Right atrial myxoma associated with portal and splenic vein thrombosis in a patient with Budd-Chiari syndrome

Budd-Chiari sendromlu bir olguda portal ve splenik ven trombozuna yol açan sağ atriyum miksoması

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Summary – Budd-Chiari syndrome (BCS) is a rare disorder characterized by hepatic venous obstruction. A 41-year-old male patient presented with right upper quadrant pain, abdominal distension, and dyspnea. He had a history of BCS that was associated with polycythemia vera. Abdominal computed tomography showed hepatomegaly and a hypodense filling defect suggestive of thrombus formation in the hepatic, splenic, and portal veins, and suprahepatic part of the inferior vena cava. Transthoracic echocardiography performed to assess the extension of this pathological process and its relation with intracardiac structures showed a mass lesion in the right atrium, about 4x3 cm in diameter. The lesion manifested as an intracardiac thrombus extending from the inferior vena cava. The patient underwent surgical treatment to remove the atrial mass. At surgery, the lesion turned out to be an atrial tumor, which was diagnosed as atrial myxoma in histopathologic examination. The symptoms of the patient resolved after surgery.

Özet – Budd-Chiari sendromu (BCS) hepatik venlerde tıkanıklık ile kendini gösteren nadir bir bozukluktur. Kırk bir yaşında erkek hasta, sağ üst kadran ağrısı, abdominal gerginlik ve nefes darlığı yakınmalarıyla başvurdu. Hastanın polisitemia veraya bağlı BCS öyküsü vardı. Abdominal bilgisayarlı tomografide hepatomegali ile hepatik, splenik ve portal venlerde ve inferiyor vena kavanın suprahepatik kısmında trombüs oluşumunu düşündüren hipodens dolum kusuru görüldü. Bu patolojik oluşumun yayılımını ve intrakardiyak yapılarla ilişkisini değerlendirmek için yapılan transtorasik ekokardiyografide sağ atriyum içinde, yaklaşık 4x3 cm çapında kitle lezyonu saptandı. Sağ atriyumdaki bu lezyonun inferiyor vena kavadan yayılan bir trombüs olabileceği düşünüldü. Hasta, atriyumdaki kitlenin çıkartılması için cerrahi tedaviye alındı. Ameliyat sırasında kardiyak tümör görüntüsü veren lezyonun histopatolojik incelemede tanısı atriyum miksoması olarak kondu. Hastanın semptomları cerrahiden sonra kayboldu.

Budd-Chiari syndrome is a rare disorder resulting from obstruction of the major hepatic veins or the suprahepatic inferior vena cava at the suprahepatic level. This syndrome develops by either hepatic vein thrombosis or mechanical venous obstruction. The etiopathogenesis of BCS includes a heterogeneous group of diseases including myeloproliferative disorders, polycythemia vera, sickle cell disease, paroxysmal nocturnal hemoglobinuria, malignancy, and congenital web. Intracardiac tumors have been rarely reported as a predisposing cause for the development of BCS and surgical resection of these tumors are generally curative for this syndrome.[1,2]

In this report, we describe a patient with BCS, who developed right atrial myxoma and presented with exacerbation of clinical symptoms despite medical therapy and peritoneovenous shunting.

Abbreviations:

BCS	*Budd-Chiari syndrome*
CT	*Computed tomography*
IVC	*Inferior vena cava*
MR	*Magnetic resonance*

CASE REPORT

A 41-year-old male patient presented to our hospital with right upper quadrant pain, abdominal distension, and dyspnea. He had a history of BCS for one year that

