

# CT-angiographic demonstration of hepatic collateral pathways due to superior vena cava obstruction in Behçet disease

Osman Temizöz, Hakan Genchellac, Ensar Yekeler, Mustafa Kemal Demir, Ercüment Ünlü, Hüseyin Özdemir

## ABSTRACT

Behçet disease (BD) is a chronic multisystemic inflammatory disorder, mainly characterized by recurrent oral and genital ulcers, skin lesions, and uveitis. Large vein thrombosis in BD is unusual; when present, it is most frequently seen in the inferior or superior vena cava (SVC). The authors describe an unusual hepatic pseudolesion caused by abnormal focal enhancement through collateral pathways to the liver in two BD patients with SVC occlusion on three-dimensional multi-detector computed tomography, using volume rendering and maximum intensity projection techniques. BD should be suspected in patients presenting a focal increased hepatic enhancement area with collaterals caused by occlusion of the SVC without evidence of a hypercoagulable state or malignant mediastinal or thoracic venous inlet obstruction.

*Key words:* • Behcet disease • vena cava, superior • computed tomography

**B**ehçet disease (BD) is a rare form of systemic vasculitis which affects all types and sizes of blood vessels. It was first described by a Turkish dermatologist, Hulusi Behçet, in 1937 (1). Its etiology is still unclear. The classical triad of the disease is recurrent oral and genital ulcerations and uveitis. The vasculitis frequently affects veins and usually results in venous thrombosis of the extremities. Superior vena cava (SVC) thrombosis is rare and accounts only for 9.8% of cases (2, 3). A focus of intense hepatic enhancement in SVC obstruction is also uncommon and has been demonstrated by venography, radioisotope study, and computed tomography (4, 5).

Multidetector computed tomography (MDCT) with maximum intensity projection (MIP) and three-dimensional (3D) volume rendering (VR) represents an advance over the classic two-dimensional CT study and MR angiography. It provides shorter acquisition times, greater anatomic coverage, superior image resolution with improved temporal and spatial resolution, and higher quality reconstructions. With reduced cost and radiation dose, MDCT also presents high-quality vascular images that are equal or superior to those of conventional angiography (6, 7).

The purpose of this study was to describe the focal increased hepatic enhancement area and the collateral pathways to the liver in two BD patients with SVC obstruction on 3D-MDCT with VR and MIP techniques.

