Case Report

Bilateral meandering pulmonary vein complex and unusual associated cardiovascular anomalies and shunt: Extremely rare entity

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ABSTRACT

Meandering pulmonary vein (MPV) is a rare entity that can be associated with an additional cardiac and pulmonary venous variations, including left-to-right shunts. Clinicians should consider further workup with dedicated cardiac imaging to evaluate for associated cardiovascular abnormalities after an abnormal pulmonary vein draining is initially identified on routine computed tomography or echocardiogram. Pulmonary venous variations in MPV represent a spectrum of disorders, and no consistent nomenclature has been established.

KEY WORDS: Cardiovascular anomalies, meandering pulmonary vein, pulmonary venous variations, shunt

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INTRODUCTION

Meandering pulmonary vein (MPV) is a rare entity and represents a "circuitous course" in the lung with <20 reported cases in the literature. MPV can be an isolated entity or occurs in combination with anomalies such as partial anomalous venous return (PAPVR) and various cardiac abnormalities. However, none of the previous reports have described about an associated complex of a bicuspid aortic valve, a fenestrated subaortic membrane, aberrant attachment of a left ventricle papillary muscle into the interventricular septum, and an anomalous return of the right upper lobe MPV into the left brachiocephalic vein. We present these unique findings in this case report.

CASE REPORT

We report the case of a young person in 4th decade who was incidentally found to have partially visualized pulmonary venous abnormalities on a computed tomography (CT) abdomen done for abdominal pain. The patient had no respiratory or cardiac symptoms or any positive findings on relevant physical examination. Further workup with a dedicated cardiac CT with pulmonary vein protocol demonstrated dilated and anomalous tortuous course of the right upper lobe pulmonary vein draining into the left brachiocephalic vein which is a systemic vein [Figures 1 and 2b, double arrow] with additional drainage of the right upper lobe pulmonary vein into

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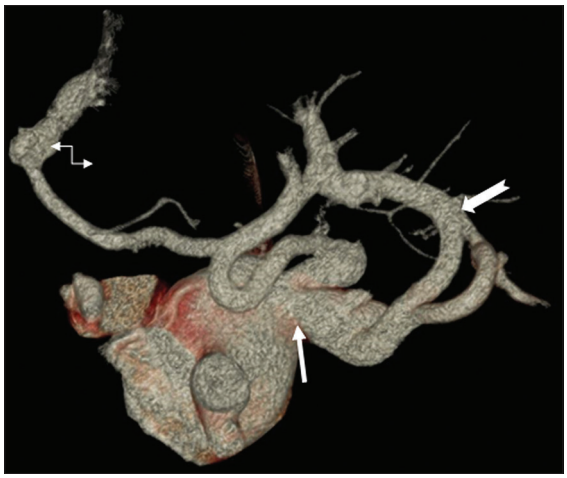

Figure 1: Dilated and anomalous tortuous course of the right upper lobe pulmonary vein draining into the left brachiocephalic vein which is a systemic vein (double arrow) with additional drainage of the right upper lobe pulmonary vein into the right middle and lower lobe pulmonary veins as common channel (notched arrow) which subsequently drained through the nonobstructed right inferior pulmonary vein ostium (white arrow) into the left atrium

the right middle and lower lobe pulmonary veins as common channel [Figures 1 and 2b, notched arrow] which subsequently drained through the nonobstructed right inferior pulmonary vein ostium [Figures 1 and 2a, white arrow] into the left atrium.

There was similar meandering tortuous course of the dilated left upper and lower pulmonary veins which formed a common channel to subsequently drain into the left atrium via single left inferior pulmonary ostium [Figure 2, black arrow]. In addition to abnormal course of MPV and single bilateral pulmonary vein ostium, CT demonstrated associated complex abnormalities of thin fenestrated subaortic membrane [Figure 3a and b], aberrant papillary muscle [Figure 3b] in the left ventricle inserting into the interventricular septum, bicuspid aortic valve [Figure 3c, black thin arrow], and anomalous draining of a right-sided pulmonary vein into the left brachiocephalic vein [Figures 1 and 2b, double arrows]. Since the patient was asymptomatic, he was asked for periodic clinical follow-up.

