Unilateral internal jugular vein phlebectasia in an adult: Management and one year follow-up

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Abstract
Phlebectasia describes an anomalous, fusiform dilatation of a vein. In the neck region, the internal and external jugular veins are mostly affected. To our knowledge, this is the first case in Saudi Arabia of internal jugular phlebectasia affecting an adult female. We describe a 61-year-old female with complaints of a neck swelling she noticed 4 years ago. Initially, the swelling increased in size and reached a stable level. It was asymptomatic and only enlarged during Valsalva maneuver. Flexible nasolaryngoscopy and computerized tomography scan showed unremarkable examination. Follow-up after 1 year with US Doppler showed no progression. Internal jugular phlebectasia is a rare disorder which is often diagnosed during childhood. More often than not, it does not cause any significant morbidity. Since it is a benign condition, observation is advised with regular monitoring. For asymptomatic lesions, surgical intervention is recommended if cosmetic or psychologic concerns are present.

Keywords
Phlebectasia, internal jugular vein, head and neck, otolaryngology, vascular anomalies

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Introduction
Phlebectasia is an aberrant, fusiform dilatation of a vein.1 It can affect any neck vein, mainly, internal and external jugular and, less commonly, anterior jugular and superficial communicus veins.2 Two-thirds of the cases arise from the internal jugular vein.3 To our knowledge, this is the first case in Saudi Arabia of internal jugular phlebectasia affecting an adult female.

Case presentation
A 61-year-old female presented with complaints of a neck swelling she noticed 4 years ago. Initially, the swelling increased in size and reached a stable level. The patient also reported that the swelling enlarged with coughing and straining. She denied stridor, dysphagia, dysphonia, neck pain, and having other neck masses. On examination, there was a mass located in the right lower region of the patient’s neck with no overlying skin changes. The mass increased in size with Valsalva (Figure 1). On palpation, the mass was compressible with transmitted pulse. Auscultation showed no bruit. Flexible nasolaryngoscopy showed unremarkable examination. Computerized tomography (CT) scan of head and neck with contrast was done (Figures 2–4). No treatment was indicated. Thus, the patient was scheduled for follow-up and observation. On patient follow-up after 1 year with US Doppler, the internal jugular vein was patent within size compared to previous CT image and no thrombus formation (Figure 5).

Discussion
Internal jugular phlebectasia is a rare disorder which can be either unilateral or bilateral, and it is diagnosed often during
childhood. Thus, adulthood onset of internal jugular phlebectasia when compared to childhood onset is extremely rare. A couple of cases were reported to have internal jugular phlebectasia presenting in adulthood; however, the neck swelling was evident since birth for both cases. Due to the rarity of this condition, no likely etiology has been found. However, it is hypothesized that a primary lack of elasticity of the venous wall can be a reason explaining mainly congenital cases. In addition, elevated pressure in the internal jugular vein is another hypothesis that can lead to phlebectasia. Trauma was the etiology in one case who presented with an internal jugular phlebectasia following a motorcycle traffic accident with a compound fracture of the right zygomatic bone. Unlike our patient, in adults, internal jugular vein phlebectasia is most common on the left side of the neck, while in children, it was commonly found on the right side. It typically presents with a benign swelling over the lateral side of the neck on the affected side, seen on effort. More often than not, it does not cause any significant morbidity which was the presentation of our patient. The chief complaint of patients is mainly due to cosmetic concerns. Other infrequent presentations reported were associated with pain and chest heaviness.

Figure 1. Right-sided neck swelling during Valsalva maneuver.

Figure 2. Sagittal contrast-enhanced CT image of the neck showing a well-defined large saccular internal jugular vein.

Figure 3. Contrast-enhanced coronal CT image is showing the extent of the dilatation of the right internal jugular vein.

Figure 4. Contrast-enhanced axial CT image showing dilated right internal jugular vein with heterogeneous contrast filling.
Dysphonia seemed to be the presenting complaint in patients having malignancies combined with internal jugular phlebectasia. Generally speaking, symptomatic presentation or developing complications such as Horner’s syndrome and thrombosis is rare. Valsalva maneuver is the most valuable clinical sign used to diagnose the dilatation of internal jugular vein. Internal jugular phlebectasia is first noticed on physical examination, subsequently color Doppler imaging confirms the diagnosis and is said to be the gold standard. Furthermore, CT scans are utilized to rule out any causative structural lesions. The differential diagnosis of a progressive swelling in the neck on straining includes tumors or cysts of the upper mediastinum, external laryngeal diverticula or laryngoceles, inflation of the apex of the lung, as well as cystic hygromas. Since internal jugular phlebectasia is a benign condition, conservative observation is advised. Surgical intervention for internal jugular phlebectasia is generally reserved for symptomatic patients together with the asymptomatic who present with cosmetic or psychologic concerns. The conventional surgical approaches include ligation of the internal jugular vein as well as longitudinal constriction suture venoplasty and encapsulation or partial resection of the phlebectasia.

**Conclusion**

Internal jugular phlebectasia presenting in adults is a very rare phenomenon as it is a disorder often seen during childhood. It typically presents with a benign swelling over the lateral side of the neck, exacerbated by Valsalva maneuver. The main complaint is predominantly cosmetic concerns. The gold standard for diagnosis is color Doppler imaging which can be aided by CT to rule out any causative structural lesions. Because internal jugular phlebectasia is a benign condition, conservative observation is advised. Surgical intervention for internal jugular phlebectasia is commonly reserved for symptomatic patients together with the asymptomatic who present with cosmetic or psychologic concerns. The conventional surgical approaches include ligation of the internal jugular vein as well as longitudinal constriction suture venoplasty and encapsulation or partial resection of the phlebectasia.

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