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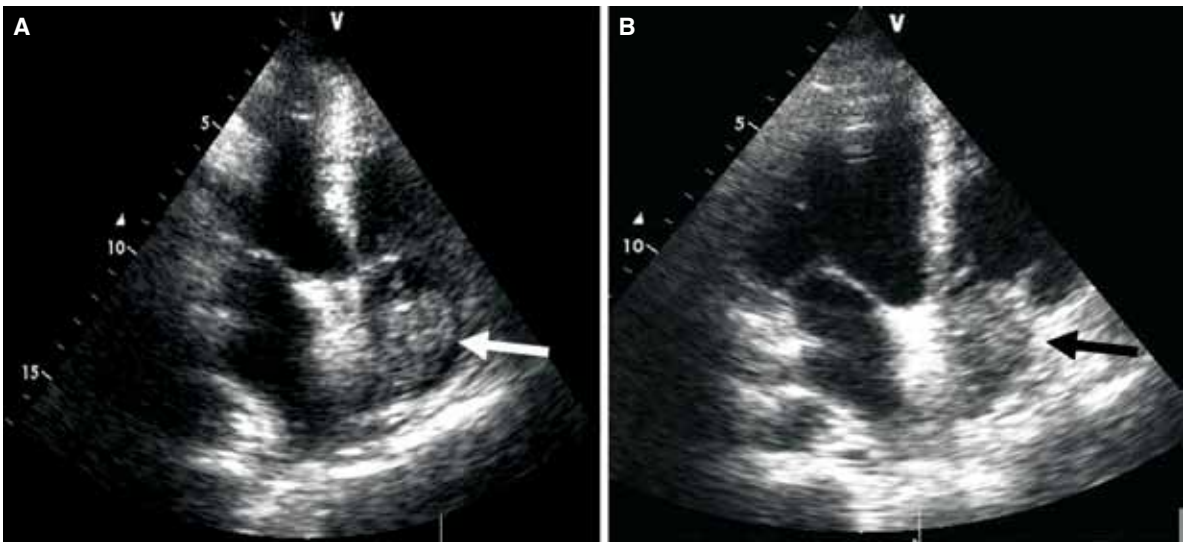

Figure 1. Preoperative transthoracic echocardiography demonstrates an atrial mass in **(A)** systole (white arrow) and **(B)** diastole (black arrow).

was associated with polycythemia vera. Four months prior to his admission, he underwent peritoneovenous shunting (Denver shunt) to decompress ascites into the systemic venous circulation and to relieve clinical symptoms. Before this operation, there was no cardiac pathology diagnosed in the preoperative work-up. The patient was taking hydroxyurea to keep the blood count within the normal range and warfarin sodium (2.5 mg per day) to prevent obstruction of the shunt catheter.

On physical examination, his pulse rate was 112 beats/minute and blood pressure was 124/84 mmHg. Respiratory and cardiac examinations were normal. The abdomen was distended with hypoactive bowel sounds. Palpation of the abdomen showed hepatomegaly of 6 cm below the right costal margin in the midclavicular line, suggesting the presence of ascites. The electrocardiogram showed sinus tachycardia without evidence for ischemia or conduction disorder. Chest roentgenogram showed the silhouette of the shunt between the right atrium and peritoneum. Cardiac index was less than 0.5. Biochemical findings were as follows: hemoglobin 14.2 mg/dl, platelet count 400,000/mm^3, white blood cell count 12,890/mm^3, aspartate aminotransferase 69 U/l, alkaline phosphatase 50 U/l, direct bilirubin 2.5 mg/dl, total bilirubin 5.7 mg/dl, albumin 3.5 mg/dl, and international normalized ratio 1.6.

During initial evaluation, the patient was consulted with general surgeons. They had the opinion that the shunt was not working properly and this could be the reason for clinical symptoms. Abdominal ultrasonography confirmed hepatomegaly and ascites in the abdomen. Doppler examination demonstrated the absence of hepatic vein flow. Abdominal contrast-enhanced computed tomography demonstrated hepatomegaly and revealed additional findings to clarify the pathogenesis of ascites. There was a hypodense filling defect suggesting thrombus formation in the hepatic, splenic and portal veins, and suprahepatic part of the IVC. Transthoracic echocardiography was performed to assess the extension of this pathologic

