



Lesions one needs to yell out for diagnosis

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Introduction

A neck mass protruding out on Valsalva manoeuvre is an infrequently encountered clinical condition in children. Three cases with such unusual neck masses were presented. In each of them the neck mass was shown up nicely by asking the child to perform some sort of Valsalva manoeuvre, e.g. yelling, to help the doctor to study it and make the diagnosis.

Case 1

A 3-year-old boy was referred from his private doctor for hoarseness of voice since infancy. He liked to scream loudly, especially when agitated. His mother noticed an anterior neck swelling in the last 6 months whenever he cried, strained, yelled or screamed. At rest, the mass disappeared. There was no noisy breathing, choking, dysphagia or respiratory distress. Birth and perinatal course was uneventful with no history of neonatal resuscitation or intubation. Physical examination showed mild hoarseness of voice but no stridor, dyspnoea, or chest insucking. There was no goiter, cervical lymphadenopathy or neck mass visible at rest or upon swallowing and the overlying neck skin was normal. An

anterior midline neck mass appeared when the boy yelled but immediately disappeared when returned to rest (Figure 1). Chest, cardiovascular, abdominal and neurological examinations were unremarkable.

Fibreoptic laryngoscopy showed screamer nodules over the vocal cords. Neck ultrasonography revealed a normal thyroid gland but with the boy shouting a well circumscribed echogenic mass was seen protruding from the infraclavicular to lower neck region. It descended back into the upper mediastinum afterwards but the lower part was not well delineated. Magnetic resonance imaging (MRI) confirmed the herniating neck mass corresponded to the superior aspect of thymus (Figure 2).



Figure 1. Anterior neck mass protruding when yelling.

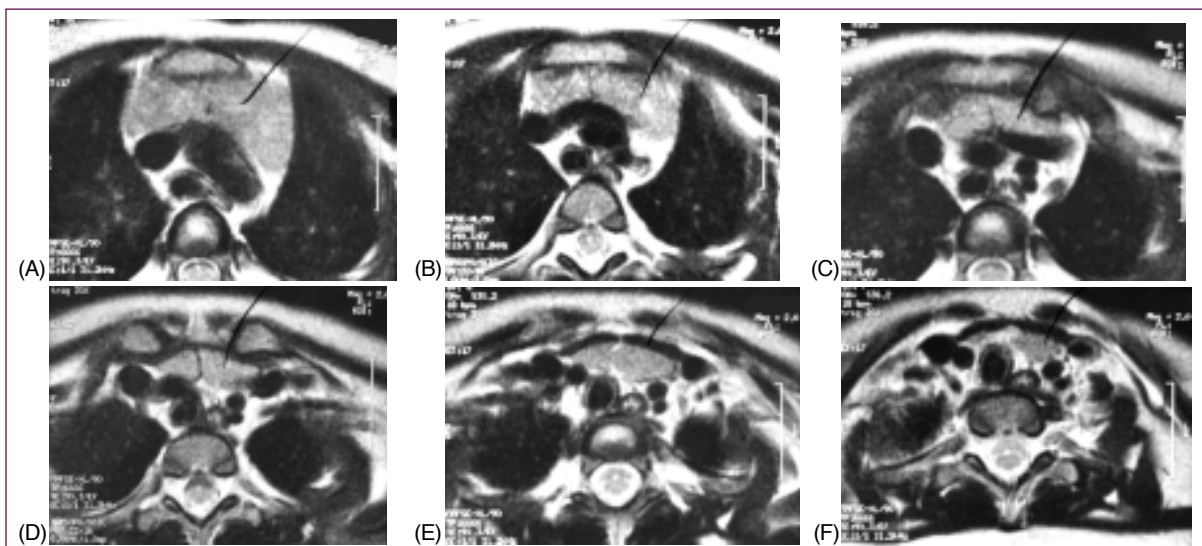


Figure 2. MRI neck and mediastinum during Valsalva manoeuvre - serial transverse images tracking the thymus (marked) from superior mediastinum (A) to lower neck level, with its left side slightly more superior than the right side (F).

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Case 2

A 11-year-old boy was referred from his private doctor for right-sided neck swelling for 3 months, suspected to be goiter. It was incidentally noted by his mother and he had no thyrotoxic symptoms, dysphagia, dyspnoea, or hoarseness of voice. He enjoyed good past health and there was no family history of thyroid problems. Physical examination showed a very small goiter with no nodules or bruit. He was clinically euthyroid. A right-sided neck swelling arose from the lower neck only during Valsalva manoeuvre (Figure 3) and it completely subsided immediately afterwards. It did not transilluminate and was non-pulsatile. Auscultation showed no venous hum or



Figure 3. Right-sided neck swelling during Valsalva manoeuvre

bruit. There was no supraclavicular or cervical lymphadenopathy and chest, cardiovascular, abdominal and neurological examinations were unremarkable.

