

Internal Jugular Phlebectasia in Children: a Diagnostic Dilemma

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Citation

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Abstract

The most common cause of a neck mass that increases in size on straining is laryngocele. Internal jugular phlebectasia, which is of unknown cause, may present similarly. Jugular vein phlebectasia, a fusiform dilatation of a vein without tortuosity, is a rare cause of cervical neck swelling in children. It commonly presents as a soft cystic mass in the neck that transiently appears during straining. Dilatation of the internal jugular vein with a Valsalva maneuver suggests a mechanical obstruction in the neck or mediastinum, but the exact cause is still unknown. Because of their rarity, jugular vein phlebectasia cases have frequently been misdiagnosed or have been managed inappropriately. We present a case of a 10-year-old male with no history of trauma and an enlarging right neck mass. No treatment is necessary for this benign, self-limiting condition. Clinical features, diagnosis, treatment options, and a review of the literature about this rare venous lesion are presented.

INTRODUCTION

Jugular vein phlebectasia (JVP) is a congenital fusiform dilatation of the jugular vein that appears as a soft, compressible mass in the neck during straining or triggered by the Valsalva maneuver. It has been described in almost all cervical veins and is usually asymptomatic. Because of its rarity, this entity is frequently ignored or misdiagnosed [1]. The swelling is not known to progress rapidly and there have been no instances of spontaneous rupture of the swelling or other serious complications [2]. The possible differential diagnosis for the swelling could include a branchial cyst, cystic hygroma, laryngocele, cavernous haemangioma and superior mediastinal cysts [2]. The possible causes of JVP are as follows: gross anatomic abnormality of the vein, mechanical compression or trauma of the vein, congenital structural defects in the vein wall, and idiopathy [3]. Dilatation of the inner jugular vein with Valsalva maneuver suggests an obstructive mechanism in the neck or mediastinum, but it has an unknown cause and several hypotheses are being proposed. Most essays have found changes in the venous wall, with a decrease in its elasticity [4]. All neck veins can be affected, and the internal and external jugular, the previous jugular and the superficial communicating ones are more common. Doppler sonogram and CT are the more useful methods in the investigation, and venography, arteriography and gamma-ray CT can also be used. The treatment is conservative when there are no

complications or deformities. Spontaneous rupture was not reported until then. The only complications described were thrombosis and Horner's syndrome. Surgical treatment, usually with aesthetic purposes, consists of the excision of the dilated portion of the vein, or a unilateral excision of the vein, usually with no further complications.

CASE REPORT

A 10-year-old boy presented with a history of a painless swelling in the right neck that increased on straining since the age of 4 years. Clinical examination revealed a cystic fluctuant swelling of 2 x 2cm in the region of the left anterior triangle of the neck, which increased in size on performing a Valsalva maneuver [figure 1] It completely regressed on sitting up or standing and was totally compressible. There was no bruit or hum over the swelling, and it was not transilluminant.

Figure 1

Figure 1: shows swelling in left carotid triangle area of the neck on Valsalva maneuver.



An ultrasound scan confirmed ectasia of the internal jugular vein (IJV). Colour flow mapping revealed an abnormally dilated proximal IJV with expansion on crying [figures 2, 3]. No surgical treatment was advised and the child was well at the time of follow-up a year later.

Figure 2

Figure 2: Colour Doppler study shows IJV phlebectasia on rest

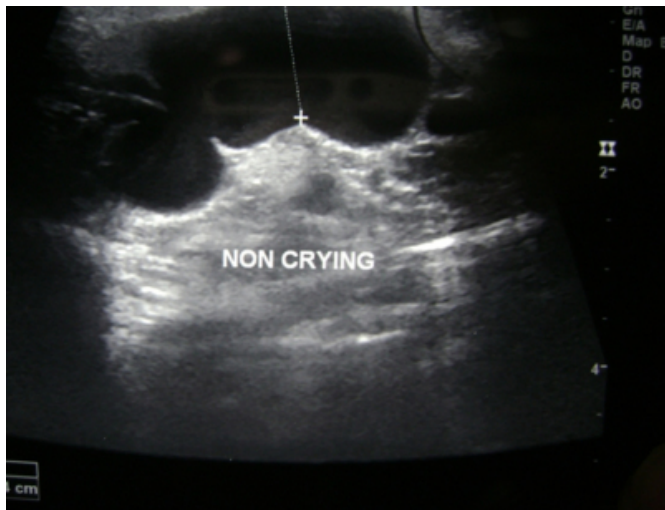
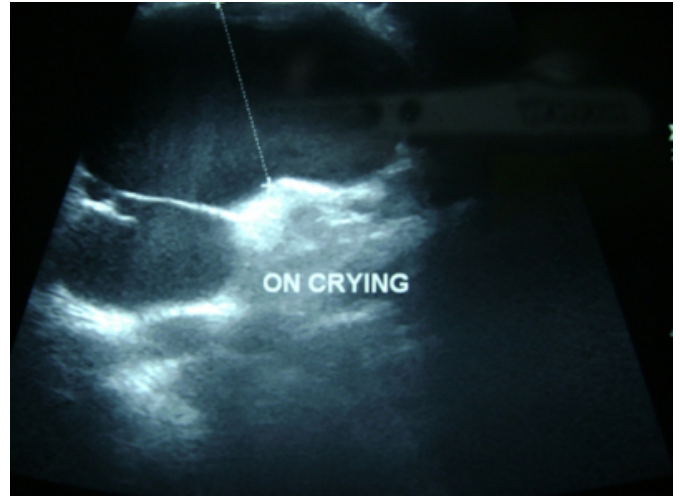


Figure 3

Figure 3: Colour Doppler study shows IJV phlebectasia on cry.



DISCUSSION

Internal jugular ectasia was first described by Zukschwerdt [3] and subsequently characterized by Gerwig [4]. A variety of etiologic hypotheses have been proposed since then; these include anomalous reduplication of the internal jugular vein [3], increased scalenus anticus muscle tone [5], congenital origin [6] and compression of the vein between the head of clavicle and the cupula of the right lung [7].

Phlebectasia of the jugular veins is a venous anomaly that usually presents in children as a soft cystic swelling in the neck during straining. The term phlebectasia indicates abnormal outward dilatation of the vein without tortuosity and differs from the term varix, which implies dilatation plus tortuosity [8]. Because there have been only sporadic reports of JVP, the exact etiology of this lesion still remains unclear. The possible causes of JVP are as follows: gross anatomic abnormality of the vein, mechanical compression or trauma of the vein, congenital structural defects in the vein wall, and idiopathy [9].

La Monte et al. [7] hypothesized that the ectasia is more common on the right side because the right innominate vein lies in contact with the right apical pleura. Hence, any increase in intrathoracic pressure could be directly communicated to the right IJV. The left vein, being placed more medially, was not subject to this stress. Diagnosis is often best made using ultrasound examination [6], with colour flow imaging also recommended to confirm vascular flow [10]. Other modalities used include computed tomography [11], venography [7], arteriography [7], nuclear scintigraphy [12] and xeroradiography [13].

Differential diagnoses of non-pulsatile soft neck masses enhanced by maneuvers that increase intrathoracic pressure include laryngocele or external laryngeal diverticulum, cupular inflation, tumours or cysts of the upper mediastinum and phlebectasias [11].

Unless complications occur or the lesions are cosmetically deforming, most authors recommend conservative management [710]. The only complications reported to date are thrombosis [11] and Horner's syndrome [14]. Spontaneous rupture has not been noted. Follow-up of patients treated conservatively ranges from 0.2 to 8.5 years with no complications.

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