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Case Report

Intraesophageal aortopulmonary collateral artery occlusion in an infant with Tetralogy of Fallot and flow-dependent major aortopulmonary collateral arteries

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ABSTRACT

Patients with flow-dependent aortopulmonary collateral arteries often exhibit tenuous clinical statuses. Here we present an infant with Tetralogy of Fallot with pulmonary atresia (TOF-PA) and major aortopulmonary collateral arteries (MAPCAs) who experienced clinically significant oxygen desaturations during feedings. These frequent episodes were attributed to gastroesophageal reflux. In preparation for possible surgical correction of her complex congenital heart condition, she was intubated for a cardiac catheterization to better characterize her anatomy, but terminally decompensated shortly after extubation and restarting feeding. On autopsy, very mild evidence of esophageal reflux was present; instead, one of her four aortopulmonary collateral arteries was unexpectedly discovered traversing through the muscular layer of her esophageal wall. This finding was confirmed microscopically with significant intimal hyperplasia compared to uninvolved collateral arteries. The remainder of the autopsy findings, including her myocardium, lungs, and esophagus, were unremarkable. Given the feeding-associated oxygen desaturations present in life and the aberrant collateral artery's comparative stenosis at death, we speculate that the repetitive trauma of esophageal peristalsis was not only significant enough to temporarily impact oxygenation but, over time, led to near-occlusion of the intraesophageal collateral artery, which may have contributed to the terminal decompensation of this already tenuous patient.

1. Introduction

The decedent was a 79-day-old female born at 33-week-4-day gestation via cesarean section. She was small for gestational age at birth and had low Apgar scores. Upon transfer to the neonatal intensive care unit, a chest x-ray demonstrated a “boot-shaped heart” and subsequent work up revealed a 22q.11 deletion, TOF-PA with MAPCAs, a closed ductus arteriosus, and severe hypocalcemia. She was kept in the NICU to mature until she was of weight suitable for cardiac surgery. During her 2 months of life, her diet was advanced from total parental nutrition (TPN) to tube feeds, her hypocalcemia was responsive to dietary supplementation, and her aortopulmonary collaterals continued to adequately shunt blood to her lungs. Her main clinical issue was persistent oxygen desaturations during feedings, which were attributed to esophageal reflux but ultimately unresponsive to anti-reflux medication. The day before her death, she was intubated and feeds were held for cardiac catheterization. The cardiac catheterization showed true confluent branch pulmonary arteries that had grown since birth. She was extubated the following day and gastric tube feeds were restarted. Later that evening, she experienced persistent stridor that rapidly progressed to respiratory failure and required re-intubation. A code event was called and she received multiple interventions to support her oxygenation and heart rate but she continued to decompensate. A chest x-ray and hemoglobin level taken during this time were unremarkable and an emergent echocardiogram showed decreased cardiac function with collateral arteries that could not be assessed due to bradycardia. Despite multiple code medications the patient ultimately expired.

2. Discussion

Pulmonary circulation in patients with TOF-PA and a closed ductus arteriosus is completely dependent upon congenital collateral arteries that arise from the aorta and supply the lungs. Such collaterals exhibit significant anatomic variability and show a higher propensity for stenosis, irregular intimal hypertrophy, and kinking than their normal pulmonary artery counterparts. While these maladaptive structural modifications may be attributed to increased blood velocity, sheer stress, and exposure to systemic pressure, recent gene expression profiling supports a unique genetic identity among collateral arteries that further explains their propensity for complications [1]. Despite their tenuous vascular integrity, children with adequate-sized aortopulmonary collaterals are often successfully managed by complete primary repair once reaching a certain age and/or weight suitable for surgery. (See Fig. 1.)

The decedent's family requested a full autopsy. Post-mortem examination revealed a small heart (17.2 g, expected reference 27 g) with a significant conoventricular septal defect, an overriding aorta, an absent pulmonary artery and valve, right ventricular hypertrophy and dilation,