## Case reports

### Case 1

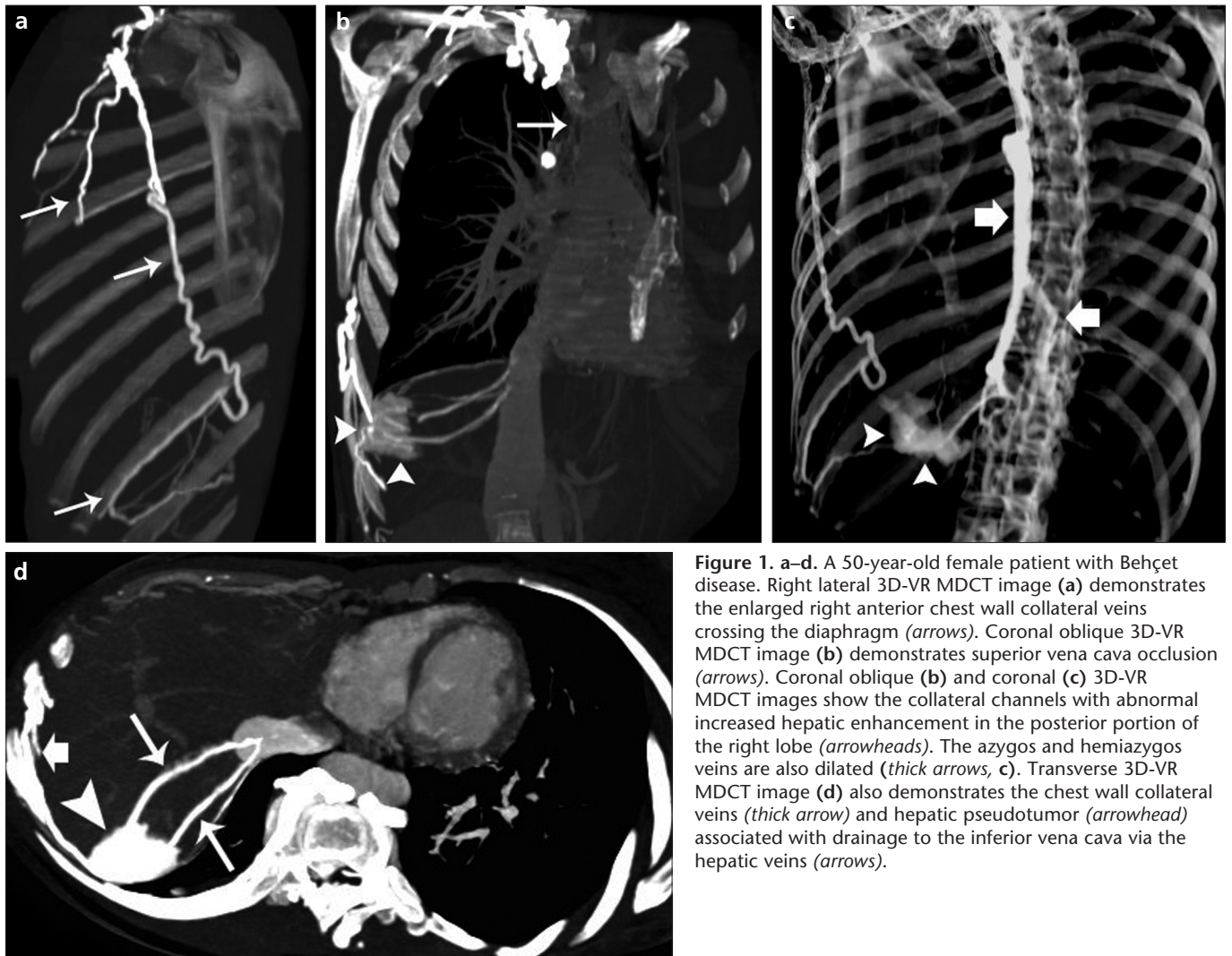
A 50-year-old female patient with BD was admitted to our hospital with a 4-week history of persistent dyspnea and chest pain. Physical examination revealed swelling of the neck and upper extremities. In addition, there were visible venous collateral channels, particularly on the right lateral side of her upper trunk. Laboratory tests including complete blood cell count, serum electrolytes, and clotting profiles were normal. A clinical diagnosis of SVC syndrome was made, and MDCT of the chest and abdomen was performed. A total of 120 mL of iohexol (Omnipaque 350, Amersham Health Inc. Princeton, New Jersey, USA) was injected at a rate of 3.5 mL/s. The area scanned extended from the supraclavicular region to just below the common iliac arteries on the arterial and venous phases. MDCT images were reconstructed using MIP and 3D-VR.

MDCT examination and reconstruction of 3D images revealed occlusion of the SVC (Fig. 1a). The dilated right lateral chest wall veins were seen as collateral channels crossing the right diaphragm on the images (Fig. 1a, b). The collateral channels were visualized with abnormal increased hepatic enhancement in the posterior portion of the right lobe, simulating a space-occupying lesion. This hepatic pseudotumor drained to the inferior vena cava via hepatic veins (Fig. 1b–d). The azygos and hemiazygos veins were also dilated (Fig. 1c).

From the Department of Radiology (O.T. ✉ [osmantemizoz@gmail.com](mailto:osmantemizoz@gmail.com), H.G., M.K.D., E.Ü., H.Ö.), Trakya University School of Medicine, Edirne, Turkey; and the Department of Radiology (E.Y.), İstanbul University School of Medicine, İstanbul, Turkey.

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**Figure 1. a–d.** A 50-year-old female patient with Behçet disease. Right lateral 3D-VR MDCT image (a) demonstrates the enlarged right anterior chest wall collateral veins crossing the diaphragm (arrows). Coronal oblique 3D-VR MDCT image (b) demonstrates superior vena cava occlusion (arrows). Coronal oblique (b) and coronal (c) 3D-VR MDCT images show the collateral channels with abnormal increased hepatic enhancement in the posterior portion of the right lobe (arrowheads). The azygos and hemiazygos veins are also dilated (thick arrows, c). Transverse 3D-VR MDCT image (d) also demonstrates the chest wall collateral veins (thick arrow) and hepatic pseudotumor (arrowhead) associated with drainage to the inferior vena cava via the hepatic veins (arrows).

