International Journal of Contemporary Pediatrics
Harsoor AR et al. Int J Contemp Pediatr. 2025 Sep;12(9):1574-1575
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20252614

Case Report

A case of paroxysmal neck swelling

Aishwarya Rani Harsoor*, P. Muralikrishnan

Department of Pediatrics, Meenakshi Medical College and Research Institute, Kanchipuram, Tamil Nadu, India

Received: 12 July 2025
Accepted: 07 August 2025

***Correspondence:**
Dr. Aishwarya Rani Harsoor,
E-mail: raniharsoor@gmail.com

ABSTRACT

Internal jugular vein ectasia is a rare congenital condition that often presents with neck swelling during straining in children. We reported a 5-year-old female with paroxysmal neck swelling noticed during activities like coughing or shouting. Imaging confirmed transient internal jugular vein ectasia. The patient was managed conservatively. Conservative management is preferred in asymptomatic cases, with surgical intervention reserved for complications.

Keywords: Internal jugular vein, Ectasia, Phlebectasia, Pediatric neck swelling, Valsalva, Conservative management

INTRODUCTION

Ectasia of the internal jugular vein (IJV) is a rare congenital disorder. It most commonly presents as unilateral or bilateral neck swelling that increases during strenuous activities like coughing and defecation.[1] The etiology remains obscure. Our case is reported in view of its rarity and to highlight the management strategies.

CASE REPORT

A 5-year-old female child, otherwise normal, was brought by her mother to the pediatric OPD with complaints of diffuse swelling over the right side of the neck, noticed during straining activities like coughing and shouting. The swelling disappeared immediately at rest. It was insidious in onset and gradually progressive over the last 3 months.

There was no history of birth trauma, fall, fever, cough, weight loss, difficulty in swallowing, breathlessness, or swelling at other body sites. Vital signs were stable. Physical examination revealed a normal neck at rest, but on Valsalva maneuver, a swelling of approximately 6×4 cm was noted anteromedial to the right sternocleidomastoid muscle. It was soft, cystic, non-pulsatile, and non-tender. Chest and neck X-rays were normal. Ultrasound of the neck revealed transient enlargement or ballooning of the right internal jugular vein during increased intrathoracic pressure (patient shouting), returning to normal size immediately, with an impression of transient IJV ectasia on the right side.

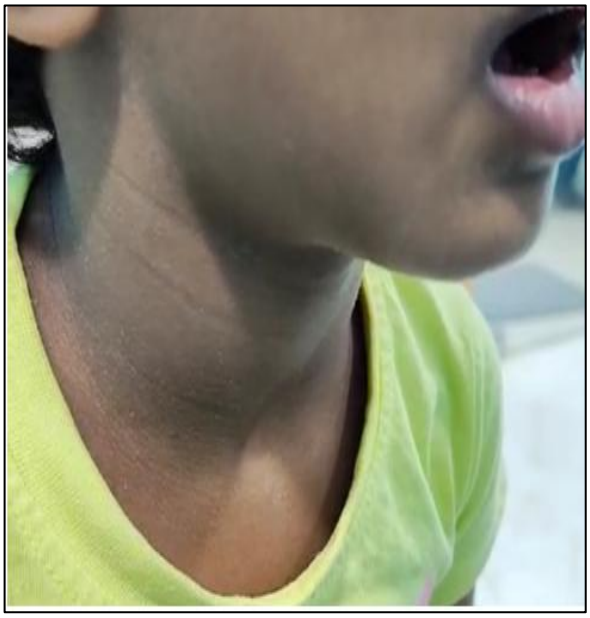

Figure 1: Normal neck without any swelling.

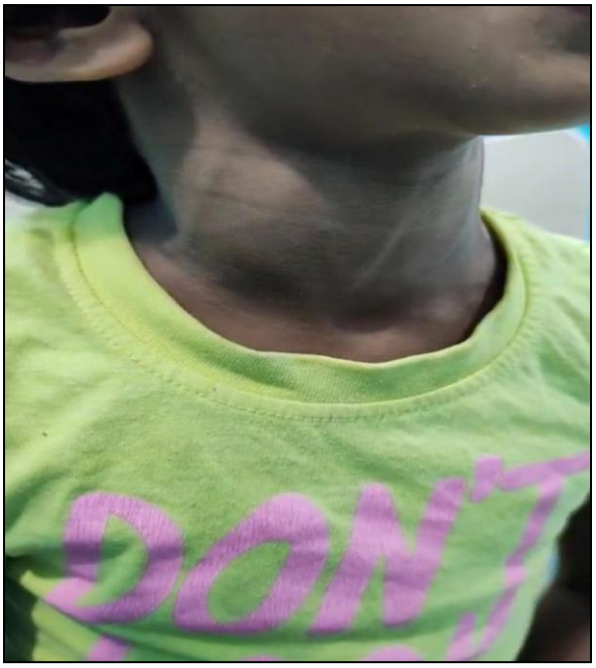

Figure 2: Neck swelling during shouting.

The patient had no complications such as hoarseness of voice, stridor, or dysphagia, and was managed conservatively with reassurance. No surgical treatment was offered. The patient has been on regular follow-up for one year, with no change in the size of the swelling and remains asymptomatic. Surgical options like IJV ligation or resection may be considered if pressure symptoms or complications develop, although this carries a risk of stroke.[2]

DISCUSSION

Ectasia of the IJV is an isolated saccular or fusiform dilatation of the vein. It is also referred to as phlebectasia, venous aneurysm, venous cyst, aneurysmal varix, or venectasia.[3] Various etiologic hypotheses have been proposed, including anomalous reduplication of the IJV, increased scalenus anticus muscle tone, compression between the clavicle and the lung apex, superior mediastinal irradiation, trauma, and congenital origin.[4] IJV ectasia is usually a childhood condition, with no proven acquired causes in the literature. A possible genetic association has been described with Menkes disease.[5]

Clinically, it presents as a soft, non-tender neck swelling that becomes prominent during Valsalva or straining maneuvers due to increased intrathoracic pressure. This pressure causes retrograde flow through the brachiocephalic vein, transmitted to the IJV.[6] Differential diagnoses include laryngocele, branchial cyst, cystic hygroma, cavernous hemangioma, and superior mediastinal cyst.[7] Most patients are asymptomatic and do not develop complications. However, hoarseness of voice, dysphagia, dyspnea, and rarely stroke can occur.[8] Management is typically conservative for uncomplicated cases, with surgical resection or ligation considered in symptomatic or complicated presentations.[9]

CONCLUSION

Internal jugular vein ectasia is an uncommon but benign condition, most often identified in childhood as a soft, intermittent neck swelling that appears during straining. Diagnosis can be established with non-invasive imaging such as ultrasound. Since most cases remain asymptomatic and carry a low risk of complications, conservative management with reassurance and regular follow-up is usually sufficient. Surgical intervention should be reserved for patients who develop pressure symptoms, cosmetic concerns, or complications. Awareness of this rare entity is important to avoid unnecessary investigations and to differentiate it from other neck swellings.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Harsoor AR, Muralikrishnan P. A case of paroxysmal neck swelling. Int J Contemp Pediatr 2025;12:1574-5.