PHLEBECTASIA OF THE INTERNAL JUGULAR VEIN- CONSERVATIVE TREATMENT: CASE REPORTS

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Abstract: introduction: Phlebectasia of the internal jugular vein is a rare clinical entity characterized by an isolated fusiform or saccular dilatation of the internal jugular vein, without torsion. Clinically, phlebectasia manifests as a cystic soft, painless, partially compressible non-pulsatile mass in the neck, which increases with Valsalva maneuver, during exertion, and spontaneously decreases during rest. To show that conservative treatment is the best modality for the treatment of phlebectasia in children. Results: The first patient presented is a 6-year-old boy who was diagnosed with phlebectasia of the right internal jugular vein by ultrasonography during Valsalva maneuver. The diagnosis was confirmed by magnetic resonance imaging of the neck vessels with contrast. The second patient is also a 10-year-old boy with an identical clinical course, so adequate diagnostics were performed, and the diagnosis of phlebectasia of the internal jugular vein on the right side was confirmed. The third case presented is a 5-year-old girl with a clinical presentation of phlebectasia of the right internal jugular vein, but in her case, radiological evaluation was postponed due to the decision of the parents. All patients presented with phlebectasia of the internal jugular vein were asymptomatic, so after appropriate diagnostics, conservative treatment and periodic monitoring in outpatient conditions were recommended. Conclusion: The decision on the modality of treatment of phlebectasia of the internal jugular vein in asymptomatic and some symptomatic patients involves conservative treatment and regular monitoring.
Key words: treatmant, ectasia, children.

Introduction

Phlebectasia of the internal jugular vein is a rare clinical entity characterized by isolated fusiform or saccular dilatation of the internal jugular vein without torsion (1). Clinically, phlebectasia of the internal jugular vein manifests as a cystic, soft, painless, partially compressible, nonpulsatile mass in the anterior triangle of the neck supraclavicularly anterior to the anterior border of the sternocleidomastoid muscle, which increases with the Valsalva maneuver, during exertion, coughing, crying, sneezing, and spontaneously decreases during rest (2,3).

It can occur in almost any cervicofacial vein, but most often affects the internal jugular vein (4). More often on the right side, in boys in the pediatric population (5). The differential diagnosis of neck swelling that increases with Valsalva maneuver includes laryngocele or external laryngeal diverticulum, jugular phlebectasia, brachial cyst, tumor and cyst of the upper mediastinum, and inflation of the apical bulla of the lung (6).

The diagnostic modality of first choice is ultrasound examination of the soft tissues of the neck at rest, and during the Valsalva maneuver. Magnetic resonance imaging with contrast angiography and venography of the main blood vessels of the neck is reserved for the definitive diagnosis of phlebectasia, especially in pediatric patients (7).

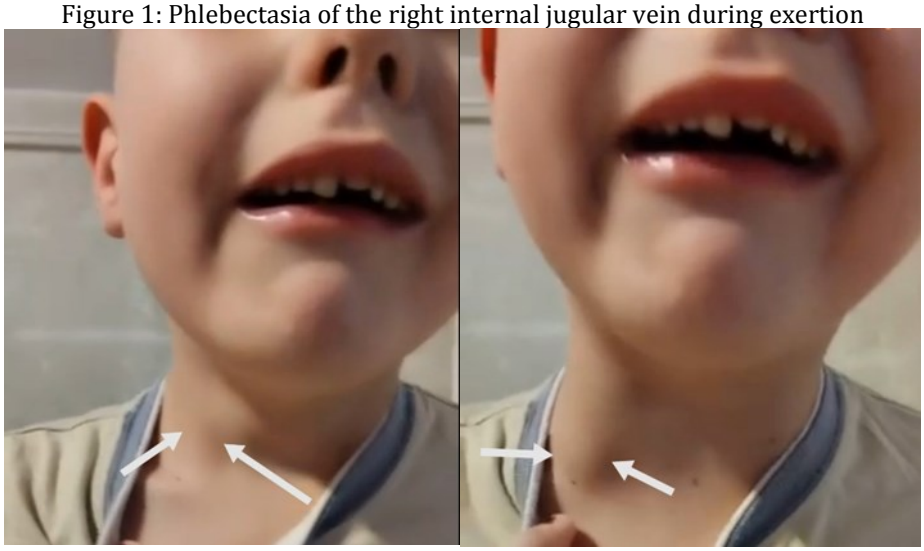
Surgical treatment is reserved for patients with complications, or for cosmetic reasons (8). The decision on the modality of treatment of phlebectasia of the internal jugular vein in asymptomatic and some symptomatic patients involves conservative treatment..

