Now you see it, now you do not!! - a case report and review of literature of bilateral jugular venous phlebectasia

Marykutty Francis[1], Pearl Mary Varughese[2*]

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ABSTRACT

Background: Jugular vein phlebectasia is a rare entity seen in children and is often an incidental finding presenting as a soft, cystic painless neck swelling that becomes prominent on coughing or straining and disappears on rest. It is usually managed conservatively.

Case Presentation: We report a case of an 8-year-old boy who visited us on an outpatient basis with a history of cough with neck swelling. Doppler ultrasound helped clinch the diagnosis of bilateral jugular venous phlebectasia.

Conclusion: It is paramount that clinicians consider this differential of jugular phlebectasia for neck swellings to avoid unnecessary investigations.

Keywords: Phlebectasia, laryngocele, case report, ultrasound.

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Correspondence to: Pearl Mary Varughese
*Consultant Pediatrician and Neonatologist, Devamatha Hospital, Kochi, India.
Email: pearlmaryvar@gmail.com
Full list of author information is available at the end of the article.

Background

Neck swellings in children are often a matter of diagnostic dilemma for pediatricians due to the broad differential diagnosis, the most common swellings being cystic hygroma, laryngocele, and cervical lymph nodes. The differentials are narrowed down when the neck mass enlarges during a Valsalva manoeuvre, mainly a tumor or cyst arising from the superior mediastinum, branchial cleft cyst, jugular vein phlebectasia (JVP), or laryngocele [1]. JVP, also known as venous aneurysm or venous dilatation, is a non-tortuous dilatation of the jugular vein [2]. Even though many singular cases have been reported worldwide, it is still a poorly understood and recognized condition.

Color-Doppler ultrasonography is a simple and valid diagnostic method to distinguish this pathology [3]. Treatment is usually conservative, with surgery reserved for cosmetic reasons and the risk of complications. We present a case of bilateral neck swelling in an 8-year-old boy who visited our outpatient department and was diagnosed with phlebectasia and is now under regular follow-up. Parental consent was obtained for the publication of this report and images.

Case Presentation

An 8-year-old boy was brought by his parents with complaints of cough for a week and mild fever. His parents also noticed a small swelling on his neck that appears on cough and disappears when he stops coughing or at rest. He did not complain of breathing or swallowing problems. There is no past history of trauma to the neck or any infections affecting the cervical region. His developmental milestones were appropriate for his age.

On general examination, there was no pallor, icterus, cyanosis, or significant lymphadenopathy. His BMI was 16.15 kg/m^2. He appeared healthy and at rest, there was no obvious swelling noticed. However, on coughing and while performing the Valsalva manoeuvre, a 3 × 2 cm ovoid soft, compressible swelling appeared in the anterior lower two-thirds of the sternocleidomastoid muscle on the right side, above the clavicle as shown in Figure 1. On his left side, there was a uniform dilatation seen alongside the lateral side of the sternocleidomastoid muscle as shown in Figure 2. There were no local signs of inflammation, no visible pulsations, and transillumination of the swelling was negative. The upper respiratory tract examination was unremarkable.

A chest X-ray was taken which showed perihilar infiltrates in the right lung (Figure 3). There was no widening of the mediastinum or lucency in the mass thus ruling out the possibility of a mediastinal tumor or laryngocele, respectively. The absence of pulsations and bruits ruled out aneurysm, arterio-venous malformation, and vascular tumor. We started the patient on oral antibiotics due to the lower respiratory tract infection.

