CASE REPORT

Uncommon presentation of adult-form scimitar syndrome associated with single left pulmonary vein in a pregnant woman

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SUMMARY

Scimitar syndrome is the constellation of malformations including an abnormal venous drainage of the right lung into the inferior vena cava, associated with the right lung and systemic supply to the right lung. The anomalous vein looks like the curved, Turkish sword (scimitar), hence the name.

The adult form of scimitar syndrome is rare, and it is usually an incidental diagnosis based on the characteristic finding on radiological imaging since the patients are usually asymptomatic or with minimal symptoms.

Our patient presented with a rare presentation of scimitar syndrome, which is tachyarrhythmia (sinus tachycardia, with episodes of supraventricular tachycardia). The diagnosis of scimitar syndrome was made based on the typical radiological finding of the anomalous venous drainage on CT angiography. Our patient does not have the full spectrum of the scimitar syndrome; therefore, she did not suffer from the usual complication (pulmonary hypertension). She was treated with ablation without surgical intervention.

CASE PRESENTATION

A 33-year-old female patient, who was 39 weeks pregnant, was admitted to the hospital under the management of the Obstetrics department. She was due to give birth by caesarean section because the fetus was in breech presentation. We were consulted because the patient was experiencing progressive shortness of breath, which had started 16 weeks previously. She also gave a history of occasional palpitation since childhood and during previous pregnancies, for which she had sought medical advice several times, but had not received any definitive diagnosis or treatment.

On physical examination, the patient was found to be mildly dyspnoeic with a respiratory rate of 22 breaths/min. SpO₂ was 88%–92% room air, heart rate was 102 beats/min, blood pressure was 113/85 and temperature was 37°C. Chest auscultation was normal, and cardiovascular examination revealed normal S1 and S2. There was no evidence of lower limb oedema or signs of deep vein thrombosis. The obstetrician was concerned about the possibility of pulmonary embolism (PE).

BACKGROUND

Partial pulmonary venous return (PPVR) is the term given to a group of cardiopulmonary malformations, in which an anomalous pulmonary vein drains part of the lungs (usually the right lung) into other sites (mostly the inferior vena cava (IVC)) rather than into the left atrium.

Scimitar syndrome, which takes its name from the distinct shape of the anomalous vein, resembling the curved Turkish sword, represents 3%–6% of cases in this group.^{1 2} Scimitar syndrome is often associated with other anomalies, especially congenital heart disease (CHD). Isolated scimitar syndrome is rare and has a more benign course.^{1 2}

Cases of scimitar syndrome are most frequently reported in infants, and reported cases in adults are rare. The adult form of scimitar syndrome tends to be asymptomatic or with minimal symptoms and has a good prognosis without surgical intervention, as patients rarely develop pulmonary hypertension or congestive heart failure (CHF).³

We report a case of a patient with adult-form scimitar syndrome presented with tachyarrhythmia which is an unusual and a rare presentation and found to be associated with a single left pulmonary vein.

INVESTIGATIONS

Chest radiograph showed non-specific right lower lobe opacity (figure 1). Initial ECG showed sinus tachycardia; later during hospitalisation, the patient developed one attack of supraventricular tachycardia, which stopped spontaneously. Blood investigations were within normal ranges (haemoglobin, 10.2 g/dL; K, 4.9 mEq/L; Na, 131 mEq/L; Ca, 8.7 mg/dL). Echocardiography showed a normal left ventricle with normal ejection fraction (68%), a mildly dilated right ventricle, normal mean pulmonary pressure and normal pulmonary artery diameter.

In order to exclude a PE, a CT angiography study (figures 2 and 3) was performed, which did not show any PE, but did show an anomalous PPVR of part of the pulmonary venous drainage in the right lower lobe into the infrahepatic IVC (that is, the basal posterior and basal medial segments of the right lower lobe were draining into the IVC). The maximum diameter of the anomalous vein was 11 mm. There was mosaic attenuation of the basal posterior and basal medial segments. The maximum diameter of the main pulmonary artery measured 25 mm (within the normal range). A

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Figure 1 Chest X-ray showing non-specific right lower lobe haziness.

single pulmonary vein on the left side was noted. No cardiac catheterisation was performed.

TREATMENT

The patient delivered a normal baby with no complications. She was later referred to a specialist centre for a cardiothoracic surgery consultation. She underwent an electrophysiology study for her palpitations, which revealed inducible and sustained common and uncommon types of atrioventricular node re-entrant tachycardia. Ablation was carried out successfully.

OUTCOME AND FOLLOW-UP

To date, no corrective surgery has been performed. The patient is undergoing regular follow-up with the Cardiothoracic Surgery/Cardiology departments.

DISCUSSION

Scimitar syndrome is a rare pulmonary venous anomaly, which consists of an anomalous partial pulmonary venous connection, right lung hypoplasia and an anomalous systemic arterial supply to the right lung. An isolated anomalous vein (scimitar vein) can also occur.^{1 2}

Dupuis *et al*³ classified scimitar syndrome into three categories:

Figure 3 Three-dimensional CT angiography showing the anomalous scimitar vein is draining into the inferior vena cava.

