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Diagnostic Challenges of Vascular Rings: Insights from a Case Report

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ABSTRACT

The vascular ring (VR) is a rare congenital anomaly that can cause compression of the trachea or esophagus, resulting in respiratory and/or gastrointestinal symptoms. The severity of symptoms depends on the type of VR, with patients who have anatomically complete VRs typically presenting earlier in life. The diagnosis of VR has advanced with the availability of improved imaging modalities. Prenatal diagnosis is possible with fetal echocardiogram, while non-invasive multidetector CT and MRA can be utilized for later diagnosis. These imaging modalities allow for the delimitation of the defect and the identification of the anatomy for treatment planning.

This case report presents the evaluation and management of a male neonate who was diagnosed with VR after initially presenting with respiratory and gastrointestinal symptoms that were suspected to be related to gastroesophageal reflux. This case highlights the importance of considering VR in the differential diagnosis of patients presenting with these symptomatologies.

KEYWORDS: Vascular Ring, Double Aortic Arch, Compression, Gastroesophageal reflux

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INTRODUCTION

A vascular ring (VR) is a congenital anomaly in which the aortic arch and its branches fully or partially encircle and compress the trachea or esophagus, resulting in a rare malformation accounting for approximately 1% of congenital cardiovascular anomalies [1]. Typical clinical manifestations of vascular ring include stridor, accompanied by a distinctive "seal-bark cough", as well as recurrent respiratory tract infections, wheezing, exertional dyspnea, and dysphagia. In some cases, affected children may experience apparent life-threatening events or apnea, and in rare instances, severe compression and tracheomalacia can lead to critical respiratory distress requiring endotracheal intubation [2]. VR can be either asymptomatic or present with respiratory and/or gastrointestinal symptoms, which are determined by the severity of the ring constriction and resulting tracheoesophageal compression [3]. The severity of symptoms and age of initial presentation vary depending on the specific configuration of the aortic arch. Patients with anatomically complete vascular rings (true vascular rings) typically present earlier in life compared to those with antomically incomplete vascular rings (partial vascular rings), which may not always exhibit noticeable symptoms and can remain undiagnosed for an extended period [4].

The diagnostic approach for patients with vascular rings has significantly advanced in recent years, including during pregnancy, due to the availability of improved imaging modalities. Fetal echocardiogram is now commonly used for prenatal diagnosis [2]. In the past, barium swallow studies were frequently employed to detect vascular rings, but they have limitations, including the possibility of a negative esophagram not ruling out the presence of a vascular anomaly and not providing comprehensive information about the type of anomaly. Furthermore, performing upper gastrointestinal studies in pediatric patients can be challenging. Consequently, more invasive modalities, such as angiography, were often used for surgical planning. Nowadays, non-invasive multidetector three-dimensional (3D) CT and 3D MRA have emerged as important tools for detecting and characterizing vascular anomalies, although each modality has its advantages and disadvantages [2,5].

In this case report, we present the evaluation and management of a male neonate who underwent a normoevolutive pregnancy and was born without any clinical evident abnormalities. However, he subsequently developed

respiratory symptoms that were initially suspected to be associated with gastroesophageal reflux. Subsequent medical evaluation by a pediatrician revealed a more complex pathology.

CASE REPORT

Male newborn patient of 36 weeks of gestation and normoevolutive pregnancy without relevant history, weighed 3215 grams, measured 51 cm, head circumference of 35 cm, apgar 9/9 and no abnormalities were detected in the initial physical examination, at 4 months of age, the patient came for review due to symptoms of cough and rhinorrhea, without fever, the mother reported symptoms compatible with gastroesophageal reflux. An esophagogastroduodenal series was performed, which revealed a diagnosis of gastroesophageal reflux grade II, with esophageal narrowing, to rule out probable aberrant subclavian artery, and treatment for reflux was started by pediatrics. Three days later, the patient went to the treating physician due to persistent cough and reflux. During the consultation, the patient presented a cyanotic episode with oxygen desaturation of 80%. Laboratory tests showed only leukocytosis of 11.2 x 10^3 mm^3, and a chest X-ray showed findings compatible with bibasal pneumonitis (Figure 1). A new esophagogastroduodenal series was performed, which reported the presence of external compression of the upper third of the esophagus (Figure 2). For that reason, an angiotomography was performed, showing the presence of a complete vascular ring (Figure 3), suggesting an anomaly in the embryonic development of the aortic arch and great vessels. Surgical correction by vascular surgery was programmed, performing the procedure successfully and reporting double aortic arch, with the right arch attached to the descending aorta below the ductus arteriosus, which had a diameter of 7 mm and was obliterated. On the eighth postoperative day, the patient was discharged due to clinical and hemodynamic improvement.

