

The louder the bigger: A case of jugular phlebectasia in a child

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Abstract

Jugular phlebectasia has been increasingly recognised with the advent of non-invasive diagnostic methods. Phlebectasia differs from varix, as it is an abnormal outward dilatation of a vein without tortuosity. It presents as a soft, compressible mass, apparent upon straining or execution of the Valsalva maneuver. The differentials for neck masses are broad, but if the swelling appears on the Valsalva maneuver, the type of mass narrows down to a laryngocele, superior mediastinal mass or phlebectasia. A simple non-invasive investigation, such as ultrasonography, is used as a diagnostic tool. We report a case of jugular phlebectasia that was suspected clinically and confirmed via ultrasound to be a vascular lesion which changed its size upon straining.

Introduction

Jugular phlebectasia (JP) is being increasingly recognized, especially with the advent of major laryngeal and cervical surgery and non-invasive diagnostic methods.¹ The term phlebectasia is used to describe an abnormal outward dilatation of a vein without tortuosity.² It can affect any neck vein, particularly the internal jugular, external jugular, anterior jugular and superficial communicus in decreasing order of frequency. The possible causes are a gross anatomic abnormality, mechanical compression, trauma and congenital structural defects in the vein wall, or it can be of idiopathic origin.³ Phlebectasia of the internal jugular vein (IJV) usually presents as an anterior neck mass, apparent upon straining. The differentials are laryngocele and superior mediastinal mass or cyst.⁴

Case Report

A 3-year-old child with thalassemia trait presented with a history of right neck swelling 2 years in duration. The amount of swelling was static and increased upon shouting, crying and straining. He did not complain of pain, facial congestion, difficulty in eating or breathing and voice changes. On physical examination, a soft, compressible swelling measuring 3 cm x 2 cm, which was apparent on shouting, was noted over the middle-third of the right sternocleidomastoid (SCM) muscle (**Figure 1**). It was non-tender upon

palpation, and there were no variations in the overlying skin and local temperature. There was no lymphadenopathy, and the swelling was not pulsatile. No bruit was heard. General examinations were unremarkable. The diagnosis of internal jugular phlebectasia was confirmed by an ultrasonography of his neck. A significant increment in the diameter of the right IJV, more marked on the Valsalva, was observed and measured as five times its original size at rest (**Figure 2**). The child is currently under observation, and the amount of swelling remained unchanged at the last follow-up 10 months later.

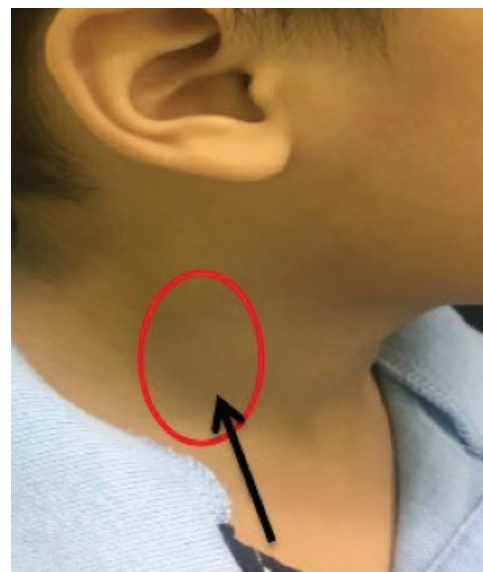


Figure 1: Swelling noted over right SCM during shouting.

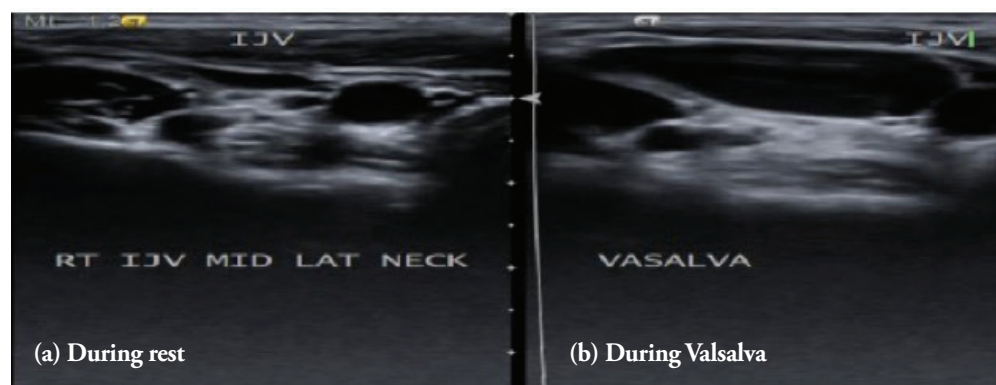


Figure 2: Ultrasound findings revealing a marked variation in size of the right internal jugular vein (IJV) (a) during rest and (b) during Valsalva maneuver.

