Rare disease

Internal jugular phlebectasia in an African adult

Godwin I Ogbale,¹ Achiaka E Irabor,² P Oladapo Adeoye,³ Bolutife P Yusuf¹¹ Department of Radiology, University College Hospital, Ibadan, Nigeria² Department of Family Medicine, University College Hospital, Ibadan, Nigeria³ Department of Surgery, University College Hospital, Ibadan, Nigeria

Correspondence to Godwin I Ogbale, gogbole@yahoo.com

Summary

Jugular vein phlebectasia is a rare venous anomaly commonly presenting as a unilateral neck swelling in children and adults. Its aetiology is unknown and its detection may have increased with availability of better imaging facilities. Due to its rarity, a high index of suspicion is required once other causes of neck swelling are excluded. The few reported cases are mainly in children and non-Africans adults. The authors report the case of a 54-year-old Nigerian woman presenting for the first time with a progressive, intermittent, painless right neck swelling, which was demonstrated on Doppler ultrasound and CT angiography to be an internal jugular phlebectasia. She had satisfactory surgical excision of the dilated segment for cosmetic reasons.

BACKGROUND

Internal jugular phlebectasia is a rare benign lesion, which mostly involves the right side. It is usually a childhood disease and believed to be of congenital origin. It can occur at any age and affects both genders. Nevertheless, to our knowledge, no single case has been reported in an African adult.

CASE PRESENTATION

A 54-year-old woman presented with complaints of painless, intermittent right neck swelling noticed initially about 2 years prior to presentation. There was an associated feeling of a mass on swallowing and feeling of heaviness in the chest 8 months before presentation. The swelling was insidious in onset, gradually progressive with prominence of the swelling seen on leaning forward or straining. There

was no history of change in voice, difficulty in swallowing or breathing. She was diagnosed hypertensive about 4 years earlier but non-compliant with antihypertensives. She had no other significant past history of illness.

Figure 1 (A) Normal appearance of neck at rest and (B) unilateral right-sided internal jugular ectasia (arrow) during the Valsalva manoeuvre.

Figure 2 Real-time B-mode ultrasonography demonstrating marked variation in the size of jugular vein between rest (A) and Valsalva manoeuvre (B).

Figure 3 (A) Contrast-enhanced axial CT image at the level of the thyroid gland shows dilated right internal jugular vein and heterogeneous contrast filling. (B) Reformatted coronal CT image shows the extent of the dilatation and a flask-shaped configuration of the right internal jugular vein (arrow).

Figure 4 Intraoperative picture shows ectatic segment (arrow) of the right internal jugular vein.

Physical examination revealed a soft, pulsatile, fusiform right lower neck swelling anteromedial to the sternocleidomastoid muscle. The swelling increased in size with Valsalva manoeuvre (figure 1) and was non-tender but compressible.

Ultrasound of the mass showed a fusiform dilatation of the right internal jugular vein measuring 4.5 cm in its widest transverse diameter at the level of the thyroid gland (figure 2). It showed normal venous spectral wave form on Doppler interrogation and was completely compressible. Turbulent flow was noted but no thrombus was seen within it. The contralateral internal jugular vein was normal in size measuring 1.1 cm in diameter at the corresponding level.

The ultrasound finding was confirmed on a carotid CT angiography. This showed the extent of the dilatation to be progressively increasing from the jugular foramen to the infraclavicular region caudally (figure 3). The CT also showed congestion of the pulmonary veins. However, there was no mediastinal mass demonstrated.

Due to cosmetic concerns and feeling of heaviness in the chest, she had excision of the phlebectatic segment through an oblique neck incision along the anterior border of sternocleidomastoid muscle (figure 4). The ends of the jugular vein were ligated with the contralateral jugular vein being available to sustain venous drainage from the brain. Histo-

logical examination of the excised segment demonstrated necrotic changes in the vessel wall with intraluminal clot.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses for a cystic neck mass that increases in size with Valsalva manoeuvre includes laryngocele, cystic hygroma and a superior mediastinal tumour.

OUTCOME AND FOLLOW-UP

The patient expressed satisfaction with the surgery and relief of her previous experience of a 'heavy chest' during her first and 6-month postoperative clinic review with no other complaints. She is presently scheduled for yearly follow-up clinic visits.

DISCUSSION

Internal jugular phlebectasia (also known as venous congenital cyst, venous aneurysm, vein ectasia or essential venous aneurysm) refers to an isolated abnormal fusiform or saccular dilatation of the internal jugular vein.¹ It presents as a soft, compressible cystic swelling in the neck that increases in size during any kind of straining that increases the intrathoracic pressure, such as coughing, sneezing or crying. It is considered as an asymptomatic benign condition but, in some cases, there are complaints of constriction, choking, tongue pain and discomfort during physical activity.² Our patient had noticed the neck swelling for over 2 years but only presented when she associated it with heaviness in her chest.

