

Meandering pulmonary vein: A case report

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ABSTRACT

Anomalies of the pulmonary venous circulation are extremely uncommon. We report a case of an anomalous unilateral single pulmonary vein also referred to as a meandering pulmonary vein. A single large tortuous pulmonary vein was seen on the right side draining into the left atrium with associated ipsilateral absence of the middle lobe bronchus and interlobar fissures. This is considered a variant of the classical scimitar syndrome. The venous anatomy was depicted with considerable accuracy using multidetector computed tomography (MDCT) non-invasively.

CASE REPORT

CASE REPORT

A 28-year-old female referred from the department of obstetrics and gynecology was undergoing work up for primary infertility. She had no complaints pertaining to the respiratory system and the physical examination was unremarkable. Her chest radiograph showed an abnormal tubular radiopacity in the para cardiac region of the right lung with mild volume loss of the right lung (Fig 1). The cardiac silhouette appeared normal. Scimitar syndrome was suspected based on the chest x-ray and contrast enhanced computed tomography scan of the thorax was done. On contrast enhanced CT (CECT) there was replacement of the normal right pulmonary veins by an abnormal single tortuous pulmonary vein with a meandering course through the right lung beginning in the right upper lobe and coursing posteriorly and medially and draining into the left atrium (Fig 2). The main pulmonary artery, right and left pulmonary arteries were found to be normal. No systemic collateral vessels were seen. In addition, there was absence of the right middle lobe bronchus, the horizontal and oblique fissures with a single fissure seen separating the apical segment of the right upper lobe from the rest of the right lung parenchyma suggesting

pulmonary hypoplasia (Fig 3). Echocardiography of the patient was found to be normal. Since the patient was asymptomatic no intervention was done and the patient has been kept on follow-up.

DISCUSSION

Meandering pulmonary vein also called pseudo-Scimitar syndrome or Scimitar variant is an extremely rare pulmonary venous anomaly which could be confused with the classical Scimitar syndrome which is a relatively more common condition with similar appearance of a linear opacity in para cardiac location on chest x-ray. Unlike the Scimitar syndrome where the abnormal pulmonary vein drains into the inferior vena cava, in pseudo-scimitar the pulmonary vein has an abnormal course but drains normally into the left atrium. The present case is an example of this condition.

Etiology & Demographics:

Congenital anomalies of the pulmonary venous system have been divided into 3 types, namely, partial anomalous

pulmonary venous drainage with or without abnormal course in the lung, anomalous venous route without abnormal connection and lastly abnormal venous diameters including varices, stenosis and atresia by Remy-Jardin et al [1]. According to this classification our case falls into the second category i.e. anomalous venous route without abnormal connection.

It has been hypothesized that these anomalies occur during the primitive splanchnic capillary stage of development. The splanchnic plexus consisting of the umbilicovitelline veins, cardinal veins and common pulmonary vein drains the immature lung. Later on, the common pulmonary vein gets incorporated into the left atrium and forms the pulmonary veins which drain into the left atrium while the umbilicovitelline veins and cardinal veins undergo regression at around 30 to 32 days [2,3]. Atresia of one of the pulmonary veins after the splanchnic circulation has separated but before pulmonary segmentation has occurred might be the cause of AUSPV formation [4].

Most of these cases are detected incidentally when imaged for some other pathology. Hence the age of detection varies significantly with cases being reported in infants and patients as old as 80 years of age. These anomalies have been reported in both males and females. In the review by Rodrigues et al a slight female predominance was seen although the authors did not mention any specific gender predilection [5].

Clinical & Imaging findings:

As the anomalous vessel drains into the left atrium, right to left shunting is not seen and hence the patients are usually asymptomatic, mostly detected incidentally. Several variants of the Scimitar syndrome have been described of which one rare variant includes a single tortuous scimitar like vein draining into the left atrium. The first of such cases was reported in 1968 by Kozuka and Nosaki [6]. The term "meandering pulmonary vein" was first used by Goodman et al to describe a single large right pulmonary vein which drained into the left atrium instead of the inferior vena cava as would be seen in a case of true scimitar vein [7]. The term anomalous unilateral single pulmonary vein (AUSPV) was used for a similar case by Goudarzi et al [2]. Unlike the true scimitar syndrome, the drainage in case of AUSPV is orthotopic.