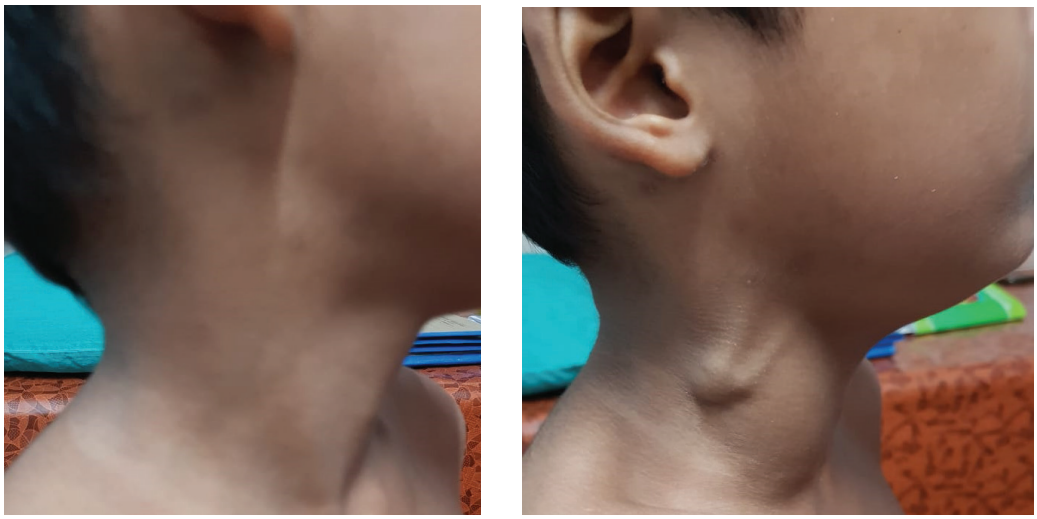

Figure 1. *Right side neck image (a) at rest and (b) during Valsalva maneuver.*

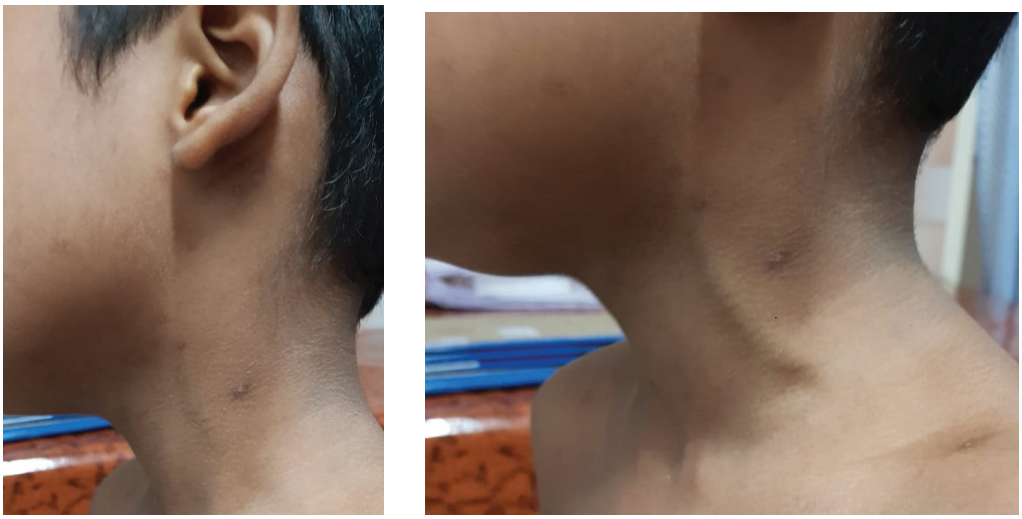

Figure 2. *Left side neck image (a) at rest and (b) during Valsalva maneuver.*

Doppler ultrasound demonstrated bilateral carotid arteries and vertebral arteries were normal. The right internal jugular vein (IJV) measured 9.6 mm at rest and 2.3 cm during the Valsalva manoeuvre at the level of the thyroid. The left IJV measures 11 mm at rest and 2.6 cm during the Valsalva manoeuvre at the level of the thyroid (Figures 4 and 5). Both IJVs are patent and showed no evidence of thrombosis (Figure 6).

As the child had no complications at present, no active intervention was done. At 2 weeks follow-up, the child's symptoms of cough had improved, he was symptomatically better and the swellings were of the same size. Parents were advised to follow up regularly and to review immediately if there was any change in the size of swelling or any new symptoms.

Discussion

The internal jugular phlebectasia was first published by Zukschwerdt, in 1929, and the anomaly was further characterized by Gerwig [4]. In 2019, a systematic review published included 97 articles that reported around 247 internal jugular phlebectasia cases in pediatric and adult patients [2]. Out of these, there were only 18 cases in the pediatric population that showed bilateral presentation [2] and we have collected the details of a few of them in Table 1.

Phlebectasia is often confused with aneurysm and varices. In aneurysm, there is a uniform dilatation of the whole vein, while in varices tortuosity is seen [5]. Some associated changes that have been described in adults are voice alterations, vocal cord paresis, and/or dysphagia, which may be attributed to the proximity of the vagus nerve and other lower cranial nerves to the IJV [1,6]. Eksioglu et al. [7] suggested using an anteroposterior diameter >15 mm as a cut-off for both sides for the diagnosis of internal jugular phlebectasia.

