A unique variant of the Scimitar Syndrome

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Abstract

A 36 year old woman was referred to the cardiology department for further evaluation of thoracic abnormalities seen on CT. She had a background of congenital abnormalities of the urogenital and gastrointestinal systems and recent haemoptysis. Cardiac MR imaging helped establish that she had a variant of the scimitar syndrome. This is the first report of scimitar syndrome associated with imperforate anus and bicornuate uterus.

Case report

A 36 year old woman was referred to the cardiology service for further evaluation of abnormalities found on a CT thorax performed to evaluate haemoptysis. She had a past history of a series of congenital abnormalities.

She was born in 1971 with an imperforate anus which required a temporary colostomy followed by refashioning of the anus. At age 17 she was admitted to another hospital after presenting with haemoptysis. Examination revealed a pansystolic murmur and right basal crepitations. Chest Xray showed reduced right lung volume with the mediastinum significantly deviated to the right. There was no significant dyspnoea and she had no further haemoptysis after being treated with a course of amoxicillin. She was subsequently discharged and outpatient bronchoscopy was performed. This showed significant abnormalities of the right bronchial tree; a right upper lobe orifice could be defined but inferiorly only one major division was seen, and the usual site of the lower lobe orifice was atrophic and occluded. Further follow-up was planned but did not occur.

During the next few years there were no further episodes of haemoptysis. During her first pregnancy, aged 21, she was identified as having a bicornuate uterus but the pregnancy was uncomplicated. A transthoracic echocardiogram was also performed at that time but did not detect any abnormalities.

She presented again in 2007 with further small episodes of haemoptysis. She also complained of lethargy and occasional non-specific palpitations. However, she had a good exercise tolerance and denied dyspnoea. Examination showed she was normotensive, the JVP was not elevated, and there were no thrills or heaves. There was a loud pulmonary component to the second heart sound but no audible murmurs. She also had a slight wheeze bilaterally and reduced air entry at the right lung base.

Initial investigations showed a normal ECG. A CXR showing changes similar to those described previously with the heart occupying the R side of the chest [figure1].

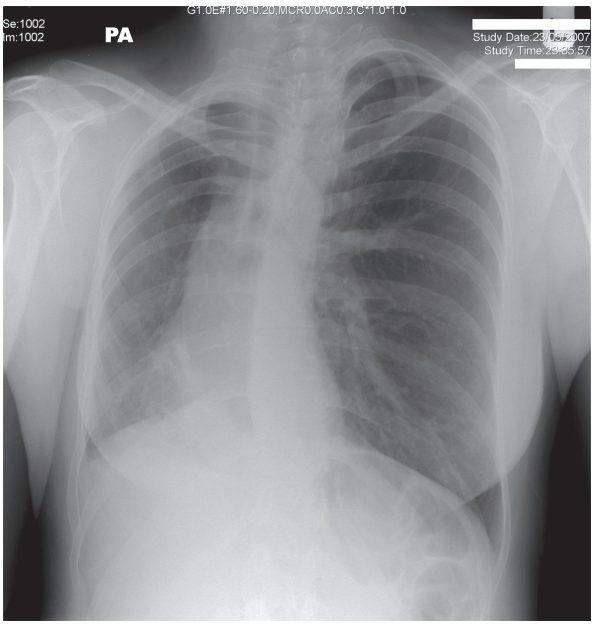

Figure 1: Chest Xray demonstrating loss of right lung volume.

A transthoracic echocardiogram again showed no gross abnormalities. CT thorax showed

significant volume loss on the right with the right lung only containing two lobes [figure 2].

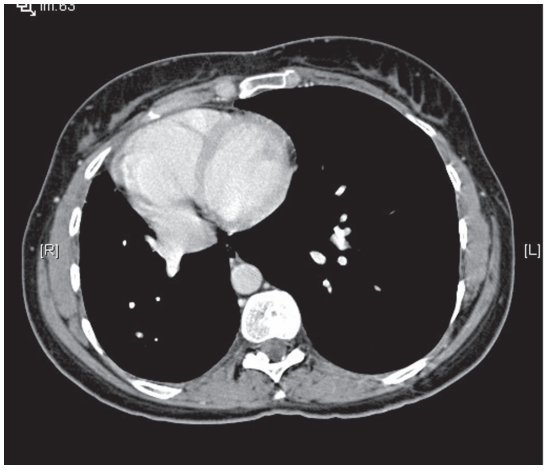

Figure 2: CT scan demonstrating the heart in the right side of thorax with the right pulmonary vein (RPV) draining into the right atrium (RA).