Our case had associated right lung hypoplasia, absence of the right middle lobe bronchus and interlobar fissures with normal pulmonary arteries. Several cases have been reported with associated anomalies in patients with a meandering pulmonary vein and hence one should be on the lookout for other findings while evaluating these cases. Tortoriello et al described a case of meandering right pulmonary vein with dual drainage both to the IVC and left atrium [8]. A case of bilateral anomalous single pulmonary veins draining into the left atrium was reported by Hidvegi and Lapin [9]. In the case described by Goodman et al hypoplasia of the right main pulmonary artery and mirror image lung was noted [7]. One of the 2 cases reported by Hasuo et al had partial anomalous

pulmonary venous drainage of the posterior basal vein into the IVC [10]. Right lung and pulmonary artery hypoplasia with bilateral left sided bronchial configuration were observed in association with AUSPV by Goudarzi et al [2]. Most of the reported cases of meandering pulmonary vein are of the right side with very few cases of left sided involvement being described such as the case reported by Aygun et al [11].

Although pulmonary angiography is the gold standard for diagnosis of pulmonary venous anomalies non-invasive techniques like MDCT and magnetic resonance angiography (MRA) can satisfactorily delineate the vascular anatomy with minimal patient discomfort, MRA having the added advantage of using non-ionizing radiation. Angiography should be reserved for doubtful cases or when intervention is required.

Treatment & Prognosis:

It is important to distinguish meandering pulmonary vein from other pathologies and make the correct diagnosis as management varies significantly. As was seen in our patient isolated AUSPVs are usually asymptomatic and they do not require any surgical intervention [9]. These patients are kept on follow up and they have a good prognosis.

Differential Diagnoses:

AUSPV or meandering pulmonary vein can mimic several other pathologies such as the true scimitar syndrome, pulmonary nodules, pulmonary arteriovenous malformations, pulmonary varices etc [12,2,3,10].

- Classical Scimitar syndrome: It consists of hypoplasia of the right lung, dextroposition of the heart, hypoplasia of the right pulmonary artery, an abnormal pulmonary vein draining into the inferior vena cava (IVC) and a systemic collateral supply to the lung [12].
- Pulmonary varix: It refers to localized congenital dilatation of the pulmonary vein. On chest x-ray it is seen as a smooth lobulated soft tissue lesion or a mass or a tubular peripheral opacity in the lung. In addition to the above findings transfissural collaterals can also be seen on CT. The angiographic criteria for the diagnosis include presence of a normal pulmonary arterial tree, drainage of the varix by the pulmonary vein, filling of the varix at the same rate as the normal pulmonary vein and drainage into the left atrium with no shunt formation between systemic and pulmonary circulations. Also, there is delayed emptying of the varix in comparison to the normal pulmonary veins [13].
- Pulmonary arteriovenous malformations: They occur as a result of abnormal communications between pulmonary arteries and veins without the intervening capillary bed. On chest x-ray they are seen as homogenous round or oval opacities along with curvilinear opacities coursing towards the hilum. CT angiography is the gold standard in which it is seen as a well-defined lesion with feeding artery and draining vein [14].
- Pulmonary nodules: The anomalous pulmonary vein can sometimes be confused with nodules due to various pathologies. These can be delineated on computed tomography which shows better depiction of the actual pathology.

TEACHING POINT

Meandering pulmonary vein is a variant of the classical scimitar syndrome in which the pulmonary vein has an abnormal course but normal drainage into the left atrium. Since this condition generally does not require any intervention it is important to distinguish this entity from the classical scimitar syndrome in which the abnormal vessel drains into the inferior vena cava.

