ABCResearch

Arch Basic Clin Res 2021; 3(3): 135–137 • DOI: 10.5152/ABCR.2021.21017

Image of Interest / Özgün Görüntü

A Rare Childhood Entity: Massive Internal Jugular Vein Phlebectasia

Nadir Bir Çocukluk Oluşumu: Masif İnternal Juguler Ven Flebektazisi

Hamit Serdar Başbuğ[1], Volkan Kızılgöz[2]

[1]Department of Cardiovascular Surgery, Kafkas University Faculty of Medicine, Kars, Turkey
[2]Department of Radiology, Erzincan Binali Yıldırım University Faculty of Medicine, Kars, Turkey

Cite this article as: Başbuğ HS, Kızılgöz V. A rare childhood entity: Massive internal jugular vein phlebectasia. *Arch Basic Clin Res.* 2021; 3(3): 135–137.

ORCID iDs of the authors: H.S.B. 0000-0002-1363-6783; V.K. 0000-0003-3450-711X.

INTRODUCTION

Phlebectasia is a word describing an abnormal fusiform or saccular dilatation of a vein that differs from the varicose veins, which implies tortuosity. The most common cause of a neck mass that becomes visible during straining is a laryngocele. However, internal jugular vein phlebectasia (JVP) also presents similarly and is a rare pathology in childhood. We present a massive internal JVP of a child on clinical examination and a computerized tomography image with its differential diagnosis.

CASE PRESENTATION

An 8-year-old boy was referred to with a swelling on his neck's right side, which quickly becomes visible during straining (Figure 1). During the Valsalva maneuver, the mass was inflated and subsided upon relaxation, leaving no abnormality with an inspection or palpation. History revealed good health and average growth with no operation or trauma. The parents stated that the swelling was noticeable for four years and showed a gradual increase. He had been unable to wear a collar as he felt uncomfortable with constriction and choking sensation whenever he would strain. Physical examination revealed no other visible pathology when he was relaxed (Figure 1a). However, when forceful expiratory efforts were made, a massive swelling appeared on the neck's right side (Figure 1b and 1c). The mass was smooth and ovoid. It was not pulsatile, and no bruit was heard. No tenderness or pain exists. Computerized tomography angiography images while the boy was straining revealed a massive right internal jugular vein dilatation (Figure 2).

DISCUSSION

A neck mass in a healthy child appearing only on straining is most likely a JVP, and it is rarely encountered.[1] Phlebectasia is a term used to describe an abnormal dilatation of a vein in fusiform shape without tortuosity[2] Although anatomic variations of the external jugular vein are often encountered, internal jugular vein is quite consistent for the developmental anomaly ocurrence. The first domestic case of internal JVP was reported in 1952 by Gerwig WH and has been described sporadically with only a few descriptions until the 1970s.[3] As the internal jugular vein is an unusual site for a phlebectasia, this case is considered to deserve a report and discussion.

The diagnosis of internal JVP is made using Color Duplex ultrasonography scanning and computerized tomography angiography.[4] The differential diagnosis of a neck mass includes laryngocele, cystic hygroma, lymphocele, thyroid swelling, cavernous hemangioma, and enterogenous cyst. A thyroglossal cyst, lymphadenopathy, branchial cleft cyst, and dermoid cyst should also be considered in the diagnosis. Distention of the neck mass during Valsalva eliminates others than a laryngocele and JVP. The absence of air inside the swelling on the X-ray further eliminates the lary ngocele.[5] Internal JVP may be associated with Menkes Disease, an X-linked recessive disorder of copper

Correspondence Author / Sorumlu Yazar: Hamit Serdar Başbuğ **E-mail / E-posta:** s_basbug@hotmail.com