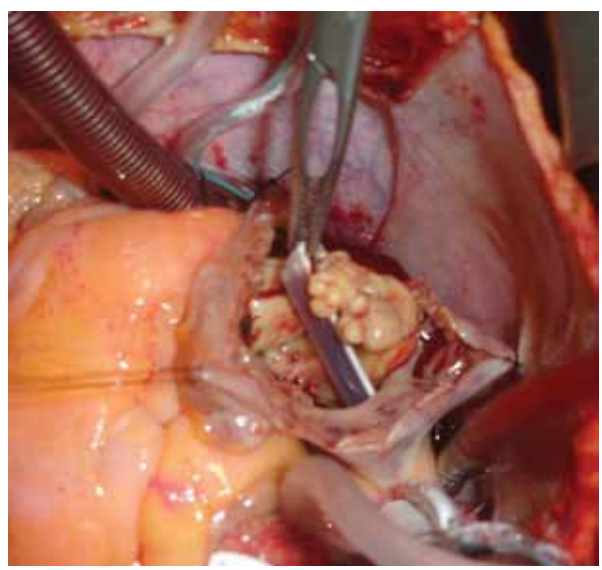

Figure 2. Operative view of the tumor in the right atrium after atriotomy incision. The tumor is relatively large and surrounds the tip of the peritoneovenous shunt catheter.

process in the vena cava and its relationship with intracardiac structures. A mass lesion was detected in the right atrium, 4 x 3 cm in diameter, that showed a close relationship with the tip of the atrial shunt (Fig. 1). The lesion was disturbing venous return to the right atrium through the IVC and was restricting diastolic filling of the right ventricle. There was no stenosis, nor insufficiency of the tricuspid valve. Open cardiac surgery was planned for resection of the atrial mass and exploration of the peritoneovenous shunt catheter.

Operation was performed under cardiopulmonary bypass with moderate hypothermia. Antegrade hypothermic blood cardioplegia was used to induce cardiac arrest. After a right atriotomy, an atrial mass was noted (Fig. 2). It was thought that the mass was a potential cause for dysfunction of the shunt due to obstruction of the orifice of the catheter in the atrium. In contrast to preoperative findings and suggestions for BCS, the morphologic appearance of the mass differed from that of thrombus formation. The mass and intra-atrial part of the shunt catheter were uneventfully resected. The tumor was gelatinous, soft, and rubbery, it originated from the lower part of the interatrial septum and measured 4 x 3 x 2 cm. There was no thrombosis within the catheter. Pathological examination yielded the diagnosis of myxoma (Fig. 3). The patient was discharged 10 days after surgery with a favorable outcome. The symptoms of the patient resolved after surgery and the shunt was working well in the follow-up visits.

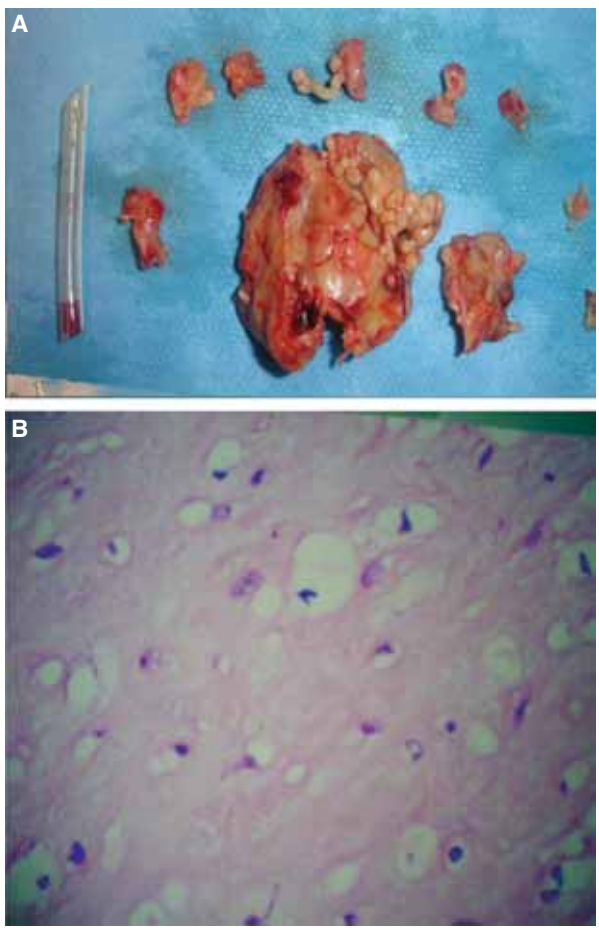

Figure 3. **(A)** Macroscopic view of the tumor measuring 4x3x2 cm and **(B)** its histopathologic appearance diagnosed as atrial myxoma (H-E x 400).

