Case Report

A rare case of waxing and waning supraclavicular swelling due to internal jugular vein saccular varix in a 79-year-old male with chronic obstructive pulmonary disease

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ABSTRACT

Supraclavicular swellings encompass a broad range of infectious, neoplastic, and vascular etiologies. A 79-year-old male with a history of chronic smoking and chronic obstructive pulmonary disease presented with a right-sided neck mass that had progressed from the size of a pea to a lemon over 3 months. The swelling exhibited a unique "waxing and waning" pattern, appearing prominently during coughing and Valsalva maneuvers and resolving completely afterward. Clinical examination revealed a 3 × 3 cm soft, non-pulsatile cystic mass. Ultrasound and computed tomography angiography confirmed a focal saccular aneurysmal dilatation of the right internal jugular vein, measuring 3.2 cm in diameter. In the absence of complications, such as thrombosis or compressive symptoms, conservative management and regular follow-up are preferred over surgical intervention. This case emphasizes the importance of considering rare vascular anomalies in the differential diagnosis of dynamic neck masses.

Key words: Chronic obstructive pulmonary disease, Internal jugular vein, Neck mass, venous dilatation, Phlebectasia, Saccular varix, Supraclavicular swelling

Supraclavicular swellings can arise from various etiologies, including infectious, inflammatory, neoplastic, and vascular causes [1]. The dynamic nature of the swelling in this case, appearing during coughing and Valsalva maneuvers and resolving afterward, suggests a vascular origin.

This report discusses a unique presentation of internal jugular vein (IJV) phlebectasia in a 79-year-old male with chronic obstructive pulmonary disease (COPD), highlighting the importance of considering vascular anomalies in the differential diagnosis of dynamic neck swellings.

CASE REPORT

A 79-year-old male with a history of chronic smoking and COPD presented to the Ear, Nose, and Throat outpatient department with a swelling on the right side of his neck persisting for 3 months (Fig. 1a). The swelling, initially the size of a pea, progressively enlarged to the size of a lemon. It was characterized by a waxing and waning pattern, appearing during coughing and Valsalva maneuvers and resolving afterward (Fig. 1b). The patient also reported a persistent cough and shortness of breath, managed conservatively by the pulmonary medicine team.

On examination, a 3 × 3 cm soft, non-pulsatile, ill-defined cystic swelling was palpable over the right supraclavicular region. The swelling became evident during coughing and Valsalva maneuvers and disappeared afterward. The overlying skin was free and pinchable, with no signs of erythema or warmth.

Ultrasound of the neck revealed a dilated right IJV measuring 3 cm in maximum caliber, suggestive of phlebectasia. Computed tomography (CT) angiography showed a focal saccular aneurysmal dilatation of the right IJV in the supraclavicular region, measuring 3.2 cm in largest diameter and involving a 3 cm length of the vessel, consistent with an IJV phlebectasia (Fig. 2). High-resolution CT thorax revealed bilateral hyperinflated lung fields with paraseptal and centriacinar emphysematous changes suggestive of COPD (Fig. 3).

Given the absence of compressive symptoms, such as dysphagia, dyspnea, or voice changes, and no signs of thrombosis, the patient was reassured and managed

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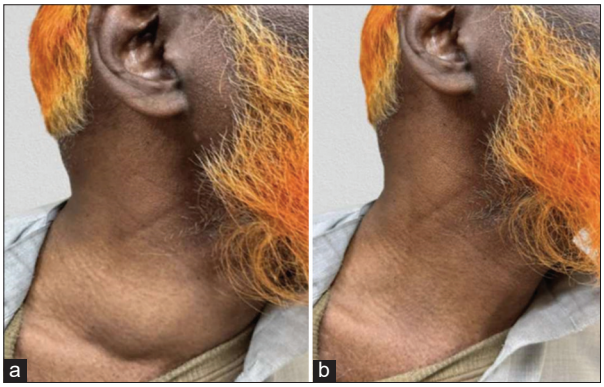

Figure 1: (a) Clinical picture of patient's right lateral neck with swelling on Valsalva (b) Clinical picture of patient's right lateral neck after swelling disappears

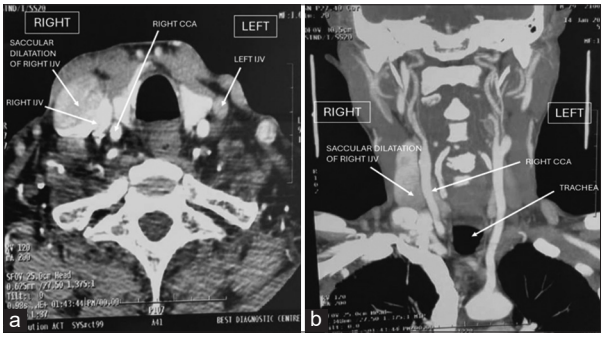

Figure 2: (a) Computed tomography (CT) angiography image showing the saccular dilatation of the right internal jugular vein (axial view) (b) CT angiography image showing the saccular dilatation of the right internal jugular vein (coronal view)

conservatively for COPD. No intervention was deemed necessary for the neck swelling. The patient was followed up after 6 months with an ultrasound of the neck. There were no new complaints by the patient. There was no interval increase in the size of the lesion clinically and radiologically.

