The cervical aortic arch is defined as an arch that is supraclavicular (1). Though often isolated, it may be associated with certain congenital aortic or cardiac anomalies, including ventricular septal defect, double outlet right ventricle, tricuspid atresia, and atrial septal defect (2). Herein, we report a case of a young girl with Turner’s syndrome presenting with a pulsatile left-sided supraclavicular swelling since birth, which proved to be the rare anomaly of a cervical aortic arch. Though elongation of the transverse aortic arch is well known in Turner’s syndrome, to the best of our knowledge, a cervical aortic arch has not been described in the literature.

Case report

An 18-year-old girl presented with a pulsatile swelling present since birth in the left side of her neck, which had gradually increased in size. There were no specific symptoms related to cardiac disease. She was also under evaluation for delayed mental and physical milestones and absence of secondary sexual characteristics.

On clinical examination, she had a short stature with webbing of the neck. A pulsatile swelling was seen in the left side of the neck. Normal blood pressure was noted with pulses equal in both upper and lower limbs. Cardiovascular examination was normal.

On echocardiography, the left carotid artery was thought to be aneurysmal. On catheter angiography, the catheter could not be advanced beyond the point of a looped segment of the aortic arch.

A contrast-enhanced 3D magnetic resonance angiogram (MRA) was then performed on a 1.5 T magnetic resonance imaging (MRI) scanner (Siemens Avanto, Erlangen, Germany). This revealed a left-sided aortic arch with the right common carotid artery arising from the proximal arch. The arch subsequently passed into the neck up to the C3-C4 level where the left internal and external carotid arteries arose directly from the arch (Fig. 1). The arch then bent acutely inferiorly passing into the upper thorax where it formed a loop from which the left subclavian artery arose. The right subclavian artery was aberrant in origin arising from the descending thoracic aorta. This artery showed a long proximal segment of diffuse stenosis (Fig. 2). The remainder of the descending thoracic and abdominal aorta and its other branches were normal in caliber (Fig. 3).

In view of the fact that her symptoms were minor, the patient was managed conservatively.

Discussion

During embryonic development, there are five or six pairs of aortic arches that connect the truncus arteriosus with the paired dorsal aortae. The normal arch is formed through progressive involution of seg-
Cervical aortic arch

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133Volume 16 • Issue 2

by Morris and Ruttley (4). Other associated anomalies of the cervical aortic arch and its branches include aneurysms, coarctation/pseudocoarctation, and stenosis of the arch or its branches, as was seen in our case.

There have been several reports of association of anomalies of the cervical aortic arch with deletions of chromosome 22q11, suggesting that the cervical aortic arch may be part of the DiGeorge spectrum of defects (5). Turner syndrome is well known to be associated with arch anomalies, including elongation of the transverse aortic arch, coarctation of the aorta, and aberrant right subclavian artery (6). Association of Turner syndrome with a cervical aortic arch as seen in our case, however, has not been described in the literature, to the best of our knowledge.

A leading hypothesis considered in the origin of cardiovascular anomalies in Turner syndrome is lymphatic hypoplasia obstruction, which affects a large number of 45X fetuses (6). Clinically, cervical arch anomalies, though rare, should be considered in the differential diagnosis of a pulsatile neck mass with the potential for being mistaken for a carotid aneurysm noted.

References