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CASE REPORT
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A rare case of obstructive supracardiac TAPVR (Total Anomalous Pulmonary Venous Return) with aberrant right subclavian artery.

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ABSTRACT… Congenital aortic arch malformations manifest a broad-spectrum of differences and abnormalities that come from disturbed embryogenesis of branchial arches. Current case was a 10 months old baby girl with length of 69 cm (less than –3 SD) and weight of 5.5 kg (less than –3 SD). The patient had history of recurrent lower respiratory tract infections since the time of birth and failure to gain adequate weight since the time of birth. The patient has been having multiple check-ups with registered medical practitioners in the nearby locality and multiple courses of antibiotics with only partial resolution of symptoms. The 2-D echocardiogram showed her to be a case of supracardiac type of "Total Anomalous Pulmonary Venous Return (TAPVR)". All pulmonary veins making a confluence and draining into the right atrium. Significant turbulence observed at the level of superior vena cava to right atrium junction. A level of obstruction was recorded at the junction of the confluence of pulmonary veins and the vertical vein. There was aberrant right subclavian artery from the aortic arch as its third branch with no obstruction or aneurysm formation, having retrotracheal and esophageal course.

Key words: Echocardiogram, Right Atrium, Supracardiac, Superior Vena Cava.

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INTRODUCTION

Congenital aortic arch malformations manifest a broad-spectrum of differences and abnormalities that come from disturbed embryogenesis of branchial arches.[1] These cases may present with or without any symptoms depending upon the usage and timing of imaging and diagnostic tools. These abnormalities may present as isolated entity or could be linked with different other types of intracrdiac or chromosomal anomalies.[2,3] Imaging is considered to be a precious tools for the identifications of these abnormalities and help assisting accurate pre-surgery decisions.[4,5]

"Total anomalous pulmonary venous return (TAPVR)" is also termed as "total anomalous pulmonary venous connection (TAPVC)". In TAPVR, all 4 pulmonary veins are unable to form usual link to the left atrium resulting into drainage of all pulmonary venous return in the systemic venous circulation.[6] The incidence of TAPVR is between 0.6-1.2/10000 live births.[6] In newborns having congenital heart defects, TAPVR occurs in 0.7-1.5% of cases.[7,8] Echocardiographic confirms the diagnosis usually whereas MRI and fast CT also assist in describing pulmonary venous drainage.[9,10]

Case Presentation

Current case was a 10 months old baby girl with length of 69 cm (less than –3 SD) and weight of 5.5 kg (less than –3 SD), resident of Karachi, who presented to the out patients department of the National Institute of Cardiovascular Diseases (NICVD). The patient had history of recurrent lower respiratory tract infections since the time of birth and failure to gain adequate weight since the time of birth. The patient has been having multiple check-ups with registered medical practitioners in the nearby locality and multiple courses of antibiotics with only partial resolution of symptoms. The initial work up in the form of chest x-ray (CXR) and electrocardiogram (ECG) categorized the patient into the ones with increased pulmonary

blood flow and pulmonary arterial hypertension. As this warranted further investigation with a transthoracic echocardiography, the patient was advised a departmental echocardiogram.

The 2D echocardiogram done in the pediatric cardiology department of NICVD showed her to be a case of supracardiac type of TAPVR. All pulmonary veins made a confluence and draining into the right atrium. The path of the anomalous channel was: confluence of pulmonary veins to the vertical vein to the innominate vein to the superior vena cava. It is important to mention the significant turbulence observed at the level of superior vena cava to right atrium junction. A level of obstruction was recorded at the junction of the confluence of pulmonary veins and the vertical vein. Figure-1 and 2 are showing 2D images of trans thoracic echocardiography.

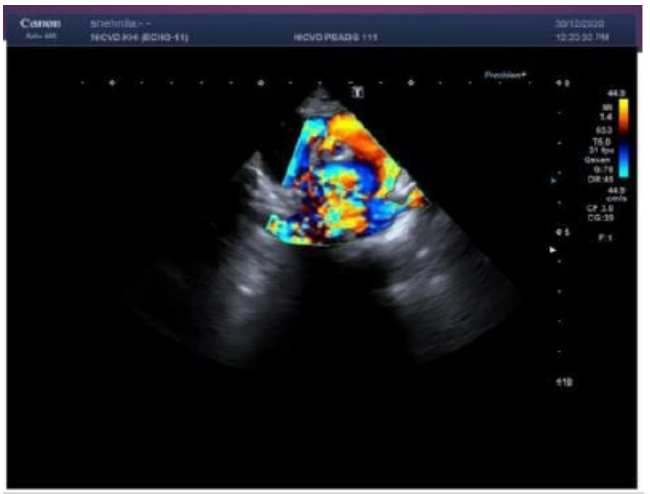

Figure-1. Transthoracic echocardiography showing path of anomalous channel.