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FIGURES

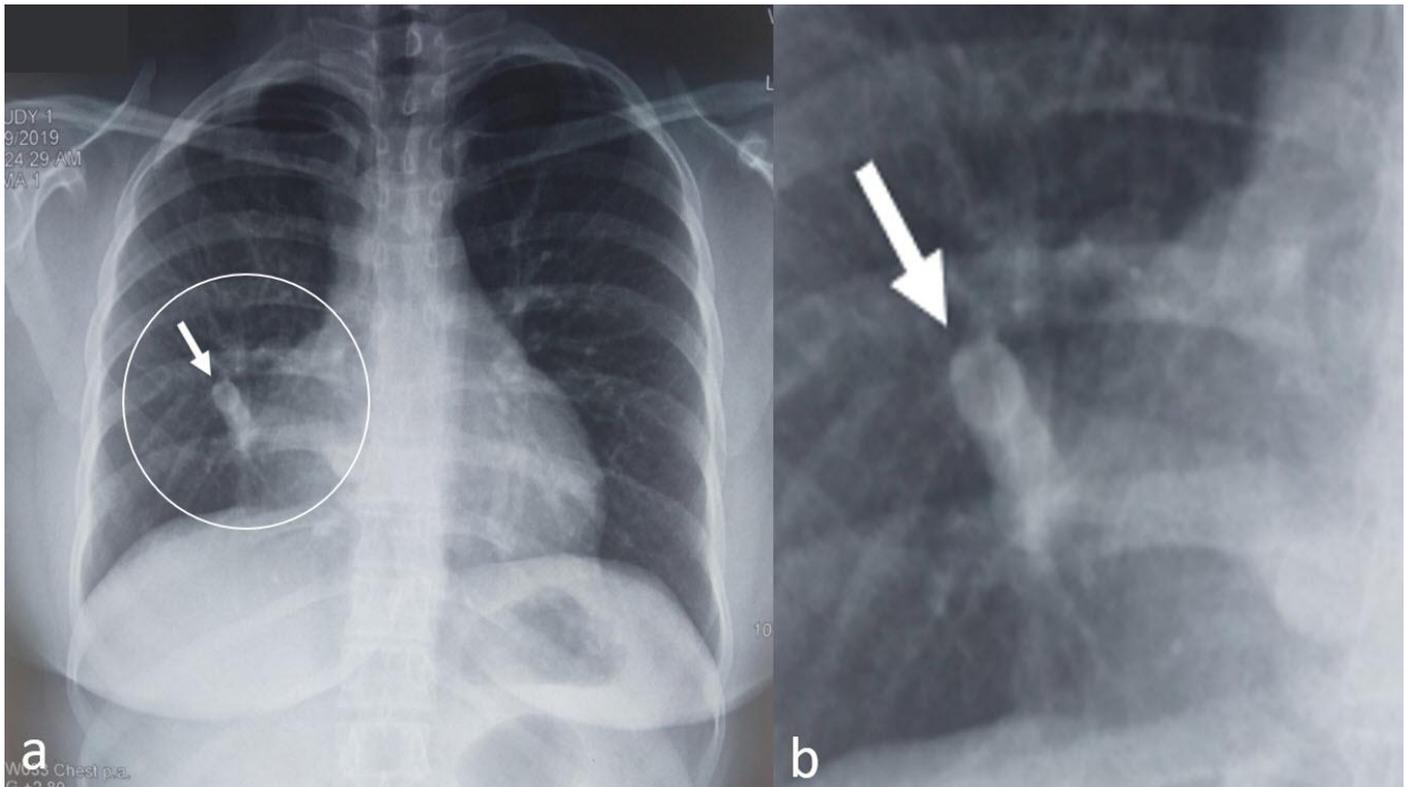


Figure 1: 28-year-old female with meandering pulmonary vein.

Findings:

Fig (a) Chest x-ray shows a tubular opacity (arrow) in the right lower zone in the para cardiac region with mild volume loss of the right lung. The left lung field is clear and cardiothoracic ratio is 0.5. Fig(b) is the magnified image showing the tubular opacity.

Technique:

Frontal chest x-ray posteroanterior view obtained using 125 kV and 1.75mAs.

