Case report

An unusual variant of scimitar syndrome predisposing to recurrent pneumonia

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ABSTRACT

Scimitar syndrome is a rare constellation of congenital conditions pertaining to partial anomalous pulmonary venous return. Radiographically, these anomalous pulmonary veins can resemble a scimitar, and have heterogeneous clinical presentations. We present an unusual case of scimitar syndrome with associated recurrent pneumonia. We will briefly review the literature on scimitar syndrome as well as discuss how a predisposition to recurrent pneumonia may develop in this unusual variant of a rare clinical entity.

1. Introduction

Scimitar syndrome is one of a multitude of rare congenital pulmonary venolobar syndromes that is named for its characteristic radiographic finding—the shadow of an abnormal pulmonary vein which resembles a Turkish sword called a scimitar [1]. With an estimated prevalence of 2 per 100,000, it is classically defined by partial anomalous pulmonary venous return (PAPVR) associated with hypoplastic lung, both of which most commonly occur on the right side [2,3]. Despite this classic definition, scimitar syndrome remains an anatomically and clinically diverse condition [4]. A number of additional anomalies can exist as part of this syndrome, including an anomalous systemic feeding artery arising from the aorta, which supplies the same area of lung that is drained by the anomalous pulmonary vein, tracheobronchial abnormalities, and even dextrocardia [5]. Most often, the anomalous pulmonary vein drains into the inferior vena cava (IVC), but there are also case reports of drainage into the hepatic vein, right atrium, and the left atrium. The heterogeneity of this syndrome is further underscored by the fact that some cases require urgent surgical intervention at birth as a result of significant left-to-right shunting, and some cases cause no symptoms and are discovered incidentally in adulthood [6]. We present an unusual case of variant scimitar syndrome with bronchoscopic findings and an intriguing clinical presentation.

2. Case report

A 37-year-old woman with a history of scimitar syndrome (incidentally discovered in her twenties), presented for evaluation of exertional dyspnea and recurrent pneumonias since childhood. She noted however, that both of these symptoms appeared to be worsening over the previous two years. Her initial evaluation at our facility included a chest X-ray (Fig. 1) computed tomography (CT) scan of the chest with intravenous contrast and three-dimensional reconstruction (Fig. 2). Her radiograph demonstrated normal heart size, slightly enlarged central pulmonary arteries and a right para-cardiac tubular opacity extending from the right hilum towards the medial right hemidiaphragm. Computed tomography revealed PAPVR of the right upper lobe and right middle lobe to the IVC. Further, the left-sided pulmonary veins were normal and drained into the left atrium. The main pulmonary arteries were enlarged. There was no evidence of lung hypoplasia. However, there was an incomplete major fissure and an absent minor fissure.

The right mainstem bronchus trifurcated immediately inferior to the enlarged right pulmonary artery into the lobar bronchi without an intervening bronchus intermedius. The right middle lobe bronchus was diminutive. Cardiac MR (obtained previously) showed right chamber enlargement and evidence of left to right shunting. (Fig. 3). Bronchoscopy demonstrated additional abnormalities in her tracheobronchial tree anatomy (Fig. 4). Her right upper lobe bronchus was noted to originate from the bronchus intermedius. She was found to have 80% obstruction of the right mainstem bronchus due to excessive dynamic airway collapse with tidal respiration. Cultures from bronchoalveolar
lavage did not identify a culprit organism and she was treated for community acquired pneumonia. As can be seen in Fig. 5, she did however appear to have RUL takeoff obstruction from her enlarged scimitar vein. Moreover, her RML takeoff was congenitally diminutive, but did not show evidence of RML syndrome (such as bronchiectatic consolidation and volume loss).

Since the time of our initial encounter, the patient underwent right heart catheterization, which revealed mild pulmonary hypertension with a mean pulmonary artery pressure of 25 mmHg and a wedge pressure of < 12 mmHg and a Qp:Qs of 1.7, suggesting WHO group 1 pulmonary hypertension from congenital heart disease with concurrent left-to-right shunt. In the months that followed, she was noted to have progressive pulmonary artery dilation and pulmonary over circulation with resultant dyspnea. In an attempt to minimize dyspnea, the patient

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Fig. 1. Postero-anterior (A) and lateral (B) chest radiographs. A curvilinear tubular opacity (arrows) is seen in the right lung base anteriorly corresponding to the anomalous pulmonary vein on cross-sectional imaging. No appreciable right lung volume loss is seen.

