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Case Report

Balanced double aortic arch demonstrated by multimodality image and 7-year follow-up in a symptomatic elderly patient: A case report ☆,☆☆

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ABSTRACT

Double aortic arch is a very rare congenital heart disease. Double aortic arch forms a vascular ring, compressing the esophagus and trachea, causing symptoms mainly in infants and young children, and symptoms rarely appear after adulthood. The management of double aortic arch depends on the severity of the symptoms, but since aging exacerbates atherosclerosis and complicates surgery, treatment in adults has many considerations. A 55-year-old woman admitted for chest discomfort, mild dyspnea and mild dysphagia. On a simple chest X-ray, dilated upper mediastinum and bilateral aortic knobs were noted. Transthoracic echocardiography revealed 2 aortic arches on suprasternal view. Contrast-enhanced computed tomography and 3-dimensional computed tomography demonstrated a balanced double aortic arch which formed a complete vascular ring and compressed the esophagus. Barium esophagogram showed marked luminal narrowing at the aortic arch level, probably due to indentation of the double aortic arch. She had several risk factors regarding progression of aortic atherosclerosis include old age, hypertension and dyslipidemia that make more severe compression of esophagus and trachea, but the symptoms were not severe, so we decided to observation while controlling the risk factors. For the next 7 years, she stayed without worsening of symptoms.

Introduction

Vascular ring are rare congenital abnormalities of the aortic arch system. Double aortic arch is a form of complete vascular rings, causing trachea-esophageal compression [1–3]. Most diagnoses of double aortic arch are performed in infancy and childhood. This is a case report of a double aortic arch diagnosed using multimodalities in an elderly patient presenting dysphagia.

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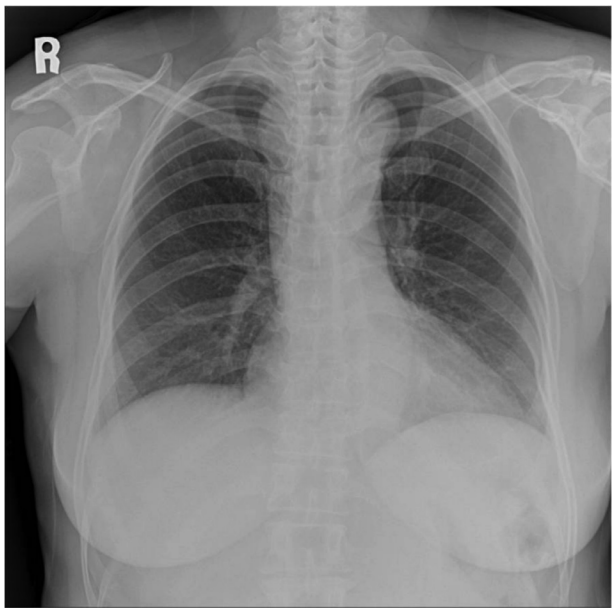

Fig. 1 – Chest radiography. Wide mediastinum and bilateral aortic knobs are shown.

Case report

A 55-year-old woman visited our hospital for chest discomfort, mild dyspnea, and mild dysphagia that began 10 years prior. She was on medication for hypertension and dyslipidemia. Physical examination and laboratory values were normal. On a simple chest X-ray, dilated upper mediastinum and bilateral aortic knobs were noted (Fig. 1). Transthoracic echocardiography revealed 2 aortic arches on suprasternal view (Figs. 2A and B). Contrast-enhanced computed tomography (CT) demonstrated a balanced double aortic arch (DAA) with a right arch diameter of 21.5 mm and a left arch diameter of 22.5 mm. They formed a complete vascular ring and compressed the esophagus (Figs. 3A-H). Three-dimensional CT demonstrated more clearly the anatomy of the DAA (Fig. 4). The right common carotid artery and subclavian artery arose from the right-side arch, the left common carotid artery and subclavian artery arose from the left-side arch. Atheromatous calcification was minimal in the aortic arch. Barium esophagography showed marked luminal narrowing at the aortic arch level, probably due to indentation of the DAA (Figs. 5A and B).

She had several risk factors of progressive atherosclerosis such as old age, hypertension, and dyslipidemia. However, since her symptoms were not severe and had not worsened over 10 years, we decided to provide conservative management without surgery. The patient has been treated with carvedilol, telmisartan, hydrochlorothiazide, aspirin, fluvastatin, and diltiazem for the next 7 years and has not experienced severe swallowing or breathing difficulties.

Discussion

DAA is a rare disease that accounts for less than 1% of congenital heart disease [1,2]. During cardiac development, failure of the right aortic arch to degenerate causes formation of a vascular ring surrounding the trachea and esophagus [3]. The 2 aortic arches are often different in size, with the right aortic arch dominant in 75% of cases and the left aortic arch dominant in 20%. The remaining 5% are similar in size and are called balanced DAA [4–8].

