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Congential Left Subclavian Steal Syndrome with Right Sided Aortic Arch

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Abstract:- The term "subclavian steal syndrome" was first suggested in 1961[1] to describe the hemodynamic changes and symptoms which occur when there is an occlusion of the intrathoracic portion of the subclavian artery, resulting in retrograde flow in the ipsilateral vertebral artery which "steals" blood from the basilar system and sometimes from the circle of Willis, resulting in symptoms of cerebral ischemia and/or claudication of the arm.

Congenital subclavian steal syndrome is a rare entity, with a reported incidence of approximately 40 cases in existingliterature[2].

We present a 33-years-old man with a 6-month history of intermittent claudication pain in his neck, who was eventually diagnosed with congenital subclavian steal syndrome.

Keywords:- *Subclavian steal, congenital subclavian steal, right sided aortic arch.*

I. INTRODUCTION

A 33-years-old manpresented with intermittent claudication pain in the neck,radiating to the left arm for 6 months, worsening with physical activity and abating with rest. He denied having any significant medical history prior to these symptoms.

Physical examination was significant for a radio-radial pulse deficit.

Blood pressure in the right arm in recumbent position was 120/80 mmHg and in the left arm was 90/70 mm Hg.

A provisional diagnosis of left subclavian steal syndrome was made and a carotid doppler was subsequently performed, which showed reversal of flow in the left vertebral artery.

The patient was advised to undergo coronary and upper limb angiogram for further evaluation.

Coronary and upper limb angiogram revealed a right sided aortic arch, with first branch being left common carotid artery, 2nd and 3rd branches beingright common carotid artery and right subclavian artery respectively.Atretic and un-opacifiedorigin of left subclavian artery was seen from the aorta, measuring approximately 29mm in length.

The left subclavian artery showed optimal opacification.

Both the vertebral arteries appeared prominent – right vertebral artery measuring 5.4mm and left vertebral artery, measuring 5.6mm (normal diameter – 2.9-3.4mm)

Additionally, there was a mild scoliotic deformity noted involving the distal cervical and proximal thoracic spine,with convexity towards the right side with partial block vertebrae involving D2, D3& D4 vertebral bodies..

➢ **FIGURE 1**

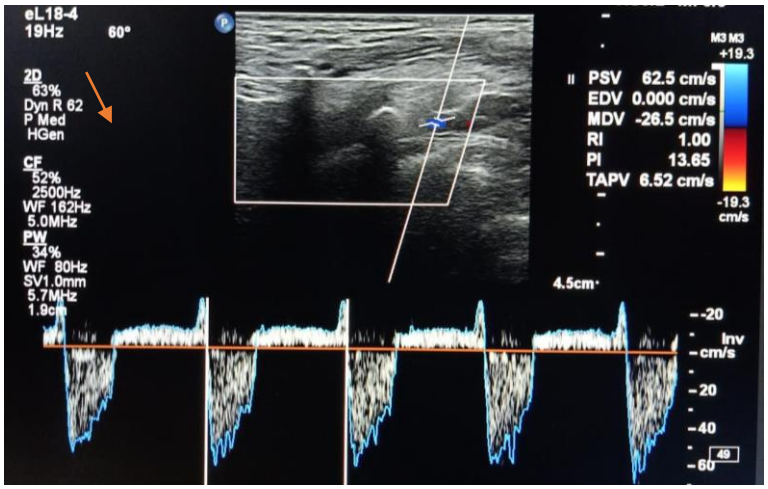

Fig. 1: Colour doppler of left vertebral artery (red arrow) showing reversal of flow, indicating "steal" phenomenon

