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CASE REPORT

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Scimitar syndrome, bronchiectasis, haemoptysis and a pneumonectomy

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Abstract

Scimitar syndrome is a rare condition characterized by partial or complete anomalous pulmonary venous drainage of the lung to the inferior vena cava, right lung hypoplasia and dextroposition of the heart. Haemoptysis is uncommon in adults, although the clinical spectrum is wide. We report a case of a 38-year-old male with scimitar syndrome who had low grade haemoptysis persisting over several years secondary to bronchiectatic changes in his hypoplastic right lung. Conservative measures to manage haemoptysis were unsuccessful and the patient proceeded to bronchial artery embolization. The post-procedure course was complicated by lung infarction and the patient ultimately required pneumonectomy. Deformities of the blood vessels and lungs are complex in scimitar syndrome. Bronchial artery embolization should be approached cautiously to protect pulmonary perfusion.

KEYWORDS
bronchial artery embolization, bronchiectasis, haemoptysis, pneumonectomy, scimitar syndrome

INTRODUCTION

Scimitar syndrome, also known as hypogenetic lung syndrome, is a rare condition comprising congenital heart and lung abnormalities. It is characterized by partial or complete anomalous pulmonary venous return to the inferior vena cava.[1] The name of the syndrome derives from the paracardial curvilinear shadow created by the anomalous pulmonary vein on imaging of the chest (scimitar: single-edged backsword, or sabre, with a curved blade), a sign that is pathognomonic of the syndrome.[1]

This case report describes a 38-year-old man with scimitar syndrome, anomalous systemic arterial supply to the right lung, an attenuated right pulmonary artery, and associated haemoptysis secondary right upper lobe bronchiectasis for which he underwent bronchial artery embolization (BAE). In our case, BAE resulted in lung infarction with cavitatory transformation and ultimately, pneumonectomy. We have no knowledge of a similar case with this trajectory.

CASE REPORT

A European male was noted to have a hypoplastic right lung at birth. He was formally diagnosed with scimitar syndrome at age 19 when he presented with life-threatening haemoptysis of more than 0.5 litres. He had a hypoplastic right pulmonary artery, no right pulmonary venous drainage to the left atrium, and systemic collaterals to a hypoplastic right lung (Figure 1A,B). He had no evidence of pulmonary hypertension on echocardiography or cardiac MRI.

Haemoptysis was attributed to right upper lobe bronchiectasis and associated hypertrophied bronchial arteries in the right upper thoracic area (Figure 1C). He remained otherwise asymptomatic of scimitar syndrome. A repair of partial anomalous pulmonary venous drainage was not indicated because of absence of left-to-right shunt and normal right ventricular size. Intermittent low-volume haemoptysis persisted without need for further hospital admissions.

At age 37, the patient had several presentations to the Emergency Department; haemoptysis became unacceptable to the patient due to impact on quality of life. He proceeded

