An Unusual Etiology of Infantile Hemoptysis

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Figure 1. (A) Chest radiograph showing mild rightward mediastinal shift with mildly decreased right lung volumes. Bilateral peribronchial thickening and patchy airspace opacities, and a suggestion of a retrocardiac scimitar vein (*arrow*), are also seen. (B) Coronal multiplanar reconstruction from computerized tomography scan with angiogram performed via a lower extremity injection, showing contrast within the lumen of the superior scimitar vein with filling defect inferiorly (*arrow*). The lack of reflux of contrast into the inferior scimitar vein despite pressure injection into the inferior vena cava also implied obstruction at the scimitar vein/inferior vena cava junction. The enhancement pattern in the superior scimitar vein (with less-dense contrast than in the left pulmonary veins) is thought to be due to the presence of downstream venous obstruction. Also seen is a hypoplastic right pulmonary artery. (C) Image from bronchoscopy showing blood welling up (*lower right portion of the image*) into the airway. (D) Anteroposterior images from cardiac catheterization. Right pulmonary artery angiogram (*left panel*) reveals normal arborization and pulmonary artery size without dilatation or tortuosity. Right pulmonary artery wedge angiogram (*right panel*) reveals preserved capillary blush in the right lower lung with tortuosity of abnormal venous collaterals and obstruction of the scimitar vein approximately 2 cm above the diaphragm (*arrow*). This correlated well with the appearances of the scimitar vein demonstrated on the computerized tomography scan with angiogram (B).

A 10-month-old female presented for evaluation of hemoptysis. She was initially crying and choking in bed and coughed up a nickel-sized amount of dark red blood on the bedsheet, with three to four additional episodes ranging from blood-streaked mucus to frank blood, including one witnessed at an outside facility. The family was adamant that these episodes were hemoptysis and not hematemesis. Examination revealed a petite infant with a systolic 2/6 ejection murmur but without respiratory distress, cyanosis, or clubbing. Hemoglobin was 10.4 g/dl and stable. Initial chest radiograph (Figure 1A) demonstrated mild rightward mediastinal shift with mildly decreased right lung volumes, bilateral peribronchial thickening, patchy airspace opacities, and, in retrospect, a suggestion of a retrocardiac scimitar vein (*arrow*). A computerized tomography scan with angiogram demonstrated right lung hypoplasia with anomalous right pulmonary venous drainage via an obstructed-appearing scimitar vein into the inferior vena cava (Figure 1B, *arrow*). Echocardiogram confirmed abnormal right-sided pulmonary venous drainage, normal left-sided pulmonary venous drainage, a ventricular septal defect, a small right pulmonary artery, and mild septal flattening. Bronchoscopy revealed edematous airways with clear secretions, and markedly erythematous and edematous right-sided bronchi (Figure 1C), and, after bronchoalveolar lavage in the right middle lobe, blood welled up into the laryngeal mask airway, and an endotracheal tube was placed. Nasal wash was positive for parainfluenza type 3. Cardiac catheterization revealed an occluded scimitar vein (Figure 1D, *arrow*), three-fourths systemic pulmonary hypertension, and a ventricular septal defect.

She underwent successful surgical graft placement from the scimitar vein to the left atrium, closure of the ventricular septal defect, creation of an atrial septal fenestration, and medical therapy with sildenafil. This case was previously presented in abstract form (1).

In a review of children with scimitar syndrome, only 18–47% of children had pulmonary hypertension (2, 3), and these patients were younger, more symptomatic, and had worse outcomes (3). Hemoptysis is an unusual presentation of scimitar syndrome (only 6% of all patients in a series) (4). In this case, an occluded scimitar vein led to pulmonary venous obstruction and pulmonary hypertension. A parainfluenza infection likely led to increased inflammation and exacerbated the underlying pulmonary hypertension, leading to hemoptysis. Pulmonary venous obstruction should always be considered as an etiology for hemoptysis. ■

Author disclosures are available with the text of this article at www.atsjournals.org.

References

1. Liptzin DR, Hoch HE, Abman, SH. "She coughed up blood": an unusual etiology of infantile hemoptysis. *Am J Respir Crit Care Med* 2015;191: A5480.
2. Vida VL, Padalino MA, Boccuzzo G, Tarja E, Berggren H, Carrel T, Ciçek S, Crupi G, Di Carlo D, Di Donato R, *et al*. Scimitar syndrome: a European Congenital Heart Surgeons Association (ECHSA) multicentric study. *Circulation* 2010;122:1159–1166.
3. Canter CE, Martin TC, Spray TL, Weldon CS, Strauss AW. Scimitar syndrome in childhood. *Am J Cardiol* 1986;58:652–654.
4. Dupuis C, Charaf LA, Brevière GM, Abou P, Rémy-Jardin M, Helmius G. The "adult" form of the scimitar syndrome. *Am J Cardiol* 1992;70: 502–507.