Submitted / Geliş Tarihi: 22.05.2021
Accepted / Kabul Tarihi: 03.09.2021

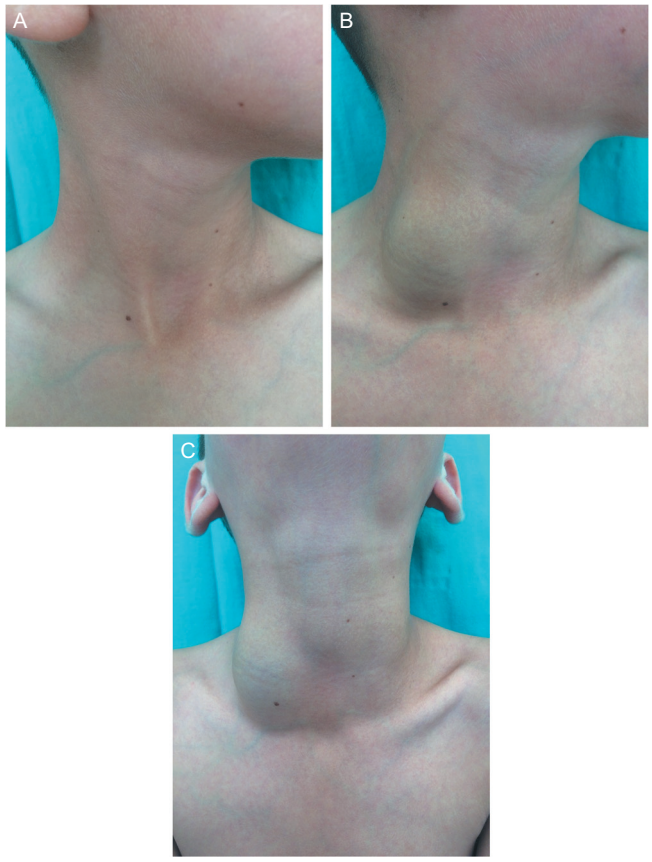

Figure 1. a-c. The neck without Valsalva (a). The right-sided massive internal jugular vein phlebectasia appearing during Valsalva: lateral (b) and anterior (c) view.

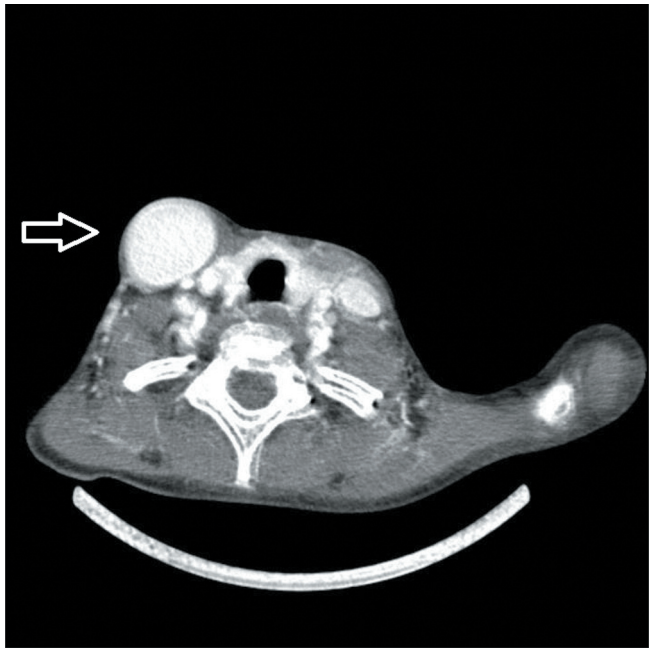

Figure 2. Computerized tomography angiography image. The arrow shows the massive internal jugular vein phlebectasia.

metabolism.[6] Massive internal JVP may also cause the classic features of Horner's syndrome that are unilateral miosis, ptosis, and anhidrosis due to the compression of the sympathic fibers.[7]

Internal JVP is a benign condition presenting with the main symptoms of constriction, choking, dizziness, coughing, tongue pain, and cessation of voice. Although these symptoms are rare, there is an absolute indication of surgical treatment in the symptomatic patients.[3] Otherwise, treatment should be conservative with routine follow-up evaluation. In asymptomatic cases, decision of the surgery depends on the growth of the lesion during follow-up, cosmetic concerns, and the emotional stress of the child caused from being taunted by their peers. In these patients, longitudinal constriction suture venoplasty plus encapsulation is recommended as a surgical technique.[8] In most surgical cases, the internal JVP has been totally ligated, cancelling the normal venous drainage pattern on that side. Resection and end-to-end anastomosing is another method for the surgical treatment of internal JVP.[9] All these options have been reported to be safe and successful in eliminating the internal JVP.[10]

Internal JVP is a rare entity. The diagnosis is considered in the presence of a neck mass enlarged by Valsalva maneuver and confirmed by imaging. Surgery is only indicated in the presence of complications.

Informed Consent: Written informed consent was obtained from the patient's parents.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – H.S.B.; Design – H.S.B.; Supervision - V.K.; Resources – V.K.; Materials – V.K.; Data Collection and/or Processing – H.S.B., V.K.; Analysis and/or Interpretation - H.S.B., V.K.; Literature Search - H.S.B.; Writing Manuscript - H.S.B.; Critical Review - V.K.

Conflict of Interest: The authors have no conflicts of interest todeclare.

Financial Disclosure: The authors declared that they have received no financial support for this study.

Hasta Onamı: Yazılı hasta onamı bu çalışmaya katılan hastanın ailesinden alınmıştır.

Yazar Katkıları: Fikir – H.S.B.; Tasarım – H.S.B.; Denetleme – V.K.; Kaynaklar – V.K.; Malzemeler – V.K.; Veri Toplanması ve/veya İşlemesi – H.S.B., V.K.; Analiz ve/veya Yorum – H.S.B., V.K.;

Literatür Taraması – H.S.B.; YazıyıYazan – H.S.B.; Eleştirel İnceleme – V.K.

Hakem Değerlendirmesi: Dış bağımsız.

Çıkar Çatışması: Yazarlar çıkar çatışması bildirmemişlerdir.

Finansal Destek: Yazarlar tarafından finansal destek almadıklar ıbildirilmiştir.

REFERENCES

1. El Fakiri MM, Hassani R, Aderdour L, Nouri H, Rochdi Y, Raji A. Congenital internal jugular phlebectasia. *Eur Ann Otorhinolaryngol Head Neck Dis*. 2011;128(6):324-326. [Crossref]
2. Başbuğ HS, Göçer H, Günerhan Y, Erdem A, Özışık K. A rare internal jugular vein phlebectasia: Review of the literature. *Turk Gogus Kalp Dama*. 2016;24(4):759-762. [Crossref]
3. Paleri V, Gopalakrishnan S. Jugular phlebectasia: Theory of pathogenesis and review of literature. *Int J Pediatr Otorhinolaryngol*. 2001;57(2):155-159. [Crossref]
4. Calligaro KD, Ahmad S, Dandora R, et al. Venous aneurysms: Surgical indications and review of the literature. *Surgery*. 1995;117(1):1-6. [Crossref]
5. Başbuğ HS, Bitargil M, Karakurt A, Özışık K. External jugular vein aneurysm in a young woman: An uncommon cause of neck mass. *Int J Cardiovasc Acad*. 2016;2(1):16-18. [Crossref]
6. Stevens KE, Price JE, Marko J, Kaler SG. Neck masses due to internal jugular vein phlebectasia: Frequency in Menkes disease and literature review of 85 pediatric subjects. *Am J Med Genet*. 2020-1377;182A(6):1364-1377. [Crossref]
7. Inci S, Bertan V, Kansu T, Cila A. Horner's syndrome due to jugular venous ectasia. *Child's Nerv Syst*. 1995;11(9):533-535. [Crossref]
8. Jianhong L, Xuewu J, Tingze H. Surgical treatment of jugular vein phlebectasia in children. *Am J Surg*. 2006;192(3):286-290. [Crossref]
9. Hung T, Campbell AI. Surgical repair of left internal jugular phlebectasia. *J Vasc Surg*. 2008;47(6):1337-1338. [Crossref]
10. Hu X, Li J, Hu T, Jiang X. Congenital jugular vein phlebectasia. *Am J Otolaryngol*. 2005;26:172-174. [Crossref]