ORIGINAL ARTICLE

MDCT venography in the evaluation of inferior vena cava in Budd-Chiari syndrome

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Abstract

Objective To assess the role of multidetector computed tomography (MDCT) venography in the evaluation of the inferior vena cava (IVC) in Budd-Chiari syndrome (BCS), its accuracy as compared to digital subtraction venography (DSV) and the potential of this technique to replace venography for the definitive diagnosis of BCS.

Methods Twenty-five suspected cases of BCS were prospectively enrolled in this study and underwent both MDCT venography and DSV. Two observers independently evaluated and graded both the axial and reformatted MDCT images for the presence, site, degree and length of IVC narrowing. The collateral pathways and the hepatic veins were also assessed in all cases. The degree of correlation between MDCT venography and DSV was expressed using Spearman’s rank correlation coefficient (Rs).

Results There was excellent correlation between MDCT venography and DSV in predicting the presence of stenosis and in grading the degree and length of IVC stenosis (Rs=0.58, p<0.05). Four patients had presence of a web within the IVC and the reconstructed MDCT venography images detected the flap of the membrane in all of them. In three cases of complete obstruction the cranial extent of the obstruction could be determined on the reconstructed MDCT venography images, while double catheter access through the femoral and jugular routes was needed to determine the same on DSV. MDCT venography was significantly more informative in depicting the presence and site of both intrahepatic and extrahepatic collaterals as compared to DSV.

Conclusion MDCT venography, in the present study, accurately provided information of both conventional CT and IVCgraphy, in the evaluation of the IVC in a non-invasive way. It helped overcome the shortcomings of CT in the evaluation of IVC and was better than DSV for the evaluation of collaterals, calcification and complete IVC obstruction. We suggest that CT venography can be used as a frontline investigation for the diagnosis of IVC obstruction and for planning surgery or percutaneous endovascular intervention.

Keywords Budd-Chiari syndrome · Venography · Hepatocellular carcinoma

Introduction

Budd-Chiari syndrome (BCS) is defined as hepatic venous outflow obstruction at any level from the small hepatic veins to the cavo-atrial junction, regardless of the cause. It can be classified according to etiology (primary or secondary), site of obstruction, manifestations or duration of disease.

The site of obstruction can be at the level of small hepatic veins, large hepatic veins, inferior vena cava (IVC), or a combined obstruction of the hepatic veins and IVC. Unlike the West, where hepatic vein thrombosis is responsible for a majority of cases with BCS, in Africa and Asia, IVC obstruction is the commonest cause of BCS. In various series, it accounted for 39–78% of patients with BCS. Hepatic vein thrombosis is commonly due to an underlying hypercoagulable state and presents more often with an acute illness. In contrast, BCS due to IVC obstruction, also known as obliterative hepatocavopathy or membranous obstruction of IVC, is frequently idiopathic and is associated with a milder, more chronic disease.
Accurate and early diagnosis of BCS is essential as it is a potentially treatable cause of liver disease, which if untreated may predispose to hepatocellular carcinoma. Knowledge of exact site, extent and degree of obstruction of the IVC is important for proper management. Though several modalities, including ultrasonography (USG), color Doppler, multidetector computed tomography (MDCT), magnetic resonance imaging (MRI) and scintigraphy have been used for evaluation of IVC obstruction, venography remains the mainstay for definitive diagnosis and for guiding surgical and/or endovascular therapy. In this prospective study, we assessed the role of CT venography in the evaluation of the IVC in BCS, its accuracy as compared to venography and explored the potential of this technique to replace venography for the definitive diagnosis of BCS.

Methods

Patients

Twenty five consecutive patients (median age 29 [range 10–58] years; 15 women), with sonographic or Doppler findings suggestive of primary BCS, or a strong clinical suspicion of BCS with equivocal sonographic findings, were studied after obtaining informed consent. Institute Board approval was taken prior to the study. Patients with an underlying hepatocellular carcinoma or renal cell carcinoma were excluded. Clinical symptoms were non-specific and included abdominal pain, abdominal distention, varicosity of veins in the abdominal wall and legs, jaundice, lower limb edema and variceal bleeding. Nine patients had associated predisposing hypercoagulable conditions, including use of oral contraceptives (n = 3), paroxysmal nocturnal hemoglobinuria (n = 2), protein C deficiency (n = 2), polycythemia (n = 1) or nephrotic syndrome (n = 1).

