

A Rare Presentation of an Entrapment in a Liver Transplant Candidate Depicted by MDCT Angiography

Karaciğer Transplant Adayında Nadir Rastlanabilecek Bir Tuzağın MDBT Anjiyografi ile Tanımlanması

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Abstract

Hypertrophic caudate lobe veins can mimic a normal venous configuration. In cases of multiple vascular collaterals, Doppler evaluations must be conducted, and the flow direction of these veins as well as the IVC should be evaluated. If the flow in the IVC is reversed, Budd-Chiari syndrome should be suspected; moreover, at the supra diaphragmatic level, which may be considered a blind spot, particularly for radiologists, a web should be searched for in the area where the IVC opens into the right atrium. In this study, we present the unique findings of multidetector computed tomography (MDCT) angiography for a liver transplant candidate with Budd-Chiari syndrome caused by a web in the proximal IVC.

Key Words: Budd-Chiari Syndrome, Liver Transplantation, MDCT

Özet

Hipertrofiye olmuş kaudat lob venleri, normal venöz konfigürasyonu taklit edebilir. Karaciğerde multipl vasküler kollaterallerin varlığında, Doppler incelemesi yapılmalı ve hem bu venlerin hem de IVC'nin akım yönü değerlendirilmelidir. IVC'de ters akım yönünün saptanması, Budd-Chiari sendromu açısından önem arz etmektedir. IVC'nin supradiafragmatik segmentinde, sağ atriüma açıldığı yerde olası bir web bu sendroma neden olabilmektedir. Bu lokalizasyon, özellikle radyologlar için bir kör nokta olarak kabul edilmektedir. Biz bu çalışmada, bir karaciğer transplant adayında proksimal IVC'deki webin neden olduğu Budd-Chiari sendromunu, multidetektör bilgisayarlı tomografi anjiyografinin (MDBTA) benzersiz bulguları ile sunmaya çalıştık.

Anahtar Kelimeler: Budd-Chiari Sendromu, Karaciğer Transplantasyonu, MDBT

Introduction

The term "Budd-Chiari syndrome" refers to the clinical manifestations of hepatic venous outflow obstruction at any level from the small hepatic veins to the junction of the inferior vena cava and the right atrium, regardless of the cause of obstruction [1]. Budd-Chiari syndrome can be either primary or secondary; the primary type is caused by an intrinsic luminal web or thrombosis, and the secondary type is caused by extraluminal compression or tumoral invasion [2]. In Asia, a membranous obstruction of the inferior vena cava (IVC) is the most common cause of Budd-Chiari syndrome [3]. We report a rare presentation of an entrapment in a liver transplant candidate with "hepatic pseudo star" identified using MDCT angiography.

Case Report

A 57-year-old female patient with a 20-year history of hepatitis B and secondary cirrhosis presented with chronic abdominal pain. While the patient was being prepared for liver transplantation, she was evaluated with multidetector computer tomography (MDCT).

MDCT angiography was performed with a 16-detector CT (Aquilion; Toshiba Medical Systems, Tokyo, Japan). The following acquisition parameters were used: 16x0.5 mm collimation, 1.0 mm slice thickness, and 1.0 mm reconstruction interval. Additionally, 120 mL of non-ionic iodine contrast matter (iohexol, Omnipaque;

Amersham Health, Cork, Ireland) was administered at a rate of 4 mL/s, followed by 40 mL of physiological serum at a rate of 2.5 mL/s. The fixed-delay technique was used to determine the starting time

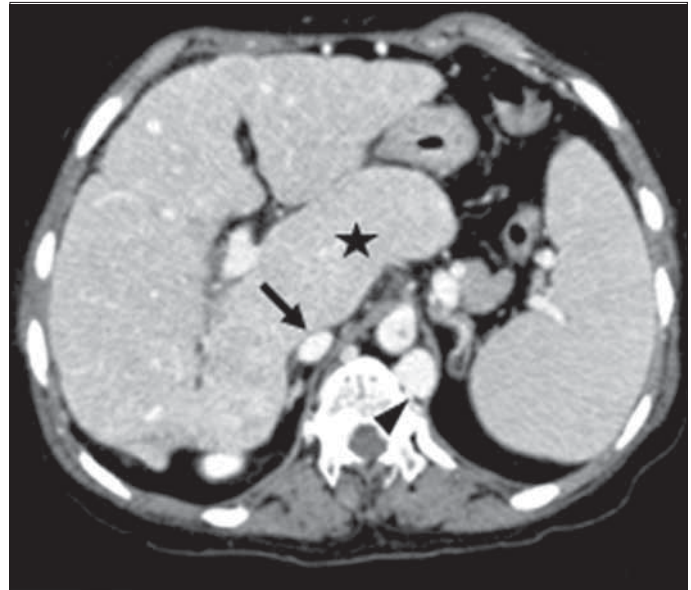


Figure 1. MDCT axial image shows the hypertrophic caudate lobe (star) and dilated azygos (arrow) and hemiazygos veins (arrowhead).

