ORIGINAL ARTICLE

Vascular ring: Role of multidetector CT with 3D reconstruction in diagnosis

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Received 5 December 2011; accepted 11 December 2011
Available online 14 January 2012

KEYWORDS
Vascular ring; Multislice CT; 3D reconstruction; Multiplanar reformation; Aortic arch malformations; Stridor

Abstract  Purpose: To assess the role of multidetector CT (MDCT) with 3D reconstruction in the diagnosis of vascular ring.

Materials and methods: We reviewed the files of 26 patients with vascular ring abnormalities at the Pediatric Hospital, Mansoura University, between March 2009 and May 2011 who had multislice CT. Patients were examined using a 16-row CT scanner. The multislice data were transmitted to a workstation for multiplanar reconstruction (MPR) and volume rendering technique (VR) reconstruction.

Results: There were 16 patients with double aortic arch; out of these patients, codominance was present in four patients and right dominance in 12 patients. Eight patients had right-sided aortic arch; six of them with aberrant left subclavian artery and the other two patients with mirror-image branching. Three patients with right-sided aortic arch were accompanied by other malformations (pulmonary artery stenosis in one patient, patent ductus arteriosus in one patient and left superior vena cava in one patient). Two patients had left-sided aortic arch with aberrant right subclavian. Two patients had associated Kommerell diverticulum, one with double aortic arch and the other one with right-sided aortic arch.

Conclusion: MDCT with 3D reconstructions is an excellent diagnostic modality in patients suspected to have a vascular ring. It not only accurately delineates the complex anatomy of these vascular
1. Introduction

The phrase “vascular ring” has been used to refer to a collection of congenital vascular anomalies that encircle and compress the esophagus and the trachea (1). A vascular ring encircling the mediastinal airway results from the failure of primitive vascular structures to fuse and regress normally during the development of the aortic arch, pulmonary arteries, and ductus arteriosus. Patients with vascular rings may present with wheezing, stridor, feeding difficulties, choking episodes, or aspiration pneumonia, depending on the degree of tracheal and esophageal narrowing (2).

With the advent of new noninvasive imaging methods, particularly multidetector computed tomography (CT) technology with increased z-axis coverage, higher spatial resolution (smaller detectors), and higher temporal resolution (faster 360° rotation times), evaluation of the aorta and its branches is possible with faster imaging times, fewer motion artifacts, and also lesser contrast material volume. This method has mostly replaced invasive angiography when aortic disease is suspected (3,4).

2. Materials and methods

We retrospectively reviewed the charts of 26 patients with vascular ring abnormalities at the Pediatric Hospital, Mansoura University, between March 2009 and May 2011 who had multislice CT. These patients were referred for CT because echocardiography did not provide adequate clinical decision.

Scanning was performed using a 16-row CT scanner. An initial scout image was obtained to determine the scan volume. Proper timing is critical to ensure that the arterial system is maximally enhanced with no contamination by the venous phase. In our study the optimum delay was calculated by the automated bolus tracking technique in all patients where the machine automatically starts scanning once the level of contrast enhancement in the artery (aortic arch) reaches a preset value of 65–70 HU. A single bolus of 2 ml/kg of scanlux (non-ionic contrast material with 370 mg iodine/mL) was administered intravenously by a power injector through a 20–22 gauge cannula located in an adequate superficial vein. Contrast material was injected at a rate of 2–4 mL/s. All patients were examined in the supine position. Included images start from the lower neck down to the upper abdomen. Scanning parameters were: section thickness, 0.65 mm; pitch, 1–1.5 and gantry rotation time, 0.5 s. About 800–1200 axial images/study were generated. The images were then transferred to an advantage workstation for display in various 3D techniques like maximum intensity projection (MIP), surface shaded display (SSD) and volume rendered images (VR). Parent’s informed consent was not required since this was a retrospective study.

