CASE REPORT

Scimitar syndrome with absent right pulmonary artery and severe pulmonary hypertension treated with coil occlusion of aortopulmonary collaterals in a term neonate

Hussain Parappil,^{1,2} Faraz Masud,³ Husam Salama,^{1,2} Sajjad ur Rahman^{1,2}

¹Department of Neonatology, Hamad Medical Corporation, Doha, Qatar

²Department of Pediatrics, Weill Cornell Medical College, Doha, Qatar

³Department of Pediatric Cardiology, Hamad Medical Corporation, Doha, Qatar

Correspondence to

Dr Hussain Parappil,
drhussainparappil@gmail.com

Accepted 30 January 2015

SUMMARY

Scimitar syndrome (SS) is a rare congenital malformation with an estimated incidence of approximately 2 in 100 000 births. A wide clinical spectrum is observed in children with this syndrome. The common clinical presentation in infancy is respiratory distress and tachypnoea due to associated pulmonary hypoplasia, pulmonary overcirculation and/or pulmonary hypertension. Babies with SS presenting with cardiac failure are prone to develop exaggerated pulmonary vascular disease. Hence early intervention, using either coil embolisation or surgical intervention, is indicated. We are reporting a case of a term baby boy who presented with respiratory failure during the first 24 h of life. Echocardiogram and CT angiogram revealed SS. The baby needed intubation due to respiratory failure. Aortopulmonary collaterals, identified on aortic angiogram, were successfully occluded with detachable coils.

BACKGROUND

Scimitar syndrome (SS) is a rare congenital malformation with an estimated incidence of approximately 2 in 100 000 births.¹ SS, first described in 1836, consists of anomalous pulmonary venous drainage from the right lung to the inferior vena cava.² There are often a constellation of findings associated with SS including hypoplasia of the right lung, pulmonary hypertension, dextroposition of the heart and anomalous systemic arterial supply (ASAS) with or without pulmonary sequestration.³

A wide clinical spectrum is observed in children with this syndrome. A classification has been proposed according to the age of presentation and the associated congenital cardiac malformations.⁴ The 'infantile' form, diagnosed in children younger than 1 year of age, is characterised by cardiorespiratory symptoms, pulmonary hypertension and severe adverse outcome. Conversely, the 'adult' form remains asymptomatic for many years and has a much more favourable outcome.¹

The common clinical presentation in infancy is respiratory distress and tachypnoea due to associated pulmonary hypoplasia, pulmonary overcirculation and/or pulmonary hypertension. The triad of respiratory distress, right lung hypoplasia and dextroposition of the heart should raise the suspicion of this syndrome.⁵

Surgical correction is recommended for patients with a pulmonary-to-systemic blood flow ratio

exceeding 2:1 because of the risk of development of pulmonary hypertension and right ventricular failure.⁶ Surgical intervention for patients with symptomatic SS can result in early as well as late surgical complications that can be frequent and devastating.⁷ The use of amplatzer vascular plugs (AVP) and tornado coils to occlude the ASAS potentially prevents the need for surgical intervention in carefully selected patients with SS.⁸ In this article, we report a case of neonatal SS that showed significant clinical improvement after successful coiling of the aortopulmonary collaterals.

CASE PRESENTATION

A term baby boy was born by normal vaginal delivery to a 21-year-old primigravida mother. The baby had a birth weight of 3200 g and APGAR scores of 9 and 10 at 5 and 10 min, respectively. The pregnancy and delivery were uneventful. The baby was admitted to neonatal intensive care unit at 24 h of life from the postnatal ward due to tachypnoea. He required respiratory support with nasal cannula initially for the first 2 days and then ventilation due to increasing oxygen requirements, chest retractions and grunting. He had right-sided heart failure. The chest X-ray showed (figure 1) radiopaque right hemithorax with mediastinal shift and dextroposition of the heart.

Two-dimensional echocardiography revealed total absence of right pulmonary artery, non-visualised

Figure 1 Chest X-ray showing radiopaque right hemithorax with right mediastinal shift and dextroposition of the heart. The left lung looks well expanded and perfused.

CrossMark

To cite: Parappil H, Masud F, Salama H, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-208743

Figure 2 Pulmonary CT showing loss of volume of the right lung with absent right upper lobe and abrupt termination of the right upper lobe bronchus.

right pulmonary veins and their drainage. However, the left pulmonary veins were draining normally into the left atrium; patent foramen ovale and small patent ductus arteriosus with bidirectional shunt were seen along with dilated right atrium and right ventricle, with depressed contractility and mild tricuspid regurgitation with right ventricular systolic pressures equalising systolic systemic pressures. These findings led to a strong suspicion for SS. An urgent CT (figures 2–4) angiogram was performed showing absent right upper lobe and right upper lobe bronchus as well as absent right pulmonary artery (figure 5). Blood supply to the right lung originated from the lower thoracic aorta. Right pulmonary veins were seen draining into inferior vena cava denoting SS.