Blood tests showed normal thyroid function and negative anti-thyroid antibodies. Neck ultrasonography revealed that the right-sided neck swelling during Valsalva manoeuvre was the abnormally dilated right internal jugular vein. At rest the internal jugular veins were already asymmetrical in caliber and the difference was amplified during Valsalva manoeuvre (Figure 4). Right internal jugular phlebectasia was diagnosed. Conservative management was recommended.

Case 3

A 2-year-and-8 month-old girl was referred from government outpatient clinic for neck swelling since age of 1 year. She was born with port-wine stain on the right side of her face, chin, neck and upper chest. Her parents noticed an anterior neck swelling since age 12 months during her vocalization and it was getting more prominent with age, easily noticeable when she shouted, laughed and strained. The mass disappeared at rest. There was no hoarseness of voice, respiratory distress or dysphagia. Her development was normal and there was

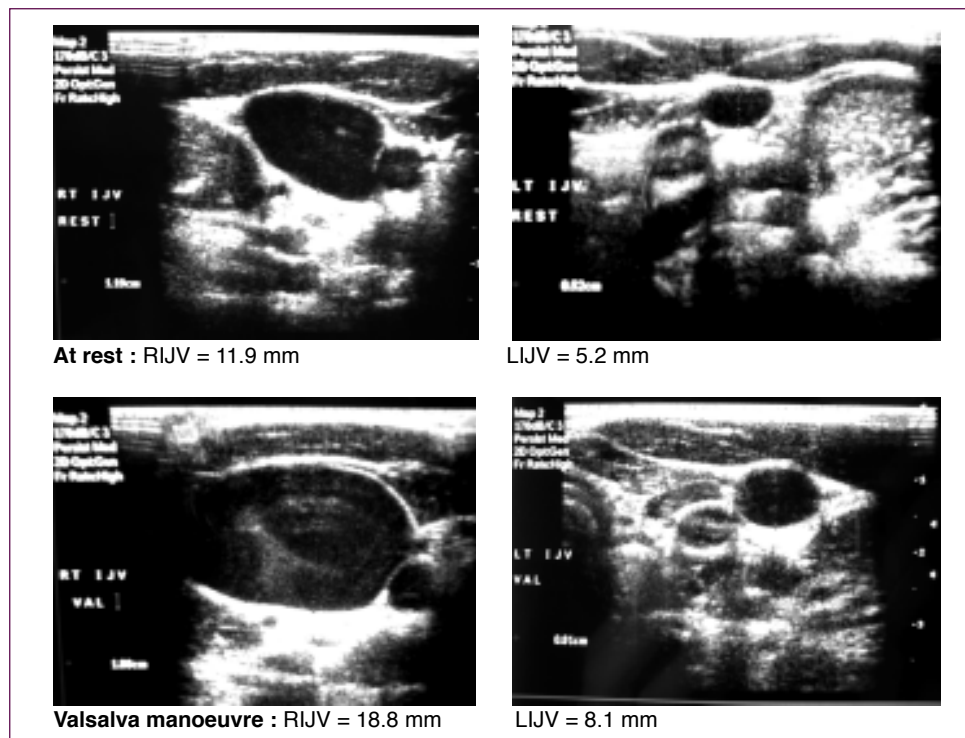


Figure 4. Anteroposterior diameters of right and left internal jugular veins (RIJV and LIJV)



no history of visual problem or convulsion. Physical examination showed extensive naevus flammeus (port-wine stain) involving right temporal scalp, right side of face, chin, neck and right upper chest regions. There was no goiter, cervical lymphadenopathy or neck mass visible at rest or upon swallowing. An anterior neck mass appeared above sternal notch when the girl yelled (Figure 5), which completely disappeared about 2 seconds after the girl stopped yelling. It did not transilluminate or pulsate. Auscultation showed no venous hum or bruit. Chest, cardiovascular, abdominal and neurological examinations were unremarkable.

Neck ultrasonography and doppler study showed a venous aneurysm with turbulent blood flow over the anterior midline lower neck region. Its axial cross-sectional diameter measured (transverse x anteroposterior) 17 mm x 12 mm at rest and 22 mm x 17 mm when laughing. On magnetic resonance imaging (MRI) and venography (MRV) the aneurysm (Figure 6) was noted to connect to left anterior jugular vein and

right side of jugular venous arch, which in turn joined the right internal jugular vein (Figure 7). It was managed conservatively upon surgical advice and laser therapy was arranged for the port-wine stain.

Discussion

Common differential diagnoses of midline anterior neck mass in children includes goitre, thyroglossal duct cyst, dermoid-epidermoid cyst, cystic hygroma, branchial cleft cyst and arteriovenous malformation. The neck swellings in our three children appeared only during Valsalva manoeuvre, therefore these were unlikely possibilities.

The most common cause of a neck mass that becomes visible or increases in size with Valsalva manoeuvre is laryngocele. Other lesions reported include saccular cyst, pharyngeal pouch, external laryngeal diverticulum, jugular phlebectasia, cavernous haemangioma, jugular



Figure 5. Anterior neck mass protruding during yelling



Figure 6. MRI neck showing the venous aneurysm (marked)



Figure 7. MRV coronal view showing the venous drainage



venous aneurysm, cupula inflation, thymic herniation and superior mediastinal cyst.

