**Introduction**

Partial anomalous pulmonary venous connection (PAPVC) is relatively uncommon congenital anomaly and covers 0.5–1% of congenital heart diseases. It is usually associated with left to right shunting at atrial level.[1] The quite rare combination (3–5%) of PAPVC with hypoplasia of the right lung and dextroposition of the heart is designated as Scimitar syndrome.[2] The term “scimitar,” referring to a Turkish sword, was first used to describe the shape of the vein by Nicholas Halasz.[1] In two-thirds of cases, the Scimitar vein (SV) provides drainage for the entire right lung, but in others for only the lower portion.[3] Via SV pulmonary veins are discharged into the inferior or superior vena cava, or directly to right atrium. So a left-to-right shunt is established and this shunt result in long-standing right ventricular volume overload and induce increased risk of right ventricular failure.[4]

Recognizing the clinical and radiographical features of this rare disease is important when dealing with patients presented with cardiac and pulmonary complaints.

**Case Report**

A 38-year-old man was referred to our hospital with abnormal findings on plain chest roentgenogram and he presented with complain of cough for 10 days. He denied dyspnea and could perform his job without limitation. His medical and family history was unremarkable and no tobacco use.

The patient appeared well on chest examination respiratory rate was 18/min and breath sound was normal. Cardiac examination revealed a regular sinus rhythm of 74/min. The remainder of the examination was normal. Chest X-ray showed a sloping opacity at the right heart border [Figure 1a]. The contrast-enhanced computed tomography (CT) demonstrated that the left pulmonary venous drainage was normal, but mild hypoplasia of right lung, mild dextroposition of heart, and left renal agenesis were detected [Figure 1b and c]. The abnormal right pulmonary venous drainage from the right lung via a large characteristically curved anomalous vein draining into inferior vena cava at just superior to diaphragm [Figure 2a] and resembling to the curved Turkish sword “scimitar” [Figure 2b]. Spirometric investigations of the patient demonstrated mild restrictive
ventilatory defect. Blood cells and biochemistry tests were unremarkable. Artery blood gas analysis revealed pH of 7.44, pO$_2$ of 72, pCO$_2$ of 37, and O$_2$ saturation of 95%. While expected Alveolar-arterial pO$_2$ gradient of the patient was 10.5 mmHg, the measured A-a gradient was 22.4 mmHg. Transthoracic and transesophageal echocardiogram revealed an ejection fraction of 65%, systolic pulmonary artery pressure of 30-35 mmHg, slight RA and RV dilatation, and a one-fourth tricuspid regurgitation were also detected.

Based on chest roentgenogram and contrast-enhanced CT findings, Scimitar syndrome was diagnosed and renal agenesis was detected. We investigated in literature but did not detect any case both with Scimitar syndrome and renal agenesis.

**Discussion**

The SV must be present in Scimitar syndrome, but rarely the only abnormal findings. Other anomalies together with the SV are: Abnormal right lung lobation and hypoplasia (almost 100%, with widely varying degrees of hypoplasia); dextroposition of the heart; hypoplasia of the right pulmonary artery (60%); systemic arterial blood supply to the right lower lung from the infradiaphragmatic aorta (60%); ASD of secundum type. In our case; SV, mild hypoplasia of right lung, mild dextroposition of heart, and left renal agenesis were determined.

Most patients are symptomatic infants, but there are sparse reports of adults with Scimitar syndrome. In 82% of adult patients, left to right shunt is less than 50% and in 77% of these patients pulmonary artery pressures are normal and in 23% are slightly elevated, and these patients lead a normal life span without surgical correction. However, in cases with shunt more than 50%, dyspnea and pulmonary artery hypertension (PAH) develop. Our male patient did not have any symptoms like dyspnea and weakness. But his artery blood gas analysis showed some hypoxemia (pO2 of 72) and spirometric investigation showed mild restrictive ventilatory defect. The corrective surgery was not performed because of his refusal and the absence of both PAH and symptoms.

Patients with Scimitar syndrome generally undergo comprehensive evaluation with CT scans and cardiac angiography to characterize the anomalous vessels. Recently cardiac magnetic resonance imaging has been proposed to assess the congenital abnormalities. In our case; chest radiograph, contrast-enhanced CT, transthoracic and transesophageal echocardiogram were sufficient to diagnose and eliminate the other anomalies.

The incidence of unilateral renal agenesis is about 1 in 1300 births and the prognosis is excellent. In renal agenesis the ureteric bud fails to differentiate to renal tubular epithelium. In humans this process starts at approximately 5th week and ceases at approximately 36th week of gestation. In normal lung development, primary blood supply is formed after the 7th week and pulmonary venous drainage to the left atrium is settled by 11th week. Since both organogenesis take place around the same period, in our case, we thought that during fetal period there might be some factor effecting both cardiovascular and urinary system. We investigated the patient history but could not get any information.

**Conclusions**

We researched the literature and did not detect Scimitar
syndrome with renal agenesis. We submitted the case because of being asymptomatic male adult patient which is seen quite rarely and for the first time to be determined Scimitar syndrome and renal agenesis together.

References

1. Halasz NA, Halloran KH, Liebow AA. Bronchial and arterial anomalies with drainage of the right lung into the inferior vena cava. Circulation 1956;14:826-46.

2. Neill CA, Ferencz C, Sabiston DC, Sheldon H. The familial occurrence of hypoplastic right lung with systemic arterial supply and venous drainage, ‘Scimitar syndrome’. Bull Johns Hopkins Hosp 1960;107:1-21.

3. Gudjonsson U, Brown JW. Scimitar syndrome. Semin Thorac Cardiovasc Surg Pediatr Card Surg 2006;9:56-62.

4. Yehia BR, Bachmann JM, Traill TA. Scimitar syndrome: A rare cause of dyspnea in adults. South Med J 2010;103:578-80.

5. Kamler M, Kerkhoff G, Budde T, Jakob H. Scimitar syndrome in an adult: Diagnosis and surgical treatment. Interact Cardiovasc Thorac Surg 2003;2:350-1.

6. Dupuis C, Charaf LA, Breviere GM, Abou P, Remy-Jardin M, Helmius G. The “adult” form of the scimitar syndrome. Am J Cardiol 1992;70:502-7.

7. Walles T, Lichtenberg A, Shiraga K, Klima U. Combined correction of an adult Scimitar syndrome and coronary artery bypass grafting. Ann Thorac Surg 2002;73:640-2.

8. Gavazzi E, Ravanelli M, Farina D, Chiari ME, Maroldi R. Scimitar syndrome: Comprehensive, noninvasive assessment with cardiovascular magnetic resonance imaging. Circulation 2008;118:e63-4.

9. Wein AJ, Kavoussi LR, Novick AC, Partin AV. Anomalies of the upper urinary tract. In: Peters CA, editor. Campbell-Walsh Urology. 9th ed. Review. Philadelphia: Saunders; 2007. p. 3269-304.

10. Quaggin SE, Kreidberg J. Embryology of the kidney. In: Brenner BM, editor. Brenner and Rector’s the Kidney. 8th ed. Philadelphia: Saunders, Elsevier; 2008. p. 3-24.

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