Fig. 2. Contrast-enhanced computed tomography. Maximum intensity projection coronal image (A) demonstrates a large anomalous pulmonary vein (arrow) which drains the right upper and middle lobes to the inferior vena cava. An incomplete right major fissure (arrow) is seen centrally (B). Volume rendered anterior (C) and posterior (D) images re-demonstrate the large anomalous pulmonary vein connecting to the inferior vena cava, absence of the right superior pulmonary vein, and presence of a right inferior pulmonary vein which drained the right lower lobe to the left atrium.
underwent surgical repair of scimitar vein with translocation of scimitar vein to the left atrium from the inferior vena cava and closure of patent foramen ovale. Approximately four months following this surgery, she has not had recurrent pneumonia, though her dyspnea still persists.

3. Discussion

The patient’s anatomical findings represent a variant of scimitar syndrome: PAPVR with bronchial and fissural anomalies without lung hypoplasia or a systemic feeding artery. Further, her associated right tracheobronchial tree anatomy, the relationship of the bronchial trifurcation to the enlarged right pulmonary artery, and the diminutive right middle lobe bronchus predisposes to recurrent pneumonia due to impaired drainage [7,8]. Due to the rarity of this syndrome, there is not a well-defined mechanism by which recurrent pneumonia in scimitar syndrome occurs. One potential mechanism for recurrent pneumonia is that the aberrant bronchial take-off angles within these hypoplastic lungs portend to diminution of radial traction, and in-turn, impairment of secretion clearance. As seen in our case, recurrent pneumonia might arise from extrinsic compression of the airway from the scimitar vein, although this may not be a unifying mechanism. Other considerations could include abnormal lymphatic drainage and impaired macrophage or lymphocyte recruitment to the affected area. Further defining this mechanism of disease will be difficult and may even vary case-to-case. Collectively however, the variants of scimitar syndrome and other hypoplastic lung syndromes represent a unique opportunity to appreciate how abnormalities within the bronchial tree and the associated blood drainage may predispose to recurrent infection.

Author contributions

All authors have contributed equally to the concept and design of this manuscript.
Funding

The authors have no financial disclosures and there was no research, pharmaceutical or industry funding provided for this report.

References

[1] C. Dupuis, L.A. Charaf, G.M. Breviere, P. Abou, M. Remy-Jardin, G. Helmius, The “adult” form of the scimitar syndrome, Am. J. Cardiol. 70 (4) (1992) 502–507.
[2] E. Gavazzi, M. Ravanelli, D. Farina, M.E. Chiari, R. Maroldi, Scimitar syndrome: comprehensive, noninvasive assessment with cardiovascular magnetic resonance imaging, Circulation 118 (3) (2008) e63–64.
[3] C.C. Wang, E.T. Wu, S.J. Chen, et al., Scimitar syndrome: incidence, treatment, and prognosis, Eur. J. Pediatr. 167 (2) (2008) 155–160.
[4] A.R. Opotowsky, G.D. Webb, A battle in the crusade to understand scimitar syndrome, Eur. Heart J. 39 (12) (2018) 1012–1014.
[5] A.A. Korkmaz, C.E. Yildiz, B. Onan, M. Guden, G. Cetin, K. Babaoglu, Scimitar syndrome: a complex form of anomalous pulmonary venous return, J. Card. Surg. 26 (5) (2011) 528–534.
[6] I. Bo, J.S. Carvalho, E. Cheasty, M. Rubens, M.L. Rigby, Variants of the scimitar syndrome, Cardiol. Young 26 (5) (2016) 941–947.
[7] T. Guðjartsson, G. Guðmundsson, Middle lobe syndrome: a review of clinicopathological features, diagnosis and treatment, Respiration 84 (1) (2012) 90–86.
[8] R.J. Albo, O.F. Grimes, The middle lobe syndrome: a clinical study, Dis. Chest 50 (5) (1966) 509–518.

Fig. 4. (A) Significant obstruction of the right mainstem bronchus with quiet breathing. (B) The right upper lobe bronchus appears to come off of the bronchus intermedius anteriorly.

Fig. 5. Contrast-enhanced computed tomography. Sagittal oblique image (A) demonstrates mass effect by the enlarged right pulmonary artery on the origin of the right upper lobe bronchus (arrow) and immediate trifurcation of the right mainstem bronchus. Coronol oblique image (B) demonstrates the proximity of the enlarged right pulmonary artery to the proximal right upper lobe bronchus inferiorly. Axial minimum intensity projection image (C) demonstrates the branching of the right mainstem bronchus with a diminutive right middle bronchus (arrow).