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Aberrant Right Subclavian Artery: A Rare Cause of Dysphagia in a Child

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Abstract

Introduction: Although aberrant right subclavian artery (ARSA) is a rare abnormality but it is the most common anomaly of aortic arch. Compressive effects of the artery are responsible for the patients' symptoms. Association of this disorder with other anomalies and necessity of treatment in symptomatic patients are the important aspects of this condition.

Case Presentation: Here we describe a 12 years old girl with dysphagia and ultimate diagnosis of ARSA. The ligation procedure of the artery was successful without any complications.

Conclusion: Dysphagia could be the main symptom of the ARSA in children as same as old adults.

Keywords: Aberrant subclavian artery, Aortic arch anomaly, Dysphagia

Introduction

The most common anomaly of aortic arch is aberrant right subclavian artery (ARSA). This is a rare condition and symptom development is not common in affected patients but it has some aspects of importance. The first aspect is the association of this abnormality with some congenital heart diseases and also increased prevalence of it in chromosomal defects. The second one is serious complications of this anomaly in untreated symptomatic patients. The last aspect is challenging coronary arteriography via radial approach in these patients.

Case Presentation

A 12-year-old girl was referred to our Pediatric Cardiology clinic from her school. At first she and her family denied any problem but delicate history taking revealed that she had suffered dysphagia for several years. Due to her suspicious transthoracic echocardiography, she was referred to aortic computed tomography (CT) angiography. Aortic CT angiography showed ARSA arising from distal portion of aortic arch but other findings were normal (Figures 1 and 2).

A barium swallow study revealed esophageal compression which was consistent with an ARSA. Due to symptomatic ARSA she was admitted for surgery. The physical examination showed the following:

Heart rate 96 beats per minute, respiratory rate 20 per minute and oxygen saturation was 100% in room air. The lungs were clear bilaterally. The heart examination was normal and the remainder of the examination findings was unremarkable. Chest radiography was normal.

Under general anesthesia great vessels were approached by left posterolateral thoracotomy incision. The orifice of the ARSA was exposed. The artery was at the posterior side of aortic arch and left subclavian artery. Esophagus was completely surrounded by the aberrant artery and then right subclavian artery was clamped for 10 minutes. Right upper limb saturation after 10 minutes was 99%-100%. With restoration of appropriate stump, ARSA was transected and sutured. The esophagus was completely released. Chest tube was then placed, hemostasis was done and the patient was transferred to intensive care unit (ICU). Postoperative course and the situation of the patient during discharging were satisfactory.

A year after discharging from hospital, the patient was followed up. This follow up showed neither complication of surgery nor symptom of right upper limb hypoperfusion.

Discussion

ARSA is the most common form of aortic arch anomaly with estimated occurrence rate of 0.5% (1). It is usually due to regression of the right forth aortic arch between the carotid and subclavian arteries; meanwhile, positioning distal to left subclavian artery is less common (2,3). In 1794, Bayford was the first who described dysphagia related to ARSA or arteria lusoria in a 62-year-old woman (4). Compression of esophagus and trachea by aberrant vessel could cause symptoms such as dysphagia, dyspnea, cough, stridor and thoracic pain. The most common symptom in adults is dysphagia while infants usually present with respiratory problems. Aspiration of food due to dysphagia and absence of tracheal rigidity are thought to be responsible for respiratory symptoms of infants. The reason of dysphagia in adult patients has not been clearly recognized. However some proposed mechanisms are aneurysmal formation, increased rigidity of the esophagus or the

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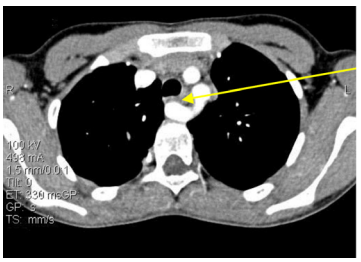

Figure 1. Cross sectional view of aberrant right subclavian artery in computed tomography (CT) scan.

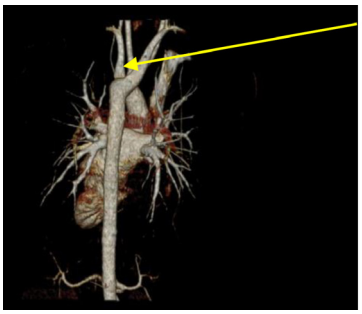

Figure 2. Re-constructed image of aortic arch and the position of aberrant right subclavian artery in computed tomography (CT) scan.

vascular wall, fibrous transformation of the paratracheal and esophageal tissues in combination with nearness of the proximal aortic branches, atheromatous process of aging, aortic elongation, and the combination of an aberrant artery with truncus bicaroticus (5). Although dysphagia as a symptom of ARSA is more common in middle aged or old patients (6) our patient also had dysphagia which reveals that children may present with this symptom.

It was shown previously that the prevalence of ARSA is higher in some syndromes such as Down and DiGeorge and also association of ARSA with other congenital heart diseases which have higher rates in these syndromes (6). It indicates the importance of physician's awareness in this disorder.

The other clinical importance of ARSA is during coronary arteriography via radial approach which may cause some technical difficulties for cardiologists. A characteristic sign in Barium swallow study of esophagus is the diagonal compression defect at the level of the third and forth vertebrae. For confirmation of diagnosis, computed tomographic scanning, angiography or both are usually needed. Magnetic resonance imaging may be helpful in situations such as the presence of an aneurysm in the proximal part of the artery for better visualization (7).

Although majority of patients are asymptomatic, symptomatic ones require treatment. One of the reasons for treatment is avoidance of rare but serious complications of untreated patients such as atrioesophageal and arteriotracheal fistula (1).

The artery usually passes through posterior side of the esophagus but occasionally it crosses between esophagus and trachea, the anterior position of the artery to the trachea is very uncommon. The first treatment for this anomaly was done by Gross in 1946 which consisted of ARSA ligation only. Derbel et al re-implanted the ligated vessel to the proximal of right common carotid artery, which caused complete restoration of right upper limb perfusion (8).

In our patient simple ligation of the artery was done without re-implantation but in follow-up evaluation, the patient did not have any symptoms related to low perfusion of right upper extremity. However it is logical to preserve blood supply of limb whenever possible, some cases of successful surgery without complication have been reported with simple resection of aberrant artery without re-implantation of it.

Ethical issues

Written informed consent was obtained from the patients for publication of this study.

Conflict of interests

Authors do not have any competing interests.

Acknowledgments

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