Case Report

Use of 64 Slice CT in Scimitar Syndrome

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Abstract

Introduction: Scimitar Syndrome is a relatively rare variety of Partial Anomalous Pulmonary Venous Connection in which the right pulmonary veins form an anomalous confluence which drains into the infra-diaphragmatic inferior vena cava. The X-ray chest in these patients shows the typical Scimitar Sign.

Case Report: We are presenting a patient who was diagnosed to have Scimitar Syndrome on the basis of X-ray chest and echocardiography. Confirmation of diagnosis and precise anatomical characterization was achieved by 64 slice CT angiography. The patient underwent successful surgical correction. Adequacy of procedure was demonstrated by the same procedure.

Discussion: Scimitar Syndrome is a type of Partial Anomalous Venous Connection. It consists of sinus venosus type of atrial septal defect, anomalous confluence of right upper and lower pulmonary veins draining into the infra-diaphragmatic inferior vena cava and right lung lower lobe hypoplasia. The X-ray chest shows the characteristic Scimitar Sign. Precise anatomical characterization in required for operative correction and cannot usually be achieved by echocardiography and requires invasive angiography, multi-detector CT angiography or cardiac MRI. We used CT angiography for diagnosis and post-operative confirmation of adequacy of correction.

Introduction

Partial Anomalous Pulmonary Venous Connection is a disorder consisting of connection of one or more, but not all, the pulmonary veins to the systemic veins or directly to the right atrium. Scimitar syndrome is a specific variety of Partial Anomalous Pulmonary Venous Connection in which the right pulmonary veins unite to form a confluence which drains into the inferior vena cava below the diaphragm, but above the drainage of the hepatic veins. We present a case of a 19 year old girl with Scimitar Syndrome who was operated.

Case Report

A 19 year old girl was referred to this hospital for advice regarding cardiac surgery. She was a diagnosed case of Sinus Venosus type of Atrial Septal Defect since six months. She had a history of recurrent respiratory tract infections since her teens. She had been asymptomatic in early childhood. There was no history of dyspnoea, chest pain, easy fatigue, cyanosis, cyanotic spells or squatting spells. On examination for respiratory infection, she was found to have the clinical features of an Atrial Septal Defect (wide fixed split of S2, early systolic murmur in the right 2nd intercostals space), with no evidence of pulmonary hypertension clinically. The X-ray chest is shown (Fig. 1). The X-ray showed mild cardiomegaly with a right ventricular type of apex, enlarged right atrium, prominent pulmonary arteries and increased pulmonary vascular markings, consistent with left-to-right shunt without significant pulmonary hypertension. A further finding of great interest is the ‘Scimitar Sign’, which is seen very clearly as a sword shaped shadow in the right lung field, running approximately parallel to the heart border. The right lung does not appear hypoplastic.

Echocardiographic examination showed the Sinus Venosus type of Atrial Septal Defect and right ventricular overload with mild pulmonary hypertension. Only the two left pulmonary veins could be seen draining into the left atrium. These findings supported the diagnosis of Partial Anomalous Pulmonary Venous Connection.

Since precise anatomic delineation is required for surgical planning, it was decided to perform a non-invasive 64 slice CT angiography study. The images obtained by the procedure are shown below (Fig. 2). As can be seen, the anomalous confluence of the right upper and lower pulmonary veins is clearly and beautifully visualized and its precise entry point into the inferior vena cava after the drainage of the hepatic veins, but below the diaphragm is accurately visualized. The absence of any obstruction in the anomalous pulmonary channel is confirmed as well. The anatomic details of the Atrial Septal Defect could be made out. The scan also demonstrated mild hypoplasia of the basal part of the right lung, which had not been seen on X-ray chest.

The patient underwent cardiac surgery under cardiopulmonary bypass. Since, the anomalous right pulmonary confluence drained into the inferior vena cava at a point quite close to its insertion into the right atrium, it was decided to avoid resection of the channel. Instead, a pericardial patch was used to create a tunnel within the right atrium, directing the flow of pulmonary venous blood from the confluence, via the tunnel, through the atrial septal defect which was enlarged surgically, into the left atrium. Complete separation of right and left circuits was obtained.