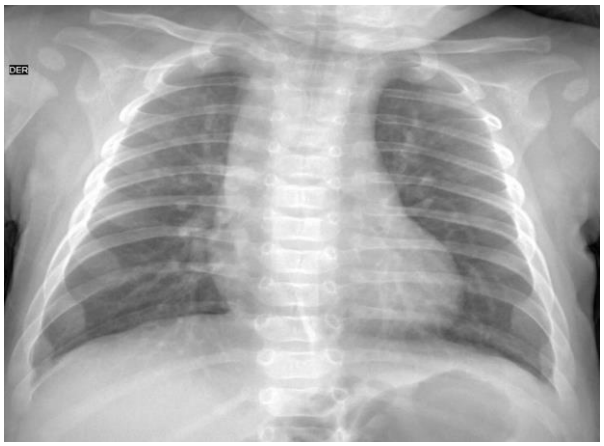

Figure 1. Anteroposterior chest x-ray. Widening of the superior mediastinum and signs of bibasal pneumonitis are identified.

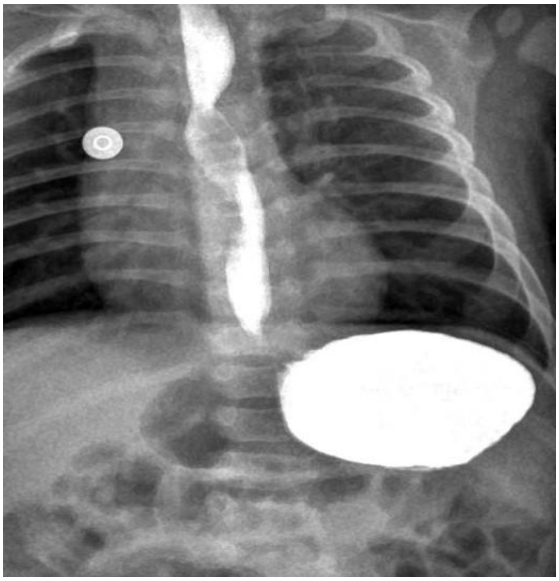

Figure 2. Esophagogastroduodenal series. A filling defect is identified at the level of the upper esophageal third compatible with extrinsic compression of the esophagus.

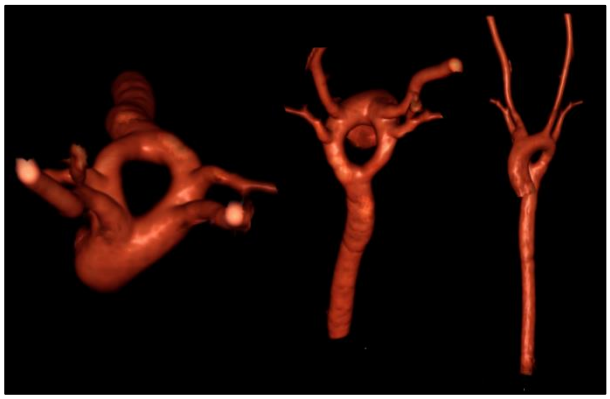

Figure 3. AngioCT of the thoracic aorta with 3D reconstruction. The presence of a complete aortic vascular ring is identified.

DISCUSSION

We reported a case of a male newborn who presented with symptoms of gastroesophageal reflux and was later diagnosed with a vascular ring anomaly, which was corrected through surgical intervention.

