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Commentary

A unique case of right cervical aortic arch with anomalous left common carotid artery and absent right common carotid artery

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HIGHLIGHTS

- An 8 year old male presented with a pulsatile swelling in right carotid triangle since birth. CT angiography revealed right sided cervical aortic arch with left common carotid artery arising from ascending aorta and the right external and internal carotid arteries originating separately from cervical arch.
- This rare anomaly arises from interruption of embryological 3rd and 4th arches with regression of fourth arch leading to persistence of right second or third brachial arches.
- This case is Haughton type A.
- Cervical aortic arch is not a very commonly encountered entity and can be quite vexing for those who are seeing such a case in OPD for the first time. Although cervical aortic arch anomalies have been reported in literature, published reports of new cases will help to increase awareness regarding this anomaly and lead to a swift and efficient diagnosis and management.

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ABSTRACT

Introduction: Cervical aortic arch is a rare anomaly where-in the ascending aorta arises normally from the left ventricle and extends in such a fashion that the aortic arch is situated high in the neck on either side. This anomaly should be suspected in any child exhibiting a pulsatile swelling in the neck.

Case commentary: An 8 year old child presented with a pulsatile swelling on the right side of the neck since birth. CT angiography revealed right sided cervical aortic arch at C4-C5 level. The left common carotid artery arose from the ascending aorta at D4 with the right external and internal carotid arteries originating separately from the cervical arch. The right and left subclavian arteries arose from the descending aorta at D1 and D4 respectively.

Discussion: Although most patients with cervical aortic arch are asymptomatic, some have dysphagia from oesophageal compression and respiratory distress from tracheal compression. There are many anatomical variations in cervical aortic arch as mentioned by Haughton. This case is Haughton type A because apart from the presence of right cervical aortic arch, 2 additional oddities in the form of leftward descending aorta and abnormal origin of the left common carotid artery from ascending aorta with absence of right common carotid artery are noted.

Conclusion: Cervical aortic arch is not a very commonly encountered entity and can be quite vexing for those who are seeing such a case in OPD for the first time. Although cervical aortic arch anomalies have been reported in literature, published reports of new cases will help to increase awareness regarding this anomaly and lead to a swift and efficient diagnosis and management.

An 8 year old child presented with a pulsatile swelling on the right side of the neck since birth. On examination, there was an approximately 2 × 2 cm pulsatile swelling in the right side of the neck anterior to the sternocleidomastoid, 4 cm cranial to the right clavicle with palpable thrill and murmur on auscultation (Fig. 1). CT angiography revealed right sided cervical aortic arch at C4-C5 level (Fig. 2a). The left common carotid artery (LCCA) arose from the ascending aorta (AA) at D4 with the right external (RECA) and internal (RICA) carotid arteries originating separately from the cervical arch. The right (RSCA) and left (LSCA) subclavian arteries arose from the descending aorta at D1 and D4 respectively (Fig. 3). At D3 the

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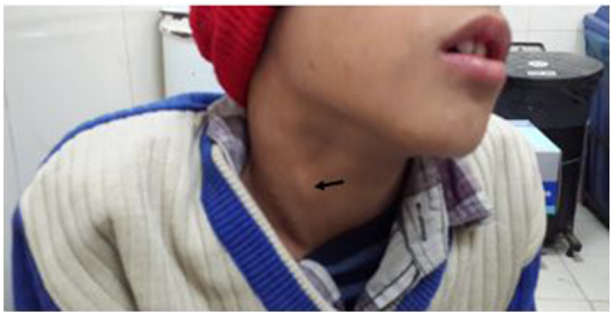

Fig. 1. Pulsatile neck swelling.

descending aorta (DA) coursed leftward and posterior to the oesophagus to form the left sided thoracic aorta at D4 (Fig. 2b). The patient was conservatively followed-up as he did not have any symptoms owing to this vascular anomaly and the diameter of the ascending aorta, aortic arch and descending aorta were within normal limits.

Cervical aortic arch is a rare anomaly where-in the ascending aorta arises normally from the left ventricle and extends in such a fashion that the aortic arch is situated high in the neck on either side.