## Case 2

A 56-year-old woman with a history of BD presented with swelling at her cervical region and face. Physical examination revealed edema of the neck associated with visible anterior chest wall collateral veins. Laboratory tests were normal. An MDCT of the chest and abdomen from supraclavicular level to the symphysis pubis demonstrated a thrombosed and narrowed SVC. The azygos, hemiazygos, and right anterior chest wall veins were dilated as collateral vessels (Fig. 2a). These anterior chest wall collateral veins communicated with the hepatic veins through the diaphragmatic venous plexus and drained to the inferior vena cava. In addition, there was an abnormal increased hepatic triangular enhancement simulating a mass in the medial segment of left lobe (Fig. 2b). A partially thrombosed abdominal aor-

tic aneurysm and infrahepatic inferior vena cava stenosis with intraabdominal venous collaterals were also detected (Fig. 2c, d). These venous collaterals drained partially to the suprahepatic inferior vena cava through the hepatic veins of posterior segment of the liver (Fig. 2b, d).

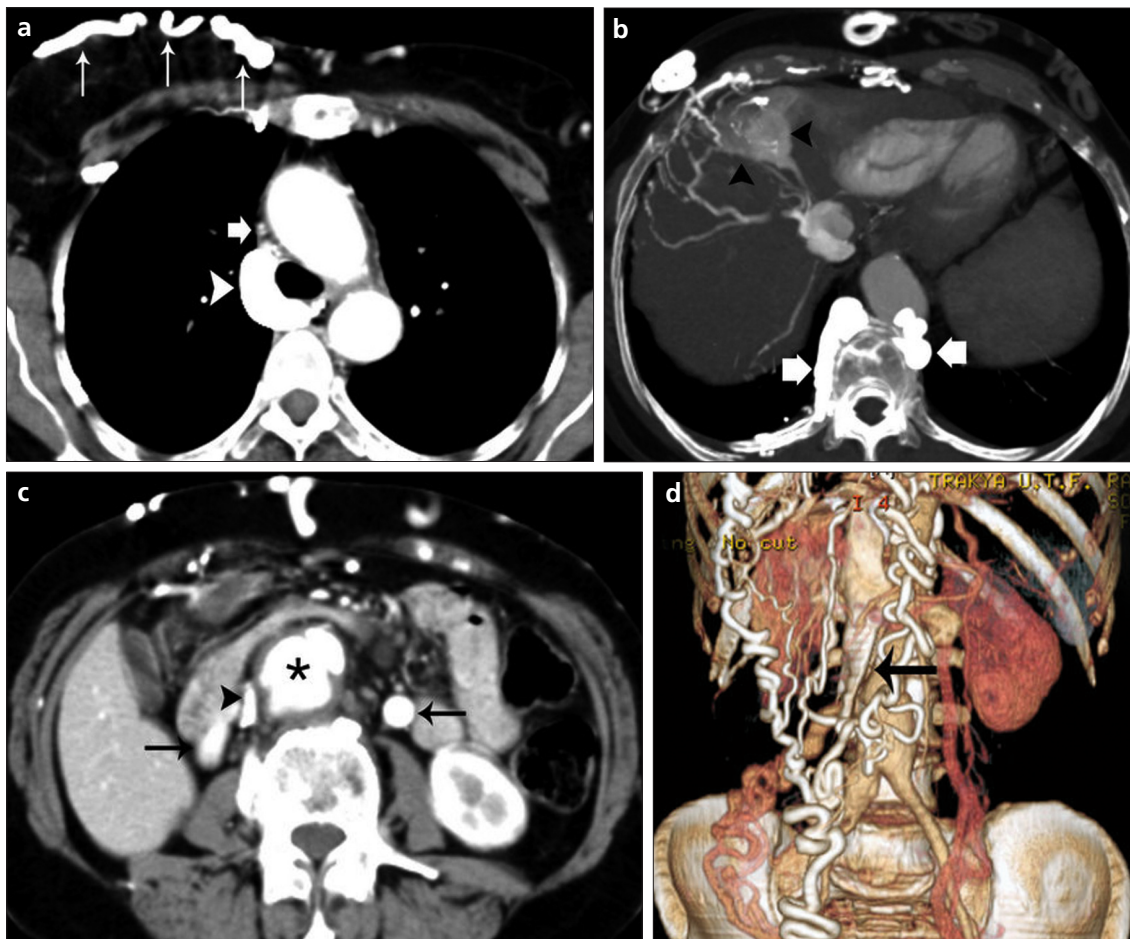
## Discussion

The major pathology in superficial and deep venous systems is the active inflammation in the endothelium in Behçet disease (8). Inflammatory vascular injury secondary to vasculitis is considered to be the most important cause for thrombosis (9). There is no cure for Behçet disease. Treatment typically focuses on reducing discomfort and preventing serious complications (10).