As a consequence of the reduced right lung volume the heart had moved to the right hemithorax. The left pulmonary veins drained into the left atrium but the right pulmonary veins seemed to drain into the right atrium. To help further evaluate the abnormalities, a cardiac MRI was performed. A multimodal study including T1 weighted, cine, phase contrast and MR angiography acquisitions was performed to evaluate fully the cardiac anatomy, function and physiology. This showed total anomalous pulmonary venous drainage of the right-sided pulmonary veins to the right atrium, at the junction with the inferior vena cava (IVC). This is most elegantly demonstrated on the MR angiogram [figure 3]. Associated with this the right pulmonary artery was hypoplastic [figure 4]. Phase contrast imaging demonstrated a small left to right shunt of 1.5:1. Similar assessments of flow in the branch pulmonary arteries demonstrated that 90% of the pulmonary flow was to the left lung. To assess differential ventilation of the lungs, a ventilation scan was performed, which showed that the right lung was contributing 35% of the ventilation. This suggested a degree of ventilation/perfusion mismatch between the lungs. These findings were consistent with a variant of the Scimitar syndrome.

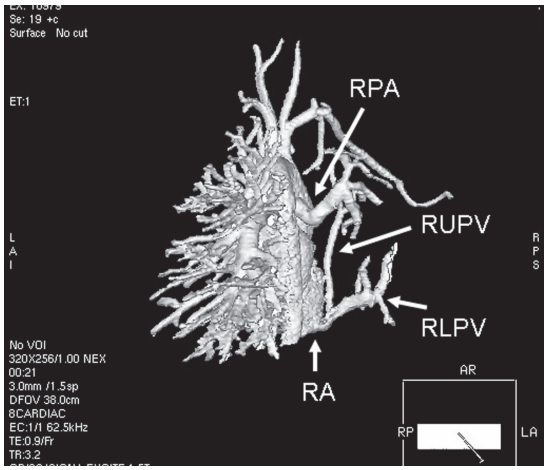

Figure 3: 3-D MPR image of venous connections demonstrating anomalous connection of RUPV and RLPV to RA.

RUPV= right upper pulmonary vein; RLPV=right lower pulmonary vein; RA=right atrium; RPA=right pulmonary artery

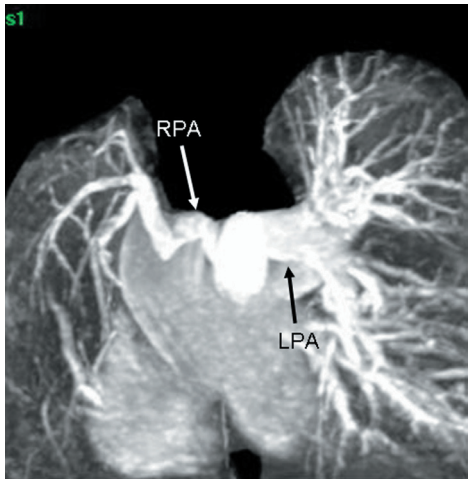

Figure 4: 3D- MIP demonstrating hypoplastic R pulmonary artery

RPA=right pulmonary artery; LPA=left pulmonary artery

Discussion

There is a large variety of anomalous pulmonary venous connections reported, of which the scimitar syndrome is a rare variant. Studies have suggested partial anomalous pulmonary venous connections are seen in approximately 0.2% of CT thorax images performed; of which an anomalous left upper lobe vein connecting to a persistent left superior vena cava is the most common abnormaility.[1] Scimitar syndrome includes a variety of abnormalities, and is thought to account for approximately 2-5% of all partial anomalous pulmonary venous connections.[2] The syndrome involves anomalous drainage of the right pulmonary veins into the IVC, with associated hypoplasia, abnormal lobation, and partial arterial blood supply of the right lung. The classical finding on CXR is of a prominent convex pulmonary vascular shadow at the right boarder of the heart, extending to the diaphragm; similar in shape to the curved Turkish sword called a scimitar. Several variations on this pattern have been described including drainage into the portal vein, involvement of the left lung, and additional abnormalities such as atrial septal defect and diaphragmatic abnormalities. The age of presentation varies, with the more severe forms presenting earlier, which are associated with higher mortality rates.[3] In adulthood, the syndrome can be identified incidentally, or present as haemoptysis, recurrent infections or dysphonea.[4,5] Corrective surgery is advocated if there are recurrent infections, or if the pulmonary-to-systemic blood flow ratio is greater then 1.5:1, as these patients have a higher risk of developing pulmonary hypertension and right ventricular failure. Surgical options vary from each patient but direct reimplantation of the scimitar vein into the left atrium can be performed, although this is not infrequently complicated by pulmonary vein stenosis.[6,7]

Our patient is asymptomatic now with a small shunt and as such is being managed conservatively with regular outpatient reviews. Although she has reduced exercise tolerance for a woman of her age she is not unduly restricted. The combination of imperforate anus and bicornuate uterus in a patient with a scimitar variant has not been described before making our patient a unique case.

References

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