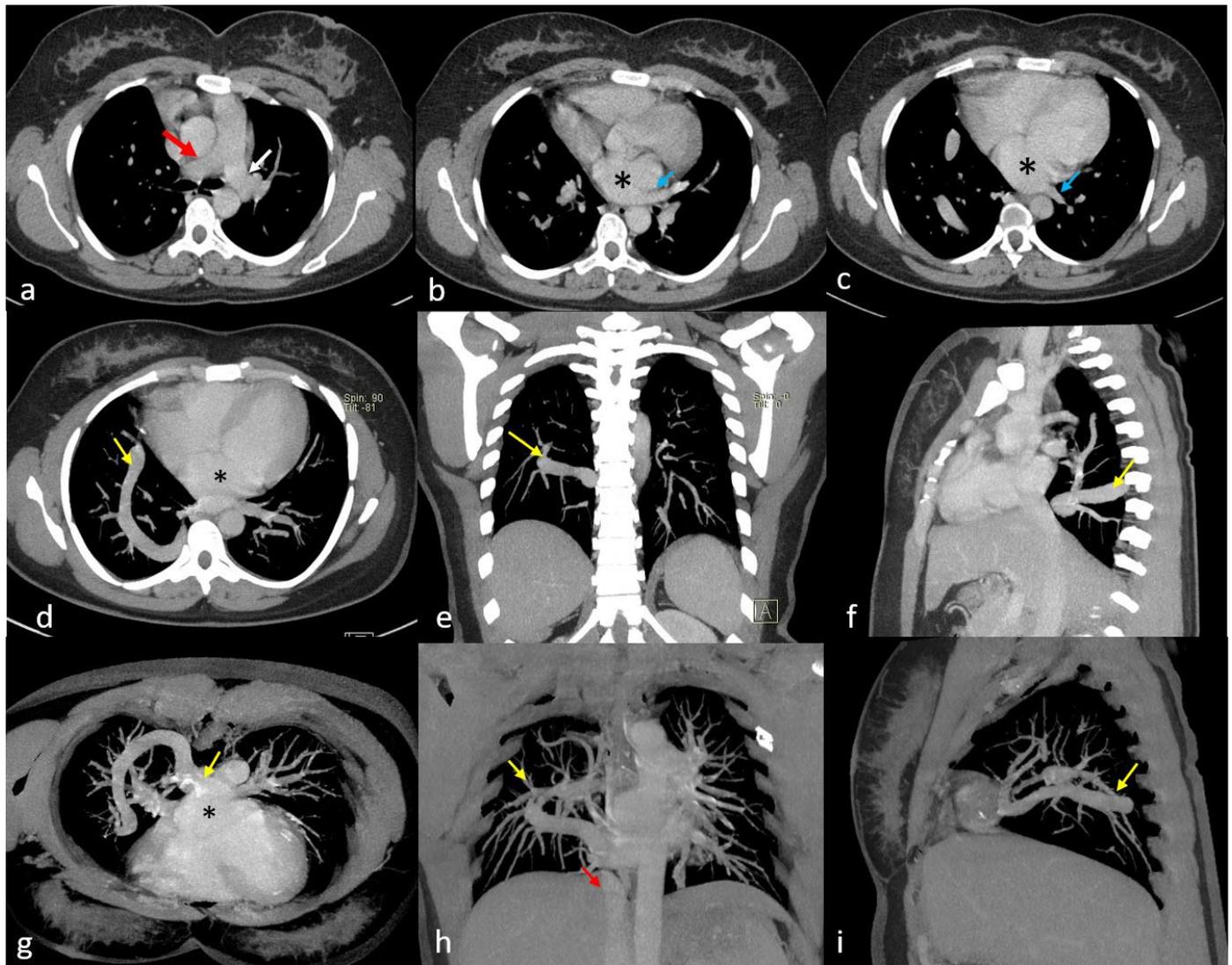


Figure 2: 28-year-old female with meandering pulmonary vein.

Findings:

Axial (a, b & c) sections of CECT thorax of a 28-year-old female from superior to inferior levels in mediastinal window show the main pulmonary artery dividing into right (red arrow in a) and left (white arrow in a) pulmonary arteries which appear normal in course and caliber. The left superior (blue arrow in b) and left inferior pulmonary veins (blue arrow in c) are seen draining normally into the left atrium (asterisk). The tortuous meandering course of the prominent anomalous right pulmonary vein is well seen in the axial (d & g), coronal (e & h), sagittal (f & i) MIP images (yellow arrows) as it courses anteroinferiorly initially, then posteriorly through the right lung and turns medially to drain into the left atrium (asterisk) which appears normal in morphology. The inferior vena cava is also seen (red arrow in h).

