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Brief Report

Scimitar syndrome associated with absence of the right pulmonary artery and a persistent primitive hepatic venous plexus

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Abstract An 18-month-old boy, referred because of an infection of the airways, was found to have a right-sided heart, a hypoplastic right lung, absence of the right pulmonary artery, and persistence of the hepatic venous plexus. The benign association of this unusual variant of the scimitar syndrome and persistence of the hepatic venous plexus needs to be recognised in order to avoid extensive investigations and surgery involving the inferior caval vein.

Keywords: Anomalous pulmonary venous connection; anomalous systemic venous connection; inferior caval vein

THE SCIMITAR SYNDROME IS A RARE CONGENITAL disorder, characterized by anomalous connection of one or more of the right pulmonary veins to the inferior caval vein. The anomalous vein appears like a scimitar-like shadow on the chest X-ray, and classically runs from the middle of the right lung to the cardiophrenic angle. The syndrome has now been described with various associations of anomalous systemic veins, pulmonary arteries, and pulmonary structure. To our knowledge, however, no one has yet reported the association with absence of the right pulmonary artery and persistence of the primitive hepatic venous plexus.

Case report

An 18-month boy was referred to our unit because of respiratory distress. At presentation, his general condition was good, with a saturation of oxygen of 97 percent. Clinical examination showed a mild deformity of the chest, tachypnoea at 50 breaths per minute, raised temperature at 38 degrees Celsius, a purulent infection of the ear, and wheezing in the right lung.

The mother had noticed; since the first months of life, such noisy respiration and, after the age of 1 year, mild dyspnoea and episodes of hypotonia at exercise.

The chest X-ray showed a right-sided heart, possible absence or atelectasia of the superior lobe of the right lung, and a normal left lung. Even in retrospect, there was no typical "scimitar" shadow to be seen.

A lung scan showed hypoplasia of the right lung, with absence of the superior and median lobes, deviation of the trachea to the right, and atresia of the right superior bronchus. It also showed absence of the right pulmonary artery, an anomalous connection between the inferior caval vein and the right atrium, a large hemiazygos vein communicating with a paravertebral venous plexus, and a large systemic collateral artery, originating from the abdominal aorta, and vascularising the right lung.

Fibroscopy showed a normal left bronchus, a globally hypoplastic right bronchus with atresia of the right lobar bronchus, and inflammation and fluid secretions in the right bronchus. Spirometry demonstrated a diminished ventilatory volume and impaired bronchodilation. Abdominal sonography showed normal biliary ducts, normal kidneys, but revealed an unusual anatomy of the intra-hepatic veins.

Echocardiography confirmed the right-sided location of the heart, but otherwise showed normal

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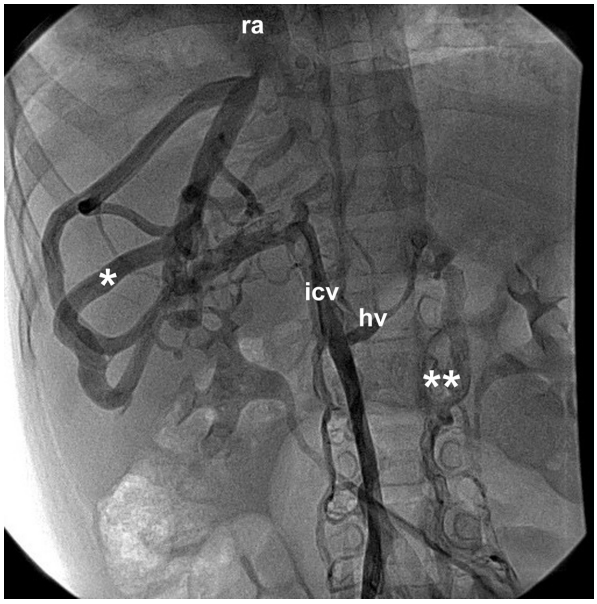

Figure 1.
A racemose network of venous channels (*) connects the intrahepatic portion of inferior caval vein (icv) to the right atrium (ra). A large hemiazygos vein (hv) is connected with the lumbar and paravertebral plexuses (**).

intracardiac anatomy, with no atrial septal defect, but confirmed the absence of the right pulmonary artery and veins. We made a preliminary diagnosis of scimitar syndrome.

Cardiac catheterisation confirmed the absence of the right pulmonary artery. The pulmonary arterial pressure was mildly elevated, with systolic, diastolic and mean pulmonary arterial pressures of 40, 9, and 23 millimetres of mercury, respectively. The saturations of oxygen were normal in the right atrium, right ventricle and pulmonary arteries, at 70 percent, but elevated in the intra-hepatic portion of the inferior caval vein, at 96 percent. The inferior caval vein itself was globally hypoplastic, being almost interrupted at its junction with the right atrium. An extensive racemose network of venous channels connected the inferior caval vein to the hepatic veins, which also had a stenotic communication with the right atrium (Fig. 1). The entirety of the right lung received its arterial supply from a large systemic artery arising from the abdominal aorta. The pulmonary venous return from the hypoplastic right lung reached the hepatic venous plexus and eventually, through the inferior caval vein, the right atrium (Fig. 2).

