An Unusual Cause of Recurrent Respiratory Distress

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Clinical Scenario

A one and a half years old boy presented with recurrent episodes of respiratory distress, accompanied by dry non spasmodic cough and characterized by appearance of a continuous stridor showing no positional variation. The child was symptomatic since early infancy.

Parents also complained of inability to gain weight as compared to other siblings. There was no history of foreign body ingestion or complaints of asthma or tuberculosis in the family. General physical examination revealed tachypnea and biphasic stridor in a malnourished and stunted child in the absence of any significant lymphadenopathy or oral thrush.

On systemic examination, bilaterally, stridorous sounds were auscultated. Primary investigations on the child showed minimal reflux on gastro-esophageal reflux isotope scan, a normal chest roentgenogram, negative sweat chloride test and negative serology for HIV (Human immunodeficiency virus).

Echocardiography performed on the child revealed a structurally normal heart. On conducting further investigations, bronchoscopy detected a bulge in the anterior tracheal wall while barium swallow displayed indentation in the upper esophagus. The MRI (magnetic resonance imaging) subsequently conducted clinched the underlying clinical condition. The MRI images are depicted below.

Q) Which of the following statements is CORRECT regarding the underlying entity?
A) Bilaterally prominent vasculature constitutes the most common variant.
B) Clinical presentations may include vomiting, feed intolerance and swallowing dysfunction.
C) Outcome is primarily poor with increased risk of mortality during surgical repair.
D) Echocardiography and Chest roentgenogram are the favored diagnostic tools.

CORRECT ANSWER: B

Explanations:

B) The first figure represents a 3D MR angiography flight image showing division of ascending aorta into right and left aortic arches with subclavian and common carotid arteries originating separately from respective arches on both sides. The second figure depicts axial MR T2 weighted angiography image in the axial plane at a level proximal to carinal bifurcation showing right and left aortic arches forming a vascular ring around trachea and rejoining together to form a common descending aorta.

Vascular ring is a term that refers to a collection of anomalous configurations of the aortic arch and its associated vascular structures that encircle and constrict the trachea and/or esophagus. Vascular rings comprise less than 1% of congenital cardiac anomalies and amongst its variants double aortic arch (shown here) constitutes the most common type responsible for 40% of the same[1].

The clinical manifestations of aortic ring are related to the tightness of the ring. Widely patent arches form a tight ring and commonly present in the first weeks of life whereas atretic arches form looser rings and are likely to present near 3-6 months ranging up to 3 years of age.

Patients may present with respiratory or gastrointestinal symptoms, with respiratory complaints being the most prevalent[2]. Amongst respiratory complaints, patients with severe obstruction may present with acute life threatening episodes and those with less severe compression may present with recurrent respiratory symptoms often treated as asthma or bronchiolitis. A classic sign of double aortic arch (DAA) is a non positional stridor. Respiratory findings usually exacerbate with crying and are resilient to bronchodilator therapy. In addition to these symptoms, patients may experience swallowing difficulties related to esophageal compression, which tend to manifest as vomiting, feeding intolerance or choking episodes[2,3,4]. Some patients may present with failure to thrive[2]. In addition swallowing dysfunction may further contribute to respiratory symptoms secondary to aspiration or compression of the membranous trachea as food boluses traverses area of esophageal obstruction.

A) Double aortic arch may exist in various forms.

In some, both arches remain patent or an atretic or a hypoplastic segment may be noted at any location along either arch. In majority (75%), a prominent right arch, typically located superiorly is reported while a prominent left arch is present in approximately 20%[5].

C) The definitive treatment for vascular rings is surgery wherein options include minimally invasive thoracoscopic or robotic-assisted repairs and open procedures like thoracotomy.

Clinical outcome is excellent with resolution of symptoms in majority of patients with low risk of morbidity and even rarer risk of death.

Between 70% and 97% of children are entirely asymptomatic following surgery while those with long-term symptoms after correction vary between 8 and 30%[6,7]. The persistence of these symptoms postoperatively can be attributed to tracheomalacia, tracheal stenosis, or both, which are because of maldevelopment of trachea[8]. Rare operative complications include thoracic duct injury, phrenic nerve injury or recurrent laryngeal nerve paralysis[8].

D) Extrinsic compression of the trachea may be suspected by the lateral indentation of the tracheal air column on the PA view or posterior indentation in the lateral view of chest radiograph. Double aortic arch may be confirmed further by bronchoscopy and barium swallow, but is best diagnosed by Computed tomography, Magnetic Resonance Imaging or angiography[9]. Barium esophagograms were once considered the single most important or the primary method of study in evaluating patients with
vascular rings\cite{4}. However, this method is no longer considered a sufficient evaluation before proceeding with treatment due to lack of insight into underlying anatomy\cite{10}.

Two-dimensional echocardiography can substantiate a diagnosis of DAA and is useful for identifying associated congenital heart diseases.

Subcostal and suprasternal views of the two-dimensional echocardiography offer diagnostic windows to assess DAA. While subcostal approach is practical only in infants younger than 12 months old, difficulty in visualization of aorta using suprasternal approach has been encountered in neonates and small children because imaging of aorta requires placement of large transducer in small suprasternal notch\cite{11}. It is likely that the diagnosis may be missed due to a lack of experience with this uncommon anomaly, perhaps also responsible for the lack of detection in our patient.

Although angiography has been the gold standard for diagnosis and classification of vascular rings, MR imaging in the coronal and axial planes has surpassed angiography as the modality of choice largely because it provides non-invasive visualization of arterial branch patterns, tracheal and esophageal location, severity of obstruction, cardiac anatomy and flow measurements\cite{10,12,13}. With recent advances in 3 dimensional reconstruction, MRI is now proving useful in pre-operative planning. It is for these reasons that MR angiography was done in our patient. Moreover catheterization, an invasive process is undesirable and many times not feasible in smaller patients with severe cardiopulmonary distress. Despite these advantages, it may not be routinely used in some locations because of its prohibitive cost, long waiting time and need for increased amounts of sedation.

**Key words:** Respiratory Distress Syndrome, Infant; Stridor; Echocardiography

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