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Total Pulmonary Artery Atresia Associated with Abnormal Pulmonary Venous Drainage as a Rare Presentation of Scimitar Syndrome

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ABSTRACT

Scimitar syndrome or pulmonary venolobar syndrome is a rare, complex, and variable malformation of the right lung characterized by an abnormal right sided pulmonary drainage into the inferior vena cava, malformation of the right lung, abnormal arterial supply, and sometimes cardiac malformation. Despite the varying degrees of pulmonary hypoplasia and pulmonary artery hypertension, about half of the patients with scimitar syndrome are asymptomatic or mildly symptomatic when the diagnosis is made. Neonates have severe symptoms and worse prognosis while older children come to light because of recurrent respiratory infections, heart murmur, or an abnormal chest radiograph.

Introduction

Scimitar syndrome is a congenital cardiac malformation in which pulmonary flow from the right lung is partially or totally drained via a venous channel that joins the inferior vena cava (IVC). Dupuis *et al*. identified two types of scimitar syndrome: the infantile and adult forms. The infantile form has a severe presentation with major associated cardiac lesions, varying degrees of hypoplasia of the right pulmonary artery and right lung bronchial anomalies, dextroposition of the heart, pulmonary hypertension, and anomalous systemic supply to the right lung.[1] This syndrome has varied presentations, from an asymptomatic state[2] to severe pulmonary hypertension and/or heart failure.[3] Those who present early in life usually have associated congenital heart disease[4] also hemoptysis[5] and pulmonary arterial hypertension (PAH) [6,7] both are uncommon presenting complaints of this rare syndrome beyond infancy.

Case History

A 15-year-old boy referred to our center with the complaint of hemoptysis and cough from 1.5 years ago. He suffered from chronic cough without fever from 3 years old. On examination, the patient was comfortable and weak. Her pulse was 72 beats/minute and regular, blood pressure 110/70 mm Hg, and respiratory rate 27 breaths/minute. Respiratory system examination revealed reduced intensity of breath sounds on the right side. P2 sound was dominant. He had received a period of empiric therapy for tuberculosis infection without response to it.

Chest PA x-ray showed cardiac dextroposition associated with several costal and vertebral anomalies as fusion of posterior arches of second and third right and left ribs and bifid anterior arch of left third rib and butterfly appearance at third thoracic vertebrae (Figure 1).

CT angiography was performed by 64 multidetector CT (MDCT) and then reconstructed via MIP and VRT techniques, and showing dilated main pulmonary artery with normal-looking left pulmonary artery and hypoplastic right pulmonary artery, a curved linear (scimitar shape) opacity running from the middle of the lung toward diaphragm. Left pulmonary vein ending into left atrium is well visualized; corresponding pulmonary vein on the right side was absent. Therefore, CT angiogram showed hypoplasia of right lung and anomalous drainage of right pulmonary veins by scimitar vein in to IVC below the diaphragm. Thickening of peribronchovascular and peripheral interstitium in favor of interstitial edema was noted (Figure 2).

Discussion

Scimitar syndrome is a rare congenital abnormality that occurs in approximately 2 of every 100 000 live births.[7] Dupuis and associates collected data from 22 university centers and demonstrated differences in patients whose diagnosis was made during the first year of life—dubbed the "infantile" form—compared with patients whose diagnosis was made after 1 year of age—called the "adult" form.[8] In contrast to more than 90% survival of those with the adult form, there was a considerably poorer prognosis for patients with the infantile form. Pulmonary hypertension is frequent in this subset of patients and is

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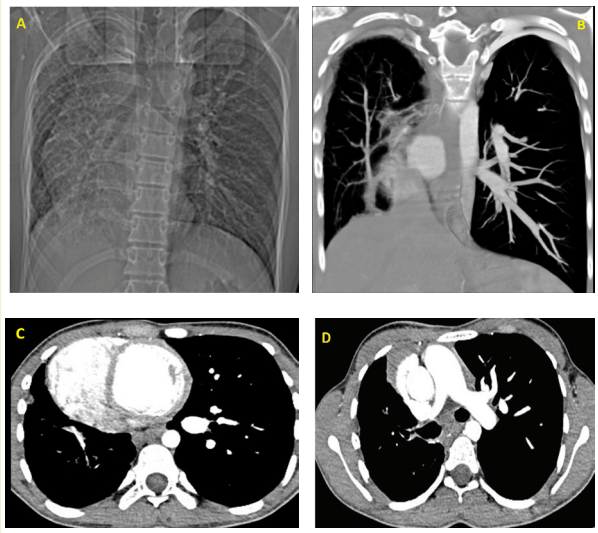

Figure 1. A) 15-year-old boy with hemoptysis and cough; A, scout view depicts cardiac dextroposition ; B, C, D) CT angiography images demanestrate hypoplastic right pulmonary artery, a curved linear (scimitar shape) opacity running from the middle of the lung toward diaphragm.

attributed to multiple factors [8,9]: 1) stenosis of the scimitar vein; 2) presence of systemic arterial supply to the right lung; 3) reduction of the pulmonary vascular bed on the right side; 4) increased pulmonary blood flow resulting from anomalous drainage or from the presence of some associated intracardiac defect. Hemoptysis as a presenting symptom is exceptionally rare in patients with scimitar syndrome.[10]

Among the five large series of patients with this syndrome comprising of total 39 subjects aged one year and above, only four has hemoptysis during their course of illness.[11,12] Possible mechanisms of hemoptysis in scimitar syndrome include rupture of hypertrophied systemic pulmonary anastomosis[13] or bleeding from some of the bronchiectatic segment in the hypoplastic lung. Clinical symptomatology

