CLINICAL REPORT

An Intriguing Case of Ectasia of External Jugular Vein Presenting as Intermittent Neck Mass

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Abstract Ectasia of external jugular vein is a rare entity presenting as an intermittent neck swelling. High index of suspicion is required to diagnose the condition. Ultrasonography with Doppler before, during and after valsalva manoeuvre is the preferred method for diagnosis. The treatment is conservative for asymptomatic patients and surgery is reserved for patients with complications.

Keywords Ectasia · External jugular vein · Phlebectasia

Introduction

Intermittent masses in neck appear during coughing, crying, straining, valsalva manoeuvre when there is increased intrathoracic pressure [1]. The differential diagnosis for a neck mass that increases in size with valsalva manoeuvre includes laryngoceles, external laryngeal diverticulae, tumors or cysts of superior mediastinum and inflation of cupula of lung. Ectasia of external jugular vein (EJV) is a rare condition in which there is abnormal sacculofusiform dilatation of the vein. The etiology is not known. We present a rare case of ectasia of EJV and its management.

Case Report

A 22 year old male presented with a right sided neck swelling since the age of 5 years. The swelling was insidious in onset, gradually progressive in size, intermittent in nature. It used to become prominent on coughing, straining and performing valsalva manoeuvre. There was no history of voice change, difficulty in swallowing or difficulty in breathing. There was no history of any trauma to the neck or any neck surgeries. No history of any medical illness (Figs. 1, 2).

Physical examination revealed no abnormality at rest. On valsalva two swellings were seen on the right side of neck. The upper one behind the angle of mandible measuring 2×2 cm and the lower one lateral to sternomastoid extending till supraclavicular fossa measuring 4×5 cm. The swellings were compressible, soft and non-tender. Skin over the swellings was normal. No pulsations were seen over the swellings. Videolaryngoscopy revealed normal larynx and hypopharynx. Examination of other systems was normal. Mediastinal mass was ruled out after Chest X-ray which showed no widening and laryngocele excluded when no air was observed at region of mass.

Fig. 1 Before valsalva

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Fig. 2 During valsalva

Fig. 3 MR venography

Fig. 4 MRV—phlebectatic segment

The diagnosis was confirmed with Ultrasonography with Doppler. MR Angiography of neck revealed segmental aneurysmal dilatation of right EJV in neck and its confluence with subclavian vein (supraclavicular fossa) suggestive of phlebectasia of EJV. The dilated segment in upper neck measured $17.5 \times 24 \times 37$ mm and the one in supraclavicular fossa was $41 \times 37 \times 36$ mm (Figs. 3, 4).

Discussion

Plebectasia as a word describes an abnormal, fusiform dilatation of vein and is different from the terms “varix” and “varicose veins” which simply imply tortuosity [3]. Intrinsic vascular malformations originating from the wall

of EJV are very rare. It may be possibly be underreported due to its benign nature [2]. The sites of occurrence of ectasia in the decreasing order of frequency are Internal, External, Anterior jugular and superficial communicans [1]. The etiology is unknown. Histologically it shows loss of elastic layer, hypertrophy of connective tissue and focal intimal thickening.

It presents as a soft, cystic, painless mass in the neck which increases in size during straining and completely disappears when patient is relaxed [1]. The differential diagnosis include laryngoceles, tumors or cysts of superior mediastinum, laryngeal diverticula and venous enlargement of superior vena caval system. Colour Doppler at rest and during Valsalva manoeuvre is the investigation of choice. It is an accurate, non-invasive imaging technique to distinguish the jugular venous enlargement. It defines the extent of the lesion and its relationship with the surrounding structures in the neck.

Most authors do not recommend surgery for this benign self-limiting condition and conservative follow-up is advised [4–6]. However for cosmetic reasons or if complications like hoarseness, stridor, dysphagia occur, surgery is advised. The standard procedure includes resection of the dilated segment after ligation of feeding vessels.

In our case, as initially the patient was asymptomatic he was advised regular follow-up. After about 9 months patient noticed increase in size of swelling associated with pain. So the patient was referred to a higher centre for further management. The patient underwent excision of the phlebectatic segment.

Conclusion

Ectasia of the external jugular vein is a rare and benign condition. It can be diagnosed by having a strong suspicion in cases presenting with intermittent neck mass. Evaluation can be done with non-invasive measures like Ultrasonography with colour Doppler and MR venography. Colour Doppler at rest and during Valsalva manoeuvre is the investigation of choice. Conservative approach is recommended in asymptomatic patients and surgery is reserved for patients with complications.

Compliance with Ethical Standards

Conflict of interest Dr. B. Ranganatha Reddy, Dr. Rakesh Vuppala, Dr. Supriya Mohan Bhat declare that they have no conflict of interest.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

Informed Consent Informed consent was obtained from the individual participated in the study. Additional informed consent was obtained from the individual for whom identifying information is included in this article.

References

1. Abdulla SA, Aldabagh MH (2008) Congenital phlebectasia of the internal jugular vein. *Dohuk Med J* 2:155–159
2. Sander S, Eliùevik M, Unal M et al (1999) Jugular phlebectasia in children: is it rare or ignored? *J Pediatr Surg* 34:1829–1832
3. Gerwig WH (1952) Internal jugular phlebectasia. *Ann Surg* 135:130–133
4. Bowdler DA, Singh SD (1986) Internal jugular phlebectasia. *Int J Pediatr Otorhinolaryngol* 12:165–171
5. Indudaran R, Quah BS, Shuaib IL (1999) Internal jugular phlebectasia unusual cause of neck swelling. *Ann Trop Pediatr* 19:105–108
6. Nwako FA, Aguagara NE, Udeh CA, Osuorji RI (1989) Jugular phlebectasia. *J Pediatr Surg* 24:303–305