CASE REPORT

Vascular ring diagnosis following respiratory arrest

Evie Alexandra Robson,¹ Alison Scott,¹ Philip Chetcuti,² David Crabbe³¹Department of Respiratory Paediatrics, Leeds General Infirmary, Leeds, UK²Department of Paediatrics, Leeds General Infirmary, Leeds, UK³Department of Paediatric Surgery, Leeds General Infirmary, Leeds, UK**Correspondence to**

Dr Evie Alexandra Robson, ea.turner@virgin.net

Accepted 9 May 2014

SUMMARY

Vascular rings can present with non-specific respiratory and/or oesophageal symptoms. Early diagnosis requires a high index of suspicion. This case report describes an uncommon acute presentation of a vascular ring. We report a thriving 14-month-old child with a long history of recurrent wheeze and 'noisy breathing'. He presented acutely with food bolus impaction in the oesophagus which led to a respiratory arrest. Oesophagoscopy and bronchoscopy suggested vascular ring anomaly. A contrast-enhanced CT scan demonstrated a right-sided aortic arch with left ligamentum arteriosum encircling the oesophagus and airway. The ligament was ligated and divided. At follow-up 6 months later, the infant had mild persistent stridor but was otherwise well.

BACKGROUND

We have produced this case report as it highlights a very unusual presentation of a vascular ring.

This infant had a significant history of chronic respiratory symptoms, which appeared refractory to treatment. At presentation to hospital with a respiratory arrest, the diagnosis was initially missed.

We feel this is an important case to alert a paediatrician to have a high index of suspicion in order to detect a vascular ring, and to also highlight the importance of repeating diagnostic investigations, including bronchoscopy, oesophagoscopy and contrast-enhanced CT.

CASE PRESENTATION

A 14-month-old toddler was admitted to hospital following a respiratory arrest at home. After eating a piece of sausage, he appeared to choke, with sudden respiratory distress. He became very wheezy, then cyanotic and apnoeic. His mother gave back slaps while awaiting the ambulance. On admission to hospital he was unresponsive with minimal respiratory effort and significant wheeze and hyperinflation. Following resuscitation the child underwent emergency rigid bronchoscopy for a suspected inhaled foreign body. No foreign body was found. A CT scan was then performed because of concern that a foreign body might have migrated distally. This scan suggested narrowing of the trachea and the child was referred to us for further evaluation.

Review of the medical history revealed 'noisy breathing' from birth, which was described as inspiratory and expiratory in nature, associated with intercostal recession. A diagnosis of laryngomalacia was made at one stage. The infant had been admitted on two occasions with bronchiolitis, requiring continuous positive airway pressure respiratory support. Following these episodes

inhaled seretide, salbutamol, atrovent and montelukast were started. The mother felt that these did not alter symptoms although the parents felt there was a good response to oral steroids. He was thriving, 'loved his food' and was particularly fond of sausages.

Mechanical ventilation proved difficult because of a persistent wheeze, despite intravenous aminophylline and steroids. Repeat bronchoscopy was performed which revealed a segment of tracheomalacia involving the distal trachea, with normal upper trachea, carina and main bronchi. Oesophagoscopy was then performed and a large piece of sausage was seen in the mid oesophagus. The oesophagus appeared angulated at this level which made passage of a rigid oesophagoscope extremely difficult. Eventually, the food bolus was pushed into the stomach with a fiberoptic gastroscope.

The combination of segmental tracheomalacia and deviation of the oesophagus suggested a vascular ring. Following removal of the foreign body a further CT scan was performed with intravenous contrast ([figure 1](#)). This revealed narrowing of the distal trachea to 6 mm secondary to a vascular ring formed by a right-sided aortic arch and a persistent left ligamentum arteriosum, with mirror image branching of the great vessels arising from the aortic arch. An echocardiogram ruled out further cardiac anomalies.

The vascular ring was divided through a left posterolateral thoracotomy. The ductus arteriosus was ligated and divided, releasing the ring around the trachea and oesophagus. The patient was extubated onto vapotherm within 4 days. He was discharged home 1 week later.

OUTCOME AND FOLLOW-UP

At follow-up, 6 months after surgery, mild inspiratory stridor was still audible but the child was otherwise well and on no regular medication. His appetite was undiminished.

DISCUSSION

Vascular rings are congenital anomalies of the aortic arch. They result from abnormal regression of parts of the branchial arch system during fetal life. The result can be complete or incomplete encirclement of the trachea, oesophagus or both. The term was first introduced in 1945 by Gross, who performed the first successful operation on a child with a double aortic arch. Fray first described a vascular ring caused by a left-sided ligamentum arteriosum and a right-sided aortic arch in 1936.

