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Brief Report

Systemic arterial supply of the right lung with venous drainage to the pulmonary arterial circuit

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Abstract In a girl suffering from "Scimitar syndrome", a rerouting of the scimitar vein was performed at the age of 6 years, but no embolisation of the aberrant systemic vessel was done. She presented with recurring respiratory problems 13 years later. An angiography revealed an invert flow from the aberrant systemic vessel via the right pulmonary artery into the left pulmonary artery. After pneumonectomy, she recovered well.

Keywords: Scimitar syndrome; pneumonectomy; thrombosis; aberrant arterial supply

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"Scimitar syndrome" is characterised by anomalous drainage of one or all right pulmonary veins into the inferior caval vein or rarely into the hepatic, portal, or azygos veins. Associated anomalies comprise various degrees of hypoplasia, malformation or agenesis of the right lung, hypoplasia of the right pulmonary artery, aberrant systemic blood supply of parts of the right lung, and a variety of intracardiac anomalies that commonly include atrial or ventricular septal defects.[1] The treatment ranges from "wait and see" in asymptomatic cases to surgical rerouting of the scimitar vein into the left atrium, or resection of the malformed lung.[1–4]

Short report

Scimitar syndrome with drainage of the whole right lung to the inferior caval vein, atrial septal defect and atypical systemic supply of the right lower lobe was diagnosed in a 6-year-old girl. Closure of the atrial septal defect and intracardiac rerouting of the anomalous vein to the left atrium was done in another hospital. The atypical systemic artery was left for later embolisation. After an uneventful course, the child was discharged. She did not present for follow-up.

The girl, who had experienced normal physical development was presented to our hospital 13 years later with dyspnoea and recurrent bronchopulmonary infections.

Spirometry showed slight restrictive and obstructive changes. There was normal ventilation but a complete lack of perfusion of the right lung as seen on a ventilation–perfusion scan. Echocardiography revealed a dilatation of the right ventricle. There was no sign of any right pulmonary veins draining into the left atrium. The right pulmonary artery, especially during a Doppler echocardiography, could not be pictured.

Aortography and pulmonary angiography showed a large systemic vessel originating from the coeliac artery, feeding the right lung. The calculated left-to-right shunt on oxymetry was 39% (pulmonary-to-systemic flow ratio 1.4). Cardiac catheterisation confirmed the complete absence of right pulmonary venous drainage into the left atrium. There were large-calibre anastomoses between the atypical systemic vessel and the right pulmonary artery permitting a reverse flow and a retrograde drainage of the right lung through the right pulmonary artery into the left one and into the left pulmonary veins (Figs 1 and 2). The pressure in the right pulmonary artery was 35 millimetres of mercury. The significant left-to-right shunt caused volume overload of the right heart.

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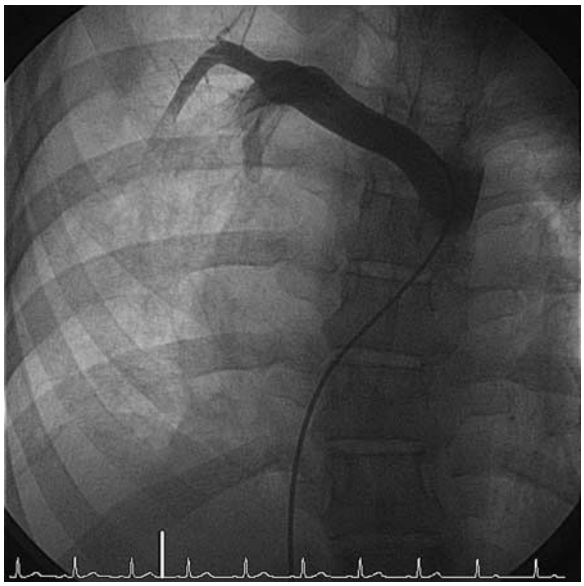

Figure 1.
Selective angiography of the right pulmonary artery demonstrating a reverse flow with washing-out effect in the right bronchial arteries.

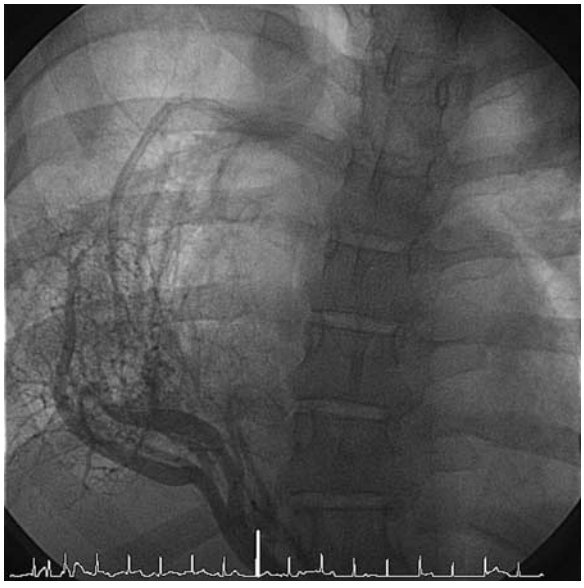

Figure 2.
Aberrant systemic collaterals feeding the right lung with drainage via the right pulmonary artery in the left pulmonary artery.

There seemed no possibility to re-establish a normal pulmonary circulation, and thus right pneumonectomy was performed. The lung showed pleuropulmonary adhesions and marked subpleural angiectasis. The right pulmonary artery was hypoplastic and lacked a truncus in the right upper lobe. No venous drainage of any kind and no signs of previous rerouting of a right pulmonary vein into the left atrium could be identified any more. Three large branches of the atypical systemic artery were dissected in the pulmonary ligament.

In the pathological specimen of the lung, which consisted of only one single lobe, signs of recurrent infection were found. The systemic as well as the pulmonary arteries showed thickening of the walls and abundant arterio-arterial anastomoses. In addition, histomorphologically, there was no sign of a venous system.

The post-operative course was uneventful, with discharge of the patient on the fourth post-operative day.

Discussion

There are two major clinical presentations of the "Scimitar syndrome": The "infantile type" manifests itself in the first month of life with right cardiac failure due to volume overload, recurrent pulmonary symptoms, or failure to thrive and is seldom asymptomatic.[2] The "adult type", however, will often run an asymptomatic course.[1]

In patients with congestive cardiac disorder or recurrent pulmonary infection, surgical or interventional procedures are suggested. Some authors claimed that embolisation of the systemic arteries when present, rather than surgical repair of the scimitar vein, was the best hope for a good clinical outcome in symptomatic infantile patients.[5] Other authors advocate straightforward lung resection or rerouting of the scimitar vein in the left atrium with baffle repair.[2,3] Owing to the aberrant arterial supply via the aortic collateral of the whole right lung, we avoided an embolisation due to feasible massive tissue decay.

After venous rerouting surgery, thrombosis of the vein eventually resulting in fibrotic stricture may develop. A recent multicentre study in Europe in 68 patients revealed that around 15% of the patients after rerouting require reintervention because of scimitar drainage stenosis.[6] Usually, this complication is seen in the early post-operative course. Although successful thrombolysis has been described, the majority of patients undergo lung resection.[1–3]

In our patient, the scimitar vein had been redirected without closure of the systemic artery; 13 years later, a venous drainage or venous system of the right lung could neither be found macroscopically nor histologically. With reverse outflow from the arterialised lung through the main stem of the right pulmonary artery, it is surprising that the patient had no symptoms of note in the interval.

Although arteriovenous fistulae have been described in Scimitar syndrome,[7] to our knowledge this is the first reported case of retrograde venous drainage of the whole lung after rerouting of a scimitar vein.

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