It is a rare benign vascular abnormality but may be under-reported³ possibly due to its benign nature. There is no known sex predilection, although one case reported that boys were affected twice often as girls.² Most reported cases are in children. They usually occur on the right side as in our patient but may be bilateral.⁴

Anatomically, phlebectasia in the neck have been described in the following sequence in frequency: internal jugular, external jugular, anterior jugular and superficial comminicans.⁵

The cause of venous ectasia is unknown. It may be congenital, although this has not been proved. Histologically, examinations of the wall have revealed a reduction in elastic fibres, intimal hyalinisation and no abnormality.⁶

Diagnosis is usually confirmed with ultrasonography, CT, MR or venography.⁷ Ultrasound will often suffice, allowing avoidance of more invasive examination, radiographic exposures and unnecessary surgical intervention. Doppler ultrasound shows an essential flat spectral wave indicating a venous structure.⁸ Ultrasound has been used to diagnose jugular phlebectasia since 1982.⁹

The differential diagnoses for a cystic neck mass that increases in size with Valsalva manoeuvre includes laryngocele, cystic hygroma, superior mediastinal tumour and other vascular lesions.¹⁰

Although cases of internal and external jugular phlebectasia have been reported mostly in children and non-African adults, as far as we are aware no case has been previously reported in the electronic literature in an African adult.^{3 11}

Nonetheless, our case showed ultrasound and CT features described in the reported cases in children.^{4 12} The swelling is not known to progress rapidly and there have been no instances of spontaneous rupture of the swelling or other serious complications.^{7 11}

The complications of this condition are rare but include thrombosis, congestive heart failure and massive haemorrhage secondary to trauma.¹²

No treatment is indicated for this benign self-limiting condition except in a few patients who complain of symptoms and require surgical resection of the affected vein or covering with muscular segment.¹³ Some authors believe that ligation of the jugular vein may produce effects of venous congestion in a small subset of patients resulting in cerebral oedema. Jugular vein ligation cannot be applied in cases with bilateral affliction.¹⁴

Our patient was essentially asymptomatic from the neck mass and the pulmonary congestion observed is most likely is due to her unattended hypertensive heart disease. Surgical excision was based on cosmetic concerns.

Learning points

- ▶ Internal jugular phlebectasia is a rare, benign, venous anomaly, commonly seen in children but can occur at any age.
- ▶ The cause is unknown but thought to be congenital, although this has not been proved.
- ▶ Ultrasound often suffices in making a diagnosis; thus, preventing more invasive examinations, radiographic exposures and unnecessary surgical intervention.
- ▶ Colour Doppler ultrasonography is a good modality for showing flow turbulence and real-time increase during Valsalva manoeuvre.
- ▶ It usually does not require surgical intervention although this can be offered for cosmetic concerns with satisfactory results.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Bowdler DA**, Singh SD. Internal jugular phlebectasia. *Int J Pediatr Otorhinolaryngol* 1999;**65**:181–84.
2. **Lugo-Vicente HL**, ed. *Jugular phlebectasia*. Pediatric surgery update 2001;**18**:p. 2.
3. **Sander S**, Eliüevik M, Unal M, *et al*. Jugular phlebectasia in children: is it rare or ignored? *J Pediatr Surg* 1999;**34**:1829–32.
4. **Gendeh BS**, Dhillon MK, Hamzah M. Bilateral internal jugular vein ectasia: a report of two cases. *J Laryngol Otol* 1994;**108**:256–60.
5. **Abdulla AS**, Aldabagh MH. Congenital phlebectasia of the internal jugular vein (case report). *Dohuk Med J* 2008;**2**:155–60.
6. **Ekim H**, Ozen S. Primary venous aneurysm of the external jugular vein. *Eastern J Med* 2002;**7**:24–25.
7. **Gordon DH**, Rose JS, Kottmeier P, *et al*. Jugular venous ectasia in children. A report of 3 cases and review of the literature. *Radiology* 1976;**118**:147–9.
8. **Lin MJ**, Chou YH, Tiu CM, *et al*. [Duplex Doppler ultrasound of internal jugular vein ectasia. Case report]. *Zhonghua Yi Xue Za Zhi (Taipei)* 1990;**46**:190–3.
9. **Chao HC**, Wong KS, Lin SJ, *et al*. Ultrasonographic diagnosis and color flow Doppler sonography of internal jugular venous ectasia in children. *J Ultrasound Med* 1999;**18**:411–6.
10. **LaMonte SJ**, Walker EA, Moran WB. Internal jugular phlebectasia. A clinicoradiographic diagnosis. *Arch Otolaryngol* 1976;**102**:706–8.
11. **Nwako FA**, Agugua NE, Udeh CA, *et al*. Jugular phlebectasia. *J Pediatr Surg* 1989;**24**:303–5.
12. **Swami SLH**, Nambiar S. Focal ectasia of internal jugular vein. *Med J Armed Forces India* 2009; **65**: 3.
13. **Lindiane Maria De Brito Macedo Ferreira**, Eric Frota Haguette. Bilateral internal jugular phlebectasia. *Int Archives Otorhinolaryngol* 2007;**11**:21.
14. **Walsh RM**, Murty GE, Bradley PJ. Bilateral internal jugular phlebectasia. *J Laryngol Otol* 1992;**106**:753–4.

This pdf has been created automatically from the final edited text and images.

Copyright 2010 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Ogbole GI, Irabor AE, Adeoye PO, Yusuf BP. Internal jugular phlebectasia in an African adult. *BMJ Case Reports* 2010;10.1136/bcr.02.2010.2724, date of publication

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow