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Case Report

Right internal jugular vein phlebectasia-a rare cause of neck swelling in paediatric population

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ABSTRACT

Reporting a case of an 8-year-old boy presenting with a right neck swelling appearing on straining which otherwise not seen with a diagnosis of right internal jugular vein phlebectasia. Paediatric population with neck swellings more commonly misdiagnosed clinically and hence with non-invasive radiological investigations like Ultrasonography with colour doppler and contrast enhanced computed tomography was done to confirm the diagnosis. Phlebectasias are at times incidental findings and stay asymptomatic, hence proper diagnosis with radiological investigations is needed as they are misdiagnosed with other neck swellings. Only few cases of internal jugular vein phlebectasias have been reported and approximately 20 cases have undergone surgery for cosmetic purposed and majority were managed conservatively like in our case report. This needs to be kept as a differential diagnosis in a paediatric patient presenting with neck swelling.

Keywords: Phlebectasia, Internal jugular vein fusiform swelling, Neck swelling rare cause, Fusiform IJV, Valsalva maneuver

INTRODUCTION

The term phlebectasia means outward dilatation of a vein without tortuosity which occurs as a neck swelling in paediatric population and serves a differential diagnosis for a swelling in the neck.[1] One such neck swelling occurred in an 8-year-old boy who presented to our Outpatient department diagnosed as phlebectasia and underwent conservative management and now being under follow up.

CASE REPORT

A 8 year old boy with informant mother presented to our ENT outpatient department with complaints of right side neck swelling since 4 months which was progressive in nature appearing only when the patient strains, speaks loudly, or while coughing. It was not associated with change in voice, difficulty in swallowing, respiratory distress or regurgitation of food. There was no history of trauma or previous surgery. No other comorbidities.

On clinical examination patient was comfortable, conscious, oriented, afebrile. No e\o respiratory distress. There was a soft cystic swelling on the right side of neck which appeared on doing Valsalva maneuverer (Figure 1) which otherwise remains undetected (Figure 2). There was no raise of temperature or tenderness\bruit. No other swelling in the neck. Flexible naso-pharyngo-laryngoscopy done which showed normal mobile vocal cords and no evidence of buldge or mass seen. Ultrasonography with colour doppler (Figure 3) was done which showed right Jugular Venous Phlebectasia with fusiform aneurysmal dilatation of internal jugular vein with aneurysmal segment measuring approx. 4.8×3.3×1.8 cm in size during valsalva maneuverer in this scan. No intra-lumen thrombus seen. CECT Neck was done which showed right internal jugular vein short segment fusiform

dilatation extending from hyoid cartilage superiorly to inferior pole of thyroid inferiorly, for a length of 4.5 cm, measuring 2.2 cm in maximum diameter. There is 1.8×1.4 cm sized well defined homogenously enhancing structure seen in left paratracheal region just above left clavicular head. The structure appears to continue as thymus in anterior mediastinum with similar density (Figure 4). Patient and parents were counselled regarding the condition of the patient and was advice for conservative management with avoidance of trauma to neck and speech therapy and avoid straining activities, as only few cases were reported insisting the need for surgery. At present patient is under follow up with conservative management and remained asymptomatic.[2]

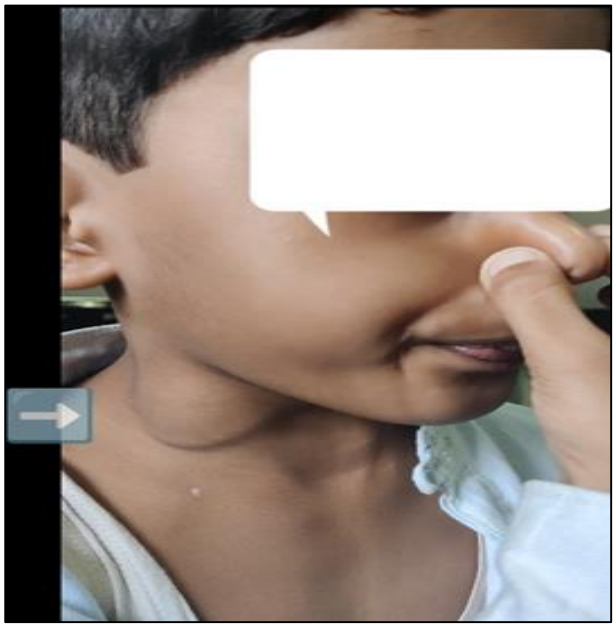

Figure 1: A 8-year-old boy with right neck swelling on valsalva maneuvere.

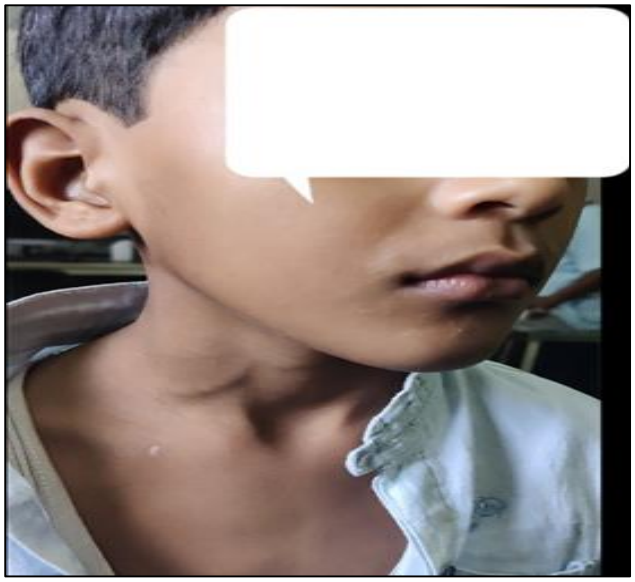

Figure 2: Clinical picture of a 8 year old boy.

