case report

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Meandering right pulmonary vein associated with retrocaval ureter and vertebral fusion anomalies

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Summary We present multimodality imaging of a meandering right pulmonary vein in a 29-year-old female patient. It was associated with right retrocaval ureter causing hydronephrosis and stone formation, vertebral fusion anomalies and corrected cardiac anomalies (patent ductus arteriosus and ventricular septal defect). To the best of our knowledge, only a few meandering right pulmonary vein have been presented in the literature until now and this is the first reported case that is associated with retrocaval ureter and vertebral fusion anomalies.

Keywords Meandering right pulmonary vein · Retrocaval ureter · Vertebral fusion · CT · MR

Gleichzeitiges Vorliegen einer mäandernden rechten Pulmonalvene mit einem retrocavalem Ureter und Wirbelfusionsanomalien

Zusammenfassung Wir berichten über eine multimodale Bildgebung einer mäandernden rechten Pulmonalvene in einer 29 Jahre alten weiblichen Patientin. Gleichzeitig lag der rechte Ureter retrocaval, was zu Hydronephrosis und Steinbildung geführt hatte. Außerdem bestanden Wirbelfusionsanomalien und korrigierte Herzanomalien (offener Ductus arteriosus und Ventrikelseptumdefekt). Soweit wir wissen, wurden bisher nur wenige mäandernde Pulmonalvenen in der Literatur beschrieben. Dieser Fall ist außerdem der erste, bei dem gleichzeitig ein retrocavaler Ureter und Wirbelfusionsanomalien vorliegen.

Schlüsselwörter Mäandernde rechte Pulmonalvene · Retrocavaler Ureter · Wirbelfusion · Computertomographie (CT) · Magnetresonanz (MR)

Introduction

Meandering right pulmonary vein (MRPV) also known as pseudo-scimitar syndrome is a rare pulmonary vein anomaly in which a winding and widened right pulmonary vein traveling through the right lung and finally draining normally to the left atrium in contrast to the inferior vena cava (IVC) as seen in scimitar syndrome [1]. MRPV is an innocent vascular anomaly and sometimes associated with other anomalies [1, 2]. In this report, we present a MRPV associated with right retrocaval ureter, vertebral fusion anomalies and corrected cardiac anomalies. To the best of our knowledge, this is the first reported case of a MRPV with retrocaval ureter and vertebral fusion anomalies.

Case

A 29-year-old female patient who had operated for patent ductus arteriosus and ventricular septal defect 22 years ago was admitted to our hospital with the complaint of coughing, palpitation, and weakness. Tubular structures in the right perihilar region was seen on chest radiograph but no marked volume difference between the two lungs was detected (Fig. 1). A contrast enhanced

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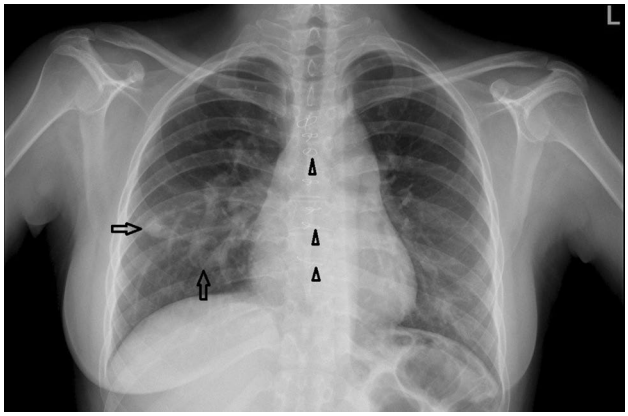

Fig. 1 Posteroanterior chest X-ray shows two dilated and curved tubular structures (*arrows*) coursing and converging toward the right pulmonary hilus, anomalously. Notice midline sternal wires (*arrow heads*) indicating previous cardiothoracic surgery

thorax computed tomography (CT) examination was performed and showed dilated right superior and inferior pulmonary veins with aberrant meandering courses. The right superior pulmonary vein coursing inferolaterally toward the diaphragm before turning upwards and the right inferior pulmonary vein coursing superolaterally and then traveling anteromedially were joining in the pulmonary hilus to form a common right pulmonary vein before draining into the left atrium (Fig. 2). No connection was seen between the right pulmonary vein and systemic venous circulation. There was calcifications in the interventricular septum and cardiac apex, and an incidental right hydronephrotic kidney containing calculus was also detected. She had no abdominal symptoms. Cardiac magnetic resonance imaging (MRI) and abdominal MR urography showed MRPV (Fig. 3a), regional wall thinning, myocardial scarring and focal aneurysmatic dilatation of the inferoseptum secondary to operation or chronic infarction. The right ureter was coursing posterior to the IVC causing high grade hydronephrosis (Fig. 3b). It was also noted that there was lower lumbar and sacral vertebral body fusion (Fig. 3c). Patient was operated for retrocaval ureter. Retrocaval segment of the ureter was identified (Fig. 3d) and excised by laparoscopic transperitoneal approach. After a double-J stent insertion, Anderson-Hynes pyeloplasty was performed.