Technique:

128 slice CT, SIEMENS SOMATOM Definition AS+

Contrast enhanced CT scan [146mAs, 120 kV, slice thickness- 1mm (a,b,c), 10 mm(d,e,f), 41.2mm(g,h), 27.2mm (i)] acquired 60 seconds after intravenous contrast injection (60 mL of nonionic contrast medium; Omnipaque 350mgI/mL, GE Healthcare, USA). Axial CECT images(a,b,c)-Window level:40 and window width: 400. MIP images (d,e,f)-Window level:64 and window width:400; MIP image(g)- Window level: 82 and window width: 271; MIP images (h,i)- Window level:100 and window width:500.



Figure 3: 28-year-old female with meandering pulmonary vein.

Findings:

Axial (a) and sagittal (b) CT images in lung window show the single fissure (arrows) separating the apical segment of the right upper lobe anteriorly from the rest of the right lung. The anomalous pulmonary vein is seen in cross section as a nodular opacity in the right lower lobe (red arrow in b). Axial (c), coronal (d) and sagittal (e) MinIP images show the right upper lobe bronchus (yellow arrows) and right lower lobe bronchi (white arrows) with non-visualization of the right middle lobe bronchus. The rounded density seen in the sagittal image (red arrow in e) represents the anomalous right pulmonary vein. The normal branching pattern of the left lung bronchi is also seen (c,d).

Technique:

128 slice CT, SIEMENS SOMATOM Definition AS+ Contrast enhanced CT scan [146mAs, 120 kV, slice thickness- 1 mm (a,b); 100mm(c,d,e)] acquired 60 seconds after intravenous contrast injection (60 mL of nonionic contrast medium; Omnipaque, GE Healthcare, USA). Window level: -600 and window width: 1200.

Etiology	Atresia of one of the pulmonary veins after the splanchnic circulation has separated but before pulmonary segmentation. *
Incidence	Unknown, rare.
Gender ratio	Unknown.
Age predilection	Usually asymptomatic and detected incidentally in children and adults.
Risk factors	Unknown.
Treatment	Conservative.
Prognosis	Good.
Findings on imaging	Replacement of the normal pulmonary veins by an abnormal single tortuous pulmonary vein with a meandering course through the lung but draining into the left atrium. **

* refer to article by Hanson et al [4]; ** refer to figure 2 for imaging findings.

Table 1: Summary table for meandering pulmonary vein

	Radiograph	Computed Tomography
Meandering pulmonary vein	A curvilinear opacity in the para-cardiac location along with other findings similar to the classic scimitar syndrome.*	An abnormal single tortuous pulmonary vein replacing the normal pulmonary vein but with orthotopic drainage into the left atrium.**
True scimitar syndrome¹	The anomalous vein is seen as a tubular opacity paralleling the right heart border similar in shape to a Turkish sword giving the 'scimitar sign'. Associated lung hypoplasia and dextroposition of the heart is seen.	Hypoplasia of the right lung, dextroposition of the heart, hypoplasia of the right pulmonary artery, an abnormal pulmonary vein draining into the inferior vena cava (IVC) and a systemic collateral supply to the lung.
Pulmonary varices²	A smooth lobulated soft tissue opacity or a mass or a tubular structure in the lung.	Localized aneurysmal dilatation of a pulmonary vein draining into left atrium.
Pulmonary arteriovenous malformations³	Homogenous round or oval opacities with curvilinear opacities coursing towards the hilum.	CT angiography is the gold standard. Well-defined lesion with feeding artery and draining vein.
Pulmonary nodules	Multiple nodular opacities.	Nodular soft tissue opacities with no vascular communication.

* refer to figure number 1; ** refer to figure number 2; ¹ Gao et al [12]; ² Shostak et al [13]; ³ Tellapuri et al [14].

Table 2: Differential diagnosis table for meandering pulmonary vein

ABBREVIATIONS

AUSPV = Anomalous unilateral single pulmonary vein
 CECT = Contrast enhanced computed tomography
 IVC = Inferior vena cava
 MDCT = Multidetector computed tomography
 MRA = Magnetic resonance angiography

KEYWORDS

Pulmonary venous anomalies; meandering pulmonary vein; Scimitar variant; anomalous unilateral single pulmonary vein (AUSPV); lung hypoplasia; multidetector CT

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