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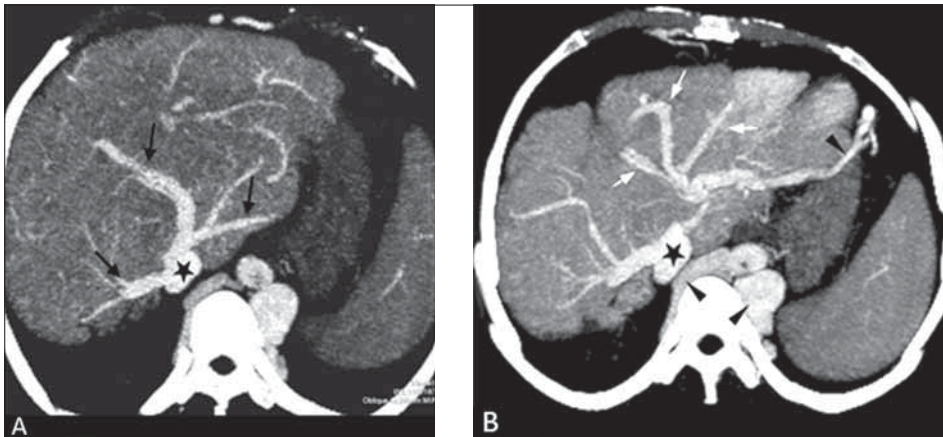


Figure 2. **A)** MDCT axial image shows three hepatic veins (right, middle, and left) (arrows) that provide drainage of the hypertrophic caudate lobe into the IVC (hepatic pseudo star) (star) **B)** MDCT axial image shows the configuration of the atrophic real hepatic veins (arrows), hepatic pseudo star (star), and collateral vessels (arrowheads).

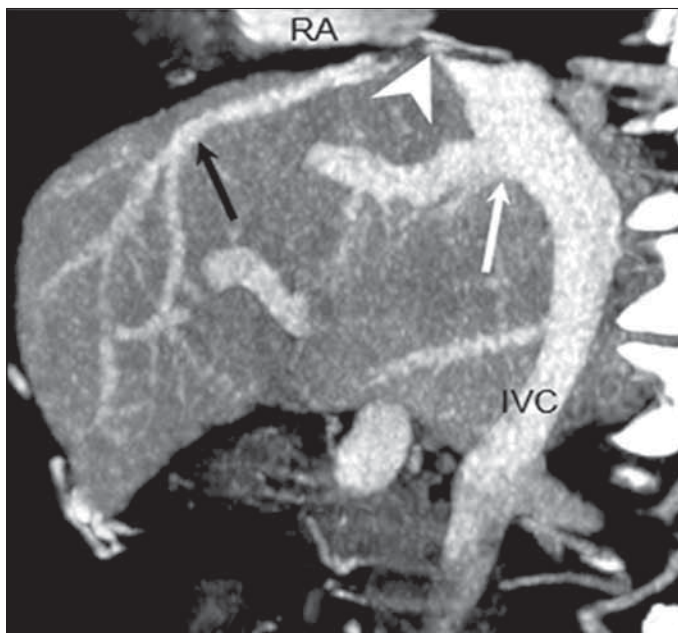


Figure 3. Coronal oblique MDCT image shows the membranous obstruction of the IVC (arrowhead), real hepatic veins (black arrow), and hepatic pseudo star (white arrow) (RA, right atrium; IVC, inferior vena cava).

of the scan. Twenty-five seconds after the injection of contrast medium, an arterial phase was taken in one breath-hold; 55 seconds post-injection, one portal venous phase was acquired; and 120 seconds post-injection, one systemic venous phase was acquired. Detailed 3D images of the celiac artery, superior mesenteric artery, portal vein, and IVC were obtained for thorough assessment using multiple plane reconstruction and maximum-intensity projection (MIP).

