Pseudocoarctation of the Aorta Associated with the Anomalous Origin of the Left Vertebral Artery: a Case Report

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Pseudocoarctation of the aorta is a rare congenital anomaly of the aortic arch, and it has been described as an elongation of the aortic arch with "kinking" at the level of the ligamentum arteriosum without a pressure gradient across the lesion. The treatment for this condition is controversial. We report here on an unusual case of pseudocoarctation of the aorta associated with the anomalous origin of the left vertebral artery and we include a review of the medical literature.

Index terms:
Pseudocoarctation
Computed tomography (CT)
Vertebral artery

DOI:10.3348/kjr.2008.9.3.283

Korean J Radiol 2008;9:283-285
Received July 30, 2007; accepted after revision September 27, 2007.

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The present research was financed by research funds of Dankook University in 2006.

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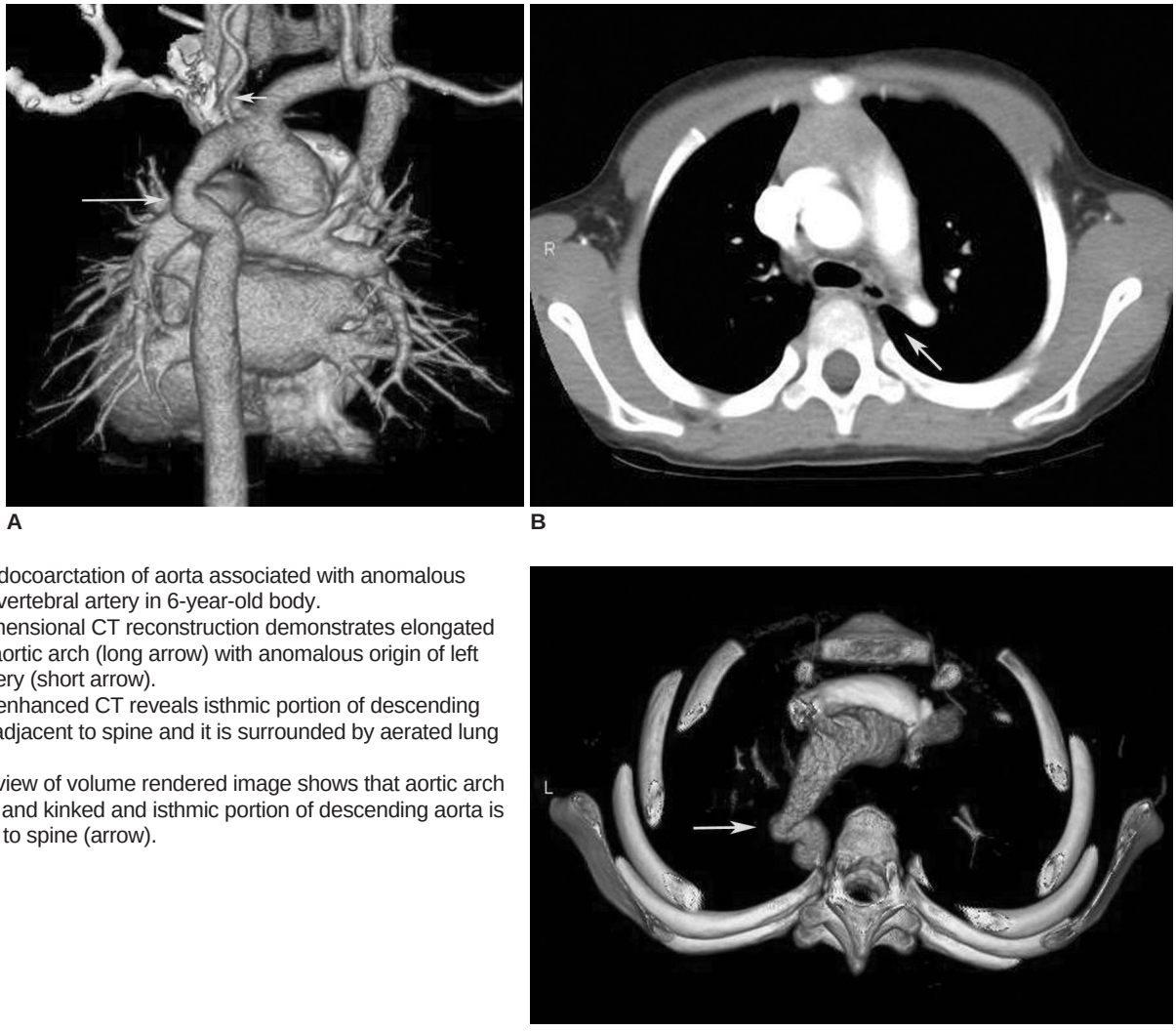
Pseudocoarctation of the aorta was first described by Dotter and Steinberg, and Souders and coworker in 1951 (1, 2). It is usually found by detecting a cardiac murmur or an abnormal superior mediastinal mass on the chest radiography. Cardiac catheterization and angiography are diagnostic for this condition. We report here on an unusual case of pseudocoarctation of the aorta associated with the anomalous origin of the left vertebral artery.

