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## Membranous IVC Obstruction Presenting with Antegrade/Retrograde Respiratory Flow in the Intrahepatic Segment in Doppler Imaging and Prostatic and Urethral Congestion

### Authors' Contribution:

- A** Study Design
- B** Data Collection
- C** Statistical Analysis
- D** Data Interpretation
- E** Manuscript Preparation
- F** Literature Search
- G** Funds Collection

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### Summary

#### Background:

Obstruction of the inferior vena cava (IVC) is infrequent, membranous obstruction of the IVC (MOIVC) being one of its rare causes. Early diagnosis is important, as it can lead to hepatic congestion, cirrhosis and Budd-Chiari syndrome (BCS) and can predispose to development of hepatocellular carcinoma (HCC) in severe cases.

#### Case Report:

We report a case of membranous IVC obstruction at the junction of hepatic and suprahepatic segments in a young male with extensive collateralization and venous aneurysms. Unique findings involved antegrade and retrograde flow during respiration in the upper part of intrahepatic IVC proximal to a large collateral vein as well as prostatic and urethral congestion leading to intermittent urinary hesitancy, which have not yet been described in such cases.

#### Conclusions:

MOIVC is a rare cause of IVC obstruction with typical radiological features. Early diagnosis and management is required due to risk of cirrhosis and HCC. Antegrade and retrograde flow may be seen in incomplete MOIVC above the level of a large collateral vein and it may lead to prostatic and urethral congestion.

#### MeSH Keywords:

Prostate • Tomography, Spiral Computed • Ultrasonography, Doppler, Color • Vena Cava, Inferior

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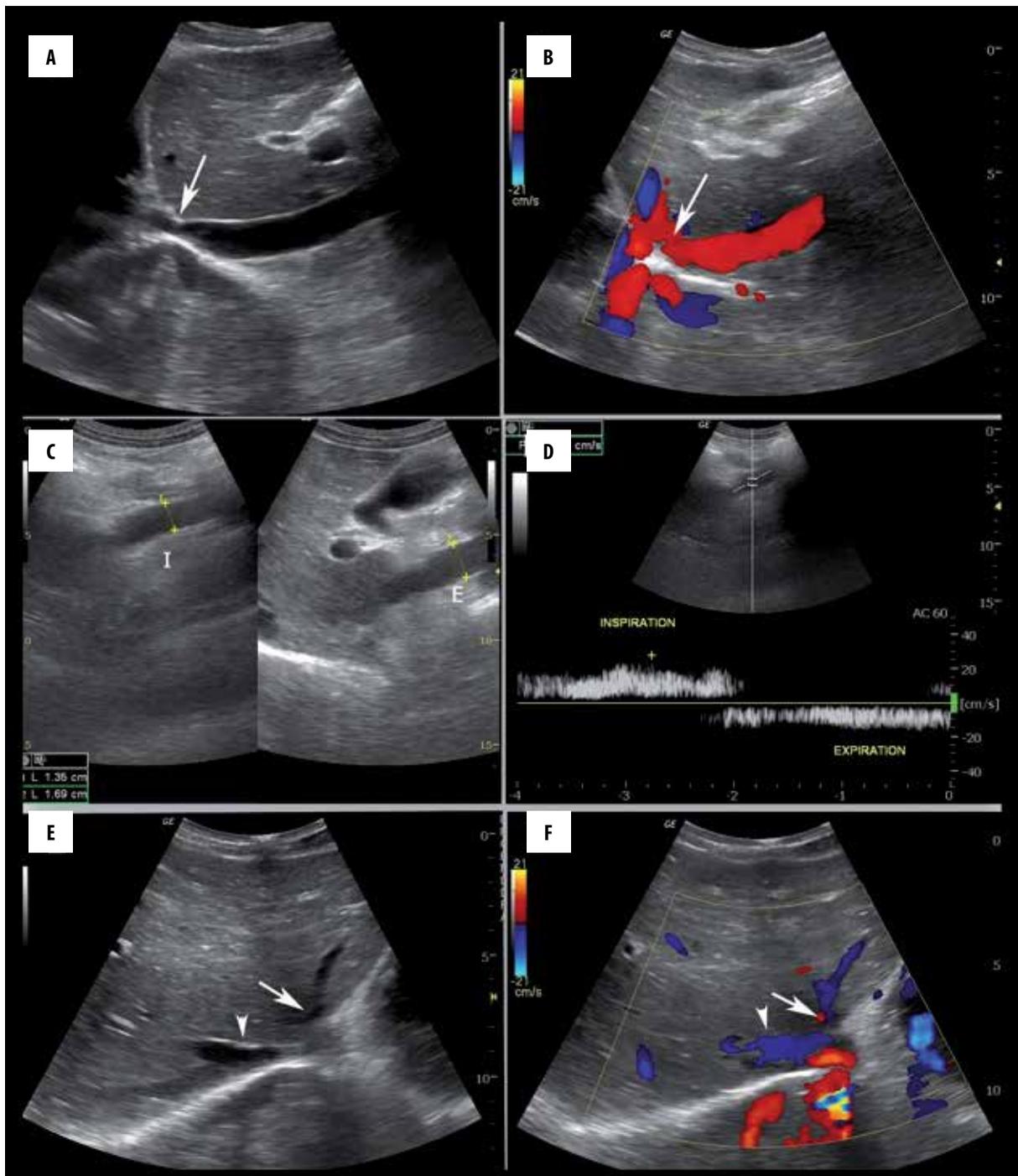
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### Background