CT venography

All MDCT venography examinations were performed on a 16-detector row scanner (Sensation 16; Siemens). The femoral vein was cannulated using a 16G intravenous cannula, and 60 mL of a non-ionic iodinated contrast medium (Ultravist 370; Schering, Berlin, Germany) was injected at a rate of 4 mL/sec at 300 psi using a pressure injector. The scan parameters used were: beam collimation 2.5 mm, pitch 0.75, reconstruction interval 0.75 mm, gantry rotation time 0.5 sec, tube current 200–250 mAs and tube voltage 120 kVp. Scanning was performed in the caudo-cranial direction with an automatic bolus tracking software (C.A.R.E. Bolus, Siemens, Germany) used for scan initiation, with the region of interest positioned at the level of infrarenal IVC and the threshold for triggering data acquisition preset at 100 HU. The scans were acquired from the level of L4 vertebra to the level of the pulmonary hilus with z-axis coverage of 32–38 cm. Subsequently, to evaluate the hepatic veins, portal vein, liver parenchyma and the associated changes of BCS, images were taken in the portal venous phase from the domes of diaphragm to the pubic symphysis in the craniocaudal direction, using slice thickness of 10 mm, pitch of 1.5 and a reconstruction interval of 2 mm.

For each data set, three series of images were systematically reconstructed, as follows: contiguous 1 mm thick transverse CT scans, oblique coronal and sagittal maximum intensity projections (MIPs) and three-dimensional volume-rendered images of the IVC.

Digital subtraction venography (DSV)

In all patients conventional venography was performed within two days of the MDCT venography using a digital subtraction technique. Venography was performed via the right transfemoral route by placing a 6F arterial sheath in the right femoral vein. Non-ionic iodinated contrast (30 mL) was injected at 12 ml/sec and 300 psi using a pressure injector. Flush IVCgrams were taken in the antero-posterior and lateral views in suspended respiration with the pigtail catheter placed in the suprarenal location. Pressure was measured at each vertebral level from the level of the cavo-atrial junction to the level of renal veins and a pressure gradient of 20 mm Hg across the stenotic site was considered significant. Where technically feasible, attempts were made to selectively catheterize all the three hepatic veins and obtain venograms.

Image analysis

The MDCT venography and the DSV were reviewed by two separate radiologists, who were blinded to the results of the other study. DSV was considered as the gold standard. Presence of significant IVC obstruction, and its site, extent and degree of obstruction was recorded. The projection that showed the most severe stenosis was used for measurement. The diameter at the site of the most severe stenosis was divided by the arbitrary normal value determined after consideration of the diameter of the proximal and distal vein beyond the stenosis; the resulting value was subtracted from one and then multiplied by 100 to yield the percentage of stenosis according to the diameter of the vessel. Stenosis was graded separately for MDCT venography and DSV as follows: 0 (1–49%) – insignificant stenosis, I (50–74%) – moderate stenosis; II (75–99%) – severe stenosis; III – complete occlusion. The length of stenosis was also graded on both CT venography and DSV as follows: I– <5 mm, II – 5–9 mm, III– 10–14 mm, IV– 15–20 mm and V– >20 mm. The intrahepatic and extrahepatic collaterals as well as the status of the hepatic veins were also studied in each case. An attempt was made to identify theazygous vein, hemiazygous vein, paravertebral and superficial abdominal wall collaterals in all the sixteen cases with significant obstruction.

Statistical assessment

Data on severity and length of stenosis on CT venography and DSV were analyzed using the Spearman’s rank correla-
tion coefficient (Rs). SPSS version 10.0 software was used, and p values <0.05 were considered significant.

**Results**

**Presence of IVC obstruction**

Four patients had a normal IVC on both venography and MDCT venography with isolated hepatic vein thrombosis being the cause of BCS. Five patients showed long segment narrowing of the IVC on the axial CT sections, but no significant obstruction on the sagittal reconstructed images on MDCT venography. This was in agreement with the findings at DSV of absence of a significant pressure gradient in the IVC (Fig. 1). Sixteen of the 25 patients showed significant obstruction on both DSV and MDCT venography.

**Site/Cause of IVC obstruction**

Of the 16 patients with significant IVC obstruction, three had stenosis at the level of cavo-atrial junction whereas the rest had obstruction of the intrahepatic IVC. Four patients had presence of a web within the IVC and the reconstructed MDCT venography images detected the flap of the membrane in all cases (Fig. 2). Three patients had complete obstruction of the intrahepatic IVC and the cranial extent of the occlusion could not be determined on DSV with a double catheter access being needed for the evaluation (both femoral and jugular venous access). In all cases of complete obstruction of the IVC, cranial extent of the obstruction could be seen on the delayed axial CT images (Fig. 3). Five patients had short segment stenosis of the IVC on both modalities.

Four patients showed significant long segment narrowing of the IVC due to caudate lobe hypertrophy. These patients had significant pressure gradient on venography and showed significant narrowing (>50%) on MDCT venography in both the AP and sagittal projections (Fig. 4).