1. Infantile form, in which the symptoms appear within the first year of life; this is usually severe with poor spontaneous prognosis.
2. Adult form, which is mostly asymptomatic until later in life and has a good prognosis.
3. Scimitar vein in association with other complex cardiopulmonary congenital anomalies such as dextrocardia, pulmonary stenosis or atrial septal defect (ASD).³

Patients with the adult form of scimitar syndrome usually present during childhood (<10 years of age)^{2 3} with minimal or unspecific symptoms. Rarely, patients present at older ages, and often the condition is discovered incidentally on chest imaging.⁴⁻⁷

In Dupuis *et al*³'s study, which included 122 patients with adult-form scimitar syndrome, 18% presented with exertional dyspnoea, 31% with recurrent chest infection, 5% with haemoptysis, 22% with other symptoms unrelated to the scimitar syndrome and only one case with paroxysmal tachycardia.³ Our patient presented with an uncommon presentation of tachyarrhythmia (supraventricular tachycardia) along with progressive dyspnoea. Though it is rare, supraventricular tachycardia is the most common type of tachyarrhythmia associated with scimitar syndrome.⁸

Scimitar syndrome is commonly associated with other anomalies, mostly cardiac (such as ASD, patent ductus arteriosus, dextrocardia) and other pulmonary/bronchial abnormalities.^{2 3} In our case, scimitar syndrome was associated with a single left pulmonary vein, which to our knowledge has not been reported previously.

Routine chest radiography or echocardiography can be diagnostic, but sometimes, as in our case, the chest X-ray is not typical. In our patient, the diagnosis was made with CT angiography, which is the standard imaging technique to confirm the diagnosis of scimitar syndrome.³

A number of cases of pregnant women with scimitar syndrome had been reported.⁹⁻¹¹ Two patients were diagnosed with scimitar syndrome before the pregnancy and had a benign course during the pregnancy which ended with uncomplicated delivery and no evidence of pulmonary hypertension or heart failure.^{9 10} Though one patient developed severe pre-eclampsia at the 34th week of pregnancy, it was not linked to the presence of scimitar syndrome.¹⁰

The third patient was diagnosed with the infantile form of scimitar syndrome at the age of 3 months and underwent right

Figure 2 CT angiography showing the anomalous vein (scimitar vein).

pneumonectomy. She suffered from severe obstructive lung disease with mild to moderate restrictive lung disease. During her pregnancy, she developed severe dyspnoea and hypoxia, which resulted in preterm delivery by caesarean section.¹¹

Our patient had a stable course during her pregnancy except for the spontaneously aborted supraventricular tachycardia and had a normal delivery without complications. The normal physiological changes during pregnancy might have helped unmask the diagnosis of scimitar syndrome. In all the three cases, CT angiography was used to establish the diagnosis.

Pulmonary hypertension is a common complication in the infantile form of scimitar syndrome, unlike the adult form. Dupuis *et al* reported that 77% of the patients had normal pulmonary pressure, and only 23% had slightly elevated pulmonary pressure (between 30 and 50 mm Hg).³ In most of the cases in that study, the scimitar vein was associated with a hypoplastic right lung, or with an anomalous arterial supply from the aorta with significant left to right shunt, leading to the complication of pulmonary hypertension.³ Our patient did not have an obvious anomalous systemic arterial supply on CT angiogram. Although CT angiography is not the optimal technique to detect an anomalous arterial systemic supply, it was helpful in our case to exclude any significant anomalous supply that can lead to a significant left to right shunt. The patient also had normal mean pulmonary pressure as measured by echocardiography.

There is no consensus on the indication for or the technique of surgical intervention for patients with the adult form of scimitar syndrome, especially those who are asymptomatic or with mild symptoms. Some authors have suggested that surgery is indicated only in patients with pulmonary hypertension or CHF.^{12 13} In addition, surgical outcome is not promising in all cases. Post-operative complications were reported by Alsoufi *et al*¹⁴ to be higher among patients with scimitar syndrome compared with patients with other partial anomalous pulmonary venous connection who underwent surgical repair. Surgical repair for scimitar syndrome carries some serious complications, as per Schramel *et al*,¹² who reported that two out of five patients who underwent surgery for scimitar syndrome developed complications, including pleural empyema, thrombosis and fibrosis. In another report, the surgical repair of a 34-year-old patient with scimitar syndrome was complicated by obstruction of the anatomised vein, which needed a revision of sternotomy.⁵

In the study by Dupuis *et al*,³ the asymptomatic patients with normal pulmonary pressure and no associated cardiac anomaly had a normal life with follow-up and no surgical intervention.³ Similarly, Vida *et al*¹⁵ performed a retrospective study on 44 patients with scimitar syndrome. None of those patients underwent correction of the anomalous scimitar vein; instead, 18% underwent correction of the associated CHD and 34% had embolisation of the anomalous systemic arterial supply if present. After 6 years of follow-up, none of the patients developed signs of CHF, and some had improvement in the respiratory symptoms after correction of the CHD.

It seems from reported studies that the patients with scimitar syndrome who underwent surgery and benefited from it had other indications such as the presence of either CHD or an anomalous arterial systemic supply, which are the main reasons behind CHF or severe pulmonary hypertension.^{2 13 14 16}

Our patient was treated with ablation, which was successful in alleviating her palpitation. Considering that she is now living

a normal life 6 months after the diagnosis and in the absence of associated anomalous arterial supply, the risks of the surgery in our case outweighed the benefits.

Learning points

- ▶ Tachyarrhythmia can be the main presenting symptom of the adult-form scimitar syndrome.
- ▶ Usually it has a benign course, as it is uncommon for the adult form to be associated with significant pulmonary hypertension.
- ▶ There is currently no consensus on the indication, time or type of surgical intervention for the adult form of scimitar syndrome in patients with no or only mild symptoms.

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