Obstruction of the inferior vena cava (IVC) is infrequent, membranous obstruction of the IVC (MOIVC) being one of its rare causes. Early diagnosis is important, as it can lead to hepatic congestion, cirrhosis and Budd-Chiari syndrome (BCS) and can predispose to development of hepatocellular carcinoma (HCC) in severe cases.

### Case Report

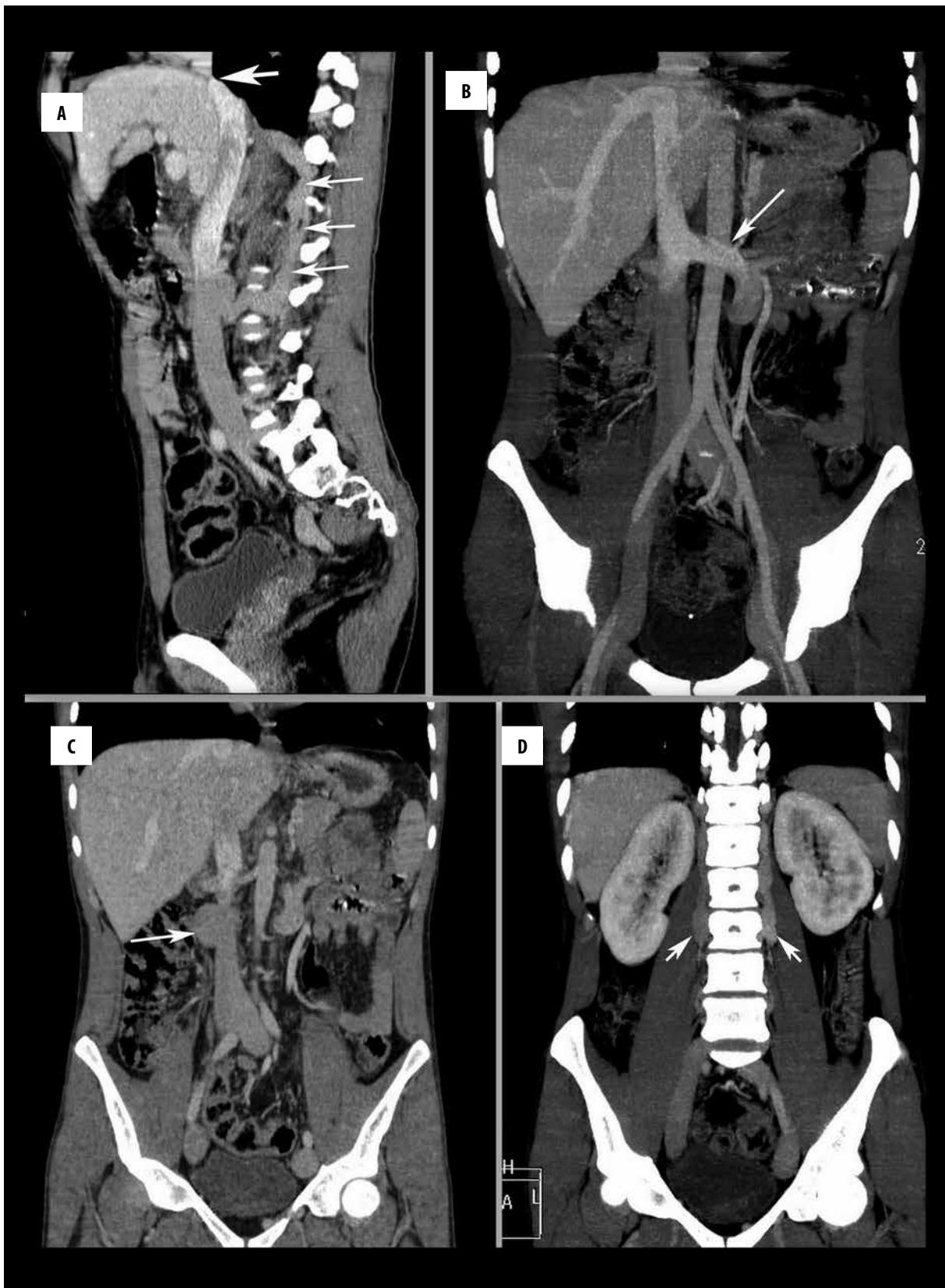
A 26-year-old Tibetan male presented with longstanding history of fatigue and breathlessness on moderate to severe exertion, such as running or hiking, for the past 6 years along with mild intermittent urinary hesitancy for 1 year. There was no history of erectile dysfunction or priapism. Hemogram, coagulogram, renal and liver function tests were normal. Serum electrolytes were unremarkable. Routine urinary examination and microscopy revealed no abnormalities.