**Degree of IVC obstruction**

There was excellent correlation between CT venography and DSV in grading the IVC stenosis. The degree of obstruction was grade 0 in nine cases, grade I in three cases, grade II in ten cases and grade III in three cases. All stenosis were classified in the same grade by both modalities (Rs=0.58) (Fig. 5).

**Length of stenosis**

The length of obstruction was grade I in four cases, grade II in one case, grade III in one case, grade IV in three cases and grade V in four cases. There was excellent correlation between CT venography and DSV in grading the length of stenosis, with all stenosis being classified in the same grade by both modalities (Rs = 0.58). Three cases had complete obstruction of the IVC and the length of obstruction could be graded only on MDCT venography as the cranial extent of obstruction could not be identified on DSV. One each of these cases was graded as III, IV and V.

**Intrahepatic and extrahepatic collaterals**

Sixteen cases had significant obstruction. Dilated hemiazygous vein was seen in seven cases on MDCT veno-

![Fig. 1 a: Digital subtraction venography (DSV) anteroposterior (AP) view shows significant long segment narrowing of the intrahepatic IVC with dilated paravertebral collaterals. b and c: MDCT venography AP and lateral volume rendered (VRT) images. The AP view, as in DSA shows significant narrowing of intrahepatic IVC. The lateral view shows normal caliber of IVC, preventing misdiagnosis of significant IVC stenosis. No significant pressure gradients were noted in this case.]
graphy and three cases on DSV, azygous vein in five cases on MDCT venography and three cases on DSV, paravertebral collaterals in ten cases on MDCT venography and five cases on DSV, whereas superficial abdominal wall collaterals were identified in six cases on MDCT venography and two cases on DSV. Tortuous, comma shaped intrahepatic collaterals could be identified in all but one case on MDCT venography; however, the typical “spider-web” collateral could be selectively cannulated in only seven of the 16 patients on DSV. Thus, MDCT venography was more informative than DSV in depicting the presence and site of both intrahepatic and extrahepatic collaterals.

Fig. 2  a and b: DSV and MDCT venography VRT, AP magnified views showing presence of web at cavoatrial junction. The flap of the membrane was well demonstrated by MDCT venography and the thickness of the web was measured as 2 mm on both modalities.

Fig. 3  a: DSV AP view showing complete occlusion of the intrahepatic IVC with grossly dilated paravertebral collaterals and azygous-hemiazygous system. The cranial extent of occlusion could not be determined. b and c: MDCT venography AP VRT and axial images. There is complete occlusion of the intrahepatic IVC with dilated collaterals showing excellent correlation with DSV. The cranial extent of obstruction could be determined on the axial image and the cavoatrial junction was demonstrated as normal.
Hepatic veins

In 8 of the 16 patients, all three hepatic veins were patent in their entire extent on MDCT venography. Only 14 of 24 hepatic veins in these eight patients, could be selectively cannulated on DSV. In the other eight patients, 10 hepatic veins could be demonstrated on MDCT venography, whereas only three could be selectively cannulated on DSV.
Discussion

BCS is not an uncommon cause of hepatic venous outflow obstruction leading to liver disease in the Far East, the Middle east, India and Africa.\textsuperscript{2, 3} IVC obstruction, with or without involvement of the hepatic veins, is the cause of BCS in the majority of the cases in these geographical areas.\textsuperscript{4}

Simson described three types of membranous obstruction of IVC: in type 1, a thin membrane is present in the IVC, in type II a variable segment of the IVC (>1 cm) is replaced by a fibrous cord with the IVC gram showing characteristic conical narrowing and in type III a variable segment of IVC below the obstruction is filled with thrombus.\textsuperscript{8} In all cases there may be a partial or complete occlusion of the IVC. In a patient with isolated hepatic vein thrombosis, extrinsic compression of the intrahepatic IVC by the hypertrophied caudate lobe can lead to erroneous clinical and radiological features of combined hepatic vein and IVC obstruction. Compression by an enlarged caudate lobe may cause narrowing of the IVC in the anteroposterior view but a lateral view on venography, along with the pressure measurements helps determine if the narrowing is causing significant obstruction.

An accurate evaluation of IVC obstruction is essential for proper surgical and radiological percutaneous management. Traditionally, surgical shunts including mesocaval, portocaval and mesoatrial shunts have been used in the treatment of BCS. A significant obstruction of the IVC helps decide the choice of surgical shunt as a significant pressure gradient in the IVC precludes the use of a mesocaval or portocaval shunt and a mesoatrial shunt, despite it’s high morbidity, is done in these cases. In the past decade, percutaneous radiological management is becoming the procedure of choice for management of BCS. For thin membranes (Type I disease) membranotomy may be done, while for short segment stenosis, percutaneous angioplasty with stenting is the preferred procedure.\textsuperscript{9, 10} The information about the severity and extent of IVC obstruction, the presence and thickness of IVC web as well as presence or absence of venous thrombosis helps select possible candidates for radiological intervention, determine the type of percutaneous management and select a stent of suitable diameter and length.