Aortic arch anomalies account for 1–2% of all cardiac anomalies, with approximately 1% due to a

CrossMark

To cite: Robson EA, Scott A, Chetcuti P, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-202164

Figure 1 Images from the intravenous contrast-enhanced CT scan showing narrowing of distal trachea by a vascular ring formed from a right-sided aortic arch and persistent left ligamentum arteriosum.

vascular ring. Other cardiovascular anomalies have been found in up to 63% of cases.^{1 2} Vascular rings are often associated with tracheal anomalies.

Klinkhamer and Stewart classified vascular rings according to embryological, pathological and radiographic criteria. Complete vascular rings encircle the trachea and oesophagus. These include double aortic arch (type 1) and right aortic arch with persistent left ligamentum arteriosum (type 2). Type 3 involves a Kommerell diverticulum, in which a ring is formed by a remnant of an incompletely regressed left or right arch, from which the left or right subclavian originates, respectively.¹ Incomplete rings formed from other vascular anomalies can cause compression but are often asymptomatic and found incidentally.

Most published reports are of observational case series of children treated in tertiary centres with surgical repair. Therefore, complete vascular rings, which are associated with more severe symptoms, represent the majority of cases. A right-sided aortic arch and left ligamentum is almost as frequent as a double aortic arch, but presentation is often later, during the first few years of life.²

Vascular rings are typically associated with non-specific respiratory and oesophageal symptoms. The severity of the symptoms and the age at diagnosis usually depend on the tightness of the vascular ring. Some children will have significant respiratory symptoms from birth, whereas others may be asymptomatic.¹ Respiratory symptoms are more common including stridor (63%), recurrent respiratory infections (47%), respiratory distress (19%) and cough (17%). Oesophageal symptoms included dysphagia (27%), reflux (6%), choking (3%) and failure to thrive (3%). Although presentation with stridor and respiratory infections appears similar for double aortic arch and right aortic arch with ligamentum, children with the latter seem to present more often with cough and dysphagia (33% and 37%, respectively).² Symptom duration is often overlooked and a high index of suspicion is required for diagnosis.

Acute presentation of a vascular ring following foreign body obstruction in the oesophagus is surprisingly unusual. Previous reports are summarised in table 1. Our case appeared to have a respiratory arrest associated with choking and severe wheeze due to the obstruction caused by food bolus impaction in the oesophagus at the level of a vascular ring, which was already causing significant ongoing asthma-like symptoms.

Anteroposterior and lateral chest radiography can reveal abnormalities including a right-sided aortic arch, tracheal displacement and posterior impression of the oesophagus.² Barium oesophagography is a simple and reliable investigation to identify or exclude a vascular ring.³

Bronchoscopy is an important investigation if tracheal obstruction is suspected. Airway endoscopy will determine the site and severity of the obstruction and identify other pathology including bronchomalacia or tracheomalacia. It is useful to document vocal cord movement prior to surgery, as vocal cord paralysis is a recognised complication of vascular ring surgery.²

Biplanar angiography has historically been the gold standard approach to delineate anatomy of vascular rings. More recently, CT and MRI have become the procedures of choice for definitive evaluation. The detail obtained by contrast-enhanced CT allows planning for surgery and assessment of airway compression.² The imaging modality of choice depends on local expertise and equipment availability.

An echocardiogram should be performed to clarify anatomy, exclude other cardiac anomalies and facilitate intraoperative management.

MANAGEMENT

Management of a child with a symptomatic vascular ring involves surgical correction. The safety and effectiveness of this has been confirmed by many authors. Early correction decreases morbidity

Table 1 Case reports of diagnosis of vascular rings following oesophageal foreign body obstruction