MDCT imaging revealed marked hypertrophy of the caudate lobe (Figure 1), and the other segments of the liver were markedly atrophic. The appearance of contrast filling and calibration of the IVC were both good. No thrombus-associated finding was detected in the hepatic veins. Multiple extrahepatic and perihepatic venous collaterals and the azygos and hemiazygos veins were markedly dilated. Numerous subdiaphragmatic collaterals and intraparenchymal small veins were observed adjacent to the eighth segment of the liver. The patent passing through the caudate lobe and opening into the IVC was well developed, and there were three veins in the hepatic vein configuration (right, middle, and left) (Figures 2A and 2B).

In a case in which liver transplantation was planned, the findings of intra- and extra-hepatic venous collaterals along with dilated azygos and hemiazygos veins were disturbing. Thus, Doppler ultrasonography was performed on the patient despite the fact that the vascular structures were unobstructed. It was observed that the large hepatic veins visualized with MDCT angiography were directed toward the IVC, but the flow of the IVC was reversed. The CT findings were thus re-evaluated to search for obstructions. The caliber of the IVC became smaller and thinner at the level of the diaphragm where the IVC opens into the right atrium. This was suggestive of the web that typically causes Budd-Chiari syndrome in Asia (Figure 3). Upon detailed evaluation, it was observed that the veins that opened into the IVC were hypertrophic caudate lobe veins mimicking a normal venous configuration.

The surgical team was informed of the possibility of a web at the point where the IVC opened into the right atrium and the consequent need for grafting in this area. The presence of a web was confirmed intraoperatively, and a bypass was performed using a cadaveric thoracic aorta graft. Currently, the patient is doing well under routine follow-up.

Discussion

Budd-Chiari syndrome was first described in 1845 by an English gastroenterologist, George Budd [4]. In 1899, an Austrian pathologist, Hans Chiari, gave the first pathological description of a liver with "obliterating endophlebitis of the hepatic veins" [5]. Most patients present in the age range of thirty to fifty years. The condition can be fulminant, acute, subacute, or chronic. Compared with patients in the chronic phase, hepatomegaly is detected more frequently in the acute phase. However, various degrees of liver atrophy are more frequent for patients in the chronic phase, during which extended congestion causes atrophy and necrosis in the hepatic parenchyma; this eventually results in cirrhosis [6].

In Budd-Chiari syndrome, the obstruction of venous structures results in the formation of multiple venous collaterals that may be extra- or intra-hepatic. In some cases, the veins of the caudate lobe and short hepatic veins may become dilated and mimic normal hepatic venous configurations.

Variations in the hepatic venous anatomy have been reported by Lafortune et al. [7]. They found three hepatic veins forming a "W" with its base on the IVC in 70% of their subjects, and they considered this to be the normal anatomy for the hepatic veins. The prevalence

of inferior right hepatic veins reported by studies employing various imaging modalities ranges between 6% and 67%. The inferior hepatic vein drains the inferior subsegments of the right hepatic liver (i.e., subsegments 5 and 6) and enters the IVC caudally to the level of the three hepatic veins (i.e., the suprahepatic hilum) [8].

There are five categories of inferior right hepatic veins depending on their number and location: Type 1, single inferior right hepatic vein; Type 2, two veins at the same level; Type 3, two veins at different levels; Type 4, more than two veins; and Type 5, truncal [9]. In our patient, the three veins were similar to the Type 4 category: anatomically, they provided the drainage of the caudate lobe and mimicked the real hepatic veins.

Because the caudate lobe drainage flows directly into the IVC through the small hepatic veins, the parenchyma in this field is not easily influenced by hepatic vein obstruction; as a result, compensatory hyperplasia may frequently be detected [10]. Evaluation of the veins that passed through and opened into the IVC in our patient showed that the veins opened into the IVC 2 cm lower than expected. Therefore, we labeled this appearance, which mimicked the configuration of the hepatic veins opening into the IVC (hepatic star), the 'hepatic pseudo star.' In additional evaluations, the probability of venous structures in the atrophic parenchyma of the eighth segment being real hepatic veins was considered. However, no thromboses were noted in these structures. At the level of the diaphragm, where the IVC opened into the right atrium, the caliber was based on a thin, small segment, suggesting the web formation that causes Budd-Chiari syndrome in Asia. The surgical team was warned of a possible obstruction in the IVC, and the presence of the web was confirmed during transplantation. A cadaveric thoracic aorta graft was used between the right atrium and the IVC.

Conflict interest statement: The authors declare that they have no conflict of interest to the publication of this article.

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