Case report

Vascular anomaly: Cause of infant respiratory distress and dysphagia

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1. Introduction

Aberrant right subclavian artery is the most common anomaly arising from the aorta. Its incidence is between 0.5% and 2% worldwide [1,2]. It usually arises distal to the left subclavian artery from a dilated segment of the proximal descending aorta, and crosses in the posterior part of the mediastinum to the right upper extremity. In majority of the cases it crosses behind the esophagus or occasionally between trachea and the esophagus [3,4]. This can result in dysphagia, respiratory distress and stridor. Here we report the case of an aberrant right subclavian artery in a baby crossing behind the esophagus leading to respiratory and digestive issues.

2. Case report

A 2 week old girl was admitted with respiratory failure. She had been born full term with no respiratory related issues, but had experienced trouble gaining weight since birth. She was initially admitted at 2 weeks of age to the hospital after an episode of perioral cyanosis and dyspnea after feeding. Her oxygen saturation in the ER was 50–60% with significant tachypnea at 80 breaths per minute. Initial chest x-ray showed diffuse bilateral haziness. Her initial complete blood count was normal. Due to her tenuous respiratory status she was intubated and ventilated for 8 days. She had a full sepsis work up which was negative.

After extubation, her respiratory rate decreased to 50–60 breaths per minute. She had chest wall retraction with bilateral lung crackles and required supplemental oxygen 0.5–1 Lpm to maintain SpO2 > 95%.

At one year of age she was finally weaned off oxygen. However, she continued to develop significant respiratory distress and hypoxemia with upper respiratory tract infections. Additionally, she had delay of her developmental milestones and mild hypotonia. There was no family history of immunodeficiency, surfactant deficiency or underlying genetic disorders. She always had trouble gaining weight and would experience coughing, choking and frequent spit up with liquid feeds. On her follow up physical exam in the clinic, she was not dysmorphic but had increased clubbing.

Due to her unusual presentation with recurrent respiratory illnesses and feeding difficulties, it was necessary for an aerodigestive approach to evaluate her. Chest x-ray showed mild central right lung haziness and bronchial wall thickening. Fig. 1.

Chest computed tomography (CT) with IV contrast confirmed a left side aortic arch with an aberrant right subclavian artery with a retro-esophageal course, Fig. 2A. There was no aneurysmal dilatation suggestive of Kommerell’s diverticulum.

The lungs show bilateral perihilar ground glass haziness in the lower

A B S T R A C T

Aberrant right subclavian artery with a left aortic arch is rare, but it is the most common congenital aortic arch anomaly. It can present as an incidental finding later in life or be symptomatic at a young age. Here, we describe a case of an aberrant right subclavian artery discovered in a 4 month old with respiratory distress and feeding difficulties. She underwent an extensive aerodigestive evaluation including bronchoscopy, both flexible and rigid, upper GI endoscopy, modified barium swallow with esophageal sweep, chest imaging, CT thorax and echocardiogram. The final decision per the management team was to observe the patient in order to allow more growth. She ultimately improved with age and remains asymptomatic.
A sweat chloride test was normal and no CFTR mutations were found on direct mutation, full gene sequencing and duplication/deletion analysis. Surfactant mutations for ABCA3, SPB and SPC were negative. Pancreatic fecal elastase was negative. FISH analysis showed normal 22q11 with no evidence of deletion or duplication.

Flexible bronchoscopy was completed which showed a pulsatile posterior membranous distal tracheal wall with a mild distal posterior tracheal bulge with copious amount of frothy white secretions in the right and left main stem bronchi. Rigid laryngoscopy did not reveal a laryngeal cleft or tracheoesophageal fistula. The right main stem bronchus did appear to be of smaller caliber than the left main stem bronchus Fig. 3(A and B).

The bronchoalveolar lavage was non-inflammatory with predominance of lipid laden macrophages.