Most cases of DAA are diagnosed in infancy or early childhood with symptoms such as respiratory distress, stridor, and feeding difficulties with recurrent lower respiratory tract infections and dysphagia. Few elderly patients experience newly developed compressive presentation. Late-onset presentation

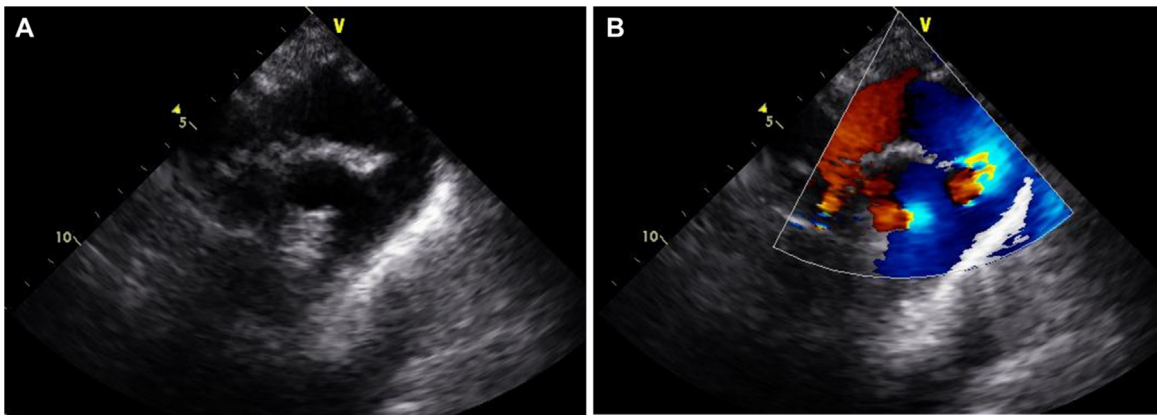

Fig. 2 – Transthoracic echocardiography. (A) Double aortic arch is shown on suprasternal notch view. (B) On color Doppler echocardiography, the patency of both left and right arches is demonstrated.

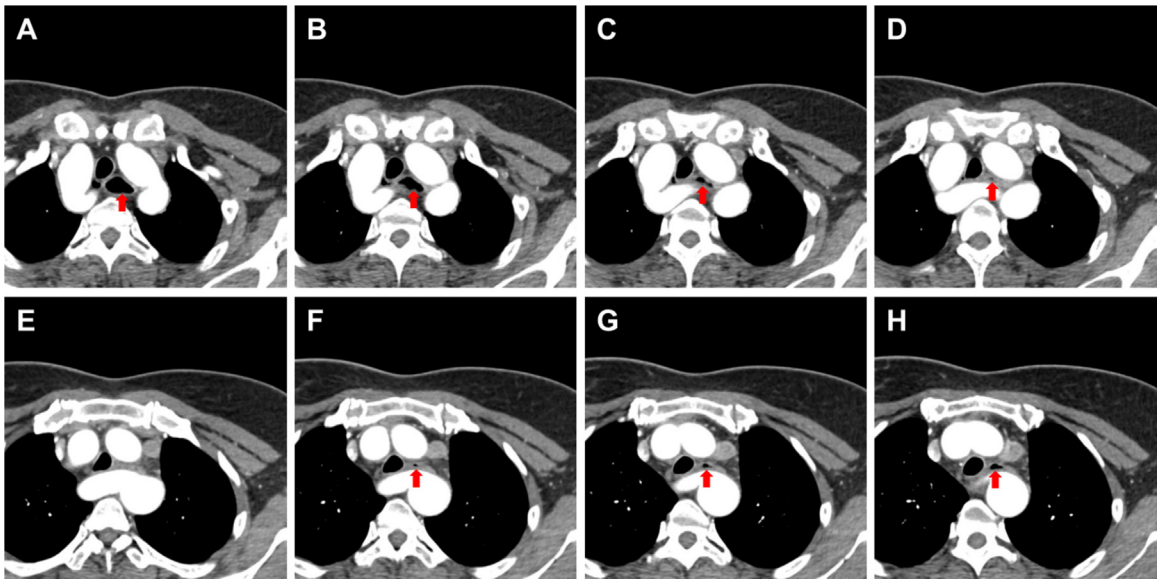

Fig. 3 – Contrast-enhanced computed tomography. Double aortic arch as a complete vascular ring encircles the trachea and esophagus. Red arrows indicate the compressed esophagus by the aortic ring.

