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Early presentation and diagnosis of aberrant right subclavian artery vascular ring: Case report

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Background:
The aberrant right subclavian artery (ARSA) is congenital vascular anomaly, the majority of ARSA involves the subclavian artery passing posterior to the esophagus.
They are usually asymptomatic and if they become symptomatic usually presented with dysphagia.
Objective:
The Aim of presenting this case is to aware a physicians about such case with rare presentation regards to age and unique clinical features
Case report:
41 days baby boy, presented with history of stridor started a few days after birth, in which became progressive with time and associated with increase respiratory effort, especially during feeding. On physical examination his vital signs were stable with normal O2 at room air. It was noticed to have audible stridor that become less prominent in supine position. Patient underwent serial lab and radiological investigation in which CT scan showed a left-sided aortic arch with an ARSA passing atypically between the trachea and the esophagus, causing flattening of the posterior tracheal contour. A subsequent CT angiography done and revealed a unique anatomical pathway of ARSA.
Conclusion:
Aberrant right subclavian artery is rare in infancy. This disorder's symptoms range from being asymptomatic to more sever dramatical symptoms in which such cases necessitate careful clinical and radiological assessment to reach final diagnosis and perhaps appropriate intervention
Key words:
Vascular ring, Right aberrant subclavian artery (RASA), stridor

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I. Introduction:

The aberrant right subclavian artery (ARSA) was first described by Hunauld in 1735, with the clinical symptom, of the so-called "dysphagia lusoria" or "difficulty in swallowing due to a quirk of nature," as described by David Bayford in 1787.[1]In Bayford's words, the symptoms "may be called lusoria, from Lusus Naturae that gives rise to it.[2] It is also known as arteria lusoria as suggested by Arkin.[3] The majority of ARSA involves the subclavian artery passing posterior to the esophagus and rarely, if ever, passing in front of the esophagus.[4]They are usually asymptomatic rarely became symptomatic and if they become symptomatic usually presented with dysphagia which is correlated with common pathway of this vascular ring type which is passing posterior to esophagus.

Here we report the case of an aberrant right subclavian artery in a baby crossing between trachea and esophagus leading to stridor, to our knowledge this is the first case of ARSA with imaging correlation of a course anterior to the esophagus, interposing between the trachea and esophagus in a infant patient presented by stridor not dysphagia and no articles have described ARSA with this presentation.

Aberrant right subclavian artery is the most common anomaly arising from the aorta. Its incidence is between 0.5% and 2% worldwide.[5,6]

II. Case Report:

41 days baby boy, full term, who has been thriving well asymptomatic, few days after birth he developed harsh, interrupted sound on inspiration that started gradually and occurs during feeding were its prolonged with increase respiratory effort than the usual feeding, the mother admits that she tends to put the baby on the lateral position because she noticed the sound while the baby is on spine position that also waking him from sleep and increase respiratory effort during feeding. On physical examination vital signs were stable in form temperature 37 c, respiratory rate 35, heart rate 144 bpm, oxygen saturation 96 on room air, For the investigation complete blood count, renal function test with electrolytes and liver function test were within normal (table 1) . Initial chest and cervical spine x-rays were normal (figure 1). For visualizing the larynx fiberoptic scope done which revealed that omega shaped laryngomalacia type 2, so further imaging was done to explore the cause, cardiac ECHO done which was normal apart from tiny patent foramen ovale, atrial septal defect secundum Chest computed tomography (CT) with intravenous contrast confirmed a left-sided aortic arch with an ARSA passing atypically between the trachea and the esophagus, causing flattening of the posterior tracheal contourCT Angio was the last imaging done which revealed that, evidence of aberrant right subclavian artery in which the right subclavian artery instead to arise from the brachiocephalic artery, it arises on its own as fourth branch from the arch distal to the left subclavian artery then it hooks back to reach the right side (figure 2, 3) . Its relation to the esophagus is not clear in this exam, directly related to the posterior aspect of the tracheal bifurcation.