Case report 1:

A 6-year-old boy. He first presented for examination due to swelling on the right side of the neck, the change appears and increases with exertion, coughing or crying for the past month. The mother describes the appearance of a round, soft, painless change in the lower third of the right side of the neck that she noticed when the child cried. Initially, the patient was examined by a pediatrician and an otolaryngologist who, after conducting diagnostics, suspected phlebectasia of the

jugular veins. The patient had no associated diseases that could be one of the causes of phlebectasia.
Clinical examination did not reveal any change in the neck at rest, but during the Valsalva maneuver, an oval change of about 3.5x 5 cm in size appeared in front of the anterior edge of the sternocleidomastoid muscle in the lower third on the right supraclavicular side, painless on palpation, partially compressible, soft consistency, non-pulsatile, the skin above the change unchanged (Figure 1).

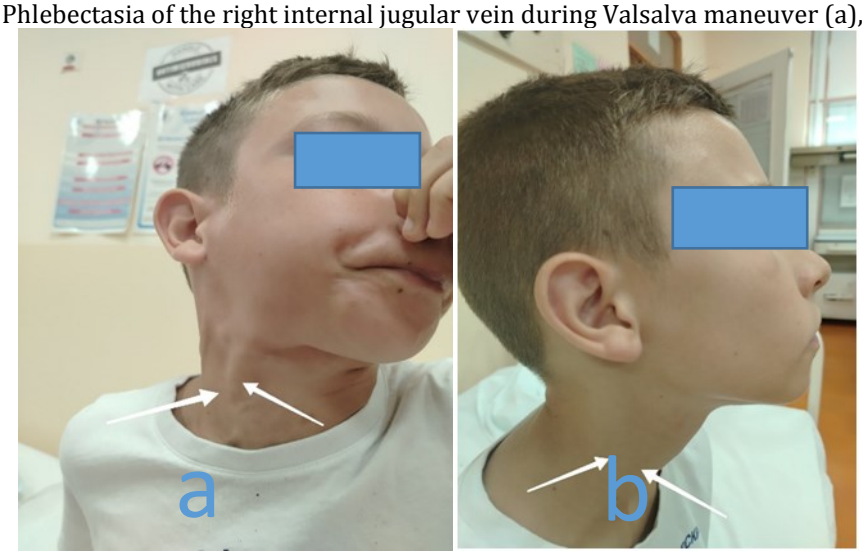
Figure 1: Phlebectasia of the right internal jugular vein during exertion



Ultrasound examination of the soft tissues of the neck at rest and during the Valsalva maneuver shows a dilated right internal jugular vein, without thrombotic masses, without tortuosity. At rest, the diameter is 12 mm, during the Valsalva maneuver it is 34 mm. The right brachiocephalic trunk is dilated to 7.5 mm. The left internal jugular vein is up to 7 mm in diameter with regular characteristics. In hospital conditions, magnetic resonance imaging of the main blood vessels of the neck with contrast was performed, which showed an ectatic right internal jugular vein predominantly in the distal part with a diameter of up to 15 mm, and a left internal jugular vein with a diameter of up to 7 mm. The jugular veins are transient, without signs of thrombosis. Due to the benign course and the absence of complications, the patient was suggested conservative treatment and a normal lifestyle. Home monitoring and periodic outpatient check-ups were suggested.