➢ **FIGURE 2**

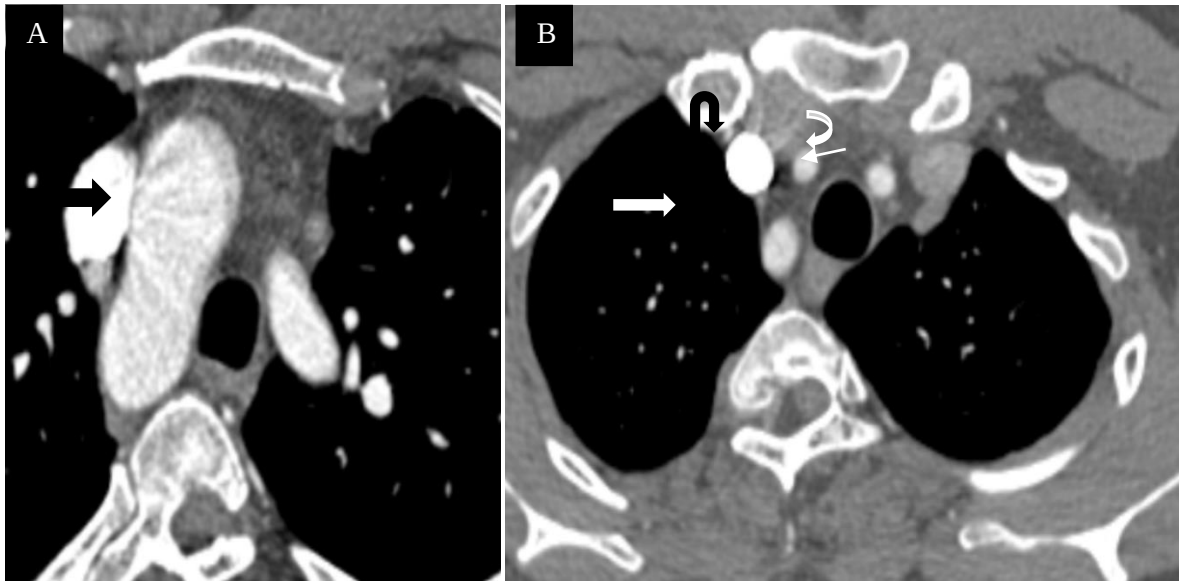

Fig. 2(A): Axial Contrast enhanced Computed Tomography (CT) image showing a right sided aortic arch (Black Arrow).

Fig. 2(B): Axial Contrast enhanced Computed Tomography (CT) image showing branches of the right sided aortic arch - First branch being Left common carotid artery (White curved arrow), second branch – Right common carotid artery (Black curved arrow) and right subclavian artery (Bold White straight arrow). Un-opacified and atretic origin of left subclavian artery is seen (White arrow).

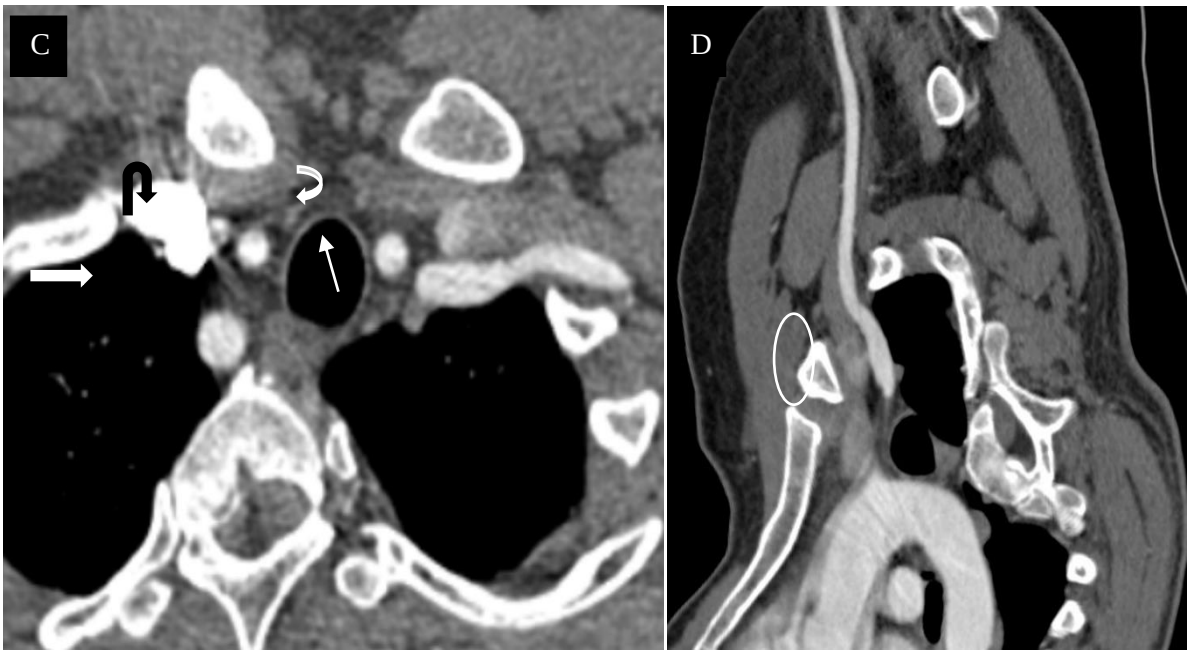

Fig. 2(C): Axial Contrast enhanced Computed Tomography (CT) image just below the level of FIGURE 2B,showing branches of the right sided aortic archas described in FIGURE 2B and right subclavian artery (White thin straight arrow). The left subclavian artery is optimally opacified (Figure 2C).