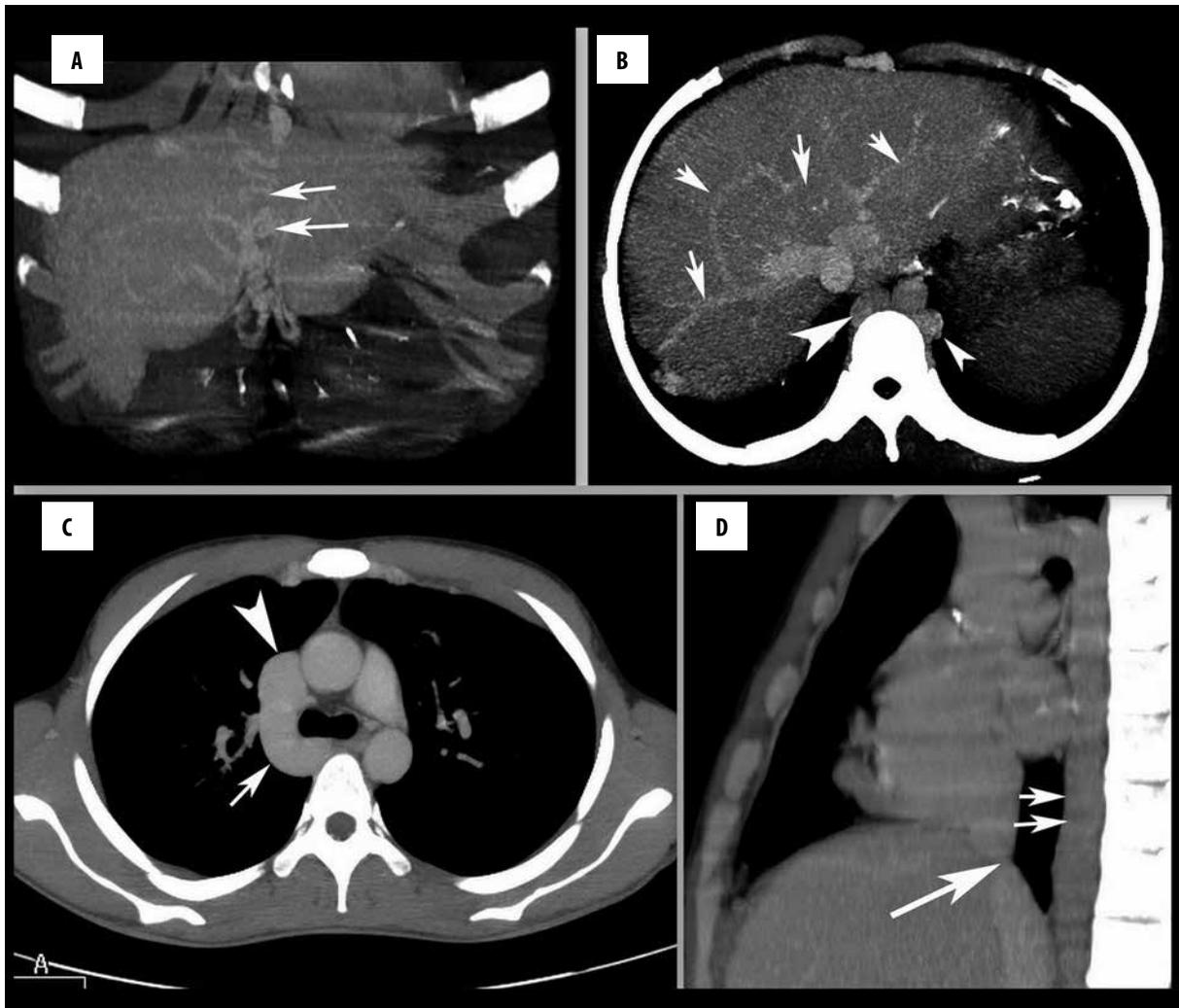
**Figure 1.** US and Doppler images of a 26-year-old male patient. Sagittal US (A) and color Doppler (B) images through the upper abdomen on inspiration showing marked narrowing of IVC at the junction of intrahepatic and suprahepatic segments (arrow in A, B). Sagittal US (C) images through upper abdomen show normal (approximately 20%) respiratory IVC collapse with IVC diameter decreasing during inspiration and increasing on expiration. Pulsed wave Doppler (D) shows cardiofugal flow on inspiration and cardiopetal flow on expiration. Oblique axial greyscale US (E) and color Doppler (F) images show narrowing of left hepatic vein (arrow) and normal middle hepatic vein (arrowhead)