Case report 2:
A 10-year-old boy. He first presented for examination due to a tumor lesion on the outer side of the right neck, the lesion appearing and increasing predominantly during exertion for the past six months. The mother describes the appearance of a round, soft, painless lesion in the lower third of the right side of the neck that she noticed when the child was straining. The patient had no associated diseases that could be one of the causes of phlebectasia. He denies any other significant complaints (Figure 2a)

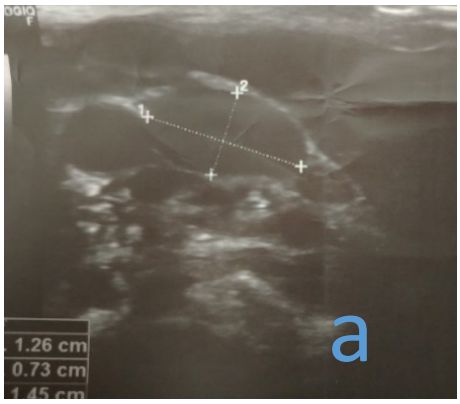
Figure 2: Phlebectasia of the right internal jugular vein during Valsalva maneuver (a), at rest (b)

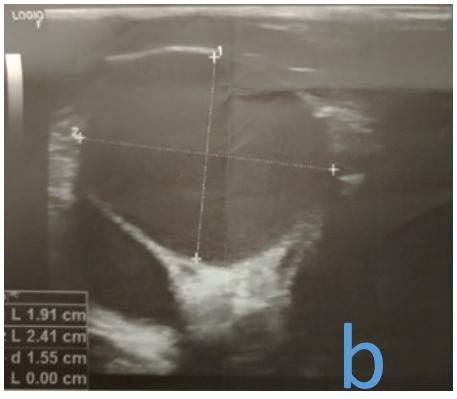


Clinical examination at rest does not reveal any changes in the neck, but during the Valsalva maneuver, an oval lesion measuring approximately 3 x 4 cm appears in front of the anterior border of the sternocleidomastoid muscle in the lower third of the right supraclavicular side, painless on palpation, partially compressible, soft consistency, non-pulsatile, the skin over the lesion unchanged (Figure 2b).

asound examination of the soft tissues of the neck at rest and during the Valsalva maneuver shows a dilated right internal jugular vein, with preserved hemodynamic flow. During the Valsalva maneuver, the maximum lumen width is 24.1 x 19.1 mm (US Figure 3a), and at rest, the lumen width is 12.6 x 7.3 mm (US Figure 3b).

Figure 3: (UZ) Ultrasonography of the main blood vessels of the neck (v. jugularis interna dex.). Cross-section during Valsalva maneuver (a), at rest (b)

