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CASE REPORT

Incidental meandering right pulmonary vein, literature review and proposed nomenclature revision

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Abstract

We report a case of an anomalous pulmonary vein on chest X-ray resembling a scimitar sign in an 80-year-old female undergoing investigation of syncope. Multislice computed tomography (CT) with multiplanar reformatting and maximum intensity projections demonstrated an aberrant right inferior pulmonary vein coursing inferomedially towards the diaphragm before turning superiorly and draining normally into the left atrium. The diagnosis of an incidental meandering right pulmonary vein was established. The case is used to review the literature on this rare pulmonary anomaly, including pathogenesis, its relationship with scimitar syndrome and scimitar variant, and diagnosis, with an emphasis on the role modern CT techniques can play in non-invasive diagnosis. A revision to the nomenclature of pulmonary vascular anomalies is proposed to help reduce confusion in the literature.

Key words: Incidental findings; Pulmonary veins/abnormalities; Scimitar syndrome/radiography; Tomography; X-ray computed

Core tip: This case highlights the chest X-ray features of scimitar syndrome are not diagnostic and a meandering right pulmonary vein (MRPV) should be considered in the differential diagnosis. The nomenclature used in the literature to describe these pulmonary vascular anomalies is inconsistent. We therefore propose a revision to the nomenclature to avoid confusion. Differentiation between pulmonary vascular anomalies is required to help decide whether treatment is necessary. Modern multislice computed tomography technology allows clear depiction of the vascular connections and associated anatomy, and has superseded invasive pulmonary angiography and cardiac catheterization as the investigation of choice for MRPV.

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INTRODUCTION

Meandering right pulmonary vein (MRPV) is a rare pulmonary vascular anomaly. Cases are often confused with the more common scimitar syndrome as both conditions consist of an anomalous right pulmonary vein, taking a circuitous route through the lung, which usually results in a scimitar sign on chest X-ray. However in contrast to scimitar syndrome, the MRPV terminates normally in the left atrium, rather than the inferior vena cava (IVC). We report a case of an 80-year-old female in which a MRPV coincided with other features of scimitar syndrome. Only a few cases of its type have been reported in the English literature.

CASE REPORT

An 80-year-old female was referred to the geriatric clinic for investigation of recurrent syncope. Her past medical history included mild postural hypotension, temporal arteritis, osteoarthritis and osteoporosis. Clinical examination revealed a small postural drop in blood pressure and an ejection systolic murmur, but was otherwise unremarkable.

A routine chest X-ray demonstrated a curvilinear structure running from the right mid zone towards the right cardiophrenic recess before curving superiorly (Figure 1), resembling a scimitar sign. In addition there was volume loss in the right hemithorax with mediastinal shift to the right suggesting cardiac dextroposition. An anomalous pulmonary vein, such as that seen in scimitar syndrome, was suspected. Contrast-enhanced computed tomography (CT) revealed a dilated right inferior pulmonary vein with an aberrant circuitous route, coursing inferomedially towards the diaphragm before turning upwards and draining normally into the left atrium (Figure 2). There was no connection to the IVC. It also confirmed the X-ray findings of right lung hypoplasia (right lung volume 1.17 L, left lung 1.66 L) and cardiac dextroposition. The right main pulmonary artery was smaller than the left in keeping with mild pulmonary artery hypoplasia. There was no evidence of anomalous systemic arterial supply to the lung. The diagnosis of an incidental MRPV was established and no further investigation or treatment of this was required.

Other investigations did not reveal any significant abnormality; routine blood tests showed an isolated mild hyponatremia (133 mmol/L). Twenty-four-hour electrocardiography monitoring demonstrated sinus rhythm throughout. Echocardiography was limited due to the abnormal positioning of the heart, but revealed mild aortic stenosis and good left ventricular function.

Neurally mediated syncope was deemed the most likely diagnosis and she was managed conservatively with advice on increasing fluid intake and taking care with postural changes.

Interestingly on questioning, the patient explained she had had an abnormal chest X-ray at the age of 5 years that showed "partial collapse" of the right lung. Having worked in the mining industry, she underwent several chest X-rays during her adult life but was told to stop having them because the appearances "always worried the doctors!"

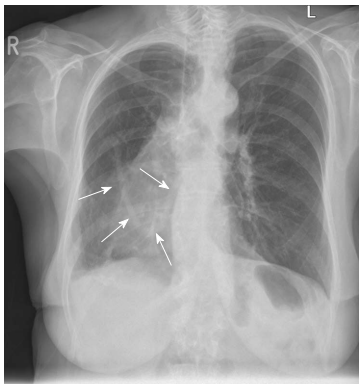

Figure 1 Posteroanterior chest X-ray demonstrating an anomalous curvilinear vessel (white arrows) running from the right mid zone inferomedially before turning superiorly. There is loss of volume within the right hemithorax and mediastinal shift to the right suggesting cardiac dextroposition. L: Left; R: Right.