In the review by Figueroa-Sanchez et al. [2], the right-to-left prevalence ratio was 4:1. Many hypotheses have been proposed for the right-sided predilection, although none have been proven. LaMonte

hypothesised that due to the anatomical proximity of the right innominate vein with the right apical pleura, increased intrathoracic pressure due to cough or sneezing would be transmitted to the right IJV predisposing to a preponderance of right IJV phlebectasia [14]. Paleri and Gopalakrishnan [15] put forth their hypothesis based on anatomical factors such as the cephalad position of the right IJV valves, the larger diameter of the right IJV when compared with the left side; the direct continuity of the superior vena cava with the right brachiocephalic vein; and the higher number of valves in the left brachiocephalic vein when compared to the right side [15].

The natural progression of internal JVP has not been established, but studies that have been reported have documented either stable size or slow progression of the lesion [16-18]. Surgery was recommended for cosmetic purposes and for other risk factors like an increase in size [2]. The ideal duration of follow-up and management has not been described so far. Studies describing surgical intervention reveal a complication rate of 6.9% in the pediatric population and 11.4% in the adult population [2]. The surgical complications include injury to adjacent vessels such as the carotid artery, or nerves such as the tenth, eleventh, or twelfth cranial nerve or the phrenic nerve, air embolism or venous thrombus formation [19]. The consensus is that conservative treatment is a safer option than surgical treatment. In cases of bilateral JVP, surgical excision would be hazardous, with a high risk of cerebral edema and its consequences [10,20].

Colour-Doppler ultrasonography is a non-invasive accurate and readily reproducible imaging technique to distinguish jugular venous enlargement. It is important to perform these investigations bilaterally and comparatively, both at rest and on strain [21]. It can also help to rule out other differential diagnoses of neck masses, such as laryngocele, mediastinal tumors, and branchial cysts. Ultrasound is the first line of investigation (72%) in a recently reported systematic review, although CT may also be used for diagnosis, is less preferred due to radiation exposure [2]. CT scan is also sufficient in identifying the initial pathology, despite being utilized much less frequently.

This case is unique from a clinical standpoint because of the rarity and the bilateral presentation.