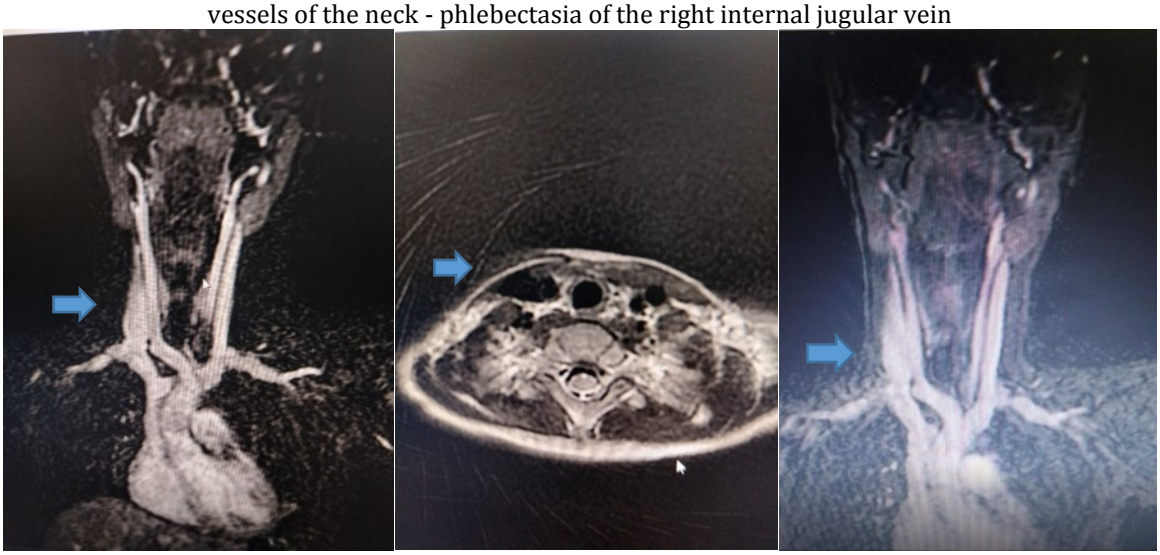




Left internal jugular vein with normal characteristics. In hospital conditions, magnetic resonance imaging of the main blood vessels of the neck was performed, which showed that the right internal jugular vein was wider along its entire length, with a maximum diameter of up to

11 mm, and the left internal jugular vein was reduced in diameter to 4 mm. The jugular veins were transient, without signs of thrombosis (MRI image 4).

Figure 4: (MR) Magnetic resonance imaging with contrast angiography and venography of the main blood vessels of the neck - phlebectasia of the right internal jugular vein



Due to the benign course and absence of complications, the patient was recommended conservative treatment and a normal lifestyle. Home monitoring and periodic outpatient check-ups were suggested.

Case report 3:

A 5-year-old girl. The parents report a change in the right side of the neck in the lower third that increases with exertion. The change is painless and does not interfere with normal life, and has been present for the past three years. The patient had no associated diseases that could be one of the causes of phlebectasia. Heteroanamnestic data are available on right-sided phlebectasia in a younger child, also female, in whom the change was seen at birth. Given the child's early infant age and the fact that the change is asymptomatic, the child has not yet had a radiological evaluation. The girl's mother was diagnosed with varicose veins in both lower legs.

Clinical examination at rest does not show any changes in the neck, but during the Valsalva maneuver, an oval change of about 2 x 3 cm in size appears in front of the anterior edge of the sternocleidomastoid muscle in the lower third on the right supraclavicular side, painless on palpation, partially compressible, soft consistency, non-pulsatile, the skin over the change unchanged.

Further diagnostics are recommended, ultrasonography of the soft tissues of the neck at rest, and during the Valsalva maneuver, as well as color Doppler of the blood vessels of the neck. It was explained to the parents that this is most often a benign condition that requires periodic monitoring in outpatient settings and parental supervision at home, in case of any complications, surgical treatment should be considered.

Discussion

Jugular vein flexure was first described by Harris in 1928 (1), and anomalous reduplication of the internal jugular vein was described by Zukschewerdh in 1929 (2). In 1952, Gerwing coined the term “phlebectasia” to describe an abnormal fusiform or saccular dilatation of the vessel (3).

Internal jugular vein phlebectasia is a rare type of vascular abnormality characterized by an isolated fusiform or saccular dilatation of the internal jugular vein without tortuosity. It is distinguished from varicosity by the absence of tortuosity, and from aneurysm by the fact that the dilatation uniformly involves the entire circumference of the vessel.