CASE REPORT

A 6-year-old boy was admitted for cardiac murmur and a tingling sensation of the lower extremities on exercise. Physical examination revealed normal pulses in all four extremities with a blood pressure of 130/60 mmHg in the right arm, 132/70 mmHg in the left arm, and 100/74 mmHg in the lower extremities on admission. A grade 3/6 harsh pansystolic murmur of the left sternal border was heard. The chest X-ray studies revealed a normal sized heart. There was no evidence of rib notching, mediastinal widening or mass, or a cervical aortic arch. Electrocardiogram showed no abnormal findings. Two-dimentional echocardiographic examination showed a small perimembranous ventricular septal defect and a suspicious coarctation of the aorta (the blood velocity at the isthmus: 2.8 m/sec). There was no left ventricular hypertrophy, aortic stenosis/regurgitation or bicuspid aortic valve. The subsequent multichannel helical computed tomographic (CT) scan revealed the follows (Figs. 1A–C): The aortic arch was kinked and elongated and the narrowest part measured approximately 7 mm. No collateral circulation was detected. The left vertebral artery arose directly from the aortic arch and the order of these vessels was as follows; right brachiocephalic, left common carotid, left subclavian and left vertebral artery. The isthmic portion of the descending thoracic aorta was not visualized to be adjacent to the spine, but rather ventral to it, and this was surrounded by aerated lung due to the elongated and kinked aortic arch. Retrograde cardiac catheterization revealed that the trans-pseudocoarctation pressure gradient was 20 mmHg. We thought the patient's symptoms were caused by dynamic aortic narrowing during exercise, although the resting pressure gradient



A B C

Fig. 1. Pseudocoarctation of aorta associated with anomalous origin of left vertebral artery in 6-year-old body.
A. Three-dimensional CT reconstruction demonstrates elongated and kinked aortic arch (long arrow) with anomalous origin of left vertebral artery (short arrow).
B. Contrast enhanced CT reveals isthmic portion of descending aorta is not adjacent to spine and it is surrounded by aerated lung (arrow).
C. Superior view of volume rendered image shows that aortic arch is elongated and kinked and isthmic portion of descending aorta is not adjacent to spine (arrow).

was not significant; we decided to perform a surgical procedure. The elongated and mildly dilated arch was resected and we performed end-to-end anastomosis. The patient was discharged without any complications. The pre-operative symptoms disappeared after surgery.

DISCUSSION

Pseudocoarctation of the aorta is a relatively rare congenital anomaly that consists of elongation and kinking of the aortic arch and narrowing of the aortic isthmus without significant obstruction (1, 2).

The exact etiology of pseudocoarctation of the aorta is not currently known. It has been proposed that the embryologic cause of pseudocoarctation of the aorta is a failure of compression of the third through the seventh segments of the dorsal aortic roots and the fourth arch segment (3). The elongation of the arch frequently produces an increased distance between the origin of the left common carotid and the left subclavian artery. This case had an elongated and kinked aortic arch, but the distance from the left common carotid to the left subclavian artery was normal. So, the presently proposed assumption does not explain the whole situation.

In our case, there was an exceptional association of a pseudocoarctation and the anomalous origin of the left vertebral artery. Embryologically, the aberrant origin of the left vertebral artery directly from the aortic arch is due to persistence of the 8th intersegmental artery (4). Vorster et al. (5) noted that the vertebral artery arose from the aortic arch in 5% of cadavers. To the best of our knowledge, there have been no previous reports of pseudocoarctation of the aorta associated with the anomalous origin of the left vertebral artery. This report will be helpful for better understanding the exact etiology of pseudocoarctation of the aorta.

Cardiac catheterization and angiography provides a definitive diagnosis for this condition. Measuring the

pressure gradient by cardiac catheterization is necessary for the diagnosis of this condition. Similarly, CT is very helpful, and the following findings are considered diagnostic of this congenital anomaly according to several case reports; 1) demonstration that the abnormal mass in the mediastinum is part of the aorta; 2) an unusually aortic arch high in the mediastinum; 3) visualization of the isthmic portion of the descending thoracic aorta that's not adjacent to the spine, but rather ventral to it, and this is surrounded by aerated lung; and 4) a more caudal origin of the subclavian artery (6, 7).

A variety of congenital heart defects have been reported in association with pseudocoarctation (1, 3, 8). Pseudocoarctation is sometimes combined with aneurysm and aneurysmal dilatation beyond the lesion.

Pseudocoarctation of the aorta has been described in the literature as a "benign" entity that warrants no specific therapy, but reports on aneurysm formation and rupture in patients with pseudocoarctation have caused some changes for its treatment strategy (3, 8, 9). Surgical treatment should be recommended for all the symptomatic patients and for the pseudocoarctation of the aorta that is associated with aneurysm formation, and close follow-up is required for the asymptomatic patients who are without any associated anomalies.

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