A CASE WITH PSEUDO-SCIMITAR SYNDROME:  
“SCIMITAR SIGN” WITH NORMAL PULMONARY  
VENOUS DRAINAGE  
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The case reported here showed a radiological appearance of hypoplasia of the right lung, dextroposition of the heart, and a curved vascular shadow in the right lower lung field known as a scimitar sign. However, a computed tomography of the chest showed this abnormal vascular shadow draining into the left atrium (pseudo-scimitar sign). Therefore, in patients with a radiological appearance of the scimitar syndrome, computed tomography of the chest should be indicated to rule out the pseudoscimitar sign.  

Chest roentgenograms provide an important clue for the diagnosis of the scimitar syndrome. This paper describes a patient, whose radiological examination showed hypoplasia of the right lung, dextroposition of the heart, and a curved vascular shadow in the right lower lung field, known as the scimitar sign, but whose computed tomography of the chest revealed that the abnormal vascular shadow drained into the left atrium.  

CASE REPORT  
A 48-year-old woman was hospitalized because of orthopnea, cough and sputum. She had a history of rheumatic fever at the age of 5 and was initially found to have a heart murmur at the age of 8. At age 24, she had a normal delivery. She led an almost normal life until 4 years ago, when she began to notice dyspnea on exertion, cough and sputum. She was admitted to another hospital where she was treated for congestive heart failure. At that time cardiac catheterization was performed and atrial fibrillation, mitral stenosis and regurgitation, and aortic stenosis and regurgitation were diagnosed. Since then she has been treated with a regimen of digitalis and diuretics.  

A month before entry, after suffering from a respiratory infection, she noticed a decrease in exercise tolerance, increasing dyspnea, cough and sputum. A week before entry, she gained 3 kg in weight and was troubled with orthopnea.  

A physical examination showed her to be thin and pale with a malar flush. She measured 156 cm in height and weighed 49 kg. The pulse was around 80 and totally irregular while the blood pressure was 110/60 mmHg. No cyanosis and clubbed fingers were noted. No mass or lymphadenopathy was found. The jugular vein was distended by 45 degrees. The right hemithorax was smaller than the left. A systolic thrust was palpable at the left lower sternal border. Decreased first heart sound, grade 3 holosystolic murmur and opening snap were audible at the apex. Both grade 3 systolic ejection and diastolic blowing murmur were  

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Fig. 1. Electrocardiogram showing atrial fibrillation, left ventricular hypertrophy and digitalis effect.

Fig. 2. X-ray film of the chest revealing moderate cardiomegaly, hypoplasia of the right lung, dextroposition of the heart, and scimitar sign.

Fig. 3. Tomography of the chest demonstrating the scimitar sign more clearly.

Fig. 4. Lung scan showing a marked decrease in the perfusion of the right lung.

heard at the second intercostal space to the right of the sternum. Crackles were heard in both lower lung fields posteriorly. The liver was felt 2 FB. There was 1+ leg edema.

Routine laboratory tests, including complete blood count, blood chemistry and urinalysis were normal. The electrocardiogram (Fig. 1) demonstrated atrial fibrillation, left ventricular hypertrophy and digitalis effect.

An X-ray film of the chest (Fig. 2) revealed moderate cardiomegaly with a cardiothoracic ratio of 0.74, hypoplasia of the right lung, dextroposition of the heart and curved vascular shadow in the right lower lung field, suggesting the scimitar sign. This was better demonstrated in tomography (Fig. 3). A lung scan showed a
marked decrease in the perfusion of the right lung (Fig. 4). The symptoms and signs of this patient subsided gradually by the routine treatment for congestive heart failure.

Echocardiography showed an enlarged left atrium (left atrial dimension = 55 mm), decreased opening of the aortic valve (11 mm) decreased EF slope (29 mm/sec), a normal left ventricular diastolic dimension (50 mm), thickened interventricular septum and left ventricular posterior wall (both 19 mm) with a slightly decreased interventricular septal movement.