Clinically, phlebectasia of the internal jugular vein manifests as a cystic, soft, painless, partially compressible, non-pulsatile mass in the anterior triangle of the neck, visible supraclavicularly in front of the anterior edge of the sternocleidomastoid muscle. It increases with the Valsalva maneuver, during exertion, coughing, crying, sneezing, and spontaneously decreases during rest. It is most often asymptomatic and benign, more often affecting boys in a ratio of 2:1. Dysphonia or aphonia caused by pressure on the laryngeal nerve is rare, there is a feeling of humming due to turbulent blood flow in the dilated venous segment, headache, difficulty swallowing, cough on exertion, shoulder pain, phlebectasia of the internal jugular vein when moving the right arm, inability to speak loudly, pain in the root of the tongue, a feeling of tightness, suffocation and discomfort on exertion, and a feeling of a foreign body in the neck. Thrombosis, phlebitis, congestive heart failure, massive bleeding due to traumatic rupture, and Horner's syndrome are rare (9). Spontaneous rupture of phlebectasia has not been reported in pediatric patients (10).

Phlebectasia of the internal jugular vein occurs more frequently on the right side in a ratio of 5.2:1. Bilateral phlebectasia of the internal jugular vein is less common, somewhat more common in boys in a ratio of 1.4:1 (11). Possible causes of venous ectasia in the neck include gross anatomical abnormality, congenital structural defects in the vein wall, mechanical compression, or trauma, but are most often idiopathic (12). The most commonly affected internal jugular vein is the internal jugular vein, followed in descending order of occurrence by the external and anterior jugular veins, the jugular bulb, the facial vein, and the superficial communicating neck veins (13). The more frequent involvement of the right internal jugular vein is explained by anatomical differences: the shorter right brachiocephalic trunk, the higher position of the bulb of the right jugular vein, and the position and size of the valves (14). La Monte et al. hypothesized that phlebectasia of the internal jugular vein generally tends to the right because the right brachiocephalic vein is in close contact with the right apical pleura, and therefore the increase in intrathoracic pressure could be transmitted to the right internal jugular vein (15). A venous valve is almost never observed in the right brachiocephalic vein, in contrast to the left where the incidence of competent valves is 4 to 8% (16). Paleri and Gopalakrishnan presented their hypothesis that the increased intrathoracic pressure is transmitted predominantly to the right internal jugular vein due to the higher anatomically positioned valve and larger diameter of the right internal jugular vein, the shorter right brachiocephalic vein that follows the course of the superior vena cava, the greater number of competent valves in the right subclavian vein, and the greater number of valves in the left brachiocephalic vein (17). Other possible less likely causes include tracheomalacia and tracheoesophageal fistula, exposure to elevated positive intrathoracic pressure, internal jugular vein cannulation, internal jugular vein duplication, congenital primary weakness of the venous muscular layer, or loss of normal connective tissue of the vein wall. An association between internal jugular vein phlebectasia and Menkes disease has been suggested (18).

Histopathological studies have shown loss of the elastic layer and hypertrophy of connective tissue with focal intimal thickening. Histologically, diffuse fibrosis and disrupted elastic tissue architecture suggest a mechanical effect (19). Histopathological studies of surgically removed specimens show a normal varicose vein pattern in most cases, but in some cases there is loss or disruption of the arrangement of smooth muscle cells, elastic fibers, and connective tissue (20). In 1962, after surgical removal of a phlebectatic portion of the internal jugular vein, Leighton observed that smooth muscle fibers were randomly distributed in the vessel wall and that there was an island of adipose tissue extending into the tunica intima between the fibers. He called the phlebectasia a vascular hamartoma (21).

Ultrasonography is the diagnostic modality of first choice, the diagnosis of phlebectasia of the internal jugular vein is confirmed by the variation in size during rest and during Valsalva maneuver - anteroposterior diameter of more than 15 mm. During Valsalva maneuver, the diameter of the affected vein can increase up to 2.2 times compared to the measurement during rest (22). Color Doppler ultrasonography confirms the presence or absence of thrombosis in the vein lumen. Neck and chest radiography, magnetic resonance imaging of the main blood vessels of the neck and contrast-enhanced computed tomography additionally provide

even more information about the size of the lesion, anatomical relationships with other structures, and are indispensable for the definitive diagnosis of phlebectasia of the internal jugular vein, especially in pediatric patients (7). Chest and neck radiography can raise suspicion of laryngocele, or exclude the presence of air, as well as changes in the upper mediastinum. Laryngoscopy is recommended to complete the diagnosis and confirm the diagnosis of laryngocele. Invasive diagnostic radiological procedures and surgical explorations are rarely used in children, as noninvasive diagnostics confirm the diagnosis of internal jugular phlebectasia.