DISCUSSION

Budd-Chiari syndrome is a rare clinical disorder characterized by a hepatic venous outflow obstruction at any level between the small hepatic veins and the junction of the IVC and right atrium. This condition results in liver congestion, fibrosis, and eventually cirrhosis with portal hypertension. The most common risk factors are hypercoagulable states associated with polycythemia vera, myeloproliferative disorders, paroxysmal nocturnal hemoglobinuria, antiphospholipid syndrome, and defects in the coagulation cascade such as protein C and S deficiency or factor V Leiden mutations. Most cases of BCS are caused by either hepatic vein thrombosis or mechanical outflow obstruction. The pathological process can be primary, when obstruction of the hepatic venous outflow tract results from an endoluminal venous lesion (thrombosis or congenital webs) or secondary, when the cause of obstruction is an extrinsic compression or nearby invasion by an abscess, cyst, benign or malignant tumor.[3,4] Tumors may cause hepatic venous obstruction by direct invasion or external occlusion. In particular, cardiac tumors are rarely associated with the development of BCS.[1,2]

Clinical presentation of BCS depends upon the extension and rapidity of venous occlusion as well as the development of collateral circulation that decompresses the hepatic veins. The syndrome may present as acute, subacute, or chronic symptoms. Abdominal pain, hepatomegaly, ascites are present in almost all BCS patients. Acute symptoms frequently include hepatomegaly, right upper quadrant pain, nausea, vomiting, jaundice, and ascites.[4] Chronic BCS is characterized by a history of vague complaints for about six months. Patients may present with complications of cirrhosis and portal hypertension. Biochemical tests generally show findings of chronic liver disease. In

our patient, the chronic form was evident before the onset of acute symptoms. The patient was stable under medical treatment and especially after peritoneovenous shunting. However, enlargement of the right atrial myxoma, with localization close to the junction of the IVC and right atrium, caused exacerbation of clinical symptoms of chronic BCS.

The diagnosis of BCS is often made by noninvasive imaging modalities. Ultrasonographic evaluation of the liver and its vasculature is useful for the diagnosis. Duplex scanning may reveal the localization of the obstruction; nonvisualization of the hepatic veins, areas of stenosis, collateral veins, and proximal dilatation may suggest BCS. Diagnostic accuracy increases if an altered or absent hepatic venous flow is demonstrated.[5] Computed tomography is also useful in evaluating the patency of hepatic veins and the liver. Hypodense filling defects in the lumen of the IVC and hepatic veins on postcontrast CT scans are considered highly specific for the diagnosis of thrombosis. Other diagnostic findings include nonvisualization of the hepatic veins or IVC and attenuation changes in the hepatic parenchyma.[5]

Atrial myxomas account for 35% to 50% of primary cardiac tumors and are usually located in the left atrium, attached to the interatrial septum. They are benign, slow-growing neoplasms that arise from the interatrial septum and extend into the left or right atrium. Only 20% of myxomas originate from the right atrium.[6] Right atrial myxoma tends to be more solid with a wider attachment to the atrial wall or septum. Very rarely, atrial myxoma may originate from the IVC or from the junction of the IVC and right atrium.[7,8] In these cases, the risk for development of BCS may be higher. A thrombus in this localization can also mimic a myxoma. Based on our literature review, there are few cases of BCS and portal vein thrombosis associated with a right atrial myxoma.[1,2] Those cases presented with BCS after occurrence of myxoma in the right atrium.

In our case, the patient was taking oral warfarin sodium in the preoperative period and had an INR of 1.6 on admission. Blood chemistry showed normal liver functions. Intraoperatively, there was no thrombosis within the shunt catheter. Despite warfarin treatment, systemic thrombosis tendency in myxoma would also be associated with occlusion of the shunt and might lead to similar clinical findings. However, it was clear that the great size of the myxoma caused occlusion of the tip of the shunt catheter and the junction of the IVC and right atrium. This tumor was the cause of aggravation of BCS symptoms and underlying pathological process in the suprahepatic veins. Although myxomas may rarely cause systemic thrombosis due to polycythemia, we believed that thrombosis in