Right Aortic Arch with Aberrant Left Subclavian Artery in Takayasu Arteritis

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ABSTRACT

A 38-year-old woman was admitted to this hospital because of intermittent claudication in the lower limbs for 2 years. Lower limb pulsations were absent bilaterally. She was evaluated and diagnosed to have Takayasu arteritis (TA). The magnetic resonance angiogram revealed a right arch of aorta with aberrant left subclavian artery (LSA). An association of TA with right aortic arch and aberrant LSA has not been reported so far.

Key words: Aberrant left subclavian artery; right arch of aorta; Takayasu arteritis

Introduction

Takayasu aorto-arteritis is a rare form of nonspecific inflammatory disease of unknown origin seen most frequently in young women that appear to be more common in Asia. The clinical manifestations are varied and related to the vessel that presents the stenotic or occlusive lesions.[1] Among Indian patients with Takayasu arteritis (TA), males have a higher frequency of hypertension and abdominal aorta involvement while females have a tendency toward the involvement of aortic arch and its branches.[2] A right-sided aortic arch is seen in 0.1% of the general population. About half of them have an aberrant left subclavian artery (LSA), which may arise either directly from the aorta or from the Kommerell's diverticulum.[3]

Case Report

A 38-year-old woman was admitted to this hospital because of intermittent claudication in both lower limbs for 2 years. She had never experienced any previous medical problems apart from obsessive compulsive disorder. Her blood pressure was 130/80 mm of Hg, and lower limb pulsations were absent bilaterally. She had microcytic hypochromic anemia. Her total leukocyte count was 9600/μl, platelet count was 2.4 × 10^9/L, erythrocyte sedimentation rate was 65 mm in 1 h, and C-reactive protein was high. There was no albuminuria or visible red blood cell in the urine. Chest X-ray and electrocardiogram were normal. Tests on liver function, thyroid function, and renal function were normal.

Arterial Doppler studies of lower limbs showed reduced flow in both lower limbs suggesting an aortoiliac disease. The echocardiogram was normal, and vasculitis workup including antinuclear antibody profile, rheumatoid arthritis, cytoplasmic antineutrophil cytoplasmic antibody, perinuclear anti-neutrophil cytoplasmic antibody, antiphospholipid antibodies, hepatitis B surface antigen, hepatitis C virus, human immunodeficiency virus, venereal disease research laboratory were all negative.

A contrast magnetic resonance angiogram of aorta and lower limb arteries showed a right aortic arch, aberrant LSA with mild dilatation of aorta at the origin of subclavian artery, long segment occlusion in infrarenal abdominal aorta and proximal popliteal artery occlusion on left side [Figures 1-6].

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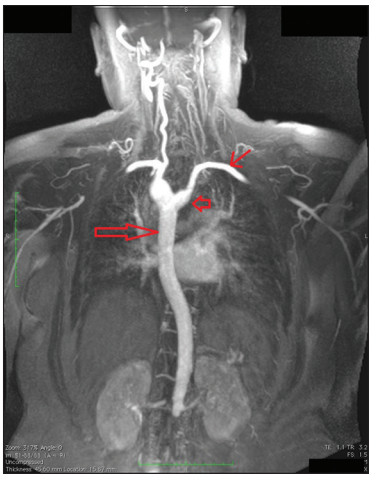

Figure 1: Coronal maximum intensity projection of an magnetic resonance angiogram unsubtracted image shows a right sided aortic arch (shown in large open arrow), an aberrant left subclavian artery (solid arrow) arising from dilated structure Kommerell's diverticulum (shown in small open arrow)