Since the arch of the aorta and its branches seemed to give a bit confusing anatomy, the patient was advised CT angiography. The CT angiogram gave almost the same findings of supracardiac TAPVR with obstruction at two levels and the associated atrial septal defect (ASD) secundum. The anatomy of the aortic arch and its major branches was delineated. As per the CT angiogram, the patient had right sided aortic arch. Of special mention is the aberrant right subclavian artery from the aortic arch as its third branch with no obstruction or aneurysm formation, having retrotracheal and esophageal course. The relevant images of the CT angiogram are shown in Figure-3.

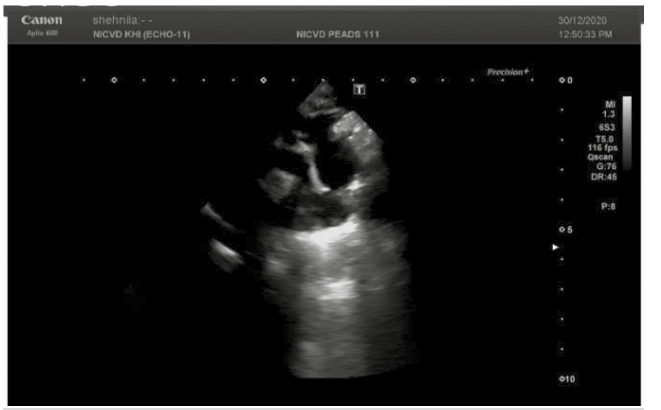

Figure-2. Transthoracic echocardiography showing obstruction at the junction of vertical vein and pulmonary venous confluence.

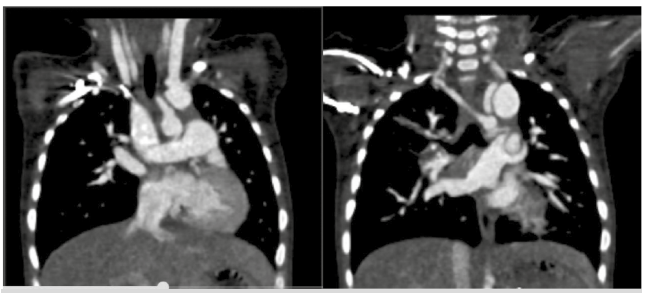

Figure-3. CT Angiogram showing right sided aortic arch.

DISCUSSION

Variety of imaging modalities are present to assist diagnosis of congenital anomalies of aortic arch. Conventional tools like barium esophagram has its own limitations in terms of its execution specifically among children.[1] Catheter angiography is another 2-dimentional tool is not usually preferred as other non-invasive options as for imaging exist. Echocardiography is generally the 1st imaging tool among children but it has its own operator dependent aspects especially among complex arch anomalies.[11] For the early and accurate description of pre-surgery anatomy, CT and MRI are so much valuable.[4,5] In the present case, 2-D echocardiogram showed supracardiac type of TAPVR. The CT angiogram gave almost the same findings of supracardiac TAPVR with obstruction at two levels and the associated ASD secundum. As per the CT angiogram, the patient had right sided aortic arch while the aberrant right subclavian artery from the

aortic arch as its third branch with no obstruction or aneurysm formation, having retrotracheal and esophageal course. As in the present case, CT delivers high spatial resolution assessment of vascular abnormalities. With emergence of newer generation scanners having modern radiation dose reduction technologies, the radiation dose has reduced significantly.[5]

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AUTHORSHIP AND CONTRIBUTION DECLARATION

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3	Waqas Ali	Literature review.	
4	Asif Ali Khuhro	Drafting, Discussion.	
5	Basharat Hussain	Literature review, Data collection.	
6	Shakeel Ahmed	Data collection, References	