Fig. 2(D): Left coronal oblique reconstructed showing proximal atretic origin of the left subclavian artery (Inside white oval) from the aorta.

➢ **FIGURE 3**

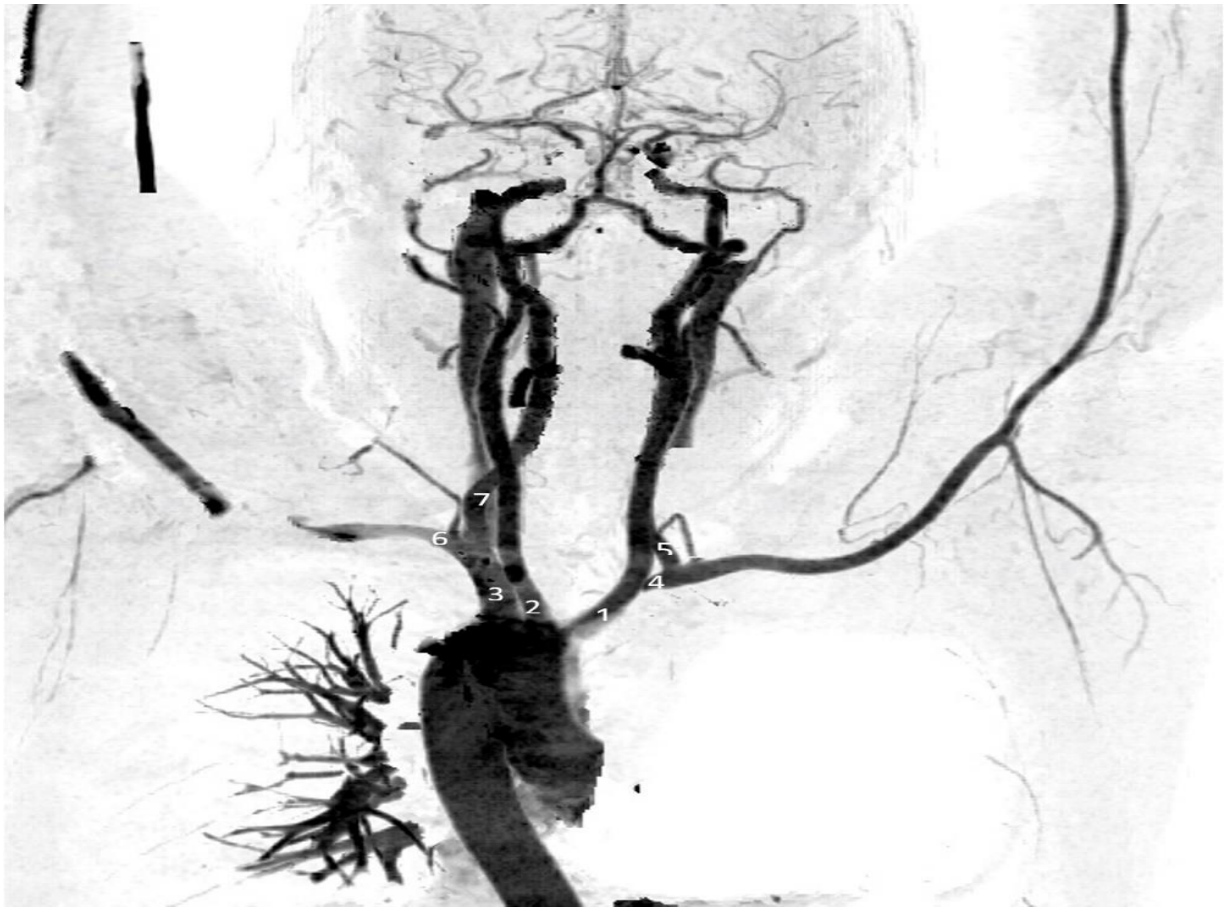

Fig. 3: Reconstructed CT angiography image showing branches of the right sided aortic arch - First branch being Left common carotid artery(1), second branch – Right common carotid artery (2) and right subclavian artery (3),left subclavian artery(4) is seen, which is showing no communication with the aortic arch, Left vertebral artery (5), Right subclavian artery (6), Right vertebral artery (7).