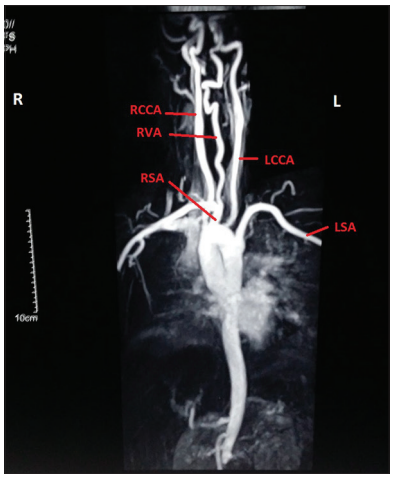

Figure 2: Coronal maximum intensity projection magnetic resonance angiogram subtracted image shows the right subclavian artery arising directly from arch giving a larger right vertebral artery. The right common carotid artery also has a direct origin from the arch. An Aberrant left subclavian artery and left common carotid artery have separate origins from the arch

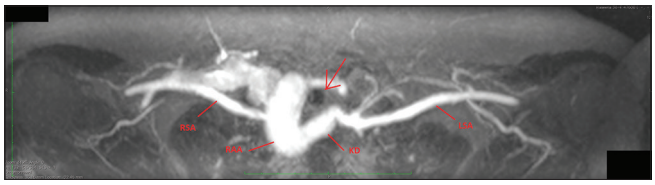

Figure 3: Axial maximum intensity projection magnetic resonance angiogram unsubtracted image showing aberrant left subclavian artery running behind the signal void of trachea-esophagus (shown in line arrow). KD: Kommerell's diverticulum, RSA: Right subclavian artery, RAA: Right aortic arch

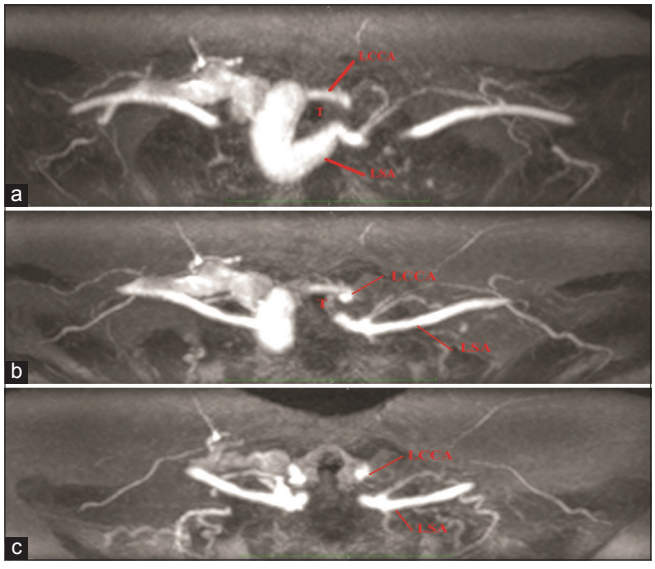

Figure 5a-c: Axial maximum intensity projection unsubtracted images in continuum showing aberrant LSA running behind the signal void of the trachea. The proximal left common carotid artery is coursing in front of the tracheal signal void. LSA: Left subclavian artery, LCCA: Left common carotid artery

A diagnosis of TA was made, and she was started on oral steroids with azathioprine.

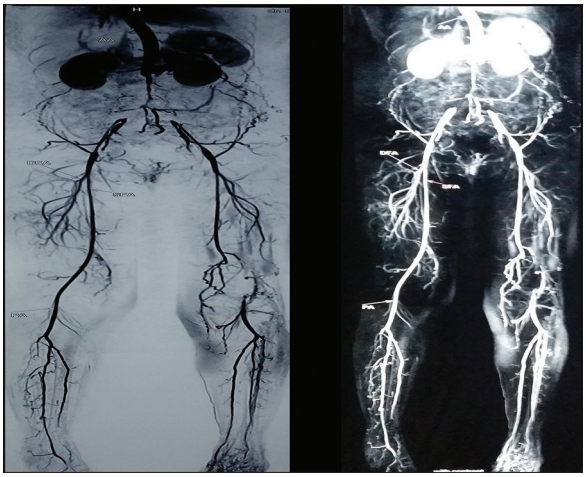

Figure 4: Contrast, magnetic resonance angiogram showing long segment occlusion in infrarenal abdominal aorta