DISCUSSION

The first report on "MPV" described it as an abnormal drainage and course pattern of pulmonary veins terminating in the left atrium without shunting.[1] Few subsequent reports expanded the definition and included any pulmonary vein demonstrating a "circuitous course" in the lung regardless of shunt presence. However, it should be noted that there is some inconsistency in the literature regarding the presence of associated left-to-right shunt.[2,3]

MPV can be an isolated finding with the aberrant pulmonary vein terminating in the left atrium or occurs

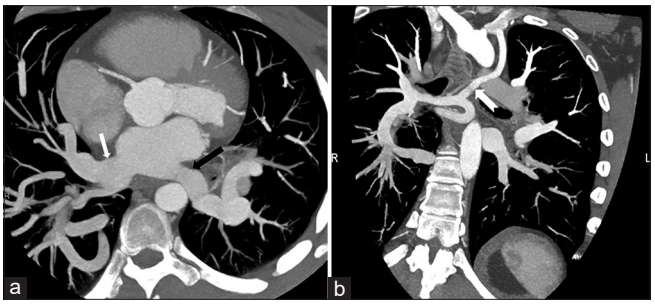

Figure 2: (a) The right middle and lower lobe pulmonary veins form a common ostium and drains as inferior right pulmonary vein (white arrow) into the left atrium. The left upper lobe pulmonary vein drains into the left lower pulmonary vein which forms a common ostium and drains as inferior left pulmonary vein (black arrow) into the left atrium. Note the tortuous course of the bilateral pulmonary veins into the lung parenchyma. (b) Dilated and anomalous tortuous course of the right upper lobe pulmonary vein (white arrow) draining into the left brachiocephalic vein which is a systemic vein

in combination with anomalies such as PAPVR and various cardiac abnormalities.[4,5] However, none of the previous reports have described about an associated complex of a bicuspid aortic valve, a fenestrated subaortic membrane, aberrant attachment of a left ventricle papillary muscle into the interventricular septum, and an anomalous return of the right upper lobe MPV into the left brachiocephalic vein. MPV can be diagnosed on routine CT chest; however, additional cardiac lesions may only be visualized on dedicated gated cardiac CT angiography (CTA).

MPV is a rare disease with <20 reported cases in the literature. Scimitar syndrome is described as a similar entity with a right-sided PAPVR draining into the inferior vena cava, but demonstrates associated right lung hypoplasia and systemic arterial supply from the aorta to the affected lung segment.[3] Both the scimitar syndrome and MPVs represent variations within the spectrum of pulmonary vein anomalies. A recent report described a case of MPV draining into the left atrium with associated right lung hypoplasia and affected lung segment arterial supply coming from the systemic vasculature.[4] These anomalies suggest that pulmonary venous variations form a continuous spectrum of related disorders rather than separate entities. Our case demonstrates a tortuous course of the bilateral dilated pulmonary veins (MPV), a left-to-right shunt, and no lung hypoplasia or abnormal systemic arterial supply to lung. It does not fit into previously classified pulmonary venous variations, highlighting the need for awareness and search pattern to look for associated congenital anomalies. In conclusion, this is the first reported case in the literature of bilateral MPVs with an associated complex of previously not reported subaortic membrane, aberrant papillary muscle in the left ventricle inserting into the interventricular septum, bicuspid aortic valve, and anomalous draining of a right-sided pulmonary vein into the left brachiocephalic vein.

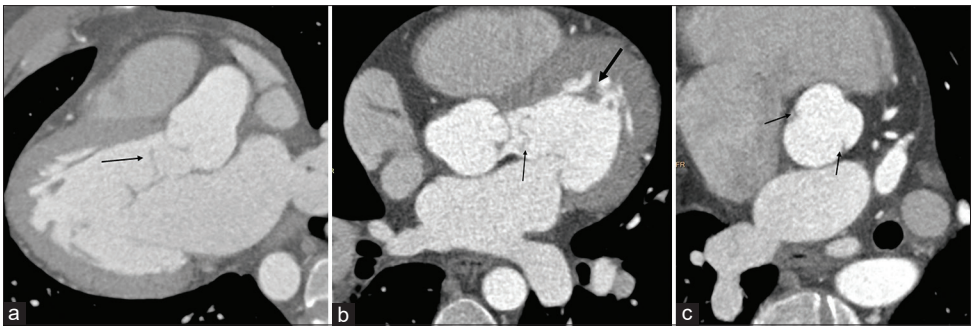

Figure 3: (a) Three-chamber view of the left ventricle showing fenestrated subaortic membrane. (b) Fenestrated subaortic membrane (thin arrow) and aberrant papillary muscle (bold arrow) in the left ventricle. (c) Bicuspid aortic valve (black thin arrow). Note anomalous right upper lobe pulmonary vein branch course left lateral to the left atrium (notched arrow) and posterior to the left central pulmonary artery which subsequently drained into the left brachiocephalic vein

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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