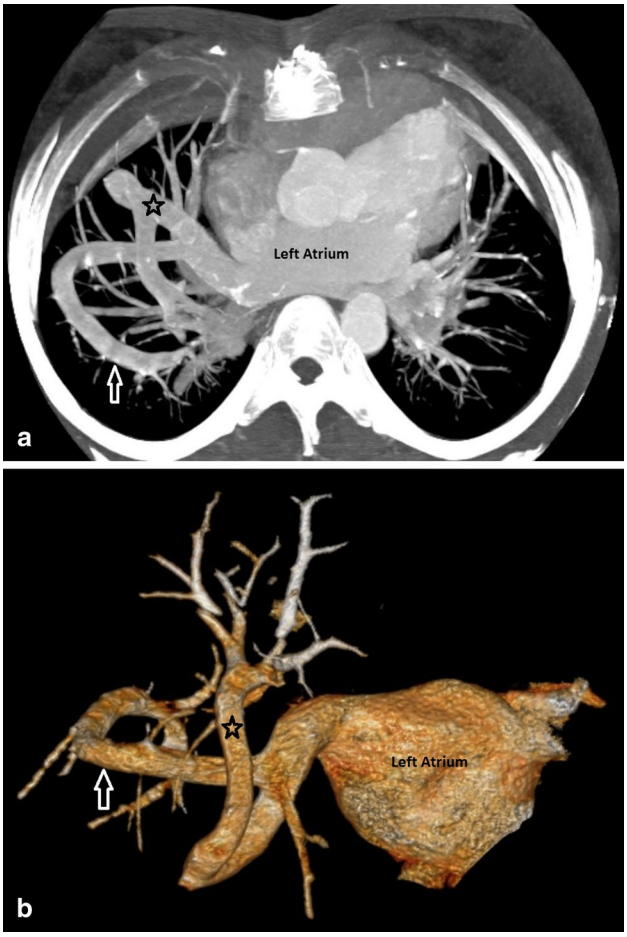

Fig. 2 Axial thick slice maximum intensity projection image (**a**) and three-dimensional volume-rendered posterior view computed tomography (CT) image (**b**) demonstrate that the right superior pulmonary vein (*asterisk*) courses inferolaterally toward the diaphragm and then turns upwards, and the right inferior pulmonary vein (*arrow*) courses superolaterally and then travels anteromedially. Meandering right pulmonary veins join in the pulmonary hilus to form a common right pulmonary vein before draining into the left atrium

Discussion

Congenital pulmonary venous return anomalies are usually classified into three categories as venous diameter abnormalities (varices, stenosis, and atresia), anomalous pulmonary venous course within the lung without abnormal drainage (MRPV), and anomalous pulmonary venous return with abnormal drainage (scimitar syndrome) [2]. MRPV is charecterised by a dilated varicose right pulmonary vein that travels within the lung in a circuitous route before draining into the left atrium. Only a few MRPV cases have been reported in the literature since its description by Goodman et al. in 1972 [1, 2]. It is hemodynamically normal, produce no symptoms and do not require any intervention [3, 4]. On the other hand, its appearance on chest radiograph is similar with the scimitar sign (Turkish sword) described for scimitar syndrome in which there are hypoplastic right lung, abnormal pulmonary venous drainage to the systemic venous circulation and systemic arterial supply of some right lung parts from the aorta [1–3]. Scimitar syndrome is hemodynamically abnormal because of left to right shunt, can lead to cyanosis and may require intervention [1]. Another subtype named as scimitar variant is used to describe connection of an anomalous right pulmonary vein to both the IVC and the left atrium [3]. That is why, these entities should be distinguished from each other by the way of imaging modalities [1, 2].