Abdominal ultrasound (US) was ordered as a part of routine assessment. It revealed dilated iliac veins and inferior vena cava (IVC) with abrupt narrowing at the junction of intrahepatic and suprahepatic IVC segments (Figure 1A) along with a narrowing of left hepatic vein (Figure 1E, 1F) and presence of multiple intrahepatic venous collaterals.

Color Doppler and pulsed wave Doppler showed cardiofugal flow in intrahepatic IVC on inspiration (Figure 1B, 1D) and cardiopetal flow on expiration devoid of transmitted cardiac pulsation (Figure 1D). Intrahepatic IVC showed normal respiratory collapse, which amounted to approximately 20% (Figure 1C). Lower IVC and iliac veins showed normal



**Figure 2.** Sagittal CT images of the abdomen in venous phase (A) showing abrupt narrowing at the junction of intra- and suprahepatic IVC (large arrow) along with dilated paravertebral collaterals (small arrows). Coronal maximum intensity projection (MIP) CT images (B–D) showing dilated IVC, iliac vein, prominent venous collateral (arrow in B), a venous aneurysm (arrow in C), and dilated paravertebral collaterals (arrows in D)



**Figure 3.** Coronal MIP image shows partially recanalized umbilical vein (arrows in **A**). Axial (**B, C**) and sagittal (**D**) maximum intensity projection (MIP) CT images show dilated azygos vein (large arrowhead in **C** and double arrow in **D**) draining into SVC (arrowhead in **C**), dilated hemiazygos vein (small arrowhead in **B**) and intrahepatic collaterals (arrows in **B**).

cardiopetal flow. Prostate enlargement was noted. Urinary bladder was of normal wall thickness and both kidneys were unremarkable.

Subsequent computed tomography (CT) venogram confirmed marked narrowing at the junction of intrahepatic and suprahepatic segments of IVC along with dilated abdominal IVC with venous aneurysm, dilated iliac veins, paravertebral veins, as well as azygos and hemiazygos veins (Figures 2, 3). Left hepatic vein was narrowed with multiple intrahepatic collaterals (Figure 3B) and partially recanalized umbilical vein (Figure 3A). Middle and right hepatic veins were normal and there were no features of cirrhosis or portal hypertension. No filling defects were seen in hepatic veins. No esophageal or gastric varices were noted. The prostate was enlarged, approximately 56 cc in volume. Multiple dilated venous channels were visualized in periprostatic region and along submucosal region of bulboprostatic urethra (Figure 4). Testicular veins were normal. The patient was considered for percutaneous transluminal balloon angioplasty of the narrowed segment of IVC.

## Discussion

Embryologically, the inferior vena cava (IVC) develops between the fifth and seventh week of gestation from three paired veins, namely the postcardinal, subcardinal and supracardinal veins [1,2]. Congenital anomalies and variations of the IVC are infrequent findings [1].