A CT scan of the thorax (Fig. 5) revealed hypoplasia of the right lung, dextro-position of the heart with a markedly enlarged left atrium and an abnormally curved right pulmonary vein draining into the left atrium.

Cardiac catheterization disclosed that the right atrial mean pressure was 8 mmHg, the right ventricular pressure was 49/8 mmHg, the pulmonary artery pressure was 49/34 (42) mmHg and aortic pressure was 110/70 (83) mmHg. The pulmonary capillary wedge pressure and left ventricular pressure were not measured due to technical problems.

A pulmonary angiography demonstrated that the abnormal pulmonary vein descended from the right upper lobe to the diaphragm, and then made a sharp turn upward to drain into the

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markedly enlarged left atrium. An aortography showed grade 3 aortic regurgitation and mitral regurgitation. The aortic orifice was small and movement of the valve was severely obstructed.

DISCUSSION

True- versus pseudo-scimitar syndrome

In almost all cases, the scimitar syndrome is diagnosed by plain X-ray films of the chest. Of the three radiological features, the anomalous vascular shadow of the pulmonary vein alongside the right second cardiac border, forming a scimitar sign, has been regarded as an indication of anomalous pulmonary venous drainage to either the inferior vena cava or right atrium and a characteristic of this syndrome.

However, as in our present case, six other cases were reported in world literature which showed the typical radiological appearance of the scimitar syndrome, in which an anomalously curved vein drained normally into the left atrium. Moreover, there are also cases where the pulmonary vein drains both the inferior vena cava and the left atrium.

Therefore, a scimitar sign does not always suggest an anomalous pulmonary venous drainage and a CT scan of the chest provides useful information concerning the return of the anomalous pulmonary vein.

Pathogenesis

Massumi et al postulated that congenital obstruction of an anomalously draining right pulmonary vein was the cardinal feature of this syndrome, leading to pulmonary venous engorgement and increasing elastic resistance of the right lung.

Neill et al proposed the same mechanism as seen in pulmonary sequestration, because the anomalous pulmonary venous drainage is the connection between the systemic circulation and the right lung.

However, cases having pulmonary venous stenosis in the scimitar syndrome are rather exceptional. Moreover, there are cases where abnormal pulmonary veins do not drain into the right side of the heart.

Therefore, a hypothesis involving a developmental anomaly of the pulmonary venous system is more convincing. The pulmonary venous system develops from the pulmonary venous plexus, common cardinal vein, sinus venosus, umbilical vein and omphalomesenteric vein.

During the course of development the latter two veins degenerate and the pulmonary venous plexus and sinus venosus combine, forming the pulmonary venous system. Koga et al and Okada et al postulated that the remains of the umbilical and omphalomesenteric vein itself is the scimitar vein.

True-, pseudo-, and the syndromes in between are brought about as a developmental anomaly during the process of forming the pulmonary venous trunk. While Goodman et al postulated that abnormal vessels drain into the coronary sinus and/or left superior vena cava when they are patent, but if neither is patent, the blood drains via the thebesian system into the left atrium and this type of drainage is an aberrant left-sided insertion into the left atrium.

Thus the true pathogenesis has not been clarified yet.

Associated cardiac lesions

Koga et al conducted a statistical survey on 91 cases from world literature and noted an associated cardiac anomaly in 22 cases (24%); 12 cases of atrial septal defect, 4 patent ductus arteriosus, 2 ventricular septal defects, 2 coarctations of the aorta, 2 tetralogy of Fallot, and one case each of multiple peripheral pulmonary stenosis, bicuspid aortic valve, persistent left superior vena cava and stenosis of the superior vena cava, respectively. No cases with mitral and aortic valve lesions as in our cases have been reported. However, the valvular lesions in our case were due to rheumatic fever and their combination with the scimitar syndrome happened only by chance.

REFERENCES

5. GOODMAN LR, JAMSHIDI A, HIPONA FA: Meandering right pulmonary vein simulating the

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