Double aortic arch in an adult

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Abstract

We present the case of a 60-year old man who complained of severe dysphagia caused by a double aortic arch (DAA) with a right-sided descending thoracic aorta. The left-sided aortic arch had a compressive segment located between the left subclavian artery and the descending thoracic aorta. Using left third thoracotomy, the segment, which caused compression of the oesophagus, was ligated and divided. After the operation, the patient was completely relieved of his symptoms. We concluded that the removal of the compressive portion of the left aortic arch and the ligation of the ligamentum arteriosum are the only treatment measures needed in such cases.

Keywords: Double aortic arch • Dysphagia • Right-sided descending thoracic aorta

INTRODUCTION

Aortic arch anomalies, which are well described in infants, cause stridor, respiratory distress or dysphagia in the first months of life [1]. The vascular ring anomaly of the aortic arch comprises 1–2% of all congenital diseases of the heart and aorta [2]. Double aortic arch (DAA) has been abundantly described in childhood and its management is well established [3]. However, symptomatic adult patients with dysphagia or dysphnoea caused by DAA are rare, because of which most cardiothoracic units have limited surgical experience in this regard [4]. Here, we report the case of a symptomatic adult with severe dysphagia associated with a DAA.

CASE REPORT

A 60-year old man visited our hospital because of severe dysphagia. He had been asymptomatic and in excellent health until ~6 months before he was referred to our hospital. The results of physical examination and laboratory evaluation were normal. Chest radiography showed an aortic knob projecting over the right side of the upper manubrium (Fig. 1f).

Esophagogastroduodenoscopy revealed no oesophageal lesions, and subsequent computed tomography (CT) scanning revealed a DAA in the form of a complete vascular ring around the trachea and oesophagus of the right and left aortic arches (Fig. 1a and b). Three-dimensional (3D) CT demonstrated the anatomy of the DAA (Fig. 1c) and the site of oesophageal compression more clearly (Fig. 1e). The right subclavian and right common carotid arteries originated from the right aortic arch, whereas the left common carotid and left subclavian arteries originated from the left aortic arch; the diameters of the aortic arches were 24 mm (right) and 14 mm (left). We assumed that the left-sided aortic arch had a compressive segment between the left subclavian artery and the right-sided descending thoracic aorta. The patient elected to undergo surgery.

At the right-side decubitus position (60°), left third intercostal space thoracotomy was performed. The compressive segment between the left subclavian artery and the right-sided descending thoracic aorta could easily be identified (Fig. 2a). However, the site where the descending thoracic aorta connected with the left-sided aortic arch (Fig. 1c; arrow) was positioned very deep, as the descending thoracic aorta was on the dorsal right. The compressive segment was 25 mm long and 12 mm wide and it had a lumen. The segment and the left subclavian artery were dissected off the oesophagus, and the left vagus nerve was mobilized anteriorly with the pleural flap. The ligamentum arteriosum was found between the descending thoracic aorta connected with the left-sided aortic arch and the pulmonary trunk. The ligamentum arteriosum was ligated with 3–0 silk suture and divided. The descending thoracic aorta was clamped on both sides of the compressive segment, and the left-sided aortic arch and left subclavian artery were clamped. Then, the compressive segment near the descending thoracic aorta was first closed with 2/0 Ti-cron (coated polyester suture, Covidien) stitches using pledgets. Secondly, the compressive segment near the left subclavian artery was ligated with 1–0 silk suture. Finally, the compressive segment was removed completely, resulting in immediate decompression of the oesophagus (Fig. 2b). The patient’s dysphagia was completely resolved. A postoperative 3D CT scan clearly showed the site at which the left arch was divided (Fig. 1d). The patient was discharged on the 12th day after the operation.
DISCUSSION

A previous study [5] has shown that failure of the normal regression of one or more segments of the six pairs of the aortic arches that arise from the truncus arteriosus leads to the formation of multiple anomalies of the aortic arch, like the vascular ring. Therefore, aortic arch anomalies could include the combination of a DAA with an equal right and left component, a smaller or compressive right or left component, and a left- or right-sided descending thoracic aorta. In the present case, the patient had a DAA with a right-sided aortic arch, compressive left component and right-sided descending thoracic aorta.

The clinical symptoms of vascular rings result from the compressive effects of the adjacent airway or part of the oesophagus. Respiratory symptoms are common in the case of vascular rings in infancy or early childhood [6]. On the other hand, adult patients complain of difficulties in swallowing rather than respiratory difficulties because of tracheal development. A DAA causing tracheal and oesophageal compression is not unusual in children. There have been some references to DAA in adults, but symptomatic and operative cases in adults have been encountered much less frequently.

The diagnosis of a DAA using chest radiography or transthoracic echocardiography is often difficult. Besides CT, magnetic resonance imaging is an important diagnostic tool for identifying anomalies of the aortic arch and its branches, and it can be considered the imaging technique of choice when planning surgical management. In particular, 3D CT aortography clearly reveals

Figure 1: Contrast-enhanced CT showing a DAA involving a complete vascular ring around the trachea and oesophagus (a and b). The 3D CT scan shows the anatomy of the DAA more clearly, and the white arrow indicates the site at which the descending thoracic aorta is connected to the left-sided aortic arch (c). The postoperative 3D CT scan clearly shows the site at which the left aortic arch is divided (d). A 3D CT scan shows the site at which the oesophagus is compressed by the compressive segment (e). A chest radiography of the posteroanterior projection shows an aortic knob projecting over the right side of the upper manubrium (f).
anomalous construction and vessels of the aortic arch. We decided on the approach to use in the operation and the point at which to divide the left arch on the basis of the 3D CT findings.

Surgical intervention involving division of the minor arch and ligamentum arteriosum is indicated for adult patients who are symptomatic. Information on the position of the compressive aortic segment, ligamentum arteriosum and descending thoracic aorta is very important for selecting the surgical approach. Thoracotomy should be carried out on the side of the compressive aortic segment and the ligamentum arteriosum because in the presence of a DAA, oesophageal compression is relieved by ligation and division of both the compressive aortic arch and ligamentum arteriosum [7]. Moreover, if the descending thoracic aorta is located on the dorsal right, the part of the descending thoracic aorta that is connected with the left-sided aortic arch is positioned very deeply. Therefore, it is more difficult to divide this portion than if the descending thoracic aorta is located on the dorsal left. Finally, in the case of DAA with a right-sided descending thoracic aorta, we consider that the operative posture and surgical approach are very important factors to perform the removal of the compressive portion of the left aortic arch and the ligation of the ligamentum arteriosum.

Conflict of interest: none declared.

REFERENCES