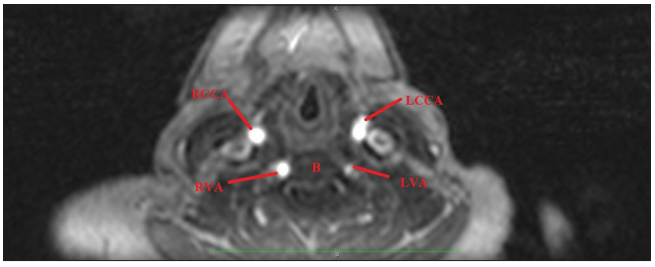

Figure 6: Axial maximum intensity projection unsubtracted image shows prominent right vertebral artery in right foramen transversarium of the vertebral body (B). LVA on the left side appears hypoplastic. RCCA: Right common carotid artery, LCCA: Left common carotid artery, RVA: Right vertebral artery, LVA: Left vertebral artery

Discussion

During development, a double aortic arch initially develops with a left and a right arch connecting the ascending with the

descending aorta. The trachea and esophagus are situated in between them. A common carotid artery and a subclavian artery arise from each arch. Normally, the right arch distal to the origin of the right subclavian artery regresses, the right common carotid and subclavian artery merge to form the brachiocephalic artery, while the left aortic arch persists and descends on the left side of the spine. Abnormal aortic arch development is seen in approximately 1–2% of human fetuses.[4] Regression occurring within the left arch, between the common and LSAs results in a right aortic arch giving rise to four vessels: The left common carotid, the right common carotid, the right subclavian, and the (aberrant) LSAs.[5] The right sided aortic arch is seen in 0.1% of the population of which half are associated with an aberrant LSA. Kommerell's diverticulum (KD) is the aneurysmal diverticulum of the descending aorta at the origin of the aberrant subclavian artery and is an embryologic remnant of the left fourth aortic arch in patients with a right aortic arch.

KD and aberrant subclavian artery can be accidentally detected in adults, but may be associated with complications due to compression of adjacent structures, dissection, or rupture.[6] Our patient had a right aortic arch with aberrant LSA with mild dilatation of aorta at the origin of subclavian artery detected incidentally.

TA is an inflammatory and stenotic disease of medium- and large-sized arteries characterized by a strong predilection for the aortic arch and its branches. The disease is a panarteritis with inflammatory mononuclear cell infiltrates and occasionally giant cells. There's marked intimal proliferation and fibrosis, scarring and vascularization of the media, and disruption and degeneration of the elastic lamina. TA is thought to result from an autoimmune process that targets large elastic-containing arteries. TA is widely recognized as a multifactorial disease, although a genetic factor has been suggested as its cause because of geographic clustering of cases. Genotyping in TA provides two independent genetic susceptibility loci in the human leukocyte antigen class I and class II regions and identifies and establishes a genetic association in the *FCGR2A/FCGR3A* and *IL12B* loci.[7] Classification is based on the vessels involved in the inflammatory process as seen on angiography.

- Type I: Branches of the aortic arch
- Type IIa: Ascending aorta, aortic arch, and branches of the aortic arch
- Type IIb: Ascending aorta, aortic arch, and its branches and thoracic descending aorta
- Type III: Thoracic descending aorta, abdominal aorta, and/or renal arteries
- Type IV: Abdominal aorta and/or renal arteries
- Type V: Features of types IIb and IV.

Right aortic arch with aberrant LSA may be associated with congenital heart defects. Conotruncal anomalies, Ventricular septal defects, Atrioventricular canal defects, atrial septal defects, and pulmonic stenosis/tricuspid atresia were found to be associated with a right aortic arch with the aberrant subclavian artery.[8] Our patient had a right aortic arch with aberrant LSA along with TA, which might be a chance coincidence. The association described, in this case, to our knowledge is the first such report.

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Conflicts of interest
There are no conflicts of interest.

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