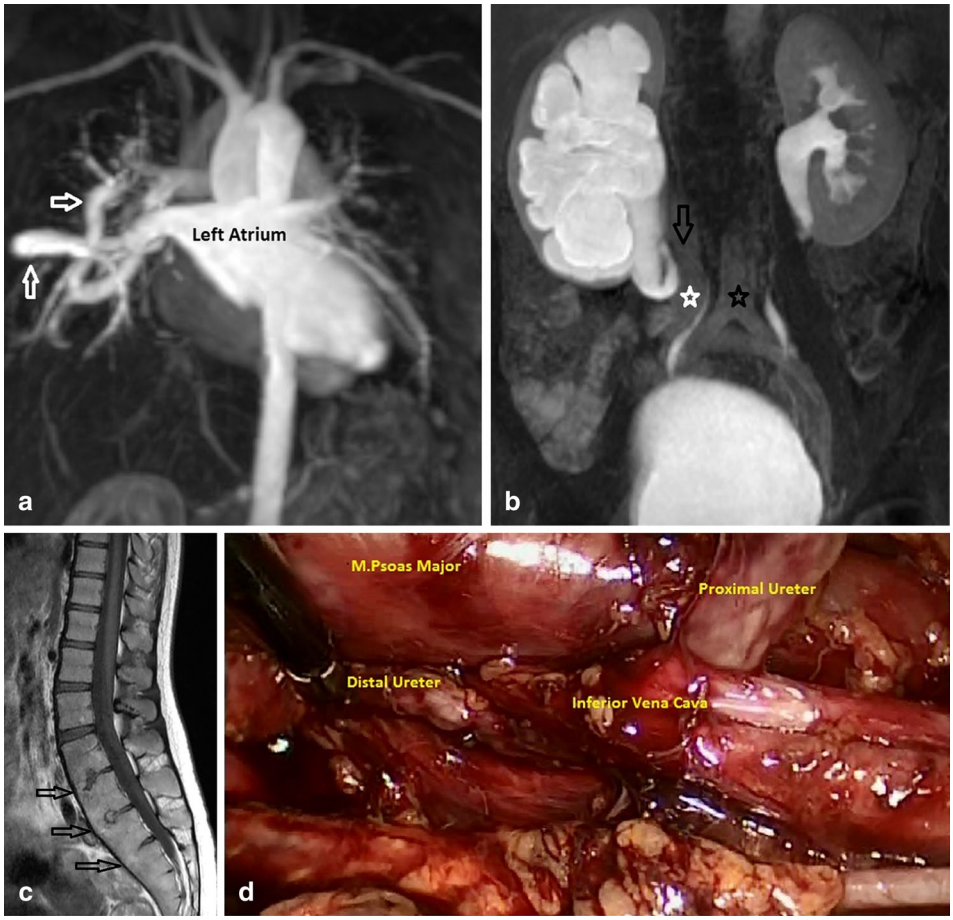

Fig. 3 Coronal view magnetic resonance (MR) angiography (**a**) shows dilated right superior and inferior pulmonary veins (*white arrows*) with aberrant meandering courses. Coronal view MR urography (**b**) demonstrates high grade right hydronephrosis and S shaped proximal right ureter (*black arrow*) which is typical for retrocaval course. Sagittal T1 weighted lumbar vertebra image (**c**) shows lower lumbar and sacral vertebral body fusion (*arrows*), anteriorly. Laparoscopic view (**d**) shows course of the right proximal ureter behind the inferior vena cava. *White asterisk* inferior vena cava, *black asterisk* aorta

Reported retrocaval ureter is around 200 cases since the first case described in 1893 in the literature [5]. It is a rare congenital anomaly and is found in 0.06–0.17 % of the autopsy series [5]. External compression of the proximal ureter coursing behind the IVC leads to obstruction of the right ureter and causes hydronephrosis and usually becomes symptomatic by the third to fourth decades of life [5, 6]. It is fortunate for our patient that thorax CT and meandering right pulmonary vein facilitated the diagnosis of retrocaval ureter by showing right hydronephrosis in the last CT slices. It is easy to diagnose this anomaly by the typical appearance of "S" or "J" shaped upper ureter and renal pelvis with intravenous urography and contrast enhanced CT or MR urography [5].

Retrocaval ureter is not usually associated with urinary obstruction. Patients who have moderate to high grade hydronephrosis with symptoms such as stone formation, infection, and worsening of renal function are candidates for laparoscopic pyeloplasty [5, 6]. Our patient was operated for high grade hydronephrosis and stone formation.

Reported associated anomalies with MRPV are right lung hypoplasia, pulmonary artery hypoplasia, systemic arterial supply to the lung, dextroposition and left thoracic isomerism [1, 2]. To the best of our knowledge, this is the first case of MRPV associated with retrocaval ureter, vertebral fusion anomalies, and operated cardiac anomalies.

In conclusion, MRPV is a rare and hemodynamically normal pulmonary vein anomaly that does not require any treatment and should be differentiated from scimitar syndrome. It can be revealed strictly by CT or MR angiography. Although, MRPV is a benign anomaly, it may be associated with serious cardiovascular, urologic and vertebral anomalies.

Conflict of interest

The authors declare that there is no actual or potential conflict of interest in relation to this article.

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