Focal Ectasia of Internal Jugular Vein

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Introduction

Internal jugular vein ectasia is a venous anomaly commonly presenting as a unilateral neck swelling in children and adults. This condition is being increasingly recognized in recent years due to better imaging facilities [1]. There is a controversy about its etiology because there have been only sporadic reports of venous ectasia in the neck. The exact cause of this lesion still remains in question. Due to the rarity of this condition, a high index of suspicion is required once the other common causes are excluded. We present a case of focal ectasia of the internal jugular vein and discuss its management.

Case Report

A 11 year old boy, presented with complaints of intermittent swelling on the right side of the neck, noticed initially at age of five years. The swelling was insidious in onset, gradually progressive with prominence of the swelling seen only on speaking loud, crying, straining or performing valsalva manoeuvre. There was no history of change in voice, difficulty in breathing or swallowing, trauma or previous surgery. Physical examination revealed normal neck at rest but on valsalva a 8x4 cm, fusiform swelling appeared on the right side of the neck anteromedial to the right sternocleidomastoid muscle. The swelling was soft, compressible, non tender, non pulsatile, not translucent and increased in size on valsalva manoeuvre (Fig. 1). The venous pressure waves were absent. Fiberoptic laryngoscopy revealed normal larynx and hypopharynx. Radiographs of the neck and chest were normal. An ultrasonography (USG) neck showed prominence of the internal jugular vein (IJV) on the right side. The swelling increased to size 6x4 cm on valsalva. The diagnosis was confirmed with doppler ultrasonography. Contrast enhanced computed tomography (CECT) neck showed a focal ectasia of the right internal jugular vein extending from clavicle to hyoid, during valsalva the length of segment was 6 cm with diameter of 4 cm. The diameter of IJV above and below the swelling was 1cm (Fig.2). The cardiovascular examination was normal.

The patient did not have any features of complications

like hoarseness of voice, stridor, dysphagia or other pressure symptoms hence parents were given reassurance and explained about the disease. Possibility of IJV ligation was entertained but it carried a risk of cerebral oedema in small subset of patients. This risk was not acceptable due to benign nature of disease presenting without any complications. No surgical treatment was offered. The patient is presently on regular follow up for the past one year without any change in size of swelling and continues to be asymptomatic.

Discussion

Ectasia of the internal jugular vein is an isolated saccular or fusiform dilatation of the vein [2]. Other terms used in literature to describe this condition include, phlebectasia, venous aneurysm, venous cyst, aneurysmal varix and venectasia [2,3]. Gruber first reported a phlebectasia of the lower part of the internal jugular vein in 1875 [4]. A variety of etiologic hypothesis have been proposed, which include anomalous reduplication of the internal jugular vein, increased scalenus anticus muscle tone, compression of the vein between the head of the clavicle and the cupula of the right lung, superior mediastinal irradiation, trauma and congenital origin [3]. Ectasia of the internal jugular vein is usually a childhood disease. No proven acquired cause has been reported in the literature. An association of this condition with certain genetic disorders of connective tissue like Menkes disease, an inherited disorder of copper transport has been suggested.

The clinical presentation is usually 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 a neck swelling that increases in size with straining, valsalva manoeuvre, coughing, bending, sneezing or after exertion. Other symptoms include cessation of voice, painful swelling, progressive enlargement, slight dyspnoea and venous hum or bruit. Incidence is higher

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Fig. 1 : Clinical presentation

Fig. 2 : Contrast enhanced computed tomography scan

on the right side [3]. The right 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. Valves are almost never seen in the right brachiocephalic vein, but 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 [3]. The complications of this condition are extremely rare. They are thrombosis, congestive heart failure and massive haemorrhage secondary to trauma [5].

In the differential diagnosis, neck swelling which have the characteristic of appearing in the neck on the valsalva manoeuvre must be considered. These include external laryngeal diverticula or laryngoceles, branchial cyst, tumours or cysts of the upper mediastinum and inflation of the cupula of the lung [6]. Besides these, other conditions like cavernous haemangioma, cystic hygroma, thyroglossal duct cyst, dermoid cyst, bronchogenic cyst, cervical adenitis and metastatic adenopathy should also be considered in the differential diagnosis. A number of diagnostic techniques have been recommended in the literature including venography, arteriography, computed tomography, ultrasonography, and colour doppler flow

imaging. USG with doppler before, during and after valsalva manoeuvre is the preferred method for diagnosis. Although it may appear clinically unilateral, internal jugular phlebectasia can occur bilaterally, so ultrasonography should be performed on both sides of the neck [7]. Histopathological studies have shown loss of the elastic layer and the hypertrophy of the connective tissue with focal intimal thickening [8].

Most authors do not recommend surgery for this benign condition and conservative follow up is required [2,5,6]. However if patient is symptomatic or when complications are present then resection of the dilated segment or covering with muscular segment is recommended [9]. The whole affected vein can be sheathed in an 8 mm poly tetra fluoro ethylene (PTFE) tube graft from the site of its emergence to the point where it disappears behind the right sternoclavicular joint.

Conflicts of Interest

None identified

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Answers to MCQ

1) a	2) b	3) c	4) c	5) c
6) a	7) d	8) c	9) a	10) a
11) b	12) c	13) b	14) a	