Causes of IVC obstruction include thrombosis, infiltration by extravascular neoplasm, neoplasm arising from the vessel wall, compression of the IVC by neoplastic or non-neoplastic masses or idiopathic obstruction referred to as membranous obstruction of the IVC (MOIVC). Despite the name, many of these idiopathic lesions do not resemble a membrane and appearance may vary from a short, web-like narrowing to a long segmental occlusion with or without narrowing of hepatic veins, therefore the term "idiopathic obstruction of the IVC" may be better suited [3,4].

This entity is rare in most parts of the world, although increased prevalence was reported in Nepal, South Africa, Japan, India, China, and Korea. The exact etiology of the



**Figure 4.** Axial (A) and sagittal (B) CT images through the pelvis showing enlarged prostate. Axial (C) MIP CT image shows multiple prominent periprostatic venous channels (arrowheads). Axial CT image through the proximal bulbar urethra (D) shows prominent submucosal vascular channels (arrows).

MOIVC is a subject of controversy and it is yet unclear whether it is a developmental abnormality or secondary to organization of a thrombus in the hepatic portion of inferior vena cava. However, in most cases onset of symptoms occurs in the early adulthood suggesting acquired etiology [3,5]. Terabayashi et al. reported a case of thrombosis transforming to membranous obstruction in a young female with lupus anticoagulant, which may further support a theory of acquired etiology [6]. However, there was no evidence of a coagulation disorder in our case. It typically affects adults with insidious onset and gradual progression, often leads to cirrhosis in a course of chronic hepatic congestion and may culminate in Budd-Chiari syndrome. MOIVC is a major cause of primary Budd-Chiari syndrome in India [4]. A longstanding obstruction and severe centrilobular fibrosis predispose to development of hepatocellular carcinoma (HCC), which is more prevalent among black South Africans and Japanese [5].

The important draining collateral veins in case of MOIVC include: accessory right hepatic vein (for intrahepatic collaterals), ascending lumbar veins (draining into the azygos vein on the right side and the hemiazygos vein on the left), left renal vein, inferior phrenic-pericardiophrenic collateral vessels, extrahepatic portosystemic collateral vessels, and superficial collateral vessels of the abdominal wall [4]. In less severe IVC involvement IVC with adequate collateralization patient may be asymptomatic or may have only mild symptoms with normal liver function tests (3). Color Doppler US, CT, MR imaging, hepatic venography, and cavography can be used for imaging of IVC obstruction [4,7].

Vijayaraghavan et al. reported a case of valvular type MOIVC without antegrade (cardiopetal) flow, with retrograde (cardiofugal) flow in the upper IVC above a large lumbar collateral vein visible on inspiration [8]. In our case we noted antegrade and retrograde flow in the upper intrahepatic IVC proximal to large collaterals, which was directed toward the heart on expiration and reversed on inspiration. To our best knowledge, such flow pattern has not yet been described in a case of MOIVC.

We also noted presence of prostatic and urethral congestion leading to prostatic enlargement and intermittent urinary hesitancy in our case, which was likely caused by obstruction of the IVC and resultant increase in venous pressures in prostatic venous plexus and periurethral veins. To our best knowledge, this phenomenon has not been described yet in association with IVC obstruction in English literature. In a large Japanese study on taxi drivers and office workers Minamiguchi (1998) attributed prostatodynia to intrapelvic venous congestion, the diagnosis of which was determined based on dilated prostatic capsular veins and/or venous plexus of the lateral ligament of the bladder on 3D magnetic resonance venography [9]. We believe that similar congestion caused lower urinary tract symptoms in our patient as well.

The length of the involved segment and thickness of the membrane are important for management of MOIVC, which should be precisely evaluated on preoperative inferior venacavogram. Cases with thin membranes are

commonly treated with percutaneous transluminal balloon angioplasty with or without stent implantation. Thicker membranes necessitate transatrial membranotomy using cardiopulmonary bypass technique [5,7].

## Conclusions

MOIVC is a rare cause of IVC obstruction with typical radiological features. Early diagnosis and management is required due to risk of development of cirrhosis and HCC. Antegrade and retrograde flow may be seen in incomplete MOIVC above the level of a large collateral vein and it may lead to prostatic and urethral congestion due to increased venous pressure.

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