➢ **FIGURE 4**

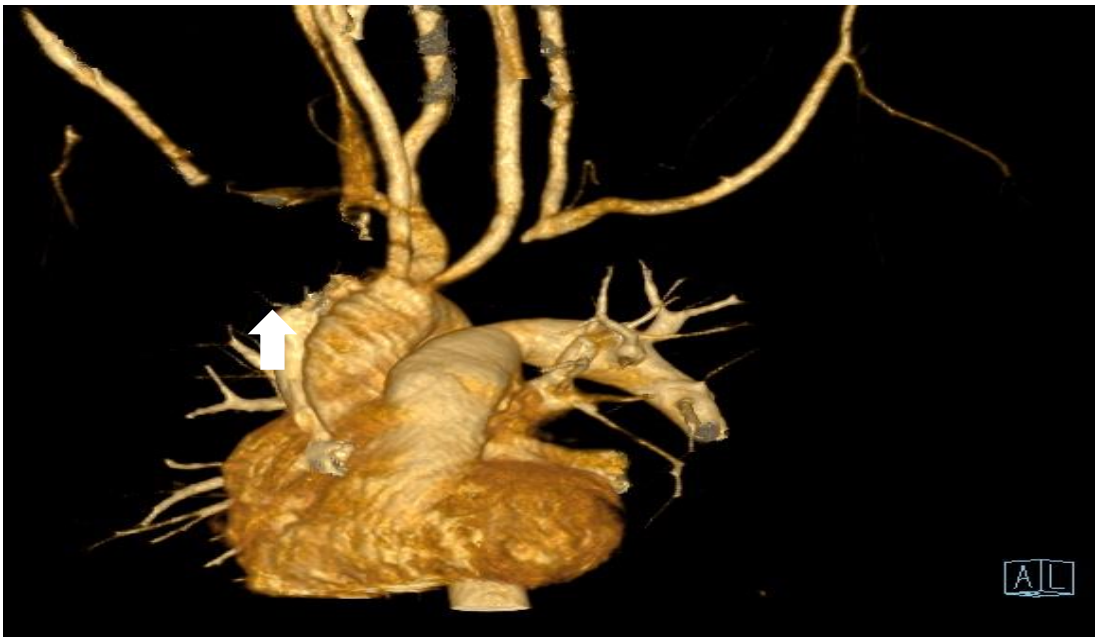

Fig. 4: 3D Reconstructed CT angiography image showing the atretic origin of the left subclavian artery which has no communication with the right sided aorta or with the left pulmonary artery

➢ **FIGURE 5**

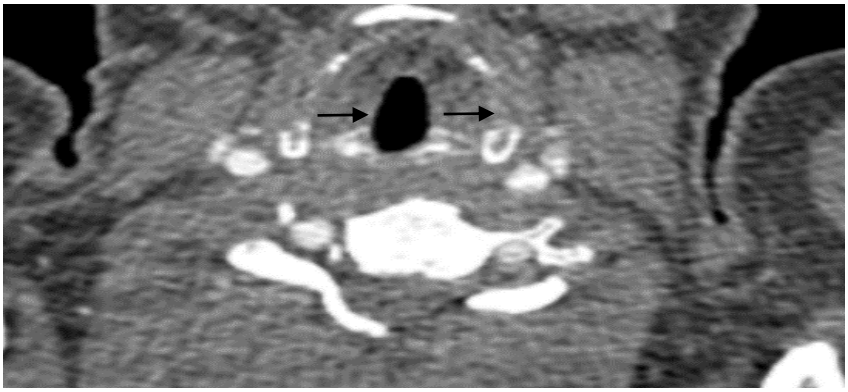

Fig. 5: Axial Contrast enhanced CT sections at the level of C7 vertebral body showing dilated bilateral vertebral arteries(Black arrows) within the foramen transversarium