Concurrent esophagogastroduodenoscopy revealed a compressed portion in the upper to mid third of esophagus. The esophagus could be minimally distended in this region, but not to the same caliber as the rest of the esophagus. Dilation or significant pressure was not required to advance distally. The mucosa was visually normal, and the proximal esophagus was not noted to be dilated. Fig. 4.

Following completion of the combined endoscopic procedures, and with continued cough related to feeding, a video fluoroscopic swallow study was performed. The study confirmed the presence of a compression, consistent with an aberrant right subclavian artery. Fig. 5.

A repeat endoscopy with bronchoscopy performed approximately one year later due to vague reports of difficulty swallowing solids, showed improved esophageal lumen in the region of the aberrant subclavian artery, without mucosal disease or proximal esophageal dilation. Fig. 6.

During the most recent pulmonary clinic follow up the patient was doing well. She showed good weight gain, was tolerating solids, and continued to be on room air with a normal respiratory exam.
cardiac defects such as conotruncal abnormalities or other
patients are asymptomatic, but 25% –
phagus and in 5% it crosses anterior to the trachea [16]. Although most
esophagus (18). In 15% it crosses in between the trachea and the eso-
0.04
the aorta and makes a U-turn to reach the right half of the body [17]. It
common arch anomalies. It arises as the last brachiocephalic branch of
is present in 0.7 –
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2% of the population with a left aortic arch and aberrant right subclavian artery
Jan et al., found that neonates with an aberrant right subclavian artery
had a smaller birth size but long term growth rates were una
some patients have congenital
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gone unnoticed in a traditional MBS without following the bolus to the
lower esophageal sphincter.

In 80% of cases the aberrant subclavian artery crosses behind the
esophagus (18). In 15% it crosses in between the trachea and the eso-
phagus and in 5% it crosses anterior to the trachea [16]. Although most
patients are asymptomatic, but 25%–37% patients have congenital
cardiac defects such as conotruncal abnormalities or other
chromosomal abnormalities [9–11,14,20].

The most commonly reported chromosomal abnormality with an
aberrant right subclavian is Trisomy 21. In a large case series of 106
fetuses with Trisomy 21, the rate of this vascular anomaly was 25% in
the second trimester ultrasound [15,20].

In a longitudinal study by Jan et [9,18], there was a slight female
gender predominance of this vascular anomaly, which has been re-
ported in many other studies. They also report that the best way to
identify this vascular anomaly is by echocardiogram, CT scan or barium
esophagram [9].

Most cases of aberrant right subclavian artery are asymptomatic,
especially if there are no additional aortic arch/vessel anomalies. It is
typical to have respiratory symptoms in infancy but dysphagia can
occur at an older age when solid or semisolids are introduced, com-
monly known as dysphagia lusoria [6,14,21]. According to Klinkhamer
[17], severe cases of tracheoesophageal compression occur if the right
and left carotid arteries arise from the aortic arch too close together or
have a common origin which can result in compression of the trachea
due to inability of the trachea to mobilize or escape the compression.
The aberrant subclavian artery can be associated with an aneurysm
al dilatation at the base known as Kommerell’s diverticulum, which can
rupture if left untreated [6,14].

Extrinsic compression of the airway and esophagus caused by an
aberrant right subclavian artery is rare in infancy. This disorder ranges
from being asymptomatic to circumstances of respiratory distress,
stridor, wheeze, dyspnea, recurrent respiratory tract infections or
feeding problems which may necessitate surgical intervention [9,17].
Some patients may experience feeding problems. This may increase
their risk for recurrent aspiration and respiratory tract infections [9].
Jan et al., found that neonates with an aberrant right subclavian artery
had a smaller birth size but long term growth rates were unaffected.
They proposed a non-surgical approach for asymptomatic infants with
isolated aberrant right subclavian vessel. They noted that larger studies
are needed to assess the long term outcomes of affected infants [9].

