Syndrome of Hypoplasia of the Right Lung and Dextroposition of the Heart:
“Scimitar Sign” with Normal Pulmonary Venous Drainage

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SUMMARY
Reported is a case in which the clinical findings of the scimitar syndrome were present but with termination of the anomalous pulmonary vein into the left atrium. There was hypoplasia of the right lung, dextroposition of the heart, and the scimitar sign in the right lower lung field. Cardiac catheterization revealed no shunt, and on pulmonary angiograms, a large common right pulmonary vein curved toward the diaphragm (as in the usual case of the scimitar syndrome), but then reversed course to enter the left atrium. No connection with the inferior vena cava was present.

Additional Indexing Words:
Anomalous pulmonary veins  Cardiac catheterization  Pulmonary angiograms  Scimitar syndrome

From its radiological appearance, the name scimitar syndrome has been given to a combination of hypoplasia of the right lung, dextroposition of the heart, and anomalous pulmonary venous drainage of the right lung to the inferior vena cava, producing a curved vascular shadow in the right lower lung field, resembling the blade of a sword or scimitar. The typical appearance on a plain chest film has been presumed to be diagnostic of this syndrome. The purpose of this communication is to describe a case with all of the typical findings of the scimitar syndrome, except that a large common right pulmonary vein curved toward the diaphragm (as in the usual case of the scimitar syndrome) and then reversed course to drain completely into the left atrium instead of into the inferior vena cava. No similar case is known in the literature.

Case Report
A 22-year-old Navy man, R. C., was referred to the Navy Hospital, San Diego, for evaluation of an abnormal chest roentgenogram. He gave a lifelong history of wheezing respirations, decreased exercise tolerance, and a persistent cough occasionally productive of clear sputum.

Physical examination revealed a well-developed, healthy-appearing, white male. The right hemithorax was smaller than the left, and cardiac pulsation was visible and palpable to the right of the sternum. The percussion note was impaired over the entire right chest. Mild inspiratory and expiratory musical wheezes were heard over the right hemithorax. There were no murmurs and normal heart sounds to the right of the sternum.
(A) posteroanterior chest X-ray, showing dextroposition of the heart, hypoplastic right lung, and a scimitar sign behind the right heart border (arrow). (B) lateral chest X-ray, with an abnormal vascular shadow posterior to the heart.

An electrocardiogram showed upright P waves in leads I and aVL, and was normal except for slightly prominent anterior orientation of the QRS loop in the transverse plane.

Chest roentgenograms (fig. 1) showed dextroposition of the heart with the entire heart rotated into the right chest. Situs solitus was present. Decreased pulmonary vascularity was present in the right lung, with normal pulmonary vascularity in the left lung. The posteroanterior film showed a vascular shadow behind the right heart border, which had the appearance of a scimitar sign, although it was better seen posterior to the heart on the lateral film.

A lung scan showed marked decrease in perfusion of the right lung (fig. 2).

Pulmonary function studies were consistent with a moderate obstructive defect, which improved slightly with a bronchodilator, and with slight hypoxemia at rest with a normal response to exercise and 3 min of oxygen.

Right ventricular and pulmonary artery systolic pressures were 35 mm Hg, with otherwise normal pressures on right and left heart catheterization. Oxygen saturation data, hydrogen inhalation studies, and Cardio-Green dye curves revealed no evidence of a left-to-right or right-to-left shunt.

A pulmonary angiogram revealed normal left pulmonary arteries, but the right pulmonary arteries were generally decreased in size. A very large single right pulmonary vein (fig. 3) descended from the upper lobe to the diaphragm, and then made a sharp turn upward to drain

Figure 1

Figure 2

Anterior lung scan with $^{131}$I macroaggregates.

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completely into the left atrium. On later films there was what appeared to be a small arterial branch from the descending aorta to the right lower medial lung field.

Discussion

Two recent excellent reviews have appeared on the scimitar syndrome.\(^1\),\(^2\) The typical scimitar sign was present in 57 of the 67 cases reviewed by Kiely et al.\(^1\) The vascular shadow of the pulmonary vein in the right medial lower lung field may be partially masked by displacement of the heart, but when it is seen alongside the right heart or behind the dextrorotated heart, it has generally been regarded as diagnostic of anomalous venous drainage.\(^3\) The plain chest radiograph of the complete syndrome with the dextrorotated heart, hypoplasia of the right lung, and the scimitar sign is considered unmistakable evidence of anomalous pulmonary venous drainage to the inferior vena cava.\(^4\) The present case demonstrates that, rarely, a false positive scimitar syndrome may be present, with dextroposition of the heart, hypoplasia of the right lung, and the scimitar sign caused by a large abnormal pulmonary vein with normal drainage to the left atrium.

Blake et al.\(^5\) previously showed a roentgenogram with a false positive scimitar sign but the other findings of the scimitar syndrome, including dextroposition of the heart or hypoplasia of the right lung, were not present. A roentgenographic shadow resembling, but higher than, the usual scimitar sign has also been observed in anomalous pulmonary venous return to a superior vena cava in association with intrapulmonary venous connections.\(^6\),\(^7\)

Other variations in pulmonary venous connection to the left atrium have been described in the scimitar syndrome. In three cases of the scimitar syndrome,\(^8\)–\(^10\) a large collateral was present to the left atrium in addition to the inferior vena cava; surgical correction required ligation only of the connection to the inferior vena cava. Mathey et al.\(^2\) reported that in 21 of 57 cases of the scimitar syndrome, two or more terminations of the anomalous pulmonary vein were present, including connection to the left atrium, right atrium, superior vena cava, and azygous vein, although most of the blood still drained into the inferior vena cava.

If the scimitar syndrome represents concomitant congenital anomalies, as assumed by

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Figure 3

Pulmonary angiogram with delayed films, demonstrating a large common right pulmonary vein descending toward the diaphragm, but then draining into the left atrium. In (A) both left pulmonary veins and the abnormal right pulmonary vein are seen emptying into the left atrium. In (B), 1 sec later, left pulmonary veins have emptied, with the right pulmonary vein and left atrium still opaque; the aorta is also opaque. One catheter had normal passage from the inferior vena cava with the tip in the pulmonary artery, and the other catheter entered from the descending aorta with the tip in the left ventricle.
most authors,\textsuperscript{11} then this case could be considered a slight variant of the scimitar syndrome, but without the connection to the inferior vena cava.

However, Massumi et al.\textsuperscript{12} proposed that the only congenital anomaly was the pulmonary venous drainage into the inferior vena cava, and that the associated anomalies—dextroposition of the heart, hypoplasia of the right lung, and attenuation of the right pulmonary artery—were the physiologic consequences of obstruction to venous return from the right lung. The present case did not have drainage into the inferior vena cava, but had an abnormal right common vein with all of the associated anomalies of the scimitar syndrome. This would seem to disprove the theory of Massumi et al.\textsuperscript{12} unless it is assumed that some obstruction occurred either at the acute angulation near the diaphragm or on entrance into the left atrium.

References


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