The diagnosis of cystic neck swelling is challenging, and the differential diagnosis in pediatric patients is broad, and in addition to phlebectasia, it includes: laryngocele, external laryngeal diverticulum, brachial cyst, cystic hygroma, cavernous hemangioma, tumors and cysts of the upper mediastinum, inflation of the pulmonary apical bulla, thyroglossal duct cyst, dermoid cyst, cervical adenopathy (23). The most common cause of a neck mass that increases with the Valsalva maneuver is laryngocele, but in children, phlebectasia of the internal jugular vein should also be considered (6).

Phlebectasia of the internal jugular vein increases in size in childhood until puberty, after which it spontaneously decreases. In asymptomatic and partially symptomatic phlebectasias, due to the benign, or self-limiting nature of the disease, the recommended treatment modality is conservative treatment with regular monitoring at home and periodic check-ups in a tertiary health care institution, most often lifelong (23).

Symptomatic phlebectasia of the internal jugular vein with complications such as thrombosis, compression of vascular structures, Horner's syndrome, or signs of rupture of the varicose vein are indications for urgent surgical intervention (24). Surgical treatment includes ligation of the varicose vein, resection of part of the phlebectatic venous wall, longitudinal venous constriction suture, coating and fixation of the varicose vein with the omohyoid muscle or an 8mm polytetrafluoroethylene tube - PTFE. (25). Cases of right-sided internal jugular phlebectasia that were treated surgically have been published, the treatment modalities are shown in the table (Table 1).

Table 1: Review of world literature - right-sided phlebectasia of the internal jugular vein.

Serial number	Author	Title of the work and year of publication	Number of patients	sex		Treatment	
1.	P.Sirivastava , V. Upadhyaya, A. Gangopadhyay, S. Sharma, R. Jaiman.	Internal Jugular Phlebectasia in Children: a Diagnostic Dilemma.The Internet Journal of Surgery. 2008	1	m	/	conservatively	/
2.	Basbug HS, Kizilgoz V.	A rare childhood entity: Massive internal jugular vein phlebectasia. Arch Basic Clin Res. 2021	1	m	/	conservatively	/
3.	Alessandro Raffaele, Marta Gazzaneo, Piero Romano, Maria Sole Prevedoni Gorone	Congenital Internal Jugular Phlebectasia: An Anomaly Still Poorly Recognized. European J Pediatr Surg 2023	1	m	/	conservatively	/
4.	Shah K. Chavan P.	Profiling of rigt Internal jugular vein phlebectasia: a rare case study. Int J Otorhinolaryngol Head Neck Surg 2024	1	m	/	conservatively	/
	Miljenko Raos, Jelica Marković	Flebektazija unutarnje jugularne vene: prikaz slučaja. 2010	1	m	/	conservatively	/
6.	Bindal SK, Vasisth GO, Chibber P.	Phlebectasia of Internal Jugular Vein. J Surg Tech Case Report 2012	1	m	/	/	Surgical (vein ligation)
7.	Dr. Pedro S. Jimenez Urueta et al.	Flebectasia de la vena jugular interna en ninos. Acta					

