Phlebectasia of the External Jugular Vein
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Citation

Abstract
Jugular phlebectasia, an entity increasingly recognised in recent years, is an isolated saccular or fusiform dilation of a vein without tortuosity. There is a controversy about etiology. This paper reports a case of localised distension of the external jugular vein in a 24-year-old male patient complaining of intermittent right neck swelling while lying down or straining (Valsalva manoeuvre). The diagnosis was confirmed by color Doppler ultrasonography, surgical excision was carried out without any complication.

INTRODUCTION
Jugular phlebectasia, a benign condition, refers to an isolated abnormal fusiform or saccular dilatation of the jugular vein of unknown etiology. Others terms have also been used in the literature including venous aneurysm, venous cyst, venous ectasia, aneurysmal varix and venectasia [1-2]. It usually present with a swelling in the posterior triangle of the neck, mostly on right side. Phlebectasia may affect any vein in the neck in the sequence of internal jugular, external jugular, anterior jugular and the superficial communicantes.

The case report intends to stress the importance of keeping in the mind the differential diagnosis of jugular phlebectasia, along with tumors or cysts of the upper mediastinum, external laryngeal diverticula or laryngoceles, in neck swelling which increases in size on coughing, sneezing and Valsalva manoeuvre. This is important in order to avoid invasive investigations which can lead to catastrophic results.

CASE REPORT
A 24-year-old male police constable was admitted in the University Hospital of Banaras Hindu University, India, with complaints of intermittent swelling of the right lower neck in the supraclavicular region on straining, of 6 months duration, that increased in volume on lying down, while it decreased in volume on sitting and standing, without any other complaint.

There was no history of change in voice, difficulty in breathing, swallowing, trauma, previous surgery, cough or facial congestion.

General and systemic examinations were unremarkable. Local examination of the right supraclavicular region showed a swelling of 2.5 x 1.5cm, oval, cystic, non-tender and localised just above the medial third of the right clavicle, anterolateral to the sternocleidomastoid muscle with normal overlying skin. The swelling increased in size on lying down and on Valsalva manoeuvre and decreased on sitting and standing.

There was no bruit. The left supraclavicular region was normal and the carotid pulse was normally palpable on both sides.

Color Doppler study revealed a collapsible cystic space in the course of the right external jugular vein with a lower end measuring approximately 22 x 10mm in maximum diameter in distended state. Color flow imaging showed low velocity pulsatile venous flow. No evidence of thrombus or abnormal wall thickening was noted. Cranially, the structure was continuous with the external jugular vein; caudally the termination of the external jugular vein was normal in caliber, behind the medial third of clavicle. Internal jugular and subclavian veins were normal on both sides. (Fig. 1a, 1b)
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Figure 1
Figure 1ab: Doppler ultrasonography of the phlebectatic external jugular vein

A diagnosis of focal ectatic dilatation of the lower part of the right external jugular vein (jugular phlebectasia) was made. Hematological and biochemical profiles were unremarkable. The patient was operated under general anaesthesia with transverse skin incision in the lower neck. Superficial saccular dilation of the right external jugular vein was identified (Fig. 2). This was carefully dissected both superiorly and inferiorly to look for any other feeding vessels or possible adhesions. After ruling out both these possibilities, the vein was double-ligated at both ends and excised. The post-operative period was uneventful. The histopathological report showed few capillary spaces stuffed with blood. The surrounding tissue was fibrous. A large vessel with flat to low cuboidal lining was seen.

Figure 2
Figure 2: The photograph shows dissection both superiorly and inferiorly of the external jugular vein and around the cystic swelling.

DISCUSSION
Gruber first reported a phlebectasia of the lower part of the internal jugular vein in 1875 [3]. Since then, more than 100 cases of phlebectasia involving the neck veins including anterior and external jugular veins have been reported in the world literature [1].

Jugular phlebectasia is usually a childhood disease. In older patients, as in our patient, it is very rare [1]. Because most of the lesions have been reported in children or have had an onset of illness that dated back to childhood, it seems quite possible that the cause is congenital. No proven acquired cause has been reported in the literature.

Because there have been only sporadic reports of venous ectasia in the neck, the exact cause of this lesion still remains in question [1,3]. Incidence is higher on the right side [1,3]. There are some hypotheses about this predominance in literature. The usual presentation is a lateral neck mass that increases in size with manoeuvres which increase intrathoracic pressure [1].

A few conditions which have the characteristic of appearing in the neck on straining, coughing, sneezing, bending, or on Valsalva manoeuvre are tumors or cysts of the upper mediastinum, external laryngeal diverticula or laryngoceles, venous enlargement of the jugular vein and inflation of the cupola of the lung [7,10]. Absence of a wide mediastinum or air in the mass on simple chest films eliminates a mediastinal tumor or laryngocele, respectively.

A number of diagnostic techniques have been recommended in the literature, including direct needle aspiration, venography, arteriography, computed tomography, ultrasonography, and color Doppler flow imaging [1]. Venography is a choice for diagnosis. However, this invasive technique is potentially dangerous, with complications such as hematoma, perforation of the lesion, thoracic duct injury and pseudoaneurysms [1]. Ultrasonography with Doppler before, during and after Valsalva manoeuvre is the preferred method for diagnosis [1].

Histopathologic studies also are distinct in the literature. Most specimens submitted for histological examination have shown no abnormality apart from dilatation and thinning of the walls. Others have shown loss of the elastic layer and hypertrophy of connective tissue with focal intimal thickening [2,5,6].

The operative procedure is quite simple but the surgeon must be on the look out for associated venules or other feeding vessels and deal with them initially, before securing the main dilated vessel.
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There is no controversy about the indication of surgical treatment in patients with symptomatic unilateral jugular phlebectasia [1]. Surgery is recommended even in asymptomatic cases, because of the tendency of the lesion to increase in size over time and because of probable emotional trauma. Most authors recommend no treatment for this benign condition [1-10], but conservative follow-up is not described exactly in the literature.

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References

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