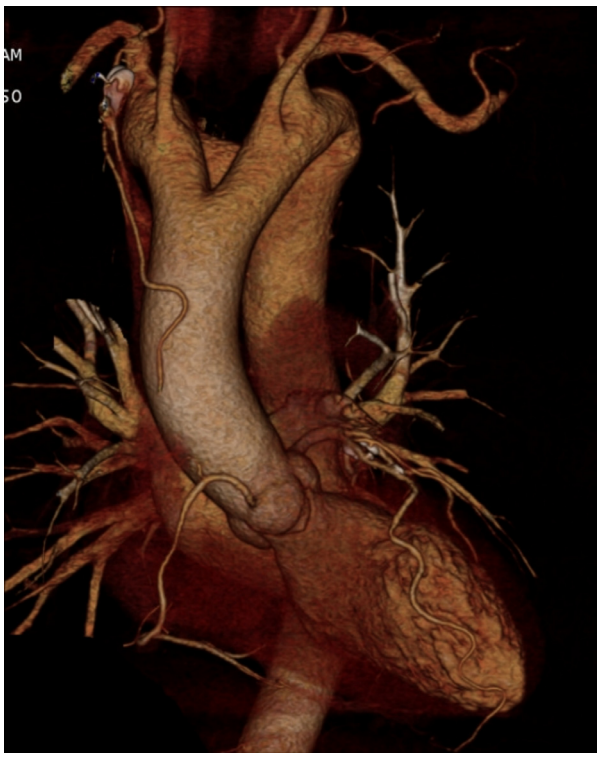

Fig. 4 – Three-dimensional volume rendering image of chest computed tomography. Balanced type double aortic arches are shown. Right common carotid artery arises from the right-side arch, and left common carotid artery from the left-side arch.

can occur due to increased esophageal rigidity, atherosclerotic changes of the major vessels, and elongation of the aorta with aging.

Diagnosis of DAA is possible using multimodalities such as CT, magnetic resonance angiography, and echocardiography. CT angiography provided the most characteristic appearance, and 3-dimensional CT is useful not only for diagnosis, but also for determination of the strategy of the operation. Although the role of echocardiography in diagnosis of DAA is often neglected, it is an important test for initial diagnosis of DAA and for detecting concomitant congenital defects [8–13].

Management of DAA depends on the severity of symptoms. Mild to moderate symptoms can be treated symptomatically, including lifestyle changes and dietary modifications. Patients with severe symptoms and those who do not respond to conservative management might require operative intervention. Aging aggravates atherosclerosis and complicates surgery. Surgical intervention with division of the minor arch is recommended for symptomatic adult patients [14].

In summary, this case is a rare balanced double aortic arch and its complication diagnosed in an elder that was successfully diagnosed using multimodalities with echocardiography, contrast enhanced CT, 3-dimensional volume rendering image of CT and esophagography. It also suggests that elderly at high risk of atherosclerosis can avoid surgery if the risk factors are actively controlled.

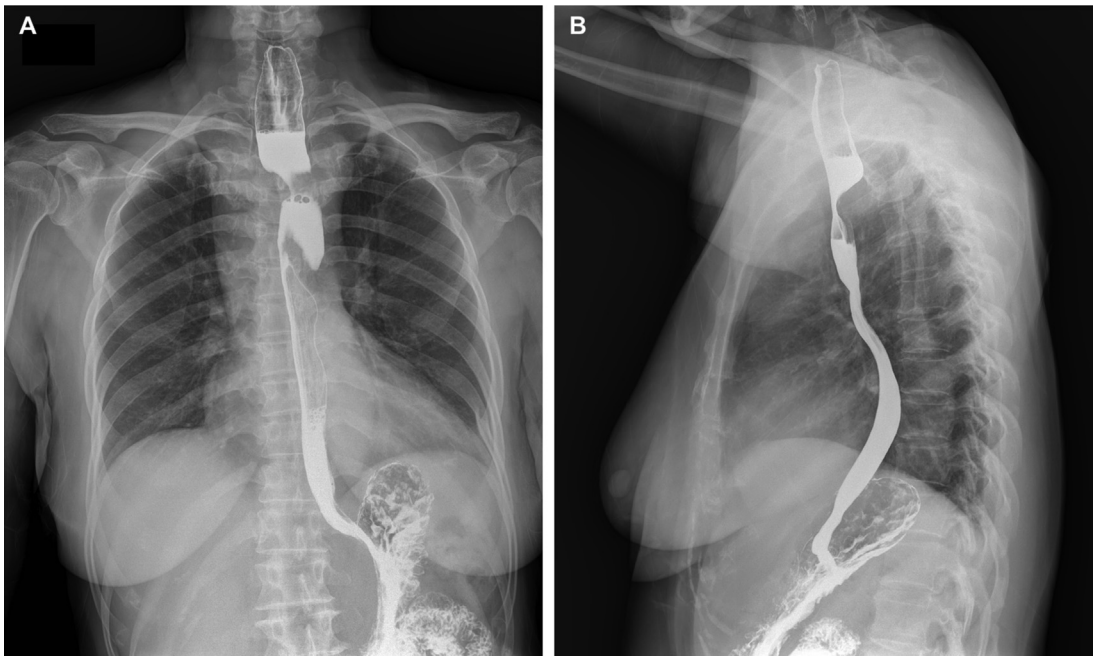

Fig. 5 – Barium esophagogram. (A) Luminal narrowing of upper thoracic esophagus with mucosal preservation is shown on antero-posterior views. (B) The lumen of the upper thoracic esophagus is observed with markedly narrowed at the level of the aortic arch on the lateral view.

Patient consent

Patient consent has been obtained.

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