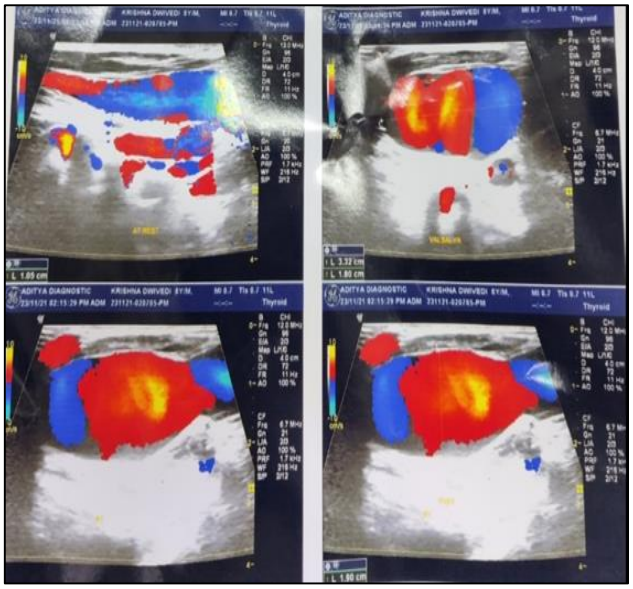

Figure 3: USG neck-showing right jugular venous phlebectasia with fusiform aneurysmal dilatation of internal jugular vein.

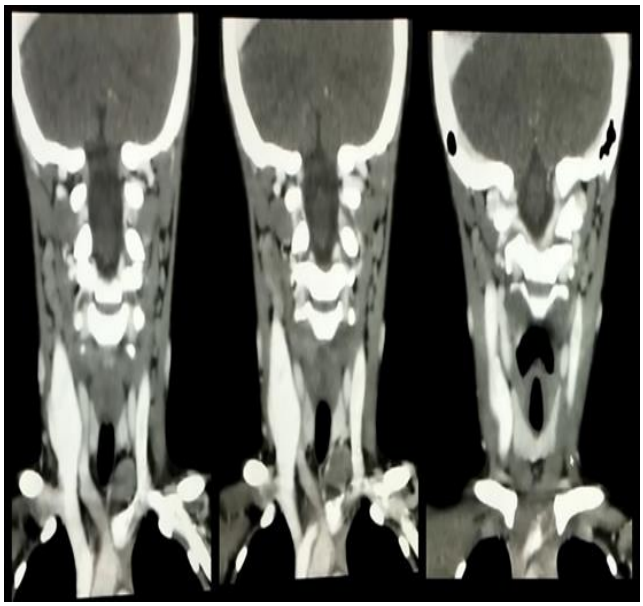

Figure 4: CECT neck showing right internal jugular vein short segment fusiform dilatation.

DISCUSSION

The term phlebectasia is different from the term varix where there is abnormal outward dilatation of the vein without tortuosity and was described first by Harris in 1928, and characterised by Gerwig in 1952 as a fusiform or saccular dilated segment of a vein. The neck veins including anterior and external jugular veins were commonly involved but there are only few cases reported with phlebectasias involving internal jugular vein which was first described by Zukschwerdt.[3,4] Right internal jugular vein more commonly involved as supported by

hypothesis given by Monte et al.[5] Males are commonly affected than females with ratio of 2:1. Only 100 cases were reported till now in the literature. The possible causes are gross anatomic abnormality, mechanical compression or trauma, congenital structural defects in the vein wall, or may be idiopathic. Most of the cases are symptomatic and brought only for cosmetic purposes. The mass usually is compressible, soft in consistency and appears when the intrathoracic pressure increases. The differential diagnosis of this swelling is more commonly confused with laryngocele, superior mediastinal mass, inflation of pulmonary apical bullae.[6] Laryngoscopy directly rules out the possibility of a laryngocele, and a thoracic computed tomography (CT) scan rules out the possibility of a mediastinal cyst or tumor. Ultrasonography during a valsalva manoeuvre easily establishes the diagnosis of jugular vein phlebectasia and this should be used as a first-line imaging test.[7] During a valsalva manoeuvre, the diameter of the affected vein may increase up to 2.2 times compared with its measurements at rest. Colour doppler imaging confirms the presence or absence of blood flow and thrombus formation in the lumen of the vein and is the gold standard for diagnosis of jugular vein phlebectasia. Most of the cases were managed conservatively with avoidance of local trauma and speech therapy and regular followup. Only approximately 20 cases have been managed by surgery with Polytetrafluoroethylene tube graft considering the complications like intramural thrombosis or cosmetic reasons.[8] Intra mural thrombosis occurs due to chronic high pressure in the dilated jugular vein.[9]

CONCLUSION

Phlebectasias are fusiform dilatation of the neck veins more commonly involving anterior and external jugular veins, but phlebectasias occurring in internal jugular vein is a rare entity in paediatric age group and oftenly misdiagnosed with other neck swellings such as laryngocele. Hence appropriate timing of diagnosis with non-invasive radiological techniques is necessary. Most of the patients are managed conservatively until some complications or deformities are coexisting.

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Ethical approval: Not required

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