Aortic arch anomalies can present as isolated defects or in conjunction with congenital heart disease and can significantly impact prognosis. However, in this case, the vascular ring was not associated with any other structural abnormalities. Various types of aortic arch anomalies can be present at birth, including left-sided, right-sided, and double aortic arches that display diverse branching patterns of the major blood vessels [6]. According to a model proposed by Edwards in 1974, malformations of the aortic arch system can be attributed to either the persistence of segments of embryonic aortic arches that should have regressed or the disappearance of segments that should have remained [7]. Certain anomalies of the aortic arch can form complete rings or true vascular rings, such as double aortic arch and right

arch/left ligament or they can be incomplete or partial vascular rings, such as anomalous innominate artery, aberrant right subclavian artery, and pulmonary artery sling [1,2,3,8]. Double aortic arch is a common form of vascular ring that accounts for 30–50% of symptomatic vascular ring cases [1]. It Is characterized by the persistence of both the right and left arches originating from the ascending aorta. These two arches encircle the trachea and esophagus, subsequently joining the descending aorta to create a true ring. The right arch gives rise to the right carotid and subclavian arteries, while the left arch is typically smaller and gives rise to the left carotid and subclavian arteries [8]. While it is infrequently accompanied by congenital heart disease, tetralogy of Fallot is the most common condition when present, followed by transposition of the great arteries [6].

The Right Aortic Arch (RAA) is also a common form of vascular ring, accounting for 12-25% of cases. It can result in two variations depending on the exact site of interruption of the left arch and the branching pattern to the left subclavian artery, left carotid artery, and ductus arteriosus. These variations are retroesophageal aberrant left subclavian artery and mirror-image branching. Retroesophageal aberrant left subclavian artery is the second most common symptomatic vascular ring, but it is commonly asymptomatic due to the vascular ring being relatively loose [3,9]. RAA is usually associated with congenital heart disease, including persistent truncus arteriosus, pulmonary atresia with ventricular septal defect, and tetralogy of Fallot [3].

True vascular rings result in complete anatomical surrounding and compression of the trachea and esophagus. Narrowing of the mainstem bronchi may also occur. Symptoms of vascular rings include biphasic stridor, wheezing, cyanosis during feeding, recurrent pneumonia or lower airway infections, and dysphagia. External compression of the esophagus from a vascular structure leads to dysphagia lusoria. Patients may report food getting stuck in the mid to upper esophagus, which can result in associated weight loss [10].

Vascular rings in newborns are typically diagnosed through imaging studies such as CT, MRI, or cardiac catheterization when symptoms are present. Asymptomatic patients with isolated vascular ring usually undergo a CT or MRI at around 2 years of age, but may be evaluated earlier if symptoms arise [11]. Early prenatal diagnosis of vascular rings is possible through fetal echocardiography. Screening for congenital heart disease is not typically advised for low-risk populations in early pregnancy [12,13], there is controversy regarding how to manage asymptomatic patients with vascular rings, as they may not exhibit symptoms for a significant amount of time. While many centers utilize echocardiography or other postnatal confirmatory tests, some have suggested that magnetic resonance imaging (MRI) may be a viable alternative due to the limitations of echocardiography [14]. However, its extended procedure time and the requirement for deep anesthesia can limit its use [15].

For symptomatic patients with vascular rings, low-dose MDCT with multiplanar and 3D volumetric reconstructions is recommended for surgical planning and predicting the need for tracheal surgery [1,2,3,9,11,14]. Esophagography and angiography are now rarely used, but may indicate previously undetected anomalies. Tola et al. has highlighted the significance of CT angiography as a diagnostic tool for evaluating vascular ring anomalies, precisely identifying the type of abnormality in most patients, and providing direct visualization of the relationship between the aortic arch and the trachea/bronchi [15].

CONCLUSION

Vascular rings require prompt diagnosis and management for a better prognosis. Understanding the different types of vascular rings and their branching patterns can aid in making an accurate diagnosis. Imaging studies such as CT, MRI, and echocardiography are useful diagnostic tools for both symptomatic and asymptomatic patients The use of these diagnostic tools can facilitate appropriate management, which may include surgical intervention. It is important for healthcare professionals to be aware of the diverse presentations of aortic arch anomalies and consider them in the differential diagnosis for patients with respiratory or gastrointestinal symptoms. Additionally, it is important to consider the possibility of structural heart disease or other chromosomal anomalies that may be linked to the aortic arch anomaly and treat the patient comprehensively.

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