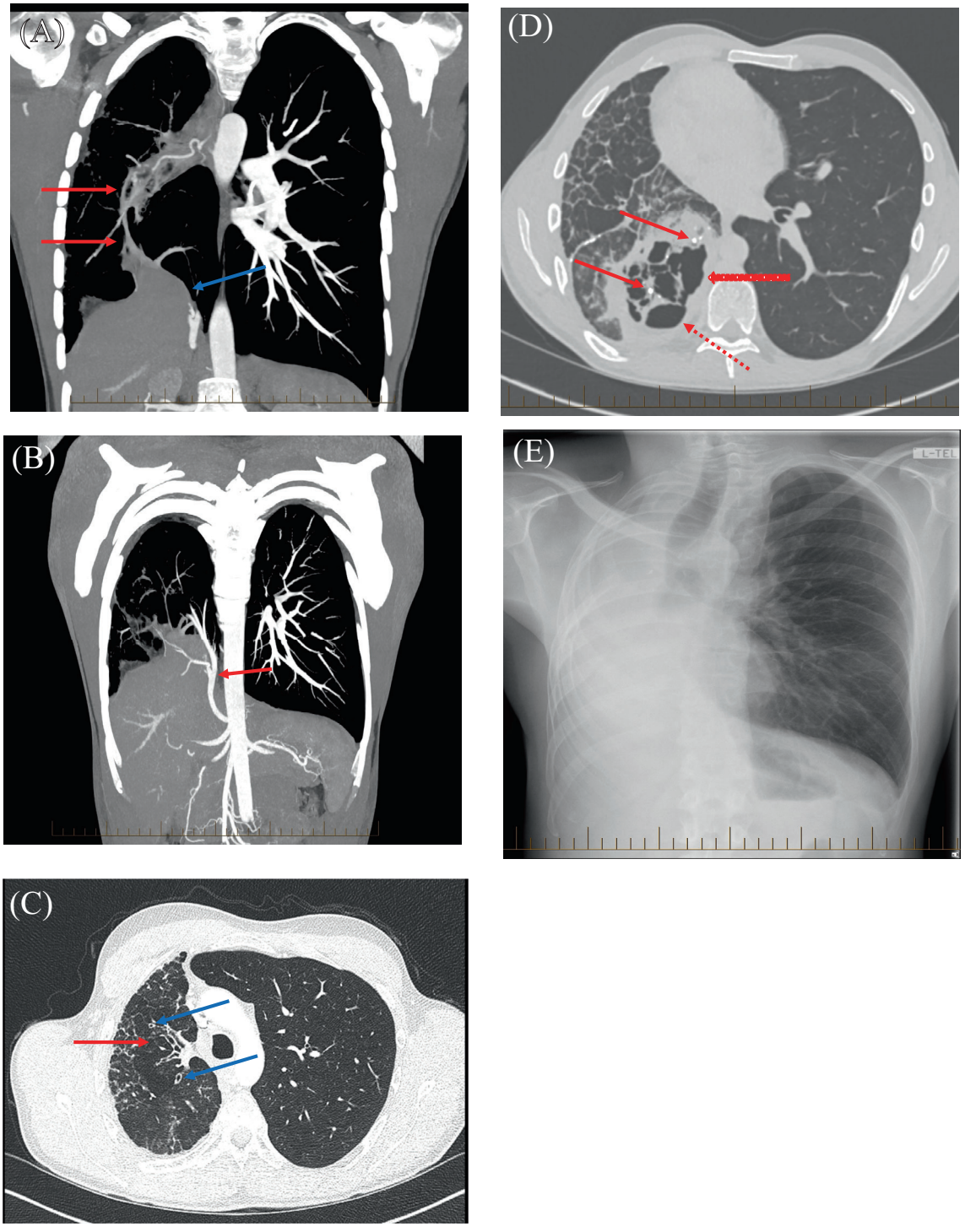

FIGURE 1 Progression of treatment. (A) CT angiography showing anomalous pulmonary venous drainage of the right lung (arrows) into the inferior vena cava (blue arrow; not clearly shown on CT scan). The left lung is normal; (B) Pulmonary angiography at the time of embolization. The arrows show the systemic collateral arteries from right inferior phrenic artery, which partially supplied the basal segment of the right lower lobe; (C) CT showing bronchiectasis in the right upper lobe (blue arrows) with associated hypertrophied arteries in the right upper thoracic area (red arrow). (D) Non-contrast CT scan of the chest showing a necrotic right lower lobe (dashed arrows) including Histoacryl-Lipiodol mixture (solid arrows) in the right bronchial artery and aberrant bronchial artery; (E) Chest x-ray 1 year following right pneumonectomy.

to BAE of the vessels supplying the segment of lung with bronchiectatic changes likely causing haemoptysis. The following arteries were embolized using Histoacryl-Lipiodol mixture: right bronchial artery, an aberrant bronchial artery from the right internal mammary artery, and the third and fourth intercostal arteries. Systemic collateral arteries from right inferior phrenic artery, which were partially supplying the basal segment of the right lower lobe, were also

TABLE 1 Lung function testing results before and after pneumonectomy.