Antibiotic therapy was started, and was combined with strict surveillance. The patient is at present doing well. Removal of the right lung is considered to be the only effective therapeutic option, but thus far we have chosen to delay surgery, hoping to avoid the early development of scoliosis.

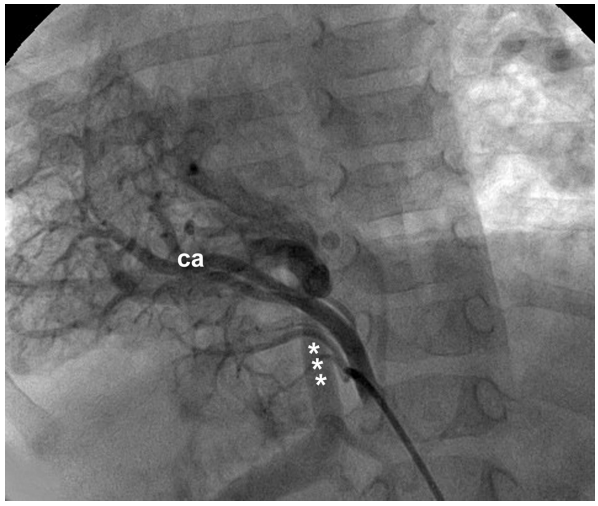

Figure 2.
The right lung is vascularised by a large systemic collateral artery (ca) originating from the abdominal aorta. The pulmonary venous return from the hypoplastic right lung drains to the hepatic venous plexus (***) and eventually, through the inferior caval vein, to the right atrium.

Discussion

Scimitar syndrome, first described by Neill et al. in 1960,[1] is considered to represent a developmental anomaly of the right lung. The clinical spectrum ranges from severely ill infants to asymptomatic adults. The true incidence of the disorder is unknown, since the syndrome may remain undetected in asymptomatic patients. Infants in whom the diagnosis is made, nonetheless, may present with severe symptoms and have a poor prognosis. Indeed, severe pulmonary hypertension, associated cardiac malformations, and large systemic collateral arteries feeding the right lung are almost invariably present.[2]

Repeated haemoptysis, and infections of the sequestrated lung, may reveal the anomaly.[3] Definitive treatment can be achieved by lobectomy, pneumonectomy, or embolisation of the collateral artery.[2–4] The diagnosis, once made by observing the typical "scimitar" shadow, is now based on the recognition of the anomalous pulmonary systemic and venous connections. Indeed, in some patients, as in our case, the chest X-ray does not show a scimitar shadow.

The scimitar syndrome has several associations. The most frequent are partial absence of right lung, horseshoe lung, and absence of the right pulmonary artery.[3] More rarely, the inferior caval vein may be hypoplastic or interrupted, and anomalous systemic venous connections have been reported.[4] A patent infrahepatic inferior caval vein, albeit underdeveloped, with a persistent hepatic venous plexus, is a distinct and very rare congenital systemic venous anomaly.

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Embryologically, the inferior caval vein can be divided into a hepatic segment, derived from the proximal part of the right vitelline vein and the hepatic sinusoids, a prerenal segment, derived from the right subcardinal vein, a renal segment, formed by anastomosis of the primitive subcardinal and supracardinal veins, and a postrenal segment, derived from the right supracardinal vein. The hepatic venous plexus may represent the precursor of the hepatic veins, and contributes to the formation of the intrahepatic portion of the inferior caval vein. Persistence of primitive hepatic venous plexus as the terminal part of the inferior caval vein is accompanied by underdevelopment of the infrahepatic portion of the inferior caval vein. It can be isolated, or associated with other anomalies.[4] The association with scimitar syndrome has been reported only a few times, with or without stenosis of the inferior caval vein.[4,5]

This rare association of the scimitar syndrome with persistence of the primitive hepatic plexus, and recognition of its benign nature, has to be held in mind in order to avoid extensive investigations of abdominal vascularisation and the hepatobiliary system. The diversion of the flow of blood away from the hepatic inferior caval vein may lead to the incorrect diagnosis of isolated inferior caval venous stenosis, and to attempted interventional or surgical procedures on the systemic veins that are doomed to failure.

References

1. Neill CA, Ferencz C, Sabiston DC, Sheldon H. The familial occurrence of hypoplastic right lung with systemic arterial supply and venous drainage: "scimitar syndrome". Johns Hopkins Med J 1960; 107: 1–21.
2. Gao YA, Burrows PE, Benson LN, Rabinovitch M, Freedom RM. Scimitar syndrome in infancy. J Am Coll Cardiol 1993; 22: 873–882.
3. Dupuis C, Remy J, Remy-Jardin M, Rey C, Brevietre GM, Kerboub A. Scimitar syndrome with anatomical and functional absence of the right pulmonary artery. Apropos of 4 cases. Arch Pediatr 1995; 2: 347–352.
4. MacDonald C, Mikhailian H, Yoo SJ, Freedom RM, Adatia I. Angiographic findings of persistent primitive hepatic venous plexus with underdevelopment of the infrahepatic inferior vena cava in pediatric patients. Am J Roentgenol 2000; 175: 1397–1401.
5. Madan N, Moore JW. Unusual angiographic appearance of scimitar syndrome associated with primitive hepatic venous plexus. Circulation 2002; 105: e78.

https://doi.org/10.1017/S1047951105000429 Published online by Cambridge University Press