3. Results

Age at the time of presentation ranged from 23 days to 10 years (mean 5.3 years). Patients were categorized according to three types of vascular anomalies into: double aortic arch (16 patients); right-sided aortic arch (eight patients); and left-sided aortic arch with aberrant right subclavian (two patients). Out of these patients with double aortic arch, codominance was present in four patients and right dominance in 12 patients. There were two configurations of right aortic arch: aberrant left subclavian artery (six patients) and mirror-image branching (two patients). Three patients with right-sided aortic arch were accompanied by other malformations (pulmonary artery stenosis in one patient, patent ductus arteriosus in one patient and left superior vena cava in one case). Two patients had left-sided aortic arch with aberrant right subclavian. Two patients had associated Kommerell diverticulum, one with double aortic arch and the other one with right-sided aortic arch Table 1.

Stridor and recurrent upper respiratory tract infection were frequent presentations among patients with double aortic arch with or without associated feeding difficulties. Feeding difficulty was also present in patients with symptomatic aberrant subclavian artery Table 2.

3D images allowed visualization of the aortic arch and its branches in a true to life 3D fashion.

4. Discussion

A vascular ring is an aortic arch anomaly in which the trachea and the esophagus are surrounded by vascular structures (5).
Vascular rings are uncommon anomalies (<1% of all congenital cardiac defects) with a similar frequency in both sexes and are caused by abnormal persistence or regression of one of the six embryonic aortic arches (6,7). The two most common types of complete vascular rings are a double aortic arch and a right aortic arch with left ligamentum arteriosum (7). Two other complete vascular rings that are rare include a right aortic arch with mirror-image branching and retro-esophageal left ligamentum arteriosum and a left aortic arch with right-sided descending aorta, right subclavian artery, and right ligamentum arteriosum (4).

The major vessels and their branches commonly overlap on conventional arteriograms, which obscure their depiction. The rapid imaging afforded by multidetector row CT provides shorter acquisition times; superior 3D renderings; and greater range of coverage, which increases the detection of vascular and nonvascular lesions (3). In the setting of vascular ring, multiplanar and 3D reconstructions can improve the understanding of the origin of the great vessels and the relationship of the vessels to the adjacent airway, compared with the understanding achievable with transverse images alone. This can help provide a more accurate assessment of airway

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Vascular anomaly</th>
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<tbody>
<tr>
<td></td>
<td>DAA (%)</td>
</tr>
<tr>
<td>Stridor</td>
<td>14 (87.5)</td>
</tr>
<tr>
<td>Recurrent chest infection</td>
<td>2 (12.5)</td>
</tr>
<tr>
<td>Feeding difficulty</td>
<td>8 (50)</td>
</tr>
</tbody>
</table>

NB: DAA = double aortic arch, RALS = right arch with aberrant left subclavian, LARS = left arch with aberrant right subclavian.
compromise and assist in surgical planning by providing a better representation of the three-dimensional anatomy (2,8).

Double aortic arch was the commonest type encountered in this series (16 out of 26 patients), 61% with right dominance in 12 patients (75%) (Fig. 2) and codominance in four patients (25%) (Fig. 1). Most of these patients (14) had stridor at presentation with associated feeding difficulties in eight patients. None of them had associated cardiovascular anomalies by echocardiography. Kussman et al. (5) reported that a double aortic arch accounts for 50–60% of the vascular rings and represents persistence of both right and left embryonic fourth arches, which are joined to the dorsal aorta. Lowe et al. (6) and Alsenaidi et al. (9) stated that the most common form of double aortic arch is one in which the right (posterior) arch is dominant in 75% of the patients, the left (anterior) arch is dominant in 20% of the patients, and the arches are of the same size in 5% of the patients. The less dominant arch is atretic in one-third of the cases. Jeeyani et al. (10) reported that the double aortic arch, although rare, is an important cause of persistent respiratory symptoms in infants. It is due to the failure of regression of the right aortic arch. The remnant of the right aortic arch becomes the right innominate artery and leaves a left arch in the normal development, freeing the trachea and the esophagus. Failure of this process of absorption produces a vascular ring that completely encircles and compresses the esophagus and the trachea, leading to severe respiratory and feeding difficulties. Usually, double aortic arch occurs without associated cardiovascular anomalies. However, anomalies like ventricular septal defect, tetralogy of Fallot, truncus arteriosus, transposition of great arteries, pulmonary atresia and complex univentricular defects are seen, with a reported incidence of 17% (11,12). Respiratory symptoms at birth or during infancy should raise the possibility of vascular ring compression. Patients with double aortic arch can present with respiratory, gastrointestinal and cardiac symptoms. In a case series of 81 patients, 91% had respiratory, 40% had gastrointestinal and 28% had cardiac symptoms (12). The classic history in a patient with double aortic arch is noisy breathing, noted by parents in the first few weeks of life. Neonates may also have episodes of acute apnea and cyanosis, which is termed as ALTE (acute life-threatening event). Gastrointestinal symptoms include choking while feeding, emesis, feeding difficulty and failure to thrive (10). The second most common type encountered in this study was the right side aortic arch.
with aberrant left subclavian in six patients (23%). The left subclavian artery arose as a fourth branch of the aortic arch from the distal part of the arch or from the descending aorta. There was associated pulmonary artery stenosis in one patient, patent ductus arteriosus in one patient and left-sided superior vena cava in another one (Fig. 3). There were two cases of right arch with mirror-image branching, one of them showed associated Kommerell diverticulum (Fig. 4).