Table 1: blood investigation

INVESTIGATION	REFERENCE	PATIENTRESULT
CBC		
Totalwhitebloodcells	4.0-11.0k/µL	7.26
Hemoglobin	13.0-18.0g/dL	10.5
Biochemistry		
Creatinine	18-35umol/L	33
BUN	1.8-6.0mmol/L	2.7
Ca+	2.25-2.75mmol/L	2.31
Na+	136-146mEq/L	131
K+	3.5-5.1mEq/L	5.1
CL-	98-107mEq/L	104
Totalserum bilirubin	0.0-20.5umol/L	14.9
Albumin	30-52g/L	37
Alkalinephosphatase	0.0-500U/L	297

Figure1:CERVICALSPINEX-RAY

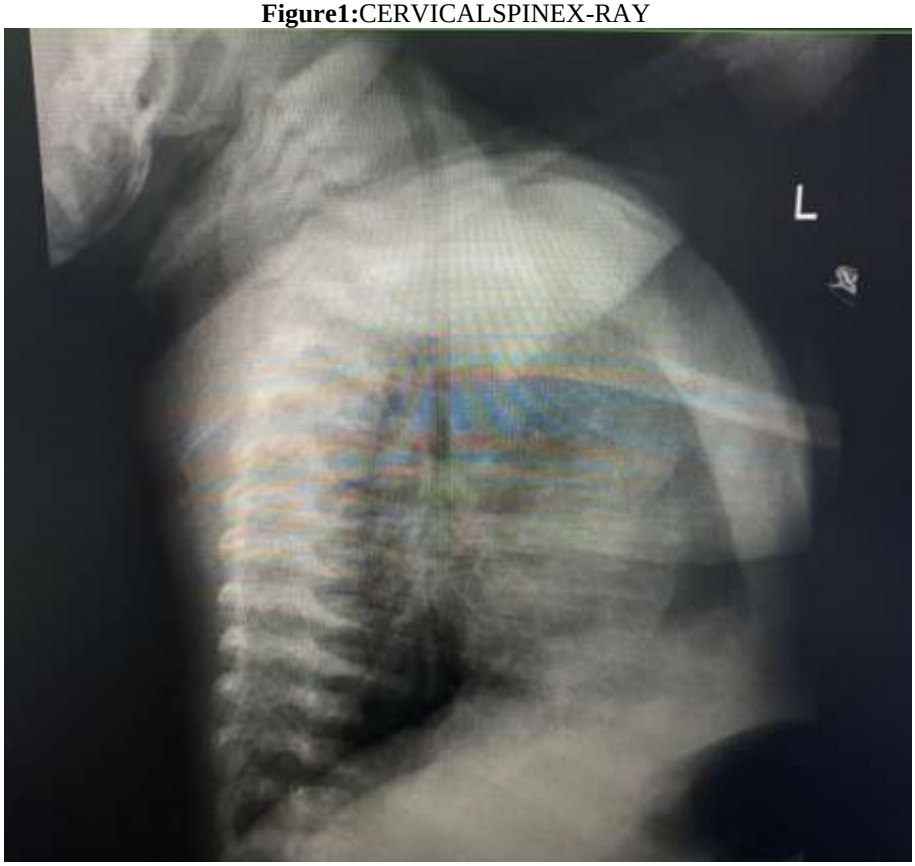


Figure2: CTANGIO

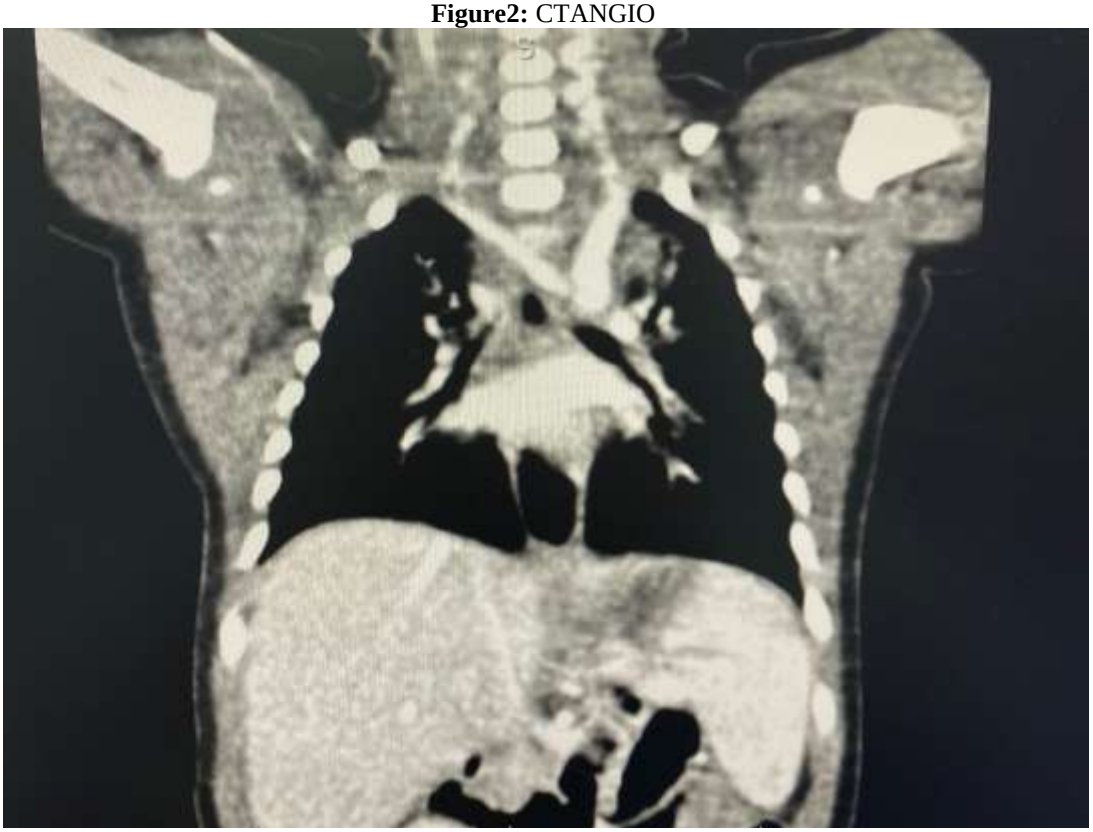
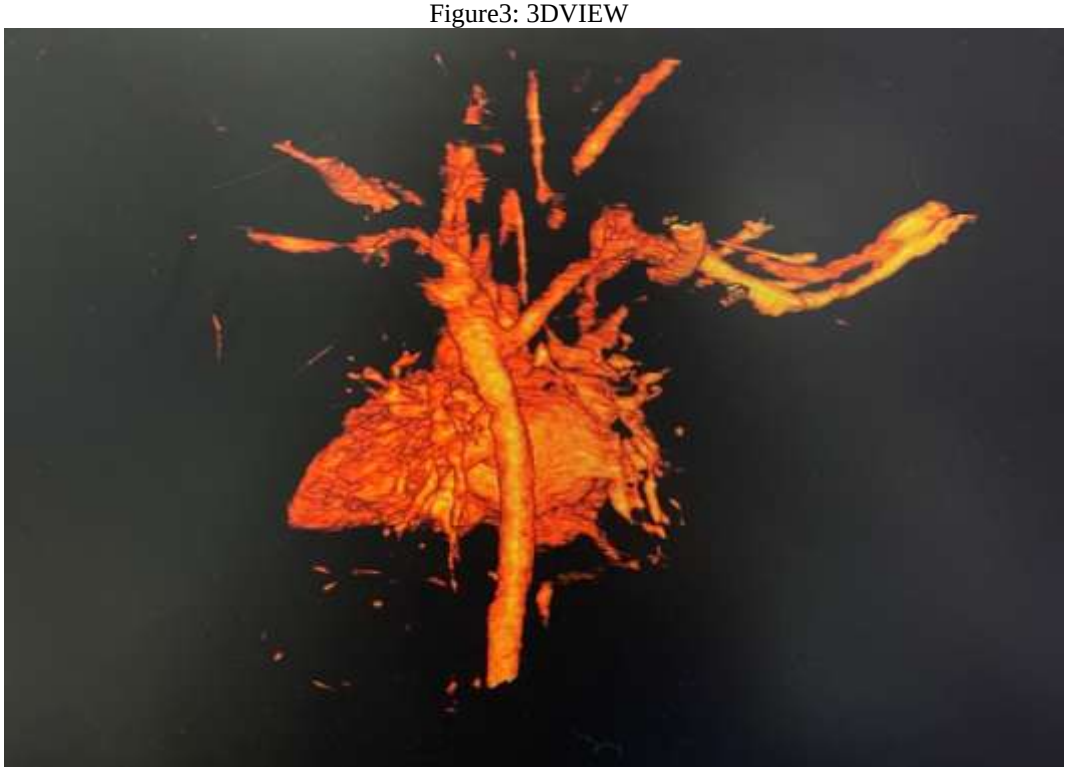


