CASE REPORT

Adult Form of Scimitar Syndrome Presenting as Severe Pulmonary Hypertension in a Child

SIVASAMBO KALPANA, SARATH BALAJI B, SELLADURAI ELILARASI AND *NEVILLE AG SOLOMON

*From Department of Pulmonology, Institute of Child Health and Hospital for Children; and *Department of Pediatric Cardiac Surgery, Apollo Children's Hospital; Chennai, India.*

Correspondence to:
Dr Sivasambo Kalpana,
Department of Pulmonology,
Institute of Child Health and Hospital for Children, Halls road, Egmore, Chennai.
drskalpana@yahoo.co.in
Received: February 21, 2015;
Initial review: April 14, 2015;
Accepted: May 29, 2015.

Background: Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies; the adult form is not usually is associated with pulmonary hypertension. **Case characteristics:** 6-year-old girl with recurrent episodes of cough and breathlessness, along with features of right heart enlargement. Computed tomography angiogram revealed right pulmonary veins draining into inferior vena cava with dextroposition of heart. **Outcome:** Successfully managed with surgical correction. **Message:** Scimitar syndrome should be considered in any child with unexplained pulmonary hypertension and dextroposed heart.

Keywords: *Congestive cardiac failure, Dextroposition, Pulmonary vein anomalies*.

Scimitar syndrome has a varied presentation from an asymptomatic state to severe pulmonary hypertension and/or heart failure. Pulmonary arterial hypertension (PAH) is an uncommon presentation of this syndrome beyond infancy.

CASE REPORT

A 6-year-old developmentally normal girl was referred to our department for evaluation of uncontrolled wheeze. There was history of recurrent episodes of breathlessness and cough since 2 years of age and exercise intolerance since 4 years of age. She had been treated with intermittent salbutamol nebulizations and oral antibiotics, with partial response. On admission, the child had no pallor, cyanosis or clubbing. Significant respiratory distress was present with bilateral severe wheeze; oxygen saturation in room air was 92 % that improved to 94% with 100% oxygen. There was a bulge on right side of chest wall with heart sounds heard well on the right; P2 was loud and heart rate was 110/min. There was no murmur. With the above clinical picture, a provisional diagnosis of Idiopathic pulmonary arterial hypertension with congestive cardiac failure was made.

Chest *X*-ray showed right sided cardiomegaly with pulmonary congestion. Electrocardiogram (ECG) demonstrated right axis deviation with right atrial enlargement. Echocardiogram revealed severe pulmonary hypertension (mean pulmonary artery systolic pressure 58 mmHg), and dilated right atrium and ventricle with a stretched patent foramen ovale. Serum N-terminal prohormone of brain natriuretic peptide was slightly raised to 144 pg/mL (normal 125 pg/mL). Computed tomography (CT) angiogram demonstrated that right pulmonary veins joined to form a single vein which was draining into the inferior vena cava (IVC) with dextroposition of heart (***Fig.* 1**). The drainage of the left pulmonary veins was normal. A persistent left superior vena cava (SVC) and atrial septal defect (ASD) were also demonstrated. The diagnosis of adult form of Scimitar syndrome presenting with severe PAH was made, based on the clinical presentation and CT angiogram findings. The child underwent intracardiac repair in view of the severe symptoms and PAH, using right posterolateral

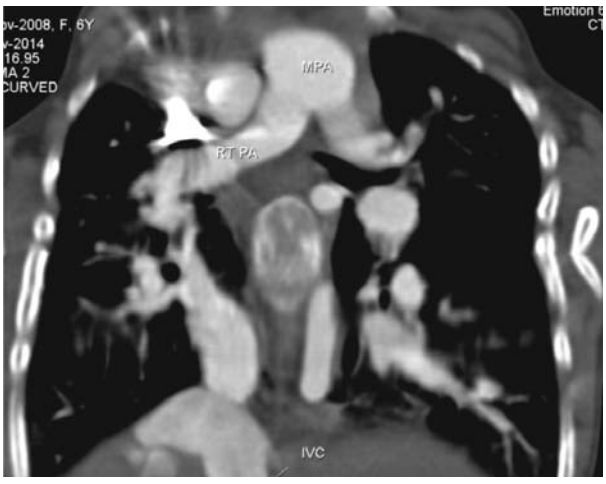

FIG. 1 *CT angiogram showing right pulmonary veins joining to form a single vein which is draining into the inferioer vna cava.*

thoracotomy. The right pulmonary vein was dissected and reimplanted onto the left atrium (LA) using a autologous pericardial patch, and closure of ASD was done. Intra- and post- operative course of the child was uneventful.

DISCUSSION

The hallmark of scimitar syndrome is an anomalous right pulmonary vein that drains part or the entire right lung into the IVC. Associated anomalies include hypoplasia of the right lung, dextroposition of the heart, hypoplasia of the right pulmonary artery and anomalous systemic arterial supply from the aorta to the right lung. It has three main forms: an infantile form with severe symptoms and pulmonary hypertension, an adult form distinguished by being asymptomatic in infancy, and a third form with associated congenital cardiac anomalies [1]. The diagnosis may be difficult, especially in children and young adults with concomitant congenital heart lesions. CT angiogram helped to clinch the diagnosis in this child and appears to be an essential investigation in all children with unexplained pulmonary hypertension, as echo-cardiography may miss the diagnosis (as in this case) [2].

Pulmonary artery hypertension is seen mostly in infants with associated congenital heart malformations or with an anomalous large systemic arterial supply to the right lung [3]. A less common cause of pulmonary artery hypertension is the presence of Scimitar vein stenosis. Although our case did not have significant hypoplasia or systemic arterial supply to the right lung, intraoperative findings demonstrated significant stenosis at entry of the scimitar vein into the IVC. This finding may explain the early presentation of this child with severe pulmonary hypertension [4]. Unrelenting pulmonary hypertension can cause irreversible damage to the pulmonary vascular bed and lead to eventual right heart failure as in this child. Associated cardiac anomalies frequently determine the clinical course of these patients. About 70% of patients with scimitar syndrome have an associated ASD. The presence of persistent left SVC, as in our patient, has been described as a rare association [5].

Indication for surgical correction is based on the severity of symptoms, pulmonary over-circulation, right ventricular dilatation and concomitant cardiac lesions [6]. The surgical repair of Scimitar syndrome consists of redirecting the pulmonary venous drainage into the LA, either baffling the anomalous drainage into the LA via a tunnel or transecting the "scimitar drainage" near its entrance into the IVC and then reimplanting it directly into the LA. The majority of patients show good clinical outcome on follow-up. Scimitar vein stenosis may occur in about 15.5% at 10 years follow- up, irrespective of surgical approach [7].

Contributors: KS: case management and writing the manuscript; SBB and SE : case management and critical appraisal of manuscript; NAGS: in case management. All authors approved the final manuscript.
Funding: None; *Competing interests*: None stated.

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