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Pediatric

Arterial tortuosity syndrome: An extremely rare disease presenting as a mimic of pulmonary sling

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ABSTRACT

Pulmonary sling is the anatomic variant defined by the aberrant origin of the left pulmonary artery from the right pulmonary artery. This patient presented with a mimic of pulmonary sling as a result of an extremely rare condition, arterial tortuosity syndrome (ATS). The patient was first diagnosed with pulmonary sling on prenatal echocardiogram performed by cardiology. Computed tomography angiography of the chest obtained at birth to evaluate respiratory depression demonstrated ATS. The early detection of ATS has been demonstrated to improve patient outcome. This case provides an overview of the typical imaging features of ATS to aid radiologists in making this uncommon diagnosis.

Introduction

Pulmonary sling is the anatomic variant defined by the aberrant origin of the left pulmonary artery from the right pulmonary artery, which courses between the trachea and the esophagus. This anatomy often results in stridor and respiratory distress because of posterior compression of the trachea. Pulmonary sling additionally results in anterior impression of the esophagus. These features differentiate the pulmonary sling from the pulmonary rings, which result in anterior compression of the trachea and posterior impression of the esophagus.

A failure of formation of the sixth aortic arch is postulated to be the cause of pulmonary sling. The course of the pulmonary artery adjacent to the right mainstem bronchus can result in compression and air trapping in the right lung. An aberrant left pulmonary artery is associated with complete tracheal rings, which may result in a long-segment tracheal stenosis, leaving patients at higher risk of respiratory compromise [1].

Alternative causes of pulmonary sling symptomatology and pathophysiology are rarely reported in the literature. One cause identified includes duplication of the left pulmonary artery, another rare vascular anomaly [2]. Symptomatic patients with pulmonary sling are surgically managed shortly after presentation because of the high mortality associated with this condition [3].

Case report

A 27-year-old G1P001 woman with a history of chronic hypertension underwent fetal echocardiogram with pediatric

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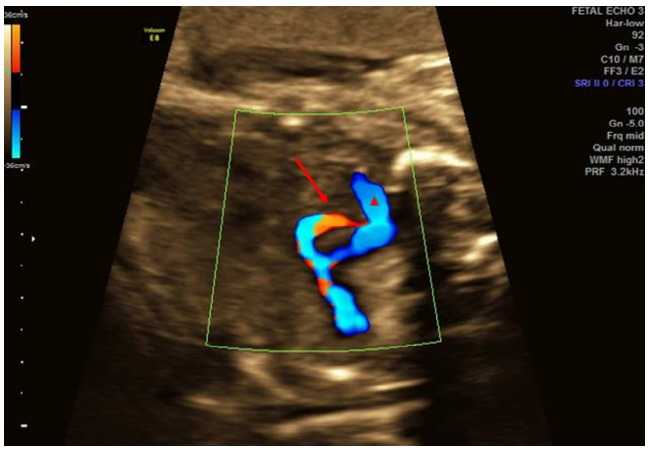

Fig. 1 – Fetal echocardiogram at 28 weeks and 4 days' gestation demonstrates an aberrant vessel with the appearance of a pulmonary sling. The arrowhead marks the main pulmonary artery, whereas the arrow marks the aberrant vessel, presumed to be the left pulmonary artery originating from the right pulmonary artery.

cardiology at 28 weeks and 4 days' gestation that demonstrated findings of a pulmonary artery sling (Fig. 1). Additional prenatal evaluation included ultrasound at 19 and 28 weeks performed by maternal fetal medicine that demonstrated a small appearance of the fetal stomach and no other evidence of fetal structural malformation.

The mother was Group B Streptococcus positive and her pregnancy was further complicated by chorioamnionitis. The mother ultimately underwent emergent cesarean section at 37 weeks and 2 days for a prolapsed cord. The neonate was a 3.21-kg male with Apgars of 5 and 7 at 1 and 5 minutes (decreased tone, little spontaneous activity, and poor respiratory effort). The patient required positive-pressure ventilation and was admitted to the neonatal intensive care unit (NICU) for continuous positive airway pressure.

Given the prenatal findings of pulmonary artery sling and respiratory depression at birth, the patient underwent a computed tomography angiography (CTA) using our congenital heart protocol on the first day of life (Figs. 2 and 3). Significant vascular findings included a markedly tortuous aorta, a dilated main pulmonary artery, and patent ductus arteriosus.

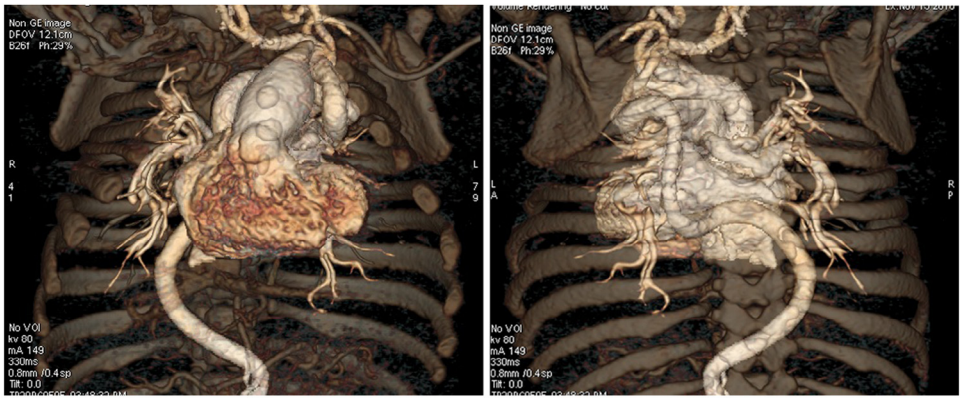

Fig. 2 – Anterior and posterior projections of the 3-dimensional reconstructions of the heart and vessels from computed tomography angiography congenital heart protocol obtained on day 1 of life demonstrate a marked tortuosity of the aorta, patent ductus arteriosus, and a dilation of the main pulmonary artery. DFOV, display field of view; GE, General Electric.

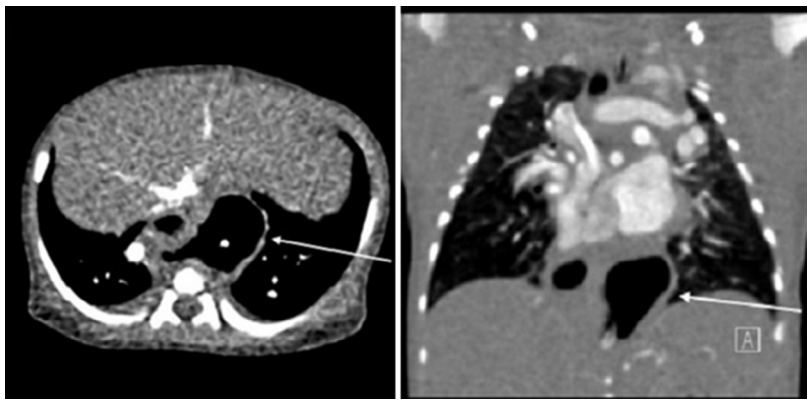

Fig. 3 – Axial and coronal images from a computed tomography angiography congenital heart protocol obtained on day 1 of life also demonstrate a large hiatal hernia with nasogastric tube in place. The hiatal hernia is marked with arrows.