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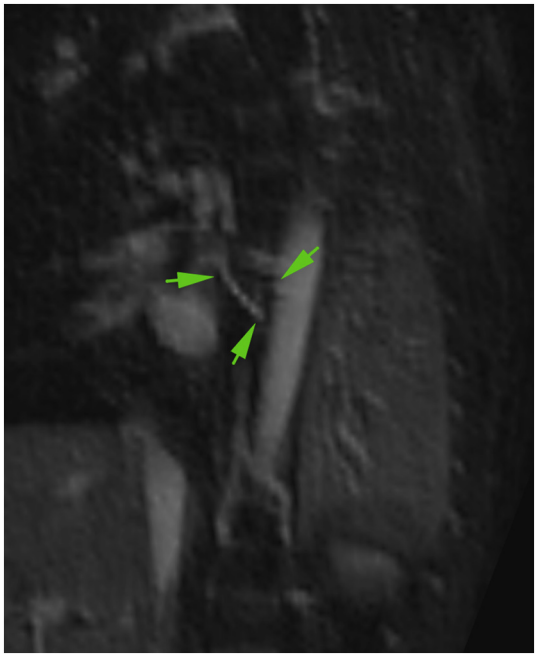

Fig. 1. 3-dimensional magnetic resonance imaging (MRI) shows major pulmonary collateral arteries coursing from the thoracic aorta toward the not-visualized pulmonary trunk. Arrows highlight the anomalous intraesophageal collateral artery appreciated on autopsy.

and a patent foramen ovale. Microscopically, the myocardium was unremarkable. A confluent pulmonary artery trunk was present posteriorly, which was fed by four major collateral arteries arising from the thoracic aorta. Notably, one collateral artery was encased within the muscularis of the esophagus (Figs. 2 and 3). Microscopically, this intraesophageal collateral artery displayed significant intimal fibromuscular hyperplasia and was 90% stenotic (Fig. 3). The three uninvolved collateral arteries did not display any degree of fibromuscular hyperplasia nor stenosis. The lungs were small (39.2 g, expected reference 74 g) and showed bilateral alveolar wall thickening, consistent with pulmonary hypertension. The pulmonary parenchyma was unremarkable. There was no evidence of aspiration. The trachea showed mild erythema and edema. The esophageal mucosa was unremarkable. (See Fig. 4.)

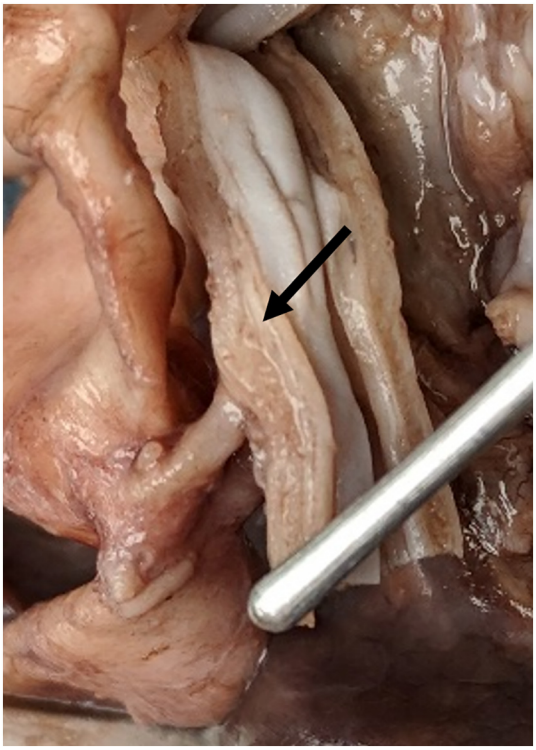

Fig. 3. Gross photograph of the posterior esophagus cut surface shows a cross section of the traversing aortopulmonary collateral artery between the muscular layers of the esophageal wall (arrow).

3. Conclusions

Patients with flow-dependent major aortopulmonary collateral arteries are at risk for a host of structural anomalies that contribute to a tenuous clinical state. A previously undescribed anomaly presented here is the traversing of a collateral artery through the muscular layer of the esophagus en route to the lungs. An intriguing possibility is that the patient's frequent oxygen desaturations during feedings were due to esophageal peristalsis, which functionally, if temporarily, restricted flow through one of her critical collateral arteries. Repetitive irritation from esophageal

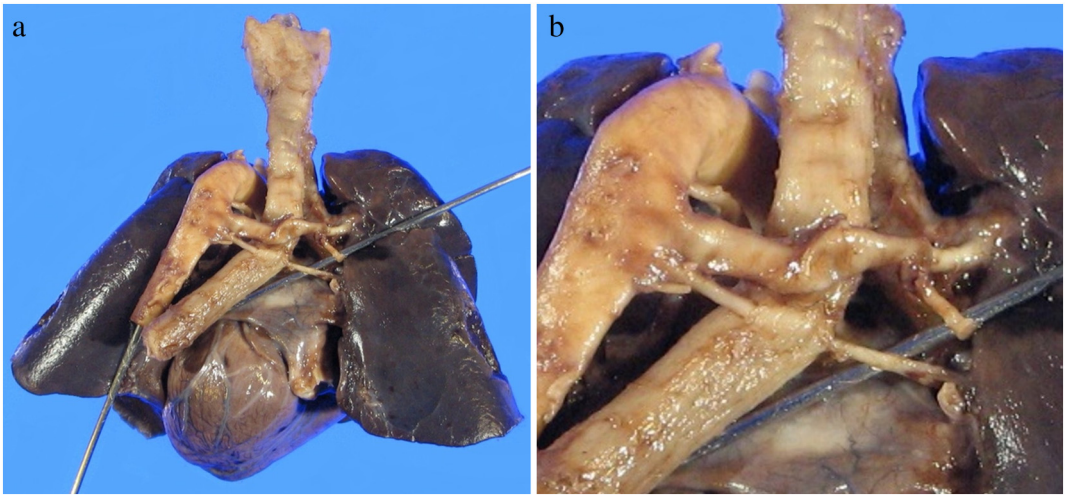

Fig. 2. Gross photographs from the posterior aspect of the excised cardiothoracic block demonstrate multiple collateral arteries arising from the descending aorta and inserting into the pulmonary trunk. The bottom-left probe in part (a) displaces the descending aorta away from the underlying esophagus while the probe on the right highlights the intraesophageal collateral artery. A close-up of the same image in part (b) shows the relationship of the collateral artery to the muscular esophageal wall.

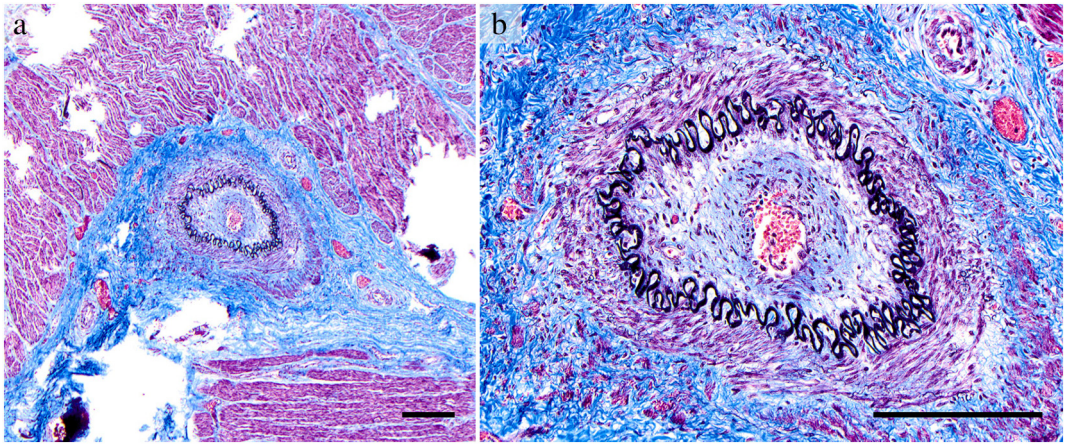

Fig. 4. Microscopic images of the intraesophageal aortopulmonary collateral artery with Masson's Trichrome stain. Low power view (a) shows a muscular blood vessel (center) with abundant surrounding fibrosis in blue, all encased within fascicles of esophageal smooth muscle (bar=200 μm). Higher power (b) demonstrates significant intimal fibromuscular hyperplasia leading to 90% luminal stenosis. The elastic lamina characteristic of arterial vessels (black) remains intact (bar=100 μm).

contraction may have resulted in maladaptive vascular remodeling as evidenced histologically by severe intimal hyperplasia and stenosis, findings not seen in the other major collateral arteries. Clinically, it was suspected that the infant's demise was related to refractory reflux and aspiration; however, the postmortem examination showed only mild airway edema and no evidence of aspiration in the lungs. The most significant pathology found at autopsy was a stenotic intraesophageal aortopulmonary collateral artery. Despite its relatively small caliber compared to the other major collateral vessels, we believe that progressive narrowing of the intraesophageal collateral artery eventually resulted in sufficient reduction of blood flow to cause respiratory failure and ultimately death.

Conflicts of interest

None declared.

Source of funding

None.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.carpath.2018.12.009.

Reference

[1] Ma X, Barboza LA, Siyahian A, et al. Tetralogy of Fallot: aorto-pulmonary collaterals and pulmonary arteries have distinctly different transcriptomes. Pediatr Res 2014; 76:341–6.