➢ **FIGURE 6**

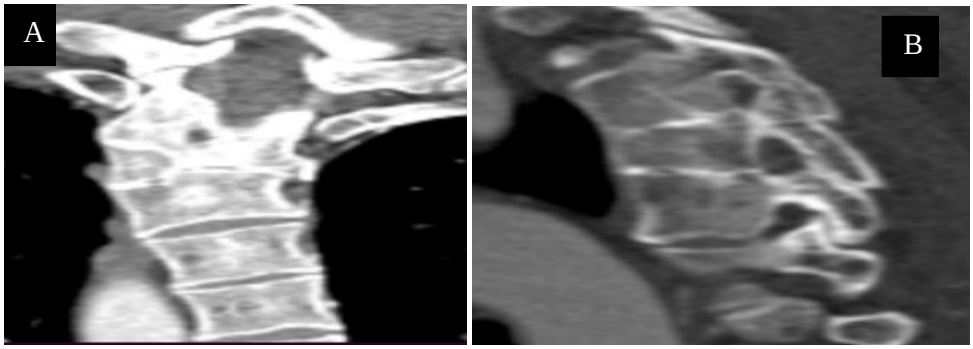

Fig. 6(A) & 6(B): Coronal and sagittal reformatted Contrast CT images at the level of upper thoracic spine showing partial block vertebrae involving D2, D3 and D4 vertebral bodies with mild scoliotic deformity towards the right side

II. DISCUSSION

A. Embrylogy

The aorta is embryologically derived from the 4th arch. The right sided aortic arch is rare with an incidence of 0.1%[3].

According to the Edward's hypothetical double aortic arch model, there is an aortic arch and a ductus arteriosus on each side, the right carotid and subclavian arteries arise from the right arch and the left carotid, and subclavian arteries originate from the left arch. The descending aorta is in the midline. Interruption of this arch system at different locations can explain the various aortic arch anomalies. The normal arch system results from interruption of the dorsal segment of the right arch between the right subclavian artery and descending aorta, with regression of the right ductus arteriosus[2,4].

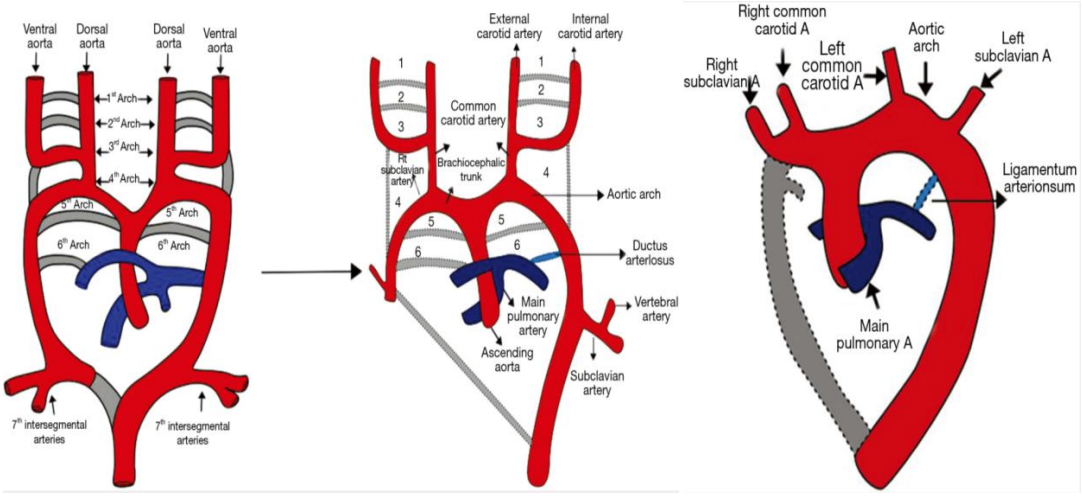

Fig. 7: Showing normal development of a left sided aortic arch from the 4th arch based on the Edwards hypothetical double aortic arch system

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Three subtypes of right sided aortic arch (RAA) based on the Edwards hypothetical double arch aortic system[5,6].

- Type I RAA with mirror image branching
- Type II RAA with aberrant left subclavian artery (Most common subtype of RAA)
- Type III RAA with isolated left subclavian artery

The type I right aortic arch with mirror image branching where the first branch is the left brachiocephalic artery, followed by right common carotid and right subclavian artery and is usually associated with intracardiac defects like truncus arteriosus, tetralogy of Fallot or tricuspid atresia[5].

Type II RAA with aberrant left subclavian artery (ALSA) is the most common subtype of RAA and the first branch arising from the aortic arch is the left carotid artery, which is followed by the right carotid artery, right subclavian arteries and ALSA[7].

RAA with isolated subclavian artery is the least common type and occurs due to regression of embryological left arch at two segments on either side of left subclavian artery[5].Theleft carotid artery arises as the first branch of the right arch, followed by the right carotid artery and right subclavian arteries. The LSA does not have a connection with the aorta, but it is connected with the pulmonary artery by a left ductus arteriosus that may be patent or closed[5,7]. The isolated left subclavian artery fills by retrograde flow from the left vertebral artery—a congenital left subclavian steal.