The patient had an uneventful post-operative course and was ready for discharge by the 7th post-operative day. A post

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operative CT angiocardiogram was obtained to confirm favorable surgical results. The study image is shown in Fig. 3. The study confirmed flow of blood from the right pulmonary confluence, into the tunnel, through the enlarged septal defect, into the left atrium. No evidence of residual shunt was found, both on CT study and post-operative echocardiography. The patient regularly follows up and is doing well.

Discussion

Scimitar Syndrome was first described by Chassàn in 1836. It is a rare, but well characterized disease accounting for 0.5-1% of all CHD. The term ‘Scimitar Syndrome’ was coined by Neill et all, in reference to the characteristic appearance of the anomalous right pulmonary venous confluence on the X-ray chest. The vein is said to resemble a curved Turkish sword called a ‘Scimitar’. The X-ray appearance is referred to as the ‘Scimitar Sign’.

The syndrome is characterized by Partial Anomalous Pulmonary Venous Connection of the right pulmonary veins, via the confluence to the infra-diaphragmatic inferior vena cava, above the drainage site of the hepatic veins. There is usually no obstruction to pulmonary venous flow. Rare cases of connection to the hepatic veins or to the portal vein have been documented.

The other components of the syndrome include hypoplasia of the lower lobe of the right lung, hypoplasia of the right pulmonary artery or its lower branch, abnormalities of tracheobronchial architecture, abnormal lobar architecture and sequestration of the right lower lobe (vascular supply from a branch of the aorta). Intact inter-atrial septum is very rare and a sinus venosus form of atrial septal defect is almost always present.

Two varieties have been described. The infantile form presents in infancy or early childhood due to other associated cardiac anomalies like Tetralogy of Fallot’s, coarctation of the aorta, patent ductus arteriosus and ventricular septal defect. The adult form presents late or may remain totally asymptomatic and is not associated with other cardiac anomalies.

Our patient had the adult form of Scimitar Syndrome with minimal hypoplasia of the right lung and no demonstrable sequestration.

Diagnosis of Scimitar syndrome is usually suspected on the X-ray chest. While echocardiography can demonstrate the atrial septal defect and absence of right pulmonary venous drainage into the left atrium, detailed anatomic evaluation of the anomalous channel requires invasive angiocardiology, multi-detector CT angiocardiology or MR angiocardiology. We used 64 slice CT angiocardiology to demonstrate the anomalous vein and plan surgery. Accurate anatomical
Information was obtained non-invasively. The same technique was also used for post-operative evaluation and adequacy of surgical procedure was clearly demonstrated.

There are three options for repair of the Scimitar Syndrome, depending on the anatomy. The first method entails the creation of an internal conduit within the right atrium to conduct right pulmonary blood from its entrance in the inferior vena cava to the left atrium through the atrial septal defect, which may or may not be enlarged. This is the technique which was used for this patient.

The second technique entails disconnection of the anomalous vein from its connection with the inferior vena cava and reimplantation higher up in the right atrium, with a baffle connecting the vein to the left atrium. The third technique consists of disconnecting the anomalous vein from the junction with the inferior vena cava and connecting it to the rightmost aspect of the left atrium, which is ‘bare’ (due to no connection of the right pulmonary veins).

The long term results of correction performed in childhood or early adult life in this disorder are generally good.

This case highlights the accuracy and ease with which 64 slice CT angiocardiography can demonstrate the anatomy of the Scimitar Syndrome along with all associated cardiac and pulmonary lesions. The case demonstrates the utility of the technique in demonstrating post-operative results.

Another point highlighted by the case was the fact that the ‘routine’ X-ray chest should always be reviewed carefully in patients with suspected or confirmed sinus venosus type of atrial septal defect.

References