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Case report

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Congenital Subclavian Artery Stenosis in a Patient with Dextrocardia and Situs Inversus: A Case Report

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ABSTRACT

Congenital right subclavian artery stenosis is extremely rare, particularly when associated with dextrocardia and situs inversus. We describe a 17-year-old boy who presented with a history of three-months of exertional right-hand discomfort and numbness. Examination revealed weak right upper limb pulses and an interarm blood pressure difference of 35 mmHg. Imaging confirmed dextrocardia with situs inversus and isolated proximal right subclavian artery stenosis from a mirror-image aortic arch. This case highlights the importance of considering congenital vascular anomalies in young patients with limb claudication and mirror-image anatomy for timely diagnosis and targeted management.

Keywords: Right subclavian artery stenosis; dextrocardia; congenital vascular anomaly

INTRODUCTION

Congenital anomalies involving the aortic arch and its branches are uncommon and often remain unnoticed until they cause hemodynamic compromise. True congenital narrowing of the subclavian artery is exceptionally rare and distinct from the more frequent acquired causes such as atherosclerosis or inflammatory arteritis. [1,2,3]

Upper limb claudication in adolescents is unusual and should prompt evaluation for an underlying structural vascular abnormality. The coexistence of subclavian artery stenosis with dextrocardia and situs inversus is an exceedingly uncommon finding. Dextrocardia with situs inversus occurs in approximately 0.01% of the population, and is usually accompanied by other developmental anomalies, with isolated vascular involvement seldom encountered. [4,5]

We present a rare case of a 17-year-old boy who developed right upper limb claudication due to congenital right subclavian artery stenosis associated with a mirror-image aortic arch, dextrocardia, and situs inversus. To our knowledge, this combination has been reported only infrequently in the literature.

CASE REPORT

A 17-year-old boy with no known comorbidities presented with a 3-month history of discomfort, coldness and numbness in the right hand, particularly during physical activity. There was no history of trauma, fever, dysphagia, weight loss or systemic symptoms. He denied resting pain, gangrene or tissue loss.

On examination, he had diminished right radial and brachial pulses, and a blood pressure difference of 35 mmHg between the right and left arms (right: 100/60 mmHg, left: 135/70mmHg). The right upper limb was cooler to touch, but capillary refill was preserved with same size as opposite limb. Neurological examination was normal. Cardiac apex was palpated on the right side with heart sounds best heard on the right precordium.

Chest radiograph showed right sided cardiac shadow with gastric bubble on the right, consistent with dextrocardia and situs inversus. Echocardiography confirmed mirror image anatomy with no structural defects.

Doppler ultrasound showed monophasic flow in the right subclavian artery. CT angiography revealed a mirror image aortic arch and isolated proximal right subclavian artery stenosis with no other vascular anomalies. The rest of the major vessels were normal and was not involved oesophagus and trachea (Fig.1-3) Given the presence of a viable limb and the absence of disability, conservative management was adopted. The patient was scheduled for regular follow-up and advised that intervention may become necessary if limb-threatening ischemic symptoms or functional impairment were to occur.