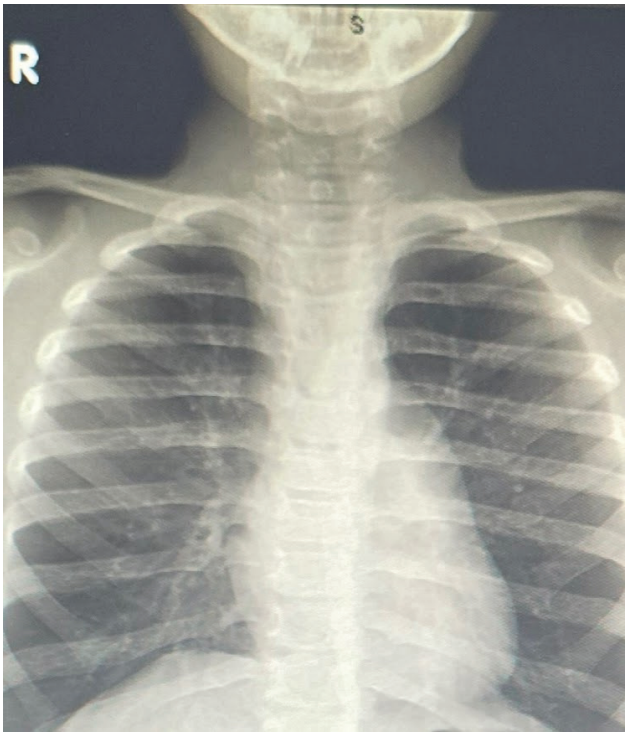

Figure 3. *Chest X-ray showing right peri-hilar infiltrates.*

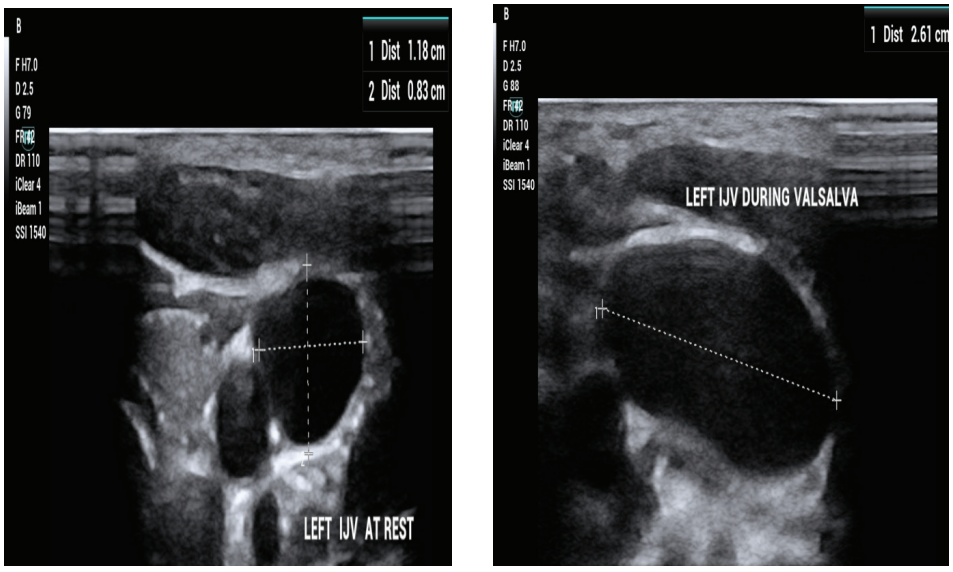

Figure 4. *(a) Left IJV at rest (b) during Valsalva maneuver.*

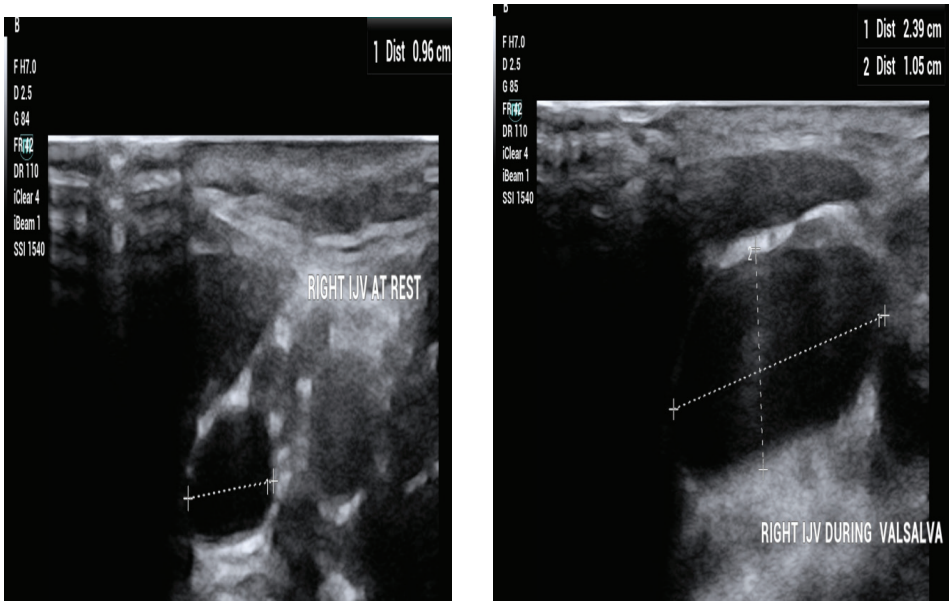

Figure 5. *(a) Right IJV at rest (b) during Valsalva maneuver.*

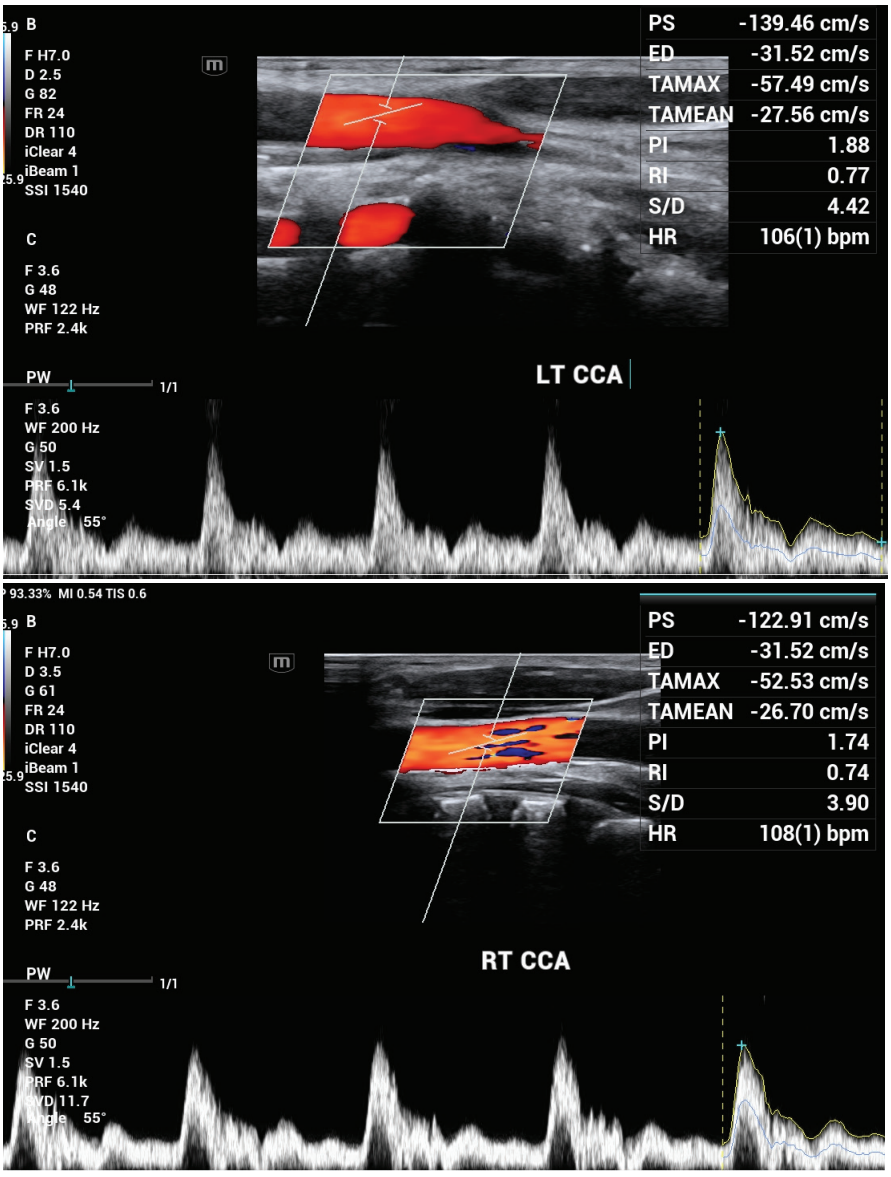

Figure 6. *Doppler images show no turbulence.*

Table 1. *Case reports of bilateral jugular phlebectasia.*

	STUDY	TREATMENT GIVEN	INVESTIGATIONS DONE	FOLLOW UP AND PROGRESS
1	Aydoğan [8]	Conservative	Color Doppler	At 2 years of follow-up, no change was observed.
2	Ferreira and Haguette Case A [9]	Conservative	Ultrasound	NA
3	Ferreira and Haguette Case B [9]	Conservative	Radiography with contrast	NA
4	Walsh et al. [10]	Conservative	Color Doppler	18 months later on follow-up, no change observed.
5	Leung et al. [11]	Conservative	Ultrasound	NA
6	Gendeh et al. [12]	Conservative	Ultrasound	At 2 years of follow-up, no change was observed.
7	Bonnet et al. Case A [13]	Conservative	Ultrasound with radiography and contrast	Venous thrombosis. Died from Respiratory complications of Menkes disease
8	Bonnet et al. Case b [13]	Conservative	Ultrasound	Death during admission for respiratory distress

Conclusion

Jugular phlebectasia should be considered in the differential diagnosis of Valsalva-dependent neck swellings in pediatric populations. Timely diagnosis can unnecessary invasive procedures and alleviate parental fear and anxiety. The condition is usually self-limited and follow-up is recommended for monitoring of changes or complications.

What is new?

JVP is a rare entity seen in children and is often an incidental finding It is usually managed conservatively. Doppler ultrasound is used to confirm the diagnosis.

List of Abbreviations

IJV Internal jugular vein
JVP Jugular vein phlebectasia

Conflict of interests

The authors declare that there is no conflict of interest regarding the publication of this article.

Funding

None.

Consent for publication

Written informed consent was obtained from the parents.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

Author details

Marykutty Francis[1], Pearl Mary Varughese[2]

1. Consultant Pediatrician, Devamatha Hospital, Kochi, India
2. Consultant Pediatrician and Neonatologist, Devamatha Hospital, Kochi, India

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Summary of the case

1	**Patient (gender, age)**	8 years, male
2	**Final diagnosis**	Bilateral jugular phlebectasia
3	**Symptoms**	Neck swellings bilaterally that increase in size on valsalva maneuver
4	**Medications**	None
5	**Clinical procedure**	None
6	**Specialty**	Pediatrics