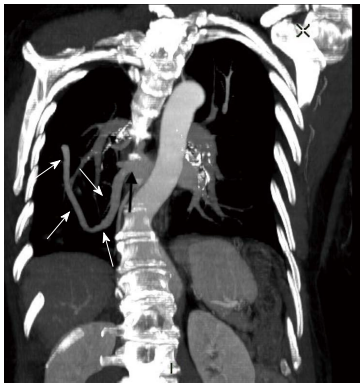

Figure 2 Contrast-enhanced maximum intensity projection multiplanar reformatted coronal image of the computed tomography chest demonstrating the path of the anomalous pulmonary vein (white arrows) and its connection to the left atrium (black arrow). R: Right.

DISCUSSION

The scimitar sign describes a curved vascular shadow on a chest X-ray, which courses along the right cardiac border towards the right cardiophrenic angle. It is so-called because the appearance resembles a Turkish sword or scimitar.

Scimitar syndrome is a rare pulmonary anomaly which consists of anomalous pulmonary venous drainage of the right lung to the IVC (giving rise to the scimitar sign), anomalous systemic arterial supply of the right lower lobe from either the thoracic or abdominal aorta, hypoplasia of the right lung, with resultant cardiac dextroposition and right pulmonary artery hypoplasia[1]. The scimitar sign was originally thought to be diagnostic of scimitar syndrome[2,3], however, a false positive scimitar sign is a rare possibility. Morgan *et al*[4] described the first case in which the scimitar sign and features of scimitar syndrome were present but the aberrant pulmonary vein ultimately drained normally into the left atrium. Other reported causes of the scimitar sign include an anomalous intrapulmonary venous connection to superior vena cava, obstruction of a major pulmonary vein with development of a distended intrapulmonary collateral and an anomalous IVC with normal pulmonary venous drainage[5,6].

The term "meandering right pulmonary vein" was subsequently coined by Goodman *et al*[7] to describe the presence of the scimitar sign and an anomalous right pulmonary vein that drains normally into the left atrium. In contrast to scimitar syndrome, there have only been a handful of cases of MRPV reported in the literature (Table 1). MRPV can occur with or without other fea-

Table 1 Published cases of meandering pulmonary vein

Ref.	Gender	Age (yr)	Symptoms	Scimitar sign	Features of scimitar syndrome	Investigations (in addition to CXR)	Anomalous pulmonary venous drainage	Original diagnosis	Proposed diagnosis
Morgan *et al*[4]	M	22	Incidental	Y	Thoracic aorta supply, right lung hypoplasia, dextroposition	Pulmonary angiography and cardiac catheterization	Single right pulmonary vein to left atrium	Scimitar syndrome with normal pulmonary venous drainage	AUSPV
Goodman *et al*[7]	F	51	Haemoptysis	Y	Dextroposition, right pulmonary artery hypoplasia	Pulmonary angiography and cardiac catheterization	Single right pulmonary vein to left atrium	MRPV	AUSPV
Kanemoto *et al*[15]	F	48	Orthopnea and productive cough	Y	Right lung hypoplasia, dextroposition	Lung perfusion, CT, ECHO, Pulmonary angiography	Single right pulmonary vein to left atrium	Pseudo-scimitar sign	AUSPV
Cukier *et al*[21]	F	27	Haemoptysis	Y	Abdominal aorta supply, right lung hypoplasia	Pulmonary angiography and cardiac catheterization	Inferior right pulmonary vein to left atrium	Scimitar syndrome	MRPV
Holt *et al*[22]	M	2	Murmur and failure to thrive	Y	Systemic supply, right lung hypoplasia, dextroposition	ECHO, cardiac catheterization	Superior and inferior right pulmonary veins to left atrium	Scimitar syndrome variant	MPVs
Tsitouridis *et al*[19]	M	41	Incidental	Y	Right lung hypoplasia, dextroposition	CT	Single right pulmonary vein to left atrium	MPV	AUSPV
Yoo *et al*[20]	F	1	Respiratory distress	Y	Right lung hypoplasia, dextroposition	CT	Single right pulmonary vein to left atrium	MRPV	AUSPV
Siu *et al*[23]	F	43	Incidental	Y	Dextroposition	ECHO, CT	Single right pulmonary vein to left atrium	Scimitar variant	AUSPV
Current case	F	80	Incidental	Y	Right lung hypoplasia, dextroposition, Hypoplasia right pulmonary artery	CT	Inferior right pulmonary vein to left atrium	MRPV	MRPV
Collins *et al*[8]	M	20	Incidental	Y	-	ECHO, cardiac catheterization, pulmonary angiography	All 4 pulmonary veins to left atrium	Idiopathic prominence of pulmonary veins	MPVs
Takeda *et al*[11]	F	28	Incidental	Y	-	Pulmonary angiography	NS	Scimitar variant	-[1]
Kriss *et al*[9]	F	12	Incidental	Y	-	CT, cardiac catheterization, pulmonary angiography	Right superior and inferior and left inferior pulmonary veins to left atrium	MPV	MPVs
Salazar-Mena *et al*[14]	F	15	Incidental	Y	-	Pulmonary angiography and cardiac catheterization	Right inferior pulmonary vein to left atrium	MRPV	MRPV
Al-Naami *et al*[24]	M	2/12	Failure to thrive, ASD, VSD	N[2]	-	ECHO, CT, cardiac catheterization	Right inferior pulmonary vein to left atrium	MPV	MRPV

[1]Unable to name due to lack of details in article. [2]Chest X-ray demonstrated cardiomegaly and congested lung fields which may have masked the scimitar sign. M: Male; F: Female; NS: Not stated in article; ASD: Atrial septal defect; AUSPV: Anomalous unilateral single pulmonary vein; CT: Computed tomography; CXR: Chest X-ray; ECHO: Echocardiogram; MPV: Meandering pulmonary vein; MRPV: Meandering right pulmonary vein; VSD: Ventricular septal defect; N: No; Y: Yes.

tures of the classic scimitar syndrome. Whilst most cases involve the right pulmonary veins, cases of anomalous right and left pulmonary veins have been described[8,9]. The scimitar sign is not always present[10].

A further anomaly, termed scimitar variant, describes the connection of an anomalous right pulmonary vein to both the IVC and left atrium[10-13].

A lack of consistency in the literature regarding nomenclature can lead to confusion. Some authors have treated MRPV and scimitar variant as synonymous[14]. Pseudo-scimitar sign has also been used to describe appearances of MRPV[15]. Anomalous unilateral single pulmonary vein (AUSPV) has been used to describe a single anomalous pulmonary vein draining the entire ipsilateral, lung regardless of whether it terminates normally in the left atrium[16,17] or elsewhere[18].

To avoid confusion we advocate using the term MRPV to describe cases in which the anomalous vein draining part of right lung terminates normally into the left atrium, reserving the scimitar variant for those with a dual connection to IVC and the left atrium. Meandering pulmonary veins (MPV) is suggested for cases that have more than one anomalous pulmonary vein draining into the left atrium. AUSPV should be used to describe cases where there is a single anomalous vein draining the entire ipsilateral lung to the left atrium or IVC.

Table 1 compares reported cases of MRPV, scimitar variant and pseudo-scimitar sign. Employing our proposed nomenclature, 2 of these cases would be classified as MRPV with features of scimitar syndrome, 1 as MPVs with features of scimitar syndrome, 2 as MRPV without features of scimitar syndrome, 2 as MPVs without features of scimitar syndrome, with 6 being reclassified as AUSPV.

Scimitar syndrome, scimitar variant and MRPV can be considered as a spectrum of pulmonary anomalies having a common embryological basis, with scimitar syndrome at one extreme, MRPV at the other, and scimitar variant somewhere in between[12,19,20]. It is likely that the stage of embryogenesis at which the anomaly occurs determines which condition develops. For example, persistence of the primitive communications between the pulmonary and systemic vascular supplies may lead to scimitar syndrome if the connection between the right pulmonary vein and left atrium is obstructed, or scimitar variant if this connection is patent[11,14,19]. Abnormally delayed obliteration of the pulmonary and systemic connections may result in a MRPV with an anomalous route in the lungs but ultimately draining normally into the left atrium.

It is important to distinguish between scimitar syndrome and MRPV. Scimitar syndrome results in a left-to-right shunt, which can lead to cyanosis and may require surgical correction. Consequently the patients are often symptomatic and present at a young age. In contrast, there is no left-to-right shunt in MRPV. As in this case, patients are usually asymptomatic, with the diagnosis made incidentally. Treatment has not been required in any reported case of MRPV.

The diagnosis of MRPV has changed drastically thanks to advances in CT technology. Several of the reported cases of MRPV are from the pre-CT era and were investigated with pulmonary angiography and cardiac catheterization, potentially hazardous invasive investigations, particularly considering MRPV is a benign condition requiring no treatment. Of those cases which underwent CT, the older CT technology at the time often did not allow detailed multiplanar reformatting (MPR), limiting the assessment of the anatomy, and therefore necessitating invasive imaging to confirm the diagnosis. This case highlights that non-invasive diagnosis is possible with modern multislice CT technology through the use of detailed MPR and maximum intensity projections, which clearly demonstrate vessel anatomy (Figure 2). Additionally, accurate assessment of lung volumes is possible with modern CT software, allowing assessment of associated lung hypoplasia.

In conclusion, this case highlights that the chest X-ray features of scimitar syndrome are not diagnostic and a MRPV should be considered in their presence. Differentiation between these conditions is required to help decide whether treatment is necessary. Modern multislice CT technology allows clear depiction of the vascular connections and associated anatomy, and has superseded invasive pulmonary angiography and cardiac catheterization as the investigation of choice for MRPV.

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