Schematic showing a right arch with isolated left subclavian artery that is explained by regression of the left arch at two locations indicated by *arrows 1* and *2*. *a.* = artery; *Ao* = aorta; *L.P.A.* = left pulmonary artery; *P.T.* = pulmonary trunk; *R.P.A.* = right pulmonary artery[2].

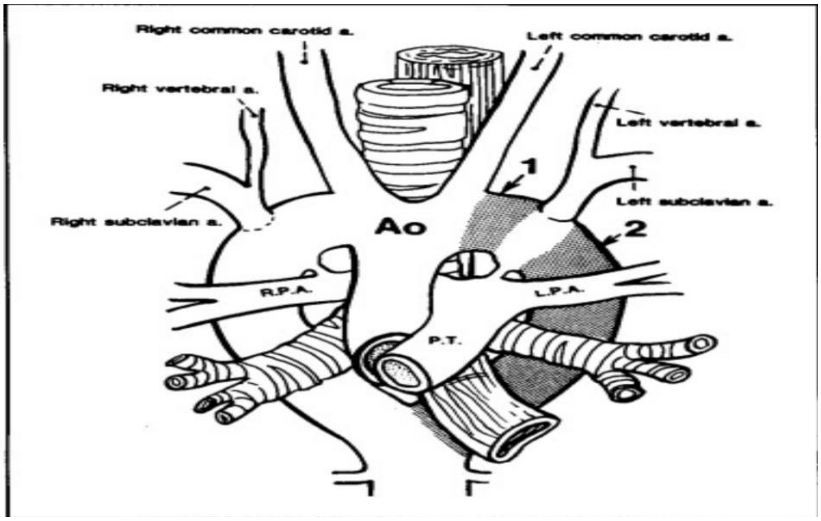

Fig. 8: showing a right arch with isolated left subclavian artery

Two variant forms of congenital left subclavian steal have been reported. If the interruption of the left arch between the left subclavian artery and the descending aorta is incomplete and an atretic cord persists, an intermediate variant between an isolated and an aberrant left subclavian artery is formed. In this variant, the atretic cord may complete a vascular ring, and an aortic diverticulum (of Kommerell) may be present at the insertion of the cord to the descending aorta[2].

Similarly, if the interruption of the left arch between the left common carotid and left subclavian arteries is incomplete and an atretic cord persists, an intermediate variant between an isolated left subclavian artery and mirror image branching is formed.Steal associated with a right aortic arch usually occurs on the left side and is caused by hypoplasia, atresia or isolation of the proximal portion of the left subclavian artery[8].

In our case, there is a right sided aortic arch with left common carotid artery as its first branch, followed by right common carotid and right subclavian arteries as its 2nd and 3rdbranches respectively. There is atretic origin of the proximal left subclavian arteryresulting inleft subclavian steal syndrome.

Congenital subclavian steal is characterized by a lack of symptoms during childhood, with most patients developing neurological steal symptoms or arm claudication with advancing age after decompensation of the collateral circulation.In the literature, only 17% of patients having congenital subclavian steal syndrome were symptomatic and the ages of the symptomatic patients at the time of diagnosis ranged from 22 to 53 years (mean, 37.3 years). Symptoms may include dizziness, vertigo, headache, disturbances of vision, dropping attacks, syncope, tinnitus, and cerebellar ataxia and can be exacerbated by exercising the involved upper extremity, which results in an increased demand for blood and an accentuation of the steal phenomenon. Symptoms of ischemia of the involved upper extremity, including pain, weakness, paraesthesia, and changes in temperature, may also occur[8].Symptoms can

often be provoked by rotating the head to the contralateral side and by arm exercise[9].

Congenital heart diseaselike Tetralogy of Fallot or tricuspid atresiais strongly associated in type I and type III right aortic arch[5]. However, no congenital heart disease waspresent in the patient we describe.

III. CONCLUSION

Congenital left Subclavian stealshould be suspected in patients having a right sided aortic arch presenting with symptoms of left arm claudication orpulse deficit/differential blood pressure on examination. The anatomical relation of the left subclavian artery to the aorta or the pulmonary artery has to be clearly demonstrated as it determines the type of congenital subclavian steal and subsequently aids in the surgical management.

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