		pediatrica de Mexico. 2005	4	m	/	/	Surgical (PTFE)
8.	Sultan Alrabea, Anoof Eshky, Thamer Albilasi, Mazyad Alenzi and Samir Bawazir.	Internal jugular phlebectasia in pediatric patients: a report of two Saudi cases. 2021	2	m	/	conservatively	/
9.	Kandiah R. Mohamed I	The louder the bigger: A case of jugular phlebectasia in a child. Malays Fam Physician. 2019	1	m	/	conservatively	/
10.	VishnuKanth, R. Manju, Jineesh Joseph, Balla Nagamali Kumar, Abhisekh Chauhan	Jugular vein ectasia presenting with hoarseness of voise: A rare association. 2017	1	/	f	conservatively	/
11.	Karthik N. Rao, Shrinivas S. Chavan et al.	IJV Phlebectasia: an approach algorithm.2017	1	m	/	conservatively	/
12.	Ayse Enise Goker, Ziya Salturk, Perihan Taskale,Enes Atac,Yavuz Uyar	Congenital jugular phlebectasia : analysis of two cases. 2015	2	m	f	conservatively	/
13.	Amber Kesarwani, Amit Goyal, Amit Kumar	Phlebectasia of Internal Jugular Vein- a Rare Differential Case of Neck Swelling With Review of Literature.Iran J. Otorhinolaryngol.2019	1	/	f	conservatively	/
14.	Deepanjan Bhattacharya, Mounika Endrakanti,Rakech Kuhar	Right Internal Jugular Vein Phlebectasia: A Rare Cause of Neck Swelling . Case reports in Pediatrics 2017	1	m	/	conservatively	/
15.	M. Afal, A Mohamed, Y Volkin	Jugular Vein Phlebectasia in a Pediatric Patient With Tracheomalacia. 2023	1	m	/	conservatively	/
16.	Neha D. Shetty et al.	A Case of Phlebectasia in a Child Presenting With Neck Mass 2023	1	m	/	conservatively	/
17.	Dr. Mehmet Demiracan et al.	Jugular Phlebectasia in Children: A Case Report.1997	1	m	/	/	Surgical (PTFE)
18.	Y. Gao et al	Diagnosis and Treatment of Internal Jugular Phlebectasia-three cases report. 1999	3	m	/	/	Surgical (excision vein)
19.	Ksim S. Kasim et al.	Internal Jugular Vein Phlebectasia in a Child : A Case Report . 2018	1	m	/	conservatively	/
20.	Alexandra Zimm	Internal Jugular Phlebectasia. 2010	1	/	f	conservatively	/
21.	V. Malk, Virender, Kumari et al.	Phlebectasia of internal jugular vein with intracranial extension. 2015	1	m	/	conservatively	/
22.	M. Safi et al.	A rare presentation of Horner syndrome due to internal jugular phlebectasia . 2021	1	m	/	conservatively	/

23.	Yoko Omata, Yochiko Takahashi,Tomo ko Nakazawa, Taku Omata	Paediatric primary internal jugular poucough headache with phlebectasia. 2021	1	m	/	conservatively	/
24.	Tamami AIN, Al Macki K.	Internal Jugular Phlebectasia: A Case Report and Literature Review. J Otolaryngol ENT Res. 2015	1	m	/	conservatively	/
25.	Jayakumar V, Arora PK	Right internal jugular vein phlebectasia-a rare cause of neck swelling in paediatric population. Int J Otorhinolaryngol Head Neck Surg. 2022	1	m	/	conservatively	/

* polytetrafluoroethylene tube- PTFE

Conclusion

Phlebectasia of the internal jugular vein should be included in the differential diagnosis of atypical neck masses in children. It can be easily diagnosed with a detailed history and physical examination. The diagnostic modality of first choice is ultrasound examination of the soft tissues of the neck at rest, and during the Valsalva maneuver, because it can be easily and effectively used for monitoring, so that the extent of the swelling can be documented. Magnetic resonance imaging with contrast angiography and venography of the main blood vessels of the neck is reserved for the definitive diagnosis of phlebectasia, especially in pediatric patients. After diagnosis, the patient should be followed up regularly. Inform the patient and his parents about the risk of possible complications. Most importantly, the patient and his parents should be reassured that this is most often a benign condition, and that it will not affect the normal life regimen. The presented patients correspond to the largest number of patients presented in the world literature in terms of clinical characteristics, diagnostic method and proposed treatment modalities..

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