Date	FEV1/FVC	Total lung capacity	DLCO	pO2 or saturations on room air
Before pneumonectomy				
11/10/2007	2.01/3.07	5.65 (83%)	26.1 (75%)	79 mmHg
14/10/2009	1.18/3.19	5.51 (83%)	22.8 (67%)	90 mmHg
06/07/2012	2.05/3.50	5.48 (82%)	24.7 (74%)	86 mmHg
22/01/2015	2.00/3.79	5.76 (86%)	25.7 (79%)	91 mmHg
11/05/2017	1.87/3.53	5.73 (86%)	23.9 (74%)	–
25/10/2019	2.06/3.55	5.72 (87%)	23.9 (82%)	–
After pneumonectomy				
12/01/2021	–	–	–	97 mmHg
16/07/2021	1.70/2.41	–	–	97%
02/11/2021	1.53/2.50	3.87 (59%)	19.7 (69%)	97%
20/12/2022	1.66/2.57	3.89 (59%)	19.5 (68%)	96%

embolized using spherical embolic agent (Embozene Microspheres 900 μm) and Histoacryl-Lipiodol mixture.

Two weeks following BAE, the patient presented febrile and with chest pain. He did not respond to intravenous cefuroxime and piperacillin/tazobactam, oral doxycycline and prednisone. CT chest 3 weeks following BAE revealed cavitatory changes in the areas of embolization representing infarction with a possible superimposed infection (Figure 1D).

The patient remained symptomatic and proceeded to a right pneumonectomy. Pneumonectomy was deemed preferable to lobectomy. A lobectomy would have been challenging in the setting of infarcted tissue; securing pulmonary venous drainage after lobectomy would have been difficult. The patient required a prolonged Intensive Care Unit admission owing to difficulties with pain management. He was discharged on day 12. The patient remains well a year following pneumonectomy and is back at work (Figure 1E). Lung function test results are presented in Table 1.

DISCUSSION

Scimitar syndrome is rare, with an estimated prevalence of one to three cases per 100,000 births per year.[1] Diagnosis earlier in life generally conveys a poorer prognosis.[2] Diagnosis after infancy is usually because of exertional dyspnoea and frequent pulmonary infections. Infections predominantly affect the right lower lobe because of the abnormal arterial supply and venous drainage.[3] Scimitar syndrome has a variable presentation ranging from asymptomatic, with an isolated finding and benign outcome, to severe, with heart failure, pulmonary hypertension and respiratory insufficiency.[2]

In our case, scimitar syndrome was diagnosed following presentation with life-threatening haemoptysis; this is a rare presentation preceding diagnosis. Possible mechanisms of haemoptysis in scimitar syndrome include rupture of hypertrophied systemic pulmonary anastomosis, or bleeding from a bronchiectatic segment in the hypoplastic lung. BAE works on the principle of selectively embolizing hypertrophied bronchial arteries and non-bronchial systemic collaterals that are recruited secondary to chronic lung inflammation.

There is a high incidence of postoperative pulmonary venous obstruction and diminished perfusion of the right lung after surgical intervention in patients with scimitar syndrome. In our case, BAE of systemic collateral arteries from the right inferior phrenic artery contributed to the compromised pulmonary perfusion and subsequent infarction. Lobectomy or pneumonectomy are indicated patients with recurrent infections, diffuse bronchiectasis, persistent haemoptysis, or marked hypoplasia of the right lung.[4] In a case series published by the European Congenital Heart Surgeons, the authors reported 68 who underwent surgery, eight patients underwent a pneumectomy with an operative mortality of 33%.[5] The outcome of a pneumonectomy was not planned, however, in this case, lobectomy or pneumonectomy could have been considered as first line management for haemoptysis without even attempting BAE.

In conclusion, malformations of the blood vessels and lungs are complex in scimitar syndrome. BAE should be approached cautiously to protect pulmonary perfusion.

AUTHOR CONTRIBUTIONS

All authors have made important intellectual contributions to manuscript draft revisions. All authors have read and approved the final manuscript.

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CONFLICT OF INTEREST STATEMENT

None declared.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ETHICS STATEMENT

The authors declare that appropriate written informed consent was obtained for the publication of this manuscript and accompanying images.

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