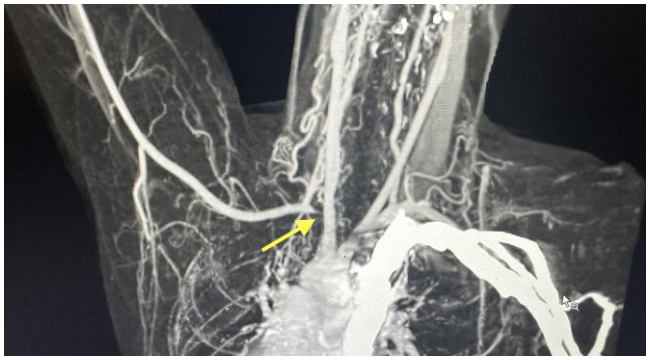

Figure 1: CTA of RSAS

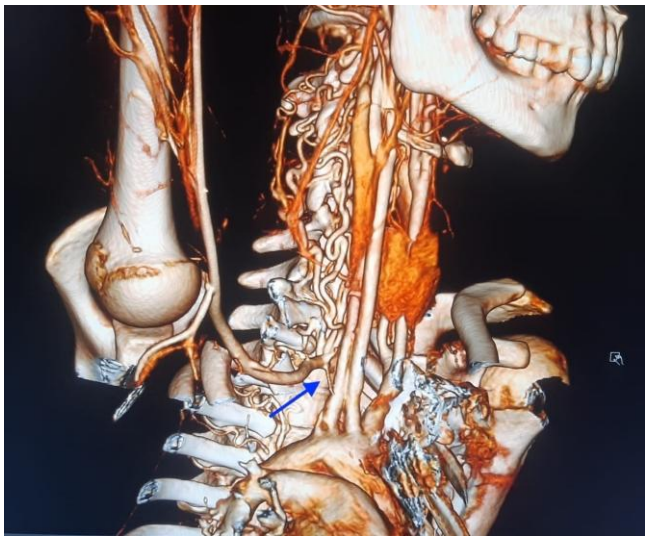

Figure 2 : CTA 3D reconstruction

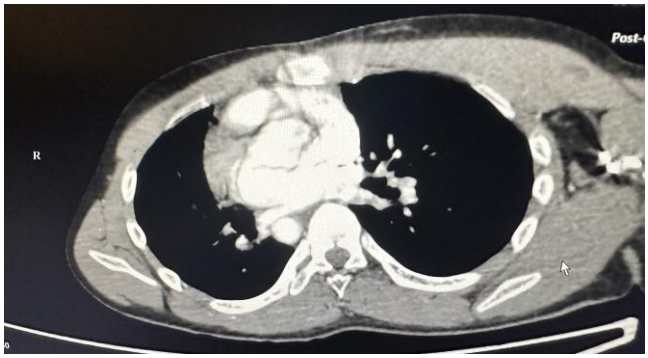

Figure 3: CTA dextrocardia

DISCUSSION

Congenital narrowing of the subclavian artery is an unusual vascular malformation and must be differentiated from more common acquired conditions such as arteritis or atherosclerosis. The subclavian arteries develop from distinct embryological components-the right subclavian artery originates from the right fourth aortic arch, the right dorsal aorta, and the right seventh intersegmental artery, while the left arises solely from the left seventh intersegmental artery. Developmental regression or malformation of any of these segments may result in focal hypoplasia or stenosis.

Most reported subclavian artery anomalies are related to variations in the origin or course rather than true congenital stenosis. Such anomalies are more frequently described in association with congenital heart disease or laterality defects. [6] In cases with dextrocardia and situs inversus, a mirror-image branching pattern of the aortic arch is typically present, and this may predispose to structural or caliber abnormalities in the subclavian arteries. [7,8,9]

In this patient, the young age, absence of vascular risk factors, and characteristic imaging findings support a

congenital origin of the lesion rather than an acquired process. Clinical features depend on the severity of the narrowing and collateral circulation. Symptoms such as upper limb claudication, inter-arm blood pressure discrepancies, or vertebrobasilar insufficiency (subclavian steal) may appear when compensatory flow is insufficient.

Detailed vascular imaging using CT or MR angiography is crucial to define the anatomy, determine the extent of the stenosis, and plan management. Recognition of the reversed thoracic anatomy is particularly important in surgical or endovascular intervention to avoid procedural complications.

CONCLUSION

This case report describes an exceptionally rare presentation of congenital right subclavian artery stenosis occurring in conjunction with dextrocardia and situs inversus. It reinforces the importance of maintaining a high index of suspicion for congenital vascular anomalies in young individuals with upper limb ischemic symptoms. Comprehensive imaging evaluation and awareness of reversed anatomical orientation are vital for correct diagnosis and safe management planning. Early identification facilitates appropriate, individualized treatment helps prevent avoidable procedures and complications.

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Author declaration

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Authors' contributions:

LAI and JA contributed to the conceptualization of the study. JA was responsible for data curation. LAI contributed to the development of the methodology. LAI prepared the original draft of the manuscript. Both authors critically reviewed and edited the manuscript and approved the final version for publication.

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The authors declare that they have no conflicts of interest.

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Ethics statement:
All patient data were anonymized and managed strictly to ensure confidentiality. Written informed consent was obtained from the patient for the publication.

Data Availability Statement
Data supporting the findings of this study are available from the corresponding author upon reasonable request.