The baby's complete blood count, serum electrolytes, blood culture, chromosomal study and microarray were normal. His brain and abdominal ultrasound and eye examination did not reveal any abnormality.

The baby was treated with frusemide, milrinone and respiratory support. At the age of 9 days, cardiac catheterisation was performed for haemodynamic studies and occlusion of aortopulmonary collaterals. Two major aortopulmonary collaterals were identified on aortic angiogram (figure 6) supplying the entire right lung; both collaterals were successfully occluded by

Figure 3 CT angiogram two-dimensional (2D) showing the right-sided pulmonary vein draining into the inferior vena cava close to its entry into right atrium.

Figure 4 CT angiogram three-dimensional reconstructed showing the right-sided pulmonary vein draining into the inferior vena cava close to its entry into right atrium.

using detachable coils (figure 7) with immediate decrease in Qp/Qs from 3.4 to 0.9, and drop in suprasystemic pulmonary artery pressures to 68% of systolic systemic pressures.

The general condition of the baby improved after occlusion by the coil. The baby was extubated to continuous positive airway pressure and then to nasal cannula in room air over the next 2 days. Feeding was started and gradually increased to full feeds in a week's time. The baby was discharged home at the age of 20 days. He had a first follow-up at 6 weeks of age and was again seen, with acceptable haemodynamics, in the outpatient ward at 4 months of age.

INVESTIGATIONS

- ▶ Echocardiogram
- ▶ CT angiogram
- ▶ Cardiac catheterisation

DIFFERENTIAL DIAGNOSIS

- ▶ Pulmonary hypoplasia
- ▶ Atelectasis of the right lung
- ▶ SS

Figure 5 Angiogram during cardiac catheterisation showing good-sized left pulmonary artery and absent right pulmonary artery.

Figure 6 Aortic angiogram (anteroposterior view) showing two major aortopulmonary collaterals arising from the descending aorta and supplying the right lung.

TREATMENT

The use of AVP and tornado coils by transcatheter intervention to occlude the aortopulmonary collaterals would be the treatment of choice over surgical intervention.

OUTCOME AND FOLLOW-UP

Our patient was ventilator dependent and on antifailure treatment. The baby showed significant clinical improvement after successful coiling of the aortopulmonary collaterals and he was extubated in 2 days.

DISCUSSION

SS is a rare type of partial anomalous pulmonary venous connection with a female preponderance of 2:1.¹ It almost always involves the right lung and its classical findings are characterised by right pulmonary vein(s) joining the inferior vena cava, hypoplasia of the right lung and bronchial anomalies (stenosis or ectasia; atresia or abnormal segmentation or branches),

Figure 7 Postcoiling angiogram showing occlusion of major aortopulmonary collateral supplying the right lung.

dextroposition of the heart (depending on the degree of hypoplasia of the right lung), anomalous arterial connection to the right lung from the aorta and hypoplasia of the right pulmonary artery,⁹ rarely with its complete absence,^{10 11} as seen in our case. Very rarely SS of the left lung has been reported.¹²

The typically coined name, Scimitar syndrome, was first described by Naill in 1960,¹³ depicting the radiological appearance of the anomalous vein appearing as tubular opacity along the right heart border resembling a curved Turkish Sword, the scimitar sign. However, this sign is usually absent in neonates.¹⁴ It has been recognised by different names including hypogenetic lung syndrome, Halasz's syndrome, congenital pulmonary veno-lobar syndrome, mirror image lung syndrome or vena cava bronchovascular syndrome.

The clinical symptomatology of respiratory and cardiac failure in the infantile type is most commonly caused by pulmonary hypertension secondary to cardiac and lung anomalies. Pulmonary hypertension holds a considerably poor prognosis during infancy,^{4 14} and is attributed to multiple factors,^{4 6} including stenosis of the scimitar vein, presence of systemic arterial supply to the right lung, reduction of the pulmonary vascular bed on the right side and increased pulmonary blood flow resulting from anomalous drainage or from the presence of some associated intra cardiac defect. In our case, there were no intra cardiac defects, but the shunt clearly contributed significantly to the patient's pulmonary hypertension and clinical presentation. After aortopulmonary collateral embolisation, there was a significant improvement in pulmonary artery pressures, proving its volume-induced mechanism of pulmonary hypertension. There may be still a vascular pathology associated with the left lung.

