

JUGULAR PHELEBECTASIA

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ABSTRACT: Phlebectasia is a term describing an isolated saccular or fusiform dilatation of a vein without tortuosity. Jugular phlebectasia is an entity that is being increasingly recognized in recent years. There is a controversy about etiology. Histopatologic studies also are distinct in literature. This paper reports a 15-year-old girl complained of intermitant non-painful right neck swelling while speaking loud or straining. The patient was followed for two years uneventfully. After two years of follow-up the patient began to complain about cosmetic problems. Surgery was offered and the patient rejected surgery.

INTRODUCTION

Phlebectasia is a term describing an isolated saccular or fusiform dilatation of a vein (1). Other terms have also been used in the literature including venous aneurysm, venous cyst, venous ectasia, aneurysmal varix and venectasia (1,2). It should not be confused with even a tortouse vein or varix, or diffuse genuine phlebectasia (1,3).

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 (2). Because there have been only sporadic reports of venous ectasia in the neck, the exact cause of this lesion still remains in question (1,2). Incidence is higher on the right side (1,2). There are some hypothesis about this predominance in literature. The usual presentation is a lateral neck mass that increases in size with manoeuvres, which increase intrathoracic pressure (4).

DISCUSSION

Jugular phlebectasia is an entity that is being increasingly recognized in recent years. There is a controversy about etiology. A variety of etiologic hypothesis have been proposed; these include anomalous reduplication of the internal jugular vein, increased scalenous anticus muscle tone, compression of the vein between the head of the clavicle and the cupoula of the right lung, superior mediastinal irradiation, trauma and, congenital origin (2). Jugular phlebectasia is usually a childhood disease. In older patients, as in our patient, its very rare (5). Because most of the lessions 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.

REPORT OF CASE

A 15-year-old girl complained of intermitant non-painful right neck swelling while speaking loud or straining. There was no history of change in voice, difficulty in

breathing or swallowing, trauma or previous surgery. Physical examination showed right neck swelling during valsalva manoeuvre (Figure 1). The mass was anteromedial to the sternocleidomastoid muscle. Radiographs of the neck and chest were normal. Computed tomography showed a 3x2,5 cm right internal jugular phlebectasia (Figure-2). The diagnosis

was confirmed with Doppler ultrasonography (Figure-3). The cardiovascular examination was normal. The patient was followed for two years uneventfully. After two years of follow-up the patient began to complain about cosmetic problems. Surgery for excision of phlebectasia and grafting of the vein was offered and the patient rejected surgery.



Figure 1. Photograph of patient showing the bulging of the neck mass during valsalva manoeuvre

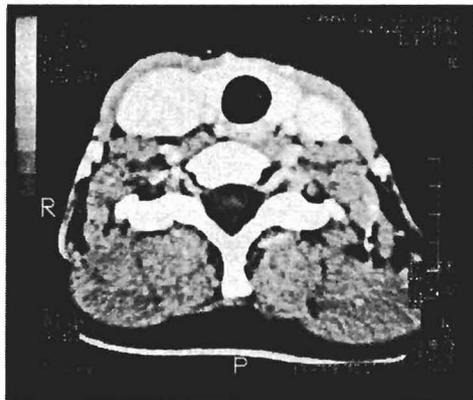


Figure 2 Axial CT image of the patient showing phlebectasia of the right internal jugular vein

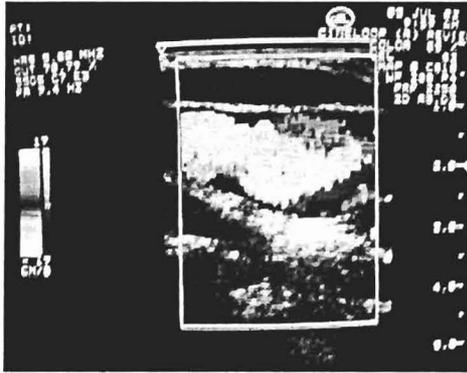


Figure 3. Doppler ultrasonography of the phlebectasic internal jugular vein

Histopathologic studies also are distinct in literature. Most specimens submitted for histological examination have shown no abnormality apart from the dilatation and thinning of the walls. Others have shown loss of the elastic layer and the hypertrophy of the connective tissue with focal intimal thickening (1,2,6,7). Manometric studies demonstrated that the increase of the intrathoracic pressure does not produce uneven pressure increase in the right internal jugular vein (6).

Internal jugular phlebectasia usually presents as a soft, round or fusiform, cyst like mass with a smooth contour that does not involve skin. It is mainly located in the lower third of the neck at the anterior border of sternocleidomastoid muscle. Typical clinical presentation is neck swelling that increases in size with straining, valsalva manoeuvre, coughing, bending, sneezing or after exertion. Other symptoms are cessation of voice (3), painful swelling (7), and slight dispnea (8), venous hum or bruit (1), progressive enlargement (1).

The possible complications of this condition, which are extremely rare, include thrombosis, secondary pharyngitis, and congestive heart failure and massive hemorrhage secondary to trauma (9).

In differential diagnosis, there are four conditions which have the characteristic of appearing in the neck on straining, coughing,

sneezing, bending, or the valsalva manoeuvre. These are tumors or cysts of the upper mediastinum, external laryngeal diverticula or laryngoceles, venous enlargement of the internal jugular vein, inflation of the cupola of the lung (7,10). Besides the four conditions which have already described, cavernous hemangioma, cystic hygroma, thyroglossal duct cyst, dermoid cyst, bronchogenic cyst, cervical adenitis and metastatic adenopathy should also be considered in differential diagnosis (7,8).

A number of diagnostic techniques have been recommended in the literature including direct needle aspiration, venography, arteriography, computed tomography, ultrasonography, and collar Doppler flow imaging (3). Venography is a choice for diagnosis. However this invasive technique is potentially dangerous, leading complications such as hematoma, perforation of the lesion, thoracic duct injury and pseudoaneurysms (8). Ultrasonography with doppler before, during and after valsalva manoeuvre is the preferred method for diagnosis (11).

Although it may appear clinically unilateral, internal jugular phlebectasia can occur bilaterally, so ultrasonography should be performed on both sides of the neck (12). In our case we used computed tomography in diagnosis, then cofirmed the diagnosis with doppler ultrasonography.

Incidence is higher on the right side (2). The right IJV (Internal Jugular Vein) valves are placed at a higher level than the left sided ones and right inferior jugular bulb is therefore larger than the left. The valves play an important role in preventing retrograde blood flow. The right IJV is larger than the left in most people. The right brachiocephalic vein is in direct continuity with the superior vena cava unlike the left brachiocephalic vein that joins the superior vena cava at the angle. The right brachiocephalic vein is also shorter than the left one. Valves almost never seen in the right brachiocephalic vein, the incidence of the valves in the left brachiocephalic vein varies from 4 - 8%, most being competent. Competent valves are present in 99% of right subclavian veins, only 97% of left sided subclavian veins have valves. Any increase in intrathoracic pressure is easily and consistently transmitted to the right jugular bulb by the presence of the above anatomical variations (2).

There is no controversy about the indication of the surgical treatment in patients with symptomatic unilateral jugular phlebectasia (3). Surgery is recommended even in asymptomatic cases, because of the tendency of the lesion to increase in size over time and probable emotional trauma. Most authors recommend no treatment for this benign condition (1,5,9,10), but the conservative follow-up is not described exactly in the literature.

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