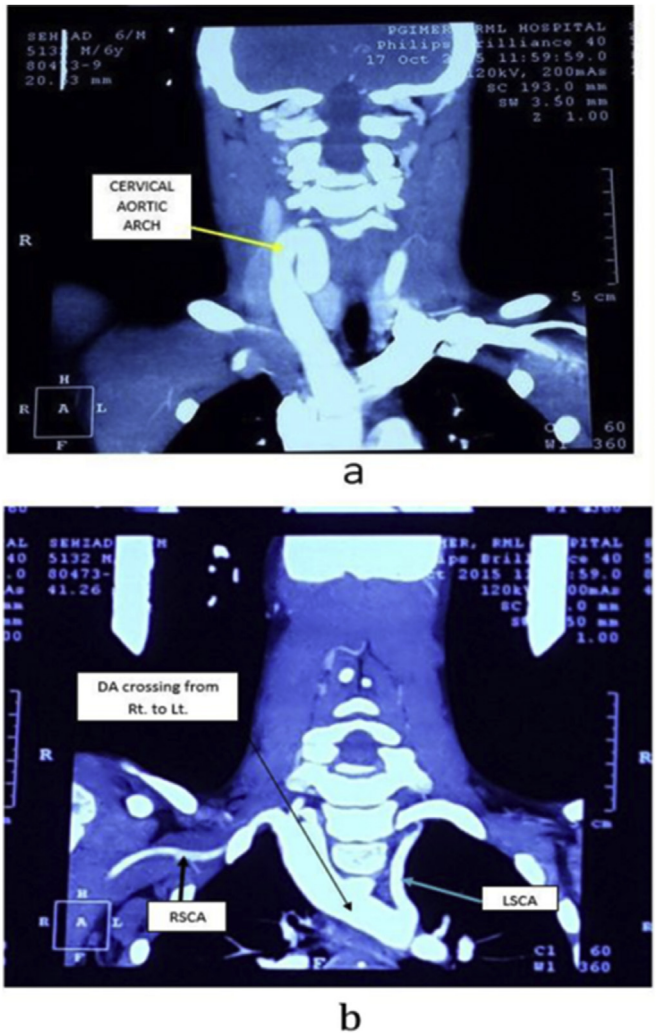

Fig. 2. Rt. cervical aortic arch (CT scan).

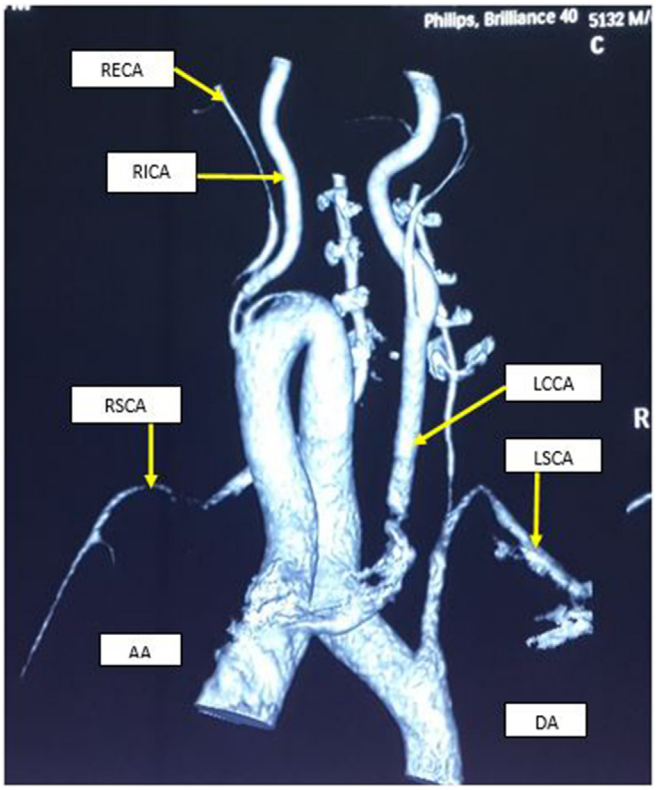

Fig. 3. Rt. cervical aortic arch and its branches.

This anomaly should be suspected in any child exhibiting a pulsatile swelling in the neck. It is considered to be due to interruption between embryological third and fourth arches with regression of fourth arch leading to persistence of right second or third brachial arches [1]. Although most patients with cervical aortic arch are asymptomatic, some have dysphagia from oesophageal compression and respiratory distress from tracheal compression [2]. Patients may even have complications in the form of aneurysms.

Haughton classification of cervical aortic arches [3]	
Type A	Contralateral descending aorta and absence of one common carotid artery (separate external and internal carotid artery branches)
Type B	Contralateral descending aorta and presence of both common carotid arteries
Type C	Contralateral descending aorta and bi-carotid trunk
Type D	Ipsilateral descending aorta with normal sequence of brachiocephalic branching
Type E	Right aortic arch and right descending aorta

There are many anatomical variations in cervical aortic arch. This case is Haughton type A because apart from the presence of right cervical aortic arch, 2 additional oddities in the form of leftward descending aorta and abnormal origin of the left common carotid artery from ascending aorta with absence of right common carotid artery are noted. Cervical aortic arch is not a very commonly encountered entity and can be quite vexing for those who are seeing such a case in OPD for the first time. Interestingly and

alarmingly an Otorhinolaryngology resident mistook the lesion for a lymph node overlying the carotid and wanted to do a FNAC (could have led to a disastrous consequence). Other differentials clinically could have been carotid body tumour, carotid or subclavian aneurysms. Although cervical aortic arch anomalies have been reported in literature, published reports of new cases will help to increase awareness regarding this anomaly and lead to a swift and efficient diagnosis and management.

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