DISCUSSION

Neck swellings are one of the most common presenting complaints in patients encountered by otorhinolaryngologists, and hence being aware of the many differentials is of utmost importance. Apart from the rare entities, such as phlebectasia, various commonly encountered differentials, such as thyroglossal cyst, dermoid cyst, branchial cyst, cystic hygroma, hemangioma, cervical lymphadenitis, laryngeal diverticula, persistent jugular lymphatic sac, and thyroid mass should be ruled out for appropriate management [1].

IJV phlebectasia (also known as saccular varix, congenital venous cyst, venous aneurysm, and venous ectasia) refers to an abnormal saccular or fusiform dilatation of part or all the layers of the IJV [2]. This term was first coined by Zukschwerdt L and later described by Gerwig [3]. IJV phlebectasias are rare benign vascular anomalies. They present as soft, compressible, cystic, dynamic neck swellings, especially during activities that increase intrathoracic pressure, such as coughing or Valsalva maneuvers. Most cases of jugular varix are seen

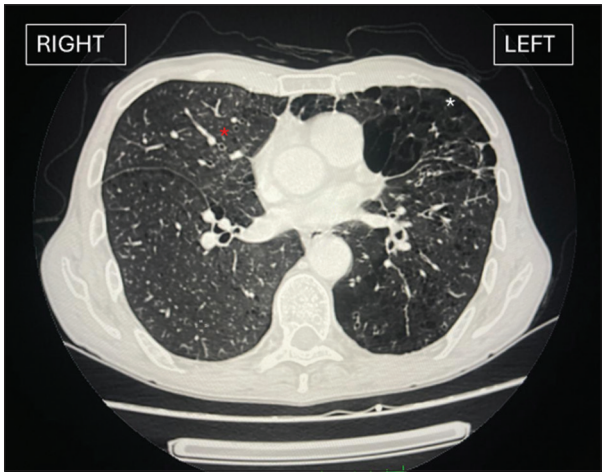

Figure 3: High-resolution computed tomography thorax image showing bilateral hyperinflated lung fields with paraseptal (white asterisk) and centriacinar (red asterisk) emphysematous changes

in children and rarely in adults. No proven acquired cause has been described for the occurrence of this anomaly, although various theories, such as radiation, increased tone of the scalene muscles and compression of the jugular vein between the lung apex and clavicle have been proposed [4]. There is a right-sided predominance for IJV phlebectasia, the right-to-left ratio being 4:1 [5]. This is hypothesized to be due to the anatomical differences between the right and left IJVs being the reason for increased transmission of intrathoracic pressure to the right IJV compared to its counterpart [6,7].

This is the first documented case of an IJV phlebectasia, potentially secondary to the persistent cough and elevated intrathoracic pressure associated with COPD, correlating with the hypothesis proposed by Paleri and Gopalakrishnan [7].

Pathophysiology includes factors, such as increased intrathoracic pressure, as in COPD, especially with chronic coughing and emphysema, which leads to persistently elevated intrathoracic pressure. Repeated Valsalva maneuvers during coughing episodes can transiently impede venous return and increase pressure in the venous system, including the IJV. Venous valve incompetence due to chronic strain can lead to valve insufficiency in the IJV. This results in retrograde flow and progressive venous dilation, manifesting as phlebectasia. Furthermore, contributing is the structural weakness due to long-term venous hypertension, which may cause thinning or loss of elastic fibers and smooth muscle in the venous wall.

The jugular vein, especially on the right side (more commonly affected), becomes distended and can form a visible neck swelling that enlarges with Valsalva or coughing. Associated features in COPD Patients are seen like hyperinflated lungs that can mechanically compress adjacent structures and further disrupt normal venous return. Polycythaemia (secondary to hypoxia) increases blood viscosity, compounding venous congestion. The Valsalva test can accentuate the swelling, hence helps in diagnosis.

Although surgical excision and histopathological examination were not done in the present case, examination

on various histopathological studies has shown focal intimal thickening, hypertrophy of the connective tissue, and loss of the elastic and muscular layer [8].

Ultrasonography with color Doppler is the most common, safe, sensitive, feasible, non-invasive, and low-cost tool used for the diagnosis of venous varix. Other non-invasive routinely used radiological investigations include Contrast-Enhanced CT, CT angiography, and magnetic resonance Venography [9]. After diagnosis, the patient should be kept on follow-up with a multidisciplinary approach, since even though rare, the associated complications include thrombosis, pulmonary embolism, congestive heart failure, massive hemorrhage secondary to trauma or spontaneous rupture, and thrombophlebitis.

There are no definitive guidelines for the treatment of IJV phlebectasias. Conservative management is generally preferred, especially in pediatric cases, due to the absence of complications, such as thrombosis or compressive symptoms. Surgical treatment is considered in severe cases with symptoms or cosmetic concerns. Surgical treatment options include excision of the involved dilated segment with primary anastomosis, longitudinal constriction suture venoplasty, partial resection of the phlebectasia, and scaffolding of the ectatic segment with an 8 mm polytetrafluoroethylene tube graft [10-12]. Alternative procedures, such as angioplasty and endoscopic resection are used only in children. Outcomes are generally good across all treatments, but surgery carries a higher complication rate (6.9% in children, 11.4% in adults), while conservative treatment has not reported any complications, making it a safer option overall [5]. The absence of thrombosis and compressive symptoms in this case supports conservative management.

CONCLUSION

This case underscores the importance of considering vascular anomalies, such as IJV phlebectasia, in the differential diagnosis of dynamic supraclavicular swellings. A thorough clinical and imaging evaluation can lead to an accurate diagnosis and guide management decisions, often favoring conservative treatment in the absence of complications.

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