Several modalities, including duplex sonography, CT, scintigraphy and MRI, have been used for the evaluation of the IVC in BCS and compared with venography for demonstration of type, extent and degree of stenosis. Duplex sonography because of its ready availability and non-invasive nature is a simple, safe and quick method for the screening and initial diagnosis for BCS. However, in a significant number of cases, body habitus and bowel gas may be limiting factors and the adequate evaluation of IVC may not be possible.\textsuperscript{11} A nodular or a shrunken and cirrhotic liver may obscure patent hepatic veins, and an enlarged caudate lobe can lead to the erroneous diagnosis of significant IVC stenosis on doppler.\textsuperscript{12} The evaluation of systemic collaterals by Doppler is not reliable. Doppler fails to provide a comprehensive overview of the whole disease and also cannot provide comprehensible anatomic images to the surgeon and the interventionist; hence, it cannot be used as a definitive investigation, replacing venography for the evaluation of IVC stenosis.

Conventional CT has been compared with venography for the depiction of IVC stenosis in primary BCS.\textsuperscript{13} CT is an excellent modality for showing the liver parenchymal changes in BCS, to detect hepatomas, to evaluate the secondary causes of BCS, in demonstrating calcification of the IVC and extrahepatic collateral vasculature. It however, remains unreliable for depiction of IVC webs and for determining the presence of significant stenosis and the exact length and degree of stenosis.\textsuperscript{14, 15} The lack of 3D visualization of the IVC, flow artifacts due to inflow of unopacified blood leading to appearance of “pseudothrombosis” and inability to assess the significance of IVC stenosis due to caudate lobe compression, all contribute to this. There may be preferential opacification of collateral vessels proximal to the site of IVC obstruction, further preventing the accurate assessment of the caudal extent of IVC stenosis on conventional CT.

MR angiography has also been used for the evaluation of IVC in BCS and with its 3D multiplanar capabilities and excellent soft tissue resolution it has given encouraging results.\textsuperscript{16} However, it is an expensive modality with limited availability and cannot be performed in acutely ill or uncooperative patients. Motion and flow artifacts may lead to limited technical success at times and detection of IVC calcification is unreliable on MRI.

To the best of our knowledge, this is the first study comparing MDCT venography with conventional venography for the evaluation of the IVC in primary BCS. MDCT venography showed excellent correlation with DSV in predicting the presence of significant stenosis and accurately classified the length and degree of stenosis in all cases. In five cases of long segment IVC compression due to caudate lobe hypertrophy, on studying the sagittal reconstructions, no significant stenosis was seen and this was confirmed on venography, in which no significant pressure gradient was found in these cases. Conversely, four cases of long segment IVC stenosis due to caudate lobe hypertrophy which were classified as moderate stenosis on MDCT venography, showed significant pressure gradient on venography. The caudal and cranial extent of IVC stenosis was accurately identified in all cases. In three cases of complete obstruction the cranial extent of the obstruction could be determined on the reconstructed axial and MIP images, while double catheter access through the femoral and jugular routes was needed to determine the same on venography. All four cases of IVC web with their exact thickness were correctly identified on the reconstructed MDCT venography images. Unlike CT, the problem of “pseudothrombosis” was not faced in any case on CT venography. CT venography was more informative than venography in showing extra and intrahepatic collateral vessels.
DSV is the gold standard for evaluation of IVC in BCS.\textsuperscript{6,17} It provides pressure measurements and thus helps guide surgery. Also, endovascular therapy can be performed at the same sitting. It is however invasive, time consuming, and needs expertise.

A limitation of the present study is that all the hepatic veins which were found to be patent on MDCT venography, could not be selectively cannulated on DSV. This may have been due to the use of femoral route in all cases. Use of double (both jugular and femoral) access could have increased the success rate of selective cannulation of hepatic veins.

In conclusion, in our study, MDCT venography provided accurate evaluation of IVC, equivalent to that obtained with a combination of conventional CT and IVCgraphy. It provided information about the liver parenchyma, cause of BCS, status of the hepatic veins and the splenoportal axis, complications of chronic BCS, length of IVC narrowing, and extrahepatic and intrahepatic collaterals. It helped overcome the shortcomings of CT in the evaluation of IVC and was better than venography for the evaluation of collaterals, calcification and complete IVC obstruction. We suggest that CT venography can be used as a frontline investigation for the diagnosis of IVC obstruction in patients with BCS, and for planning surgery or percutaneous endovascular intervention, with venography being reserved for therapeutic purposes.

References

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