Discussion

Jugular phlebectasia (JP) is a congenital fusiform dilatation of the jugular vein that appears as a soft, compressible mass in the neck and is triggered by the Valsalva maneuver or straining. Although it might affect the anterior, external and internal jugular veins, it affects the IJV in the majority of patients.³

JP in children occurs most commonly on the right side, as the right internal jugular bulb is at a higher level and larger than its counterpart on the left side. In addition, the right brachiocephalic vein is directly continuous with the superior vena cava (SVC), unlike the left brachiocephalic vein, which joins the SVC at an angle.¹

The possible causes of venous ectasia in the neck are congenital structural defects in the venous wall, mechanical trauma, or it can be of idiopathic origin.⁴ JP is a benign lesion which commonly affects boys during childhood at a rate twice that for girls. The appearance of a neck mass upon straining can be due to laryngocele, jugular phlebectasia, the hyperinflation of the cupula of the lung and neoplasms or cysts of the upper mediastinum. Simple procedures, such as a laryngoscopy and thoracic computed tomography, will rule out laryngocele and a mediastinal mass, respectively.⁴

The gold standard in diagnosing JP is ultrasonography (USG) or color Doppler flow imaging (CDFI), both non-invasive modalities. CDFI is helpful in determining the presence or absence of blood flow as well as its direction and thrombus formation.^{3,4} We used USG of neck in our case, as it is fast, safe, conducted easily, reliable and can be used to visualize the IJV with or without Valsalva

maneuver. In USG, the cross-sectional area (CSA), anteroposterior (AP) diameter and transverse diameter are usually measured at rest and during the Valsalva maneuver. The CSA is used for differentiating normal veins from instances of phlebectasia and has a specificity and sensitivity of approximately 90%, but the measurement of the AP diameter during the Valsalva maneuver is more practical and highly accepted by sonographers (specificity and sensitivity of approximately 85%). An AP diameter of more than 15 mm (mean value of the right and left sides) is sufficient for diagnosing phlebectasia in daily practice.⁵ In our patient, the diameter of the right IJV increased significantly during the Valsalva maneuver, measuring 20 mm or five times the original size at rest. Eksioğlu et al. concluded that the cut-off values for IJV measurements were same for all patients, as there were no significant correlations between USG measurements and the age, sex, height and weight of participants in their study.⁵

After the introduction of USG, surgical exploration, invasive contrasted radiography (xeroradiography, angiography) and fine needle aspiration and cytology (FNAC) were no longer needed to diagnose JP. Modalities such as digital subtraction angiography (DSA) and computer tomography (CT) are very sensitive in terms of demonstrating vascular lesions but are not preferred choices for investigation because patients are exposed to radiation. Nuclear scintigraphy detects the dilatation of the jugular vein from the origin to the SVC. However, it is more costly, and the procedure is time consuming.⁶ Invasive contrasted angiography and DSA run the risk of complications such as a vascular perforation, hematoma, pseudoaneurysm of brachiocephalic arteries and thoracic duct injury. Hence, these complications and adverse effects can be

avoided by using USG.⁶ However, USG has its limitations when the lesion has extended distally into the thoracic cavity or has an intracranial extension. Thus, in those instances, a CT or magnetic resonance imaging will be the modality of choice.

The treatment of JP varies according to the symptoms. It is either observe first (conservative) or treated surgically. Asymptomatic patients are treated conservatively with timely follow up, as per our case, when the swelling is static.⁷ Surgical treatments are suggested for patients with masses which are growing with or without cosmetics defects, thrombus formation, phlebitis, Horner syndrome and rupture of the lesion.^{8,9} Hence, we observe our patient first and only opt to conduct surgery if the mass is increasing in size and causing some of the symptoms mentioned above. Surgery will depend on the type of dilatation and consist of either excision or venorrhaphy.

Complications of surgery include injury to the any of these nerves; phrenic, vagus or brachial plexus, plus an air embolism. Excision is not advisable for bilateral lesions because it is associated with complications such as cerebral edema and massive facial edema.⁸ Spontaneous resolution, especially in children, has been reported.⁹

In conclusion, JP can be diagnosed easily with a physical examination, particularly when a prominent neck mass is apparent upon conducting the Valsalva maneuver or straining. An USG of the neck is sufficient to confirm the diagnosis, and treatment depends on the patient's symptoms and consists of either observation or surgical intervention.

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