A right aortic arch is rare in normal individuals and usually is associated with congenital cardiac malformations such as persistent truncus arteriosus, tetralogy of Fallot, and pulmonary atresia with ventricular septal defect. If the left fourth aortic arch regresses proximal to the left subclavian artery, a right aortic arch with aberrant left subclavian artery as the last branch results. The aberrant left subclavian artery commonly originates from an aortic diverticulum, passes

Fig. 3  Right aortic arch with aberrant left subclavian artery causing complete vascular ring with associated left SVC. (A) Axial post contrast CT showing the right side aortic arch and the origin of the small aberrant subclavian artery passing posterior to the trachea. (B and C) 3D volume rendered contrast-enhanced CT images showing right side arch with small aberrant left subclavian artery (long arrow) as well as opacified persistent left superior vena cava (SVC) seen also in (D) coronal MIP image (short arrow).
behind the esophagus and forms a complete vascular ring together with a left-sided ductus ligament (13). Exceptional cases of vascular rings formed by a right arch with mirror-image branching and left-sided ductus ligament extending from the left pulmonary artery to the right descending aorta have been described (14).

In patients with either a double aortic arch or a right aortic arch with a left ligamentum, there are four separate brachiocephalic vessels (instead of the normal three) in the superior mediastinum grouped around the trachea. This is called the “four-vessel” sign (6). There were two cases with left-sided aortic arch and aberrant right subclavian (Fig. 5), one of them presented with stridor and dysphagia. The anomalous right subclavian artery is always described as passing behind the esophagus. David Bayford (15,16), in his report in 1794, reported a 63-year-old woman with lifelong swallowing problems in whom the necropsy specimens showed the artery to be passing between the trachea and the esophagus. Isolated posterior oblique indentation of the esophagus from an anomalous right subclavian artery should not be accepted as the cause of stridor in an infant. Although it is occasionally associated with swallowing problems (dysplagia lusoria), it is usually asymptomatic and occurs in approximately one in 200 individuals (17). In this study, two cases with right aortic arch showed a diverticulum arising from the descending aorta (Fig. 2). Kommerell (18) in 1937 described this condition in an adult patient who had an anomalous right subclavian artery with a left arch. Since then, Kommerell diverticulum has been used by most authors, although not as originally described, to define any anomalous subclavian artery, including the origin of an aberrant left subclavian artery in association with a right aortic arch (17). Although a ductus ligament usually cannot be directly imaged, its location may be implied by the presence of a diverticulum of the aorta, subclavian artery or pulmonary artery. Such diverticula are caused by the large blood flow volume through the fetal ductus arteriosus (19).

In confirmation with previous reports (20–23), we found that the ability to image the vascular anomalies has been greatly enhanced by the use of 3D reconstruction of contrast-enhanced CT scans.

The limitations of this study were the small number of patients, which precluded statistical analysis, and its retrospective method. Also, we did not compare the contrast CT imaging to other noninvasive imaging modalities.
5. Conclusion

MDCT with 3D reconstructions is an excellent modality in patients suspected to have a vascular ring. It not only accurately delineates the complex anatomy of these vascular structures but also allows the evaluation of possible airway compromise or esophageal impingement and provides evidences concerning the presence and location of ligamentous structures. These information can eliminate the need for other imaging studies as barium study, echocardiography and angiography.

References

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