The management of symptomatic infants with SS is complex. Surgical intervention is indicated in the presence of associated severe cardiac malformations, lung sequestration and recurrent right-sided infections. Surgery usually involves partial or total resection of the right lung, redirection of the Scimitar vein into the left atrium, correction of the intra cardiac defects and division of the abnormal arterial supply to the right lung.¹⁵ Recently, coil embolisation of the ASAS has been reported as an alternative to surgical ligation in symptomatic infants before cardiac surgery,^{16 17} or to control infection in patients with pulmonary sequestration. Coil embolisation thus offers the best hope for a good clinical outcome in symptomatic infantile patients (although survival was still approximately 50%).⁴

In conclusion, early intervention with coil embolisation is an important therapeutic option for symptomatic infantile patients with SS especially when a significant left to right shunt contributes to the patient's clinical status.

Learning points

- ▶ Early transcatheter (or surgical) intervention to eliminate aortopulmonary collateral flow should be considered in symptomatic infants with Scimitar syndrome.
- ▶ Coil embolisation of the anomalous systemic arterial supply is an alternative to surgical ligation in symptomatic infants.
- ▶ Absence of the right pulmonary artery is a rare anomaly associated with Scimitar syndrome.

Acknowledgements The authors are grateful to Dr Ashraf Mansour (Consultant Neonatologist), Dr Sabry Nasr (Specialist Neonatologist) and Dr Hesham Al-Saloos

(Sr Consultant Cardiologist) for their involvement in the management of the baby and their support in writing this paper.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- Gudjonsson U, Brown JW. Scimitar syndrome. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2006;9:56–62.
- Cooper G. Case of malformation of the thoracic viscera consisting of imperfect development of the right lung and transposition of the heart. *London Med Gaz* 1836;18:600–1.
- Brown JW, Ruzmetov M, Minnich DJ, *et al.* Surgical management of Scimitar syndrome: an alternative approach. *J Thorac Cardiovasc Surg* 2003;125:238–45.
- Dupuis C, Charaf LA, Breviere GM, *et al.* 'Infantile' form of the Scimitar syndrome with pulmonary hypertension. *Am J Cardiol* 1993;71:1326–30.
- Midyat L, Demir E, Askin M, *et al.* Eponym. Scimitar syndrome. *Eur J Pediatr* 2010;169:1171–7.
- Najm HK, Williams WG, Coles JG, *et al.* Scimitar syndrome: twenty years' experience and results of repair. *J Thorac Cardiovasc Surg* 1996;112:1161–9.
- Mok Q, Ross-Russell R, Mulvey D, *et al.* Phrenic nerve injury in infants and children undergoing cardiac surgery. *Br Heart J* 1991;65:287–92.
- Weems C, Peuster M, Trivedi K. One plug may not be enough: a novel technique for the occlusion of high-flow vascular connections: combined AGA vascular plug II and coil occlusion of a sequester artery in a patient with Scimitar syndrome. *Catheter Cardiovasc Interv* 2011;78:687–91.
- Hayashi S, Hirai S, Naito M, *et al.* Scimitar vein descending from the right inferior lobe to the inferior vena cava beneath the diaphragm: a rare case. *Anat Sci Int* 2014. Published Online First: 15 March 2014. doi 10.1007/s12565-014-0230-5
- Muktachand L, Rokade RV, Devdas S, *et al.* Scimitar syndrome. *Ind J Paediatr* 2005;72:245–7.
- Patel AM, Joshi R, Vaghela D, *et al.* A face of Scimitar syndrome. *Ind J Radiol Imag* 2004;14:401–4.
- Rutledge JM, Hiatt PW, Wesley Vick G III, *et al.* A sword for the left hand: an unusual case of left sided Scimitar syndrome. *Paediatr Cardiol* 2001;22:350–2.
- Neill CA, Ferencs C, Sabiston DC. The familial occurrence of hypoplastic right lung with systemic arterial supply and venous drainage "Scimitar syndrome". *Bull Johns Hopkins Hosp* 1960;107:1–21.
- Canter CE, Martin TC, Spray TL, *et al.* Scimitar syndrome in childhood. *Am J Cardiol* 1986;58:652–4.
- Shinohara G, Morita K, Uno K, *et al.* Scimitar syndrome in an infant with right lung hypoplasia, ventricular septal defect and severe pulmonary hypertension. *Gen Thorac Cardiovasc Surg* 2010;58:524–7.
- Muta H, Akagi T, Lemura M, *et al.* Coil occlusion of aortopulmonary collateral arteries in an infant with Scimitar syndrome. *Jpn Circ J* 1999;63:29–31.
- Sinan S, Ahmet C, Yalim Y, *et al.* Embolization of the systemic arterial supply via a detachable silicon balloon in a child with scimitar syndrome. *Cardiovasc Intervent Radiol* 2005;28:249–53.

Copyright 2015 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