Article	History	Investigations	Diagnosis	Outcome
Pumberger <i>et al</i> ⁴	A 2-year-old girl presented following ingestion of sausage which led to excessive drooling, vomiting, dysphagia and respiratory difficulty History of frequent respiratory tract infections, dysphagia, vomiting, regurgitation, failure to thrive, no stridor or respiratory distress	Foreign body removed during oesophagoscopy Barium study and MRI with angiography performed due to history	Right descending aorta with a ligamentum arteriosum and mirror branching	Postoperatively oesophageal symptoms ceased A 5-year follow-up revealed reduction in respiratory symptoms
Swischuk ⁶	A 3-year-old boy swallowed a penny. Presented with slightly hoarse voice but examination normal Previous similar episode aged 6 months	AP and lateral chest X-rays (CXR) showed penny lodged above aortic knob and anterior displacement of trachea. CXR postremoval showed tracheal deviation and indentation to left. Barium swallow and MRI also performed	Right aortic arch with aberrant left subclavian artery	No surgical intervention
O'Connor and Cooney ⁷	An 8-month-old boy with a history of choking episode 2 days prior to presentation with wheeze, biphasic stridor and cough, but with no swallowing difficulties History revealed subtle respiratory symptoms over last 6 weeks	Rigid bronchoscopy showed significant tracheomalacia and posterior tracheal compression. Rigid oesophagoscopy revealed metallic foreign body in upper oesophagus. CT also performed	Double aortic arch	Discharged postforeign-body removal. Symptoms returned Repeat bronchoscopy showed residual posterolateral tracheal compression
Currarino and Nikaidoh ⁸	An 11-month-old presented following ingestion of earring History revealed dysphagia with solid feeds but no respiratory symptoms A 11-month-old with a history of mild inspiratory stridor since birth Presentation of recent croup like illness with choking and respiratory distress related to feeds A 17-month-old presented with a 2 day history of cough, dyspnoea gagging, difficulty with secretions and low-grade fever A 3-year-old, ingested a coin 2 days prior to presentation with cough, vomiting and low-grade fever	Lateral CXR showed round foreign body and trachea bowed forward. Barium swallow showed extrinsic defect of posterior oesophageal wall and narrowing of trachea Lateral neck X-ray revealed round foreign body in proximal oesophagus. Barium oesophogram showed large extrinsic defect in posterior wall of oesophagus and slight narrowing of trachea at aortic arch level	Right aortic arch	Symptoms resolved by repair 5 years later
		CXR showed coin in proximal oesophagus. Oesophagoscopy revealed corroded oesophagus. Barium oesophagram showed extrinsic deformity of aortic arch	Double aortic arch with small patent left arch	Dysphagia resolved Mild stridor for few months postrepair
		AP and lateral CXR showed coin in proximal oesophagus and long segment of tracheal narrowing and anterior displacement. Oesophagoscopy revealed large erosion. Barium oesophagram showed extrinsic defect in posterior wall of oesophagus	Aberrant right subclavian artery Aberrant right subclavian artery	Asymptomatic Pseudodiverticulum 1 year later Asymptomatic Pseudodiverticulum 8 months later

and allows for normal tracheal growth. Complications of surgery include vocal cord paralysis, pneumothorax and chylothorax.³

Respiratory function has been found to normalise in 90% of children during postoperative follow-up.⁴ Relief of preoperative symptoms is reported in 43% of children immediately after surgery, and a further 57% within 4 years.¹ Eklof *et al* showed that persistent symptoms appear to include mild stridor and/or dysphagia (6/30 patients) at 5-year follow-up.⁵

Learning points

- ▶ Bolus obstruction in the oesophagus is a very unusual presentation of vascular ring. A high index of suspicion and detailed background history are required.
- ▶ A finding of a foreign body in the oesophagus, in the context of more chronic symptoms, may alert a team to the possibility of a vascular ring.
- ▶ If one suspects a foreign body has been aspirated then bronchoscopy is mandatory. If bronchoscopy is normal then oesophagoscopy must be considered.
- ▶ CT without contrast will miss a vascular ring.

CONCLUSION

Vascular rings are rare and diagnosis can be difficult and delayed in infants who present with non-specific respiratory symptoms. Our case is unusual because of the acute presentation following a food bolus obstruction in the oesophagus.

This case highlights the value of repeat diagnostic investigations including bronchoscopy and CT with contrast to establish the diagnosis.

Acknowledgements The authors would like to thank Dr N Tahir (Consultant Radiologist) and Dr H Woodley (Consultant Radiologist) who have contributed radiology images for use in the report.

Contributors EAR contributed to the planning, conduct and reporting of the article, and was also involved in writing the case report, discussion, investigations and management sections of the case report. AS has contributed to the planning and conduct, writing the case report and producing table 1. PC has contributed to the conduct of work in the article and reviewed the completed text. DC has contributed to the conduct and reporting of the article, provided bronchoscopy images and reviewed the completed text.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Bakker DA, Berger RM, Witsenburg M, *et al*. Vascular rings: a rare cause of common respiratory symptoms. *Acta Paediatr* 1999;88:947–52.

- 2 Shah RK, Mora BN, Bacha E, *et al.* The presentation and management of vascular rings: an otolaryngology perspective. *Int J Pediatr Otorhinolaryngol* 2007; 71:57–62.
- 3 O'Connor TE, Cooney T. Oesophageal foreign body and a double aortic arch: rare dual pathology. *J Laryngol Otol* 2009;123:1404–6.
- 4 Pumberger W, Voitl P, Göpfrich H. Recurrent respiratory tract infections and dysphagia in a child with an aortic vascular ring. *South Med J* 2002; 95:265–8.
- 5 Eklöf O, Ekström G, Eriksson BO, *et al.* Arterial anomalies causing compression of the trachea and/or the oesophagus. A report of 30 symptomatic cases. *Acta Paediatr Scand* 1971;60:81–9.
- 6 Swischuk LE. Swallowed penny. *Pediatr Emerg Care* 2003;19:59–61.
- 7 O'Connor TE, Cooney T. Oesophageal foreign body and a double aortic arch: rare dual pathology. *J Laryngol Otol* 2009;123:1404–6.
- 8 Currarino G, Nikaidoh H. Esophageal foreign bodies in children with vascular ring or aberrant right subclavian artery: coincidence or causation? *Pediatr Radiol* 1991;21:406–8.

Copyright 2014 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.

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