Cardiology in the Young (2016), **26**, 615–618
doi:10.1017/S1047951115001687

Brief report

Meandering right pulmonary vein associated with severe and progressive "idiopathic-like" pulmonary hypertensive vascular disease

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Abstract Congenital anomalies of the pulmonary veins are rare. Meandering right pulmonary vein, considered a part of the Scimitar syndrome spectrum, is often an incidental finding during chest imaging. We present the case of a 4-year-old girl diagnosed with meandering pulmonary vein, who developed pulmonary hypertensive disease with an aggressive course, in spite of absence of hypoxia or elevated pulmonary wedge pressure.

Keywords: Meandering pulmonary vein; scimitar syndrome; pulmonary hypertension

Received: 22 June 2015; Accepted: 4 August 2015; First published online: 16 September 2015

Congenital anomalies of lung and vascular structures are rare. A varied spectrum of pathologies has been described[1–5] within the syndrome called "congenital pulmonary venolobar syndrome". Pseudo-scimitar syndrome, or "meandering" right pulmonary vein, consists of an anomalous right pulmonary vein, which takes a tortuous route through the lung, but terminates with normal drainage into the left atrium. This termination differentiates the meandering pulmonary vein from other entities such as Scimitar syndrome. Although meandering right pulmonary vein has been described in some cases with an association with right lung hypoplasia, it is often an incidental finding.[3] In the Panama classification of Paediatric Hypertensive Pulmonary Vascular Disease,[6] pulmonary vein anomalies were considered to be a part of pulmonary hypertensive vascular disease associated with CHD. In the National Institute for Health and Care Excellence updated classification,[7] pulmonary hypertensive vascular disease associated with abnormalities in lung development was included in group III – pulmonary hypertension associated with lung disease or hypoxia.

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We report the case of a 4-year-old girl with an isolated meandering right pulmonary vein, with no other structural lung abnormalities, whose progressive pulmonary hypertensive vascular disease could neither be explained by the haemodynamic effect of the meandering pulmonary vein on pulmonary wedge pressure nor by hypoxia.

Case report

A 4-year-old girl was referred to the Paediatric Cardiology Department of our hospital, diagnosed with pulmonary hypertension. She had a history of repeated spastic bronchitis from the age of 1 year, which led to hospitalisations at the ages of 2 and 4 years. She had no other symptoms and had experienced normal growth and development.

Her physical examination was unremarkable except for a split-second heart sound, with slightly increased P2 sound. Oxygen saturation was higher than 95% in room air; electrocardiogram presented sinus rhythm and 90° QRS axis. Echocardiography showed mild flow acceleration in both superior pulmonary veins, type I interventricular septum with paradoxical movement, and mild tricuspid insufficiency, with an estimated systolic right ventricular

pressure of 48 mmHg – 45% of systemic blood pressure. Sleep breathing disorders, thyroid or immune anomalies, hepatic or portal abnormalities, and human immunodeficiency virus infection were ruled out. Frontal chest radiograph revealed a shadow along the right heart border (Fig 1), identified in a CT pulmonary angiogram as an anomalous venous structure connecting the superior and inferior right pulmonary veins, describing a tortuous route through the lung (Fig 1). The dilated inferior pulmonary vein drained into the left atrium. The left pulmonary veins were normal, except for a mild stenosis of the upper left pulmonary vein. Cardiac catheterisation found mildly elevated pulmonary pressure, mild left upper pulmonary vein stenosis – 2 mmHg mean gradient – and normal bilateral pulmonary wedge pressure (Supplementary table 1). Flow was quantified by MRI, with 35% flow to the left lung and 65% to the right lung. Right pulmonary artery hypoplasia and aberrant arterial systemic lung supply were ruled out.

Treatment of her bronchitis was intensified, adding maintenance inhaled budesonide, with significant decrease in bronchoconstriction episodes. During the next 3 years, in spite of dramatic improvement of her respiratory symptoms, her pulmonary hypertension progressed rapidly, with effort dyspnoea unrelated to bronchoconstrictive episodes – NYHA functional class IIIa. Echocardiography showed increased pulmonary pressure, hypertrophy, dilation, and dysfunction of the right ventricle. Repeated cardiac catheterisation revealed severe pulmonary hypertension unresponsive to nitric oxide, normal pulmonary wedge pressure (Supplementary table 1), pruning, and loss of peripheral vessels on angiography, showing progression compared with the angiographies obtained previously (Fig 2). Treatment with sildenafil was initiated, and was increased up to 40 mg t.i.d., with poor improvement in her functional class. Ambrisentan 7.5 mg o.d. was included 6 months later, with significant improvement in her functional class (NYHA I), which had remained stable after 4 years of combined therapy.

Discussion

Congenital pulmonary venolobar syndrome refers to a wide spectrum of pulmonary developmental anomalies that involve abnormal connections of the pulmonary parenchyma, the pulmonary and systemic vasculature, and, rarely, the gastrointestinal tract. Anomalies involved in this syndrome may appear in isolation or in combination, and thus the clinical presentation of this syndrome is variable, depending mainly on the presence of associated cardiac abnormalities.[1–5] "Scimitar syndrome", described in 1960, consists of variable combinations of hypoplasia of the right lung and right pulmonary artery, dextroposition of the heart, anomalous arterial supply to the right lower lobe, and anomalous pulmonary venous drainage of the right lung into the inferior caval vein[4] causing the characteristic "scimitar sign" – a Turkish sword-shaped shadow along the right heart border on chest X-ray. Infant forms, presenting with respiratory insufficiency, heart failure, or pulmonary hypertension, are usually associated with systemic arterial supply (sequestrum), severe right lung hypoplasia, and abnormal drainage of the right pulmonary vein into the inferior caval vein. In the "adult form", patients can remain asymptomatic and have an incidental diagnosis through a chest X-ray.

On the contrary, meandering right pulmonary vein – pseudo-scimitar syndrome, scimitar variant[3,5] – which can also associate the classical right parasternal shadow in the chest X-ray, is rarely associated with severe lung or heart malformations.

Our patient presented progressive pulmonary hypertension, with normal pulmonary wedge pressure and without sustained hypoxia or hypercarbia, behaving like idiopathic pulmonary hypertension, which responded to combined therapy with phosphodiesterase-5 inhibitors and endothelin receptor antagonist. Several publications[8,9] have reported pulmonary hypertension associated with scimitar syndrome and horseshoe lung associated with significant heart anomalies; however, to

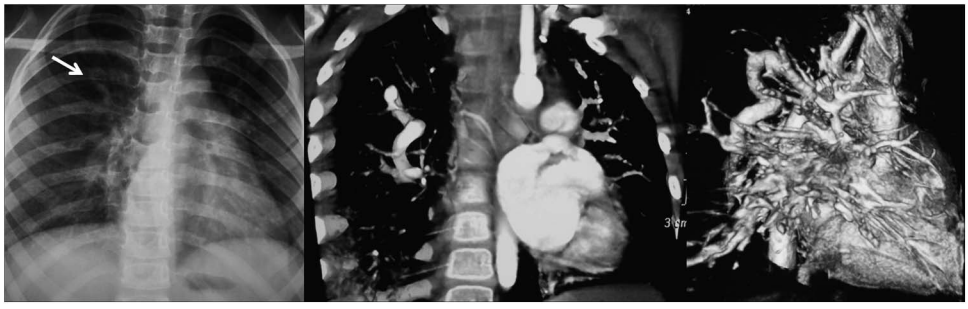

Figure 1.
Left: chest X-ray showing the shadow of the meandering right pulmonary vein. Centre and right: CT scan images of the meandering right pulmonary vein.

https://doi.org/10.1017/S1047951115001687 Published online by Cambridge University Press

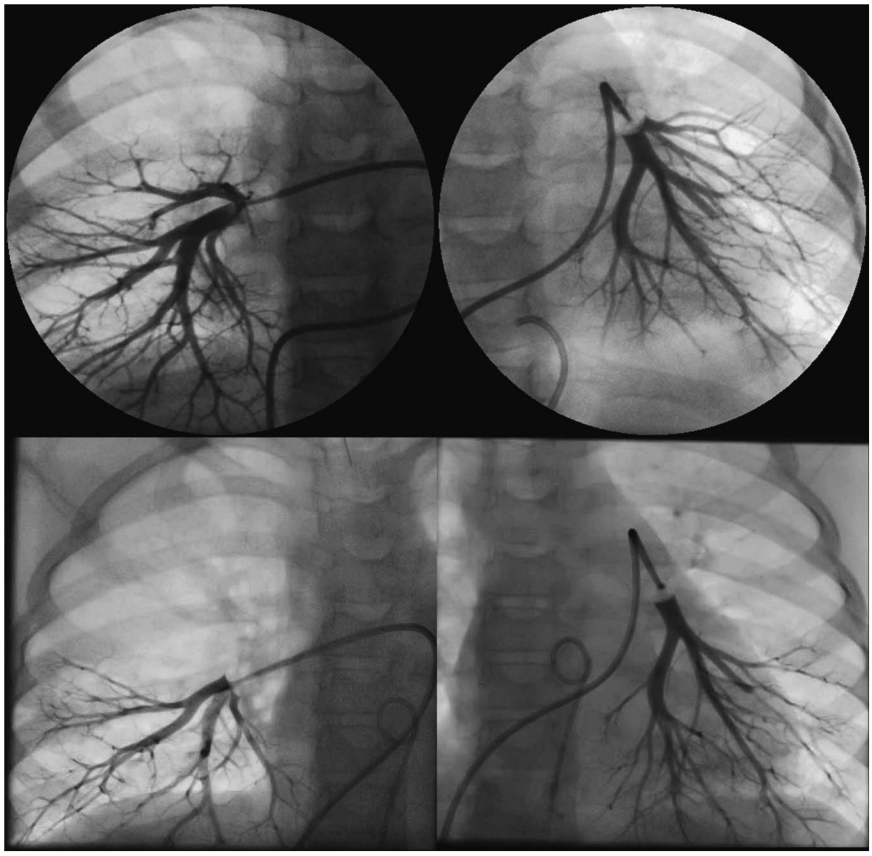

Figure 2.
Pulmonary wedge angiographies at 4 (up) and 7 (down) years of age, showing worsening of the tapering and pruning of the distal vessels, with remarkable loss of distal vascular bed at the age of 7 years.

the best of our knowledge, there are no reports of isolated meandering pulmonary vein associated with severe pulmonary hypertension. Mechanisms linking congenital abnormalities of the pulmonary veins and the development of pulmonary hypertension are not completely understood.[10] We hypothesise that the prenatally conditioned morphological abnormalities in the pulmonary veins and lung development could be associated with histological abnormalities of the arterioles, capillaries, lymphatics, and venules and/or to disrupted molecular pathways causing progressive vascular re-modelling, although this cannot be proved without lung tissue evaluation.

Congenital anomalies of the pulmonary vessels can go unnoticed without the use of an imaging technique (MRI or CT scan), which should be included in the initial evaluation of every pulmonary hypertension case. In light of cases like ours, clinical and echocardiographical follow-up of these patients is mandatory, even if the initial evaluation does not suggest the severity of the pulmonary hypertension. Some patients with pulmonary hypertension associated with congenital anomalies of the pulmonary veins[6] can benefit from treatment with phosphodiesterase-5 inhibitors and endothelin receptor antagonists.

Acknowledgement

None.

Financial Support

This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Conflicts of Interest

None.

Ethical Standards

The authors assert that all work reported complies with the ethical standards of the Helsinki convention, and consent has been granted by the patient's family.

Supplementary material

To view supplementary material for this article, please visit http://dx.doi.org/10.1017/S1047951115001687

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https://doi.org/10.1017/S1047951115001687 Published online by Cambridge University Press