Although subclinical posterior buckling of the cervical trachea due to thymic herniation has been previously reported in infants,¹ clinically detectable thymic herniation is rare in older children. With such a dramatic, clinically visible and palpable thymic herniation in the neck, our patient in Case 1 is the youngest child reported in Hong Kong to date. Diagnosis can be confirmed by ultrasonography and MRI,² and biopsy is unnecessary. Little has been described in the literature regarding its natural history and it is believed to resolve with the regression of the thymus but the age of resolution is unclear. It has been reported to persist into mid-childhood.² Concerns, if any, would be cosmetic, psychosocial disturbance or parental anxiety.

Jugular phlebectasia is an entity that is being increasingly recognized in recent years. Phlebectasia was the term first used by Gerwig³ to describe an abnormal fusiform dilatation of a vein. The internal jugular vein is the most commonly affected and in most cases it is unilateral right side involvement. Diagnosis is confirmed by ultrasonography at rest and during Valsalva manoeuvre comparing both internal jugular veins.^{4,5} The cause is idiopathic. To date, locally reported cases were associated with childhood asthma^{6,7} but our patient in Case 2 was a non-asthmatic. Surgical exploration and resection was common in the past for diagnosis and management^{8,9} but associated with potential complications. Clinical course appears benign and conservative management is recommended for asymptomatic patients.¹⁰⁻¹⁴

Venous malformations in general are rare in children. Opinions on their aetiology vary and classification is not universal. Most common lesions in the head and neck region involve the internal jugular veins.¹⁵ Ultrasonography and Doppler study is diagnostic in the majority of patients, while computed tomography, angiography have also been used in the assessment of these lesions. Recently developed fast sequences have enabled MRI and MR angiography to provide three-dimensional vascular imaging of the jugular venous system as well as more central venous structures in the mediastinum.¹⁶ These non-invasive techniques without ionising radiation have proven successful and reliable

even in children as young as our patient in Case 3. The natural history of venous aneurysms depends on their anatomic location and those in the head and neck region, including the internal jugular vein, appear to have a benign course. Treatment strategies vary. Some recommended surgical excision while others adopt conservative management as serious complications have been rarely reported.¹⁵

References

1. Mandell GA, Bellah RD, Boulden ME, Sherman NH, Harcke HT, Padman RJ, McNicholas KW. Cervical trachea: dynamics in response to herniation of the normal thymus. *Radiology* 1993;186:383-6.
2. Wong KT, Lee DL, Chan MS, Tsang RK, Yuen EH, Ahuja AT. Unusual anterior neck mass visible only during Valsalva's maneuver in a child. *Am J Roent* 2005;185:1355-7.
3. Gerwig WJ. Internal jugular phlebectasia. *Ann Surg* 1952;135:130-3.
4. Jasinski RW, Rubin JM. CT and ultrasonographic findings in jugular vein ectasia. *J Ultrasound Med* 1984;3:417-20.
5. Jeon CW, Choo MJ, Bae IH, Shin SO, Choi YS, Lee DW, et al. Diagnostic criteria of internal jugular phlebectasia in Koreana children. *Yonsei Med J* 2002;43:329-34.
6. Ng DKK, Kwok KL, Lam HS. Unilateral internal jugular phlebectasia. *HK Med J* 2000;6:431.
7. Kwok KL, Lam HS, Ng DKK. Unilateral right-sided internal jugular phlebectasia in asthmatic children. *J Paed Child Health* 2000;36:517-9.
8. Sander S, Elicevik M, Unal M, Vural O. Jugular phlebectasia in children: is it rare or ignored? *J Ped Surg* 1999;34:1829-32.
9. Gurpinar A, Kiristioğlu I, Dogruyol H. Jugular phlebectasia. *Eur J Ped Surg* 1999;9:182-3.
10. Al-Dousary S. Internal jugular phlebectasia. *Int J Ped Otorhinolaryngol* 1997;38:273-80.
11. Lubianca-Neto JF, Mauri M, Prati C. Internal jugular phlebectasia in children. *Am J Otolaryngol* 1999;20:415-8.
12. Paleri V, Gopalakrishnan S. Jugular phlebectasia: theory of pathogenesis and review of literature. *Int J Ped Otorhinolaryngol* 2001;57:155-9.
13. Hopsu E, Pitkaranta A. Jugular vein aneurysm or phlebectasia. *Am J Surg* 2004;188:622.
14. Li J, Jiang X, Hu T. Surgical treatment of jugular vein phlebectasia in children. *Am J Surg* 2006;192:286-90.
15. Gillespie DL, Villavicencio JL, Gallagher C, Chang A, Hamelink JK, Fiala LA, et al. Presentation and management of venous aneurysms. *J Vasc Surg* 1997;26:845-52.
16. Fitoz S, Atasoy C, Yagmurlu A, Erden I, Akyar S. Gadolinium-enhanced three-dimensional MR angiography in jugular phlebectasia and aneurysm. *Clin Imaging* 2001;25:323-6.