Figure3: 3DVIEW



III. Discussion:

ARSA is one of the most common intrathoracic major arterial anomalies, with a reported incidence between 0.4% and 2% in the general population.[4,5]According toMolz and Burri,ARSA predominates in female patients, whereas aberrant left subclavian arteries predominate in male patients.[6] Aberrant subclavian artery is one of the most common arch anomalies. Around 0.7–2% of the population with a left aortic arch and 0.04–0.4% with a right aortic arch and aberrant right subclavian artery.[7,8]It arises as the last brachiocephalic branch of the aorta and makes a U-turn to reach the right half of the body. In 80% of cases the aberrant subclavian artery crosses behind the esophagus.[9]In 15% it crosses in between the trachea and the esophagus and in 5% it crosses anterior to the trachea.[10]In our case ARSA passing atypically between the trachea and the esophagus symptoms of ARSA occur with different manifestations depending on the age.Infants mostly present with wheezing, frequent pulmonary infections, feeding problems, due to absence of tracheal rigidity or stridor just like our case[11]

Most cases of aberrant right subclavian artery are asymptomatic, especially if there are no additional aortic arch/vessel anomalies. It is typical to have respiratory distress in infancy but dysphagia can occur at an older age. Extrinsic compression of the airway and esophagus caused by anAberrant right subclavian artery is rare in infancy. This disorder ranges from being asymptomatic to circumstances of respiratory distress, Stridor, wheeze, dyspnea, recurrent respiratory tract infections or feeding problems which may necessitate surgical intervention.[8]

In our case, baby presented with history of expiratory stridor started on first week of life which is not common at this age of presentation and as clinical presentation. Although most patients are asymptomatic, but 25%–37% patients have congenital cardiac defects such as conotruncal abnormalities or other chromosomal abnormalities.[7,8,12,13]

Majority of retro esophageal ARSA cases are also clinically silent, with symptoms in 10% to 33% of cases, which may be either due to abnormal course of the anomalous vessel or due to the presence of a diverticulum ofKommerell.[9,19]The aberrant subclavian artery can be associated with an aneurysmal dilatation at the base known as Kommerell'sdiverticulum, which can rupture if left untreated.[22] Dysphagia and tracheal compression are common in children.[20] Adults have a completely formed cartilaginous trachea, so respiratory symptoms are rare, but they may exhibit the aforementioned "dysphagia lusoria."[21]

The most commonly reported chromosomal abnormality with an aberrant right subclavian is Trisomy 21.[14] In literature review study of 106 fetuses with Trisomy 21, the rate of this vascular anomaly was 25% in the second trimester ultrasound.[13,14] In our case the examination shows no apparently dysmorphic feature and karyotype not done.

In the presence of respiratory symptoms, the evaluation normally begins with chestradiography.[24]In

our case Initial chest and cervical spine x-rays were normal (figure 1). For visualizing the larynx, flexible airway endoscopy is the procedure of choice if the patient has stridors , noisybreathing .Fiberoptic scope was done which revealed that omega shaped laryngomalacia type 2, so further imaging was done toexplore the cause and because the stridor started at first week of life.

CT or MRI angiography is the gold standard diagnostic tool which has been replaced conventional angiography. It not only confirms the diagnosis but also helps to exclude aneurysm of the aorta, presence of vascular ring or other associated anomalies.[23]CT Angio was the last imaging done which revealed that evidence of aberrant right subclavian artery arises fromdistal part of descending aorta instead to arise from the brachiocephalic artery, it arises on its own as fourth branchfrom the arch distal to the left subclavian artery then it hooks back to reach the right side (figure 2, 3). Its relation tothe esophagus is not clear in this exam, directly related to the posterior aspect of the tracheal bifurcation for thisreason our patient presented with expiratory stridor.

The treatment depends on the symptoms, age comorbidity, and concomitant vascular abnormalities of each patient.[15]There is no indication of operative intervention for incidentally detected, asymptomatic, non-aneurysmal ASA.[16]Surgical approach is indicated when ARSA is symptomatic or has evidence of aneurysm.[17] And in the present ofaneurysm the right subclavian artery ligature is recommended when the patient symptomaticVarious surgical approaches can be used, each with its own advantages and limitations.[18]For the pediatrics age group, we considered operative management for them when they have signs of airway,esophageal compression, recurrent chest infection, FTT, recurrent ARDS or dysphagia.

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