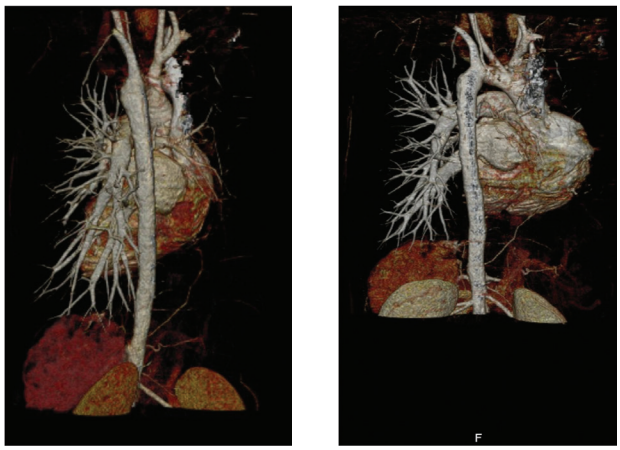

Figure 2. VRT images: Left pulmonary vein ending into left atrium is well visualized; corresponding pulmonary vein on the right side is absent.

of the scimitar syndrome is governed by the age at which the patient presents. Infants having scimitar syndrome present with cyanosis, poor growth, PAH and often complex cardiac defects, many of them need surgical intervention having a high mortality.[14,15] Disease in older children and adults commonly presents with recurrent pulmonary infections and/or exertional dyspnea. This group of the patients usually has a benign course.[5] Diagnosis of this syndrome is straightforward in presence of characteristic radiological sign (scimitar sign) on conventional chest radiography. However, when scimitar vein is masked by cardiac shadow, diagnosis can be documented by one or more traditional modalities, for example angiography[16], CT scan[17] and echocardiography.[18] Currently available MR technology also provides excellent visualization of vascular anatomy of this complex congenital defect noninvasively.

Ethical issues: This study was reviewed and confirmed by the ethics committee of Tabriz University of Medical Sciences.

Conflict of interests: The authors declare no conflicts of interest.

References

1. Canter CE, Martin TC, Spray TL, Weldon CS, Strauss AW. Scimitar syndrome in childhood. **Am J Cardiol** 1986; 58: 652-4.
2. Oakley D, Naik D, Verel D, Rajan S. Scimitar vein syndrome: report of nine new cases. **Am Heart J** 1984; 107: 596-8.
3. Gikonyo DK, Tandon R, Lucas RV Jr, Edwards JE. Scimitar syndrome in neonates: Report of four cases and review of the literature. **Pediatr Cardiol** 1986; 6:193-7
4. Gikonyo DK, Tandon R, Lucas RV Jr, Edwards JE. Scimitar syndrome in neonates: Report of four cases and review of the literature. **Pediatr Cardiol** 1986; 6:193-7.
5. Canter CE, Martin TC, Spray TL, Weldon CS, Strauss AW. Scimitar syndrome in childhood. **Am J Cardiol** 1986; 58:652-54.
6. Seaton D, Seaton A. Developmental disorders of the lungs. In: Seaton A, Seaton D, Leitcha G, eds. Crofton and Douglas's respiratory diseases. 5th ed. Oxford: Blackwell Science; 2000. pp. 1309-29.
7. Brown JW, Ruzmetov M, Minnich DJ, Vijay P, Edwards CA, Uhlig PN, et al. Surgical management of scimitar syndrome: an alternative approach. **J Thorac Cardiovasc Surg** 2003; 125:238-45.
8. Dupuis C, Charaf LA, Breviere G, Abou P. Infantile form of the scimitar syndrome with pulmonary hypertension. **Am J Cardiol** 1993; 71:1326-30.
9. Najam HK, Williams WG, Coles JG, Rebeyka IM, Freedom RM. Scimitar syndrome: twenty years' experience and results of repair. **J Thorac Cardiovasc Surg** 1996; 112:1161-9.
10. Reddy R, Shah R, Thorpe JA, John G. Scimitar syndrome: A rare cause of hemoptysis. **Eur J Cardiothorac Surg** 2002;22:821.
11. Najm HK, William WG, Coles JG, Rebeyka IM, Freedom RM. Scimitar syndrome: Twenty years' experience and results of repair. **J Thorac Cardiovasc Surg** 1996; 112 :1161-8.
12. Canter CE, Martin TC, Spray TL, Weldon CS, Strauss AW. Scimitar syndrome in childhood. **Am J Cardiol** 1986; 58:652-54.
13. Haroutunian LM, Neill CA. Pulmonary complications of congenital heart disease: Hemoptysis. **Am Heart J** 1972; 84:540-59.
14. Huddleston CB, Exil V, Canter CE, Mendeloff EN. Scimitar syndrome presenting in infancy. **Ann Thorac Surg** 1999; 67:154-9.
15. Saha K, Iyer KS. Intracardiac repair of obstructed right sided Scimitar syndrome. **Indian Heart J** 1995; 47:378-9.

16. Schramel FM, Westermann CJ, Knaepen PJ, van den Bosch JM. The scimitar syndrome: Clinical spectrum and surgical treatment. **Eur Respir J** 1995; 8:196-201.
17. Wang CC, Wu ET, Chen SJ, Lu F, Huang SC, Wang JK, *et al*. Scimitar syndrome: Incidence, treatment and prognosis. **Eur J Pediatr** 2008; 167:155-60.
18. Mordue BC. A case series of five infants with scimitar syndrome. **Adv Neonatal Care** 2003; 3:121-32.