Although the most common cause of obstruction of the SVC is malignant neoplasia, it may also be caused by

benign diseases such as mediastinal fibrosis, irradiation, thrombosis induced by transvenous devices, and vasculitis. Vascular involvement in BD caused by systemic vasculitis can affect all vessels in the body, including the SVC. Abnormal enhancement of the hepatic parenchyma in SVC obstruction with collateral pathways on abdominal CT is extremely rare (5, 11, 12); it has never been reported in BD.

In the presence of SVC obstruction, blood from the arm is diverted into various pathways by main and accessory collateral routes (13, 14). Well-known collateral routes include the azygos-hemiazygos veins, internal mammary vein, vertebral venous plexus route, and the lateral thoracic and superficial thoracoabdominal vein. The superficial lateral thoracic wall veins may communicate with the right hepatic vein through the subscapular vein. This



**Figure 2. a–d.** A 56-year-old woman with a history of Behçet disease. Transverse MDCT image (a) through the level of upper mediastinum reveals a thrombosed and narrowed superior vena cava (thick arrow) with dilated azygos vein (arrowhead) and right anterior chest wall veins (arrows). Transverse 3D-VR MDCT image (b) demonstrates the communication of the dilated anterior chest wall collateral veins with the hepatic veins through the diaphragmatic venous plexus and drainage to the inferior vena cava. Additionally, there is an abnormal increased hepatic triangular enhancement simulating a mass in the medial segment of left lobe (arrowheads). The azygos and hemiazygos veins are markedly dilated (thick arrows). Transverse abdominal MDCT image (c) demonstrates a partially thrombosed abdominal aortic aneurysm (asterisk) and infrahepatic inferior vena cava stenosis (arrowhead) together with intraabdominal venous collaterals (arrows). Coronal 3D-VR MDCT image (d) clearly demonstrates the dilated collateral veins of thoracoabdominal region in the presence of superior vena cava and inferior vena cava occlusions (arrow).

anastomosis is rarely associated with abnormal enhancement of segment VIII of the liver, simulating a mass (5) as in case 1. However, in the case of SVC obstruction, the appearance increased enhancement in the cephalic portion of segment IV by contrast medium injected through an upper extremity via the internal thoracic vein, superior epigastric vein, and superior vein of Sappey is more common than a pseudolesion of the posterior portion of the right hepatic lobe (8, 15).

The reason for increased abnormal enhancement in segments IV and VIII of the liver in SVC obstruction is ob-

secure. The theories based on anatomical anastomoses failed to explain this phenomenon due to the presence of numerous accessory collateral routes. In this sense, the proposed association of the focal hepatic enhancement of the segment IV and existence of anastomoses between the paraumbilical and portal veins were not present in our case 1 (5). However, we agree that venous stasis in these areas due to excessive venous flow through collaterals may lead to the occurrence of focal increased abnormal enhancement in the segments IV and VIII of the liver in SVC obstruction (16).

Finally, understanding and diagnosing these pseudolesions in the liver are important for radiologists, because misinterpretation of them may lead the patients to undergo unnecessary investigations and treatments. Vascular malformation, hypervascular metastasis, hepatoma, focal nodular hyperplasia, and hemangioma are the most common focal liver lesions that must be considered in the differential diagnosis. The presence of related collateral vessels along the thoracic or abdominal wall is easily recognized on MDCT examination, specifically by using reconstruction techniques, and it

is helpful in the determination of the pathogenesis of these pseudolesions.

As a conclusion, BD should be suspected in patients presenting with focal increased hepatic enhancement area with collaterals due to occlusion of the SVC without evidence of a hypercoagulable state and malignant mediastinal or thoracic venous inlet obstruction.

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