary tissue that does not communicate with the tracheobronchial tree or the pulmonary blood supply. Its incidence has been reported to be approximately 0.15-6.4% [1]. Extra-lobar sequestration accounts for approximately 25% of all sequestrations and has been reported to be infra-diaphragmatic in approximately 10% of cases [2, 3]. Pulmonary sequestration is present in 15-30% of patients with congenital diaphragmatic hernia (CDH) [4-7]. Congenital diaphragmatic hernias typically present in the newborn as respiratory distress and feeding difficulties. Herein, we describe an infant with the rare presentation of an asymptomatic left sided congenital diaphragmatic hernia with associated supra-diaphragmatic extra-lobar pulmonary sequestration intermittently sliding through the diaphragm.

Case Presentation

A full-term infant (39 weeks 0 days) born via spontaneous vaginal delivery was admitted to the NICU following planned intubation for prenatal ultrasound and MRI revealing a mass suggestive of left sided CDH (Figure 1). At birth, the infant weighed 3280 grams, presented with no respiratory distress and Apgar scores of 9 and 9 at 1 and 5 minutes, respectively. Post intubation X-ray showed the orogastric tube in the stomach, well below the diaphragm. Postnatal ultrasound was obtained to evaluate for the suspected CDH; however, it revealed an intact

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diaphragm, without herniation of any viscus, as well as the presence of a 3 x 2 x 2 cm intra-abdominal mass with radiographic features suggestive of a pulmonary sequestration (Figure 2). As the infant required minimal ventilator support and ultrasound was negative for CDH, the child was extubated. At one and two months postnatal follow up, the infant had remained asymptomatic with no reported hospitalizations, pneumonias, growth delay, or respiratory or feeding difficulty. Preoperative CT imaging was obtained at six months, demonstrating a left extra-lobar bronchopulmonary sequestration, now pictured above the diaphragm, with arterial supply arising from the abdominal aorta at the level of the celiac artery, and suspected venous drainage from the left renal vein running superiorly to the mass (Figure 3). The infant was taken to the operating room for elective resection of the suspected supra-diaphragmatic sequestration via thoracoscopy. Intraoperative findings demonstrated a supra-diaphragmatic extra-lobar pulmonary sequestration obscuring the undiagnosed CDH. Inspection of the specimen revealed adjacent diaphragm tissue. A left sided CDH was confirmed (Figure 4) and repaired primarily thoracoscopically. The infant recovered well and post-operatively remains asymptomatic and free of complications. At 6-month follow-up there is no evidence of recurrent CDH, and the toddler is thriving.

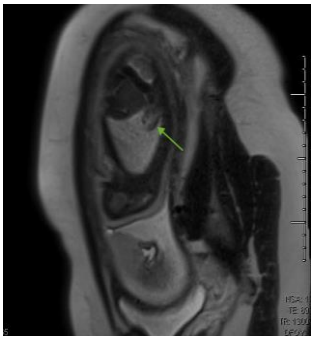

Figure 1: Saggital cut of prenatal MRI abdomen/pelvis demonstrating the presence of a mass suggestive of a left sided CDH.

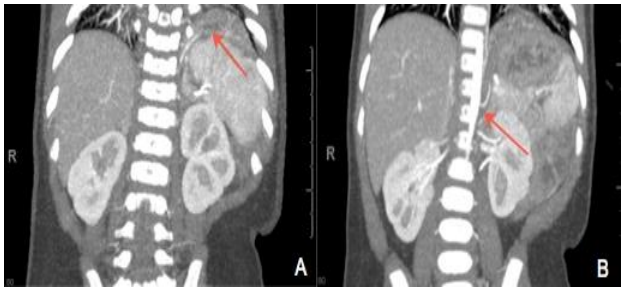

Figure 2: Postnatal abdominal ultrasound, longitudinal view, showing presence of 3 x 2 x 2 cm mass below the diaphragm and adjacent to spleen, suggestive of pulmonary sequestration.

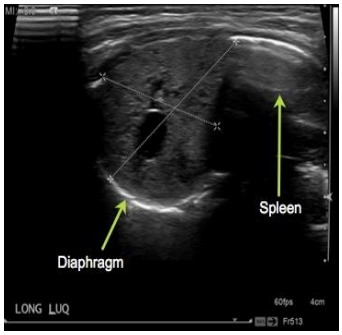

Figure 3: (A) Coronal cut of CT abdomen/pelvis at 6 months, demonstrates partially cystic mass consistent with a left supra-diaphragmatic extra-lobar bronchopulmonary sequestration.

(B) Arterial supply arising from the abdominal aorta

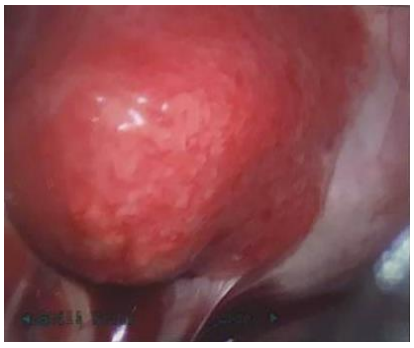

Figure 4: Thoracoscopic view of left extra-lobar pulmonary sequestration overlying a congenital diaphragmatic hernia.

Conclusions

The purpose of this report is to describe a rare presentation of a supra diaphragmatic pulmonary sequestration seen on imaging studies to be intermittently herniating through an unrecognized asymptomatic CDH. Pediatric surgeons should be prepared for potential intra-operative findings of bronchopulmonary foregut malformation with associated congenital diaphragmatic hernia despite pre-operative radiographic findings.

Abbreviations

CDH - congenital diaphragmatic hernia

Ethics approval and consent to participate

No UMC-IRB approval was required for publication of this case report.

Consent for publication

Written consent for publication obtained from the patient's guardians.

Availability of data and materials

No data set used for this article.

Conflict of interest

The authors have no conflicts of interest.

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Each author has made substantial contributions to the information or material submitted for publication.

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