The extrinsic compression of the esophagus causing a bulge to the
posterior aspect of trachea can cause wheezing, stridor, recurrent
pneumonia, cough and cyanosis [5,16].

It is important to consider a complete aerodigestive work up, CT
scan, upper GI tract endoscopy, and airway evaluation.

According to Naqvi et al., aberrant right subclavian artery may have
dysphagia lusoria, which was described by Bayford in 1787 [7,17] as
‘difficulty in swallowing due to quirk of nature’. Dysphagia may be
initially with solids but progress to liquids as well. This is more
common in adults when the tracheal rings have more cartilage and are
less compressive [17]. The presence of Komerrell’s diverticulum may
worsen the dysphagia and needs to be evaluated [6]. It is also important
to exhibit caution when considering a thickened liquid trial given
clinical symptoms of coughing and congestion with feeding. In this
case, a thickened liquid would have exacerbated respiratory symptoms.

Barium esophagram is a simple and very valuable test in identifying
many vascular anomalies such as pulmonary slings, complete aortic
ring or aberrant vessels compressing the esophagus. It typically shows a
lateral indentation in the case of an aberrant subclavian artery [8,12].
Given the nonspecific symptoms of coughing and congestion with
feeding, a modified barium swallow study with esophageal sweep
should be considered. Given the known link between pharyngeal and
esophageal dysphagia, their simultaneous investigation can prove more
efficacious for our patients in relation to time, cost and length of work
up to diagnosis. Watts et al., [19] reported that one in four patients
(26%) had an esophageal cause of their dysphagia that would have
gone unnoticed in a traditional MBS without following the bolus to the
lower esophageal sphincter.

There is a case report by Still et al. [7] of a patient with history of
TEF repair presenting with recurrent pneumonia and chronic barky
cough due to possible post-surgical retrotracheal course of the aberrant
right subclavian artery resulting in anterior wall esophageal and

3. Discussion

Developmentally, aberrant subclavian artery is one of the most
common arch anomalies. It arises as the last brachiocephalic branch of
the aorta and makes a U-turn to reach the right half of the body [17]. It
is present in 0.7–2% of the population with a left aortic arch and
0.04–0.4% with a right aortic arch and aberrant right subclavian artery
[9,14].

In 80% of cases the aberrant subclavian artery crosses behind the
esophagus (18). In 15% it crosses in between the trachea and the eso-
phagus and in 5% it crosses anterior to the trachea [16]. Although most
patients are asymptomatic, but 25%–37% patients have congenital
cardiac defects such as conotruncal abnormalities or other
posterior tracheal wall compression. There is a case report of a development of a TEF in an adult from an aberrant right subclavian following a retrooesophageal course with esophageal occlusion and small perforation of the esophagus into the trachea leading to chronic malnutrition and aspiration pneumonitis in the absence of a diverticulum [6].

The treatment of an aberrant right subclavian artery can vary by age and additional associated vascular malformations. In healthy infants and young children with less severe disease, where malformations are not likely obvious, the multidisciplinary approach of the aerodigestive team will lead to early and accurate diagnosis, and dramatically improve outcomes.

In older adults treatment options vary from esophageal dilatations to the use of proton pump inhibitors and occasionally thoracic vascular surgery based on the location and any other nerve or thoracic duct anomalies [13,22].

This case highlights the importance of an aerodigestive team in the care of a young infant with an unusually severe lung disease and oxygen dependence. The patient’s symptoms of dysphagia and poor weight gain improved and she was able to tolerate solid foods without aspiration.

Funding source
No funding was secured for this study.

Potential conflicts of interest
The authors have indicated they have no potential conflicts of interest to disclose.

Author disclosure
Drs Baig, Fortner, Rivera, Gupta, Sher, Mortelitti and Ms. Merrow have disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

Acknowledgements
The authors would like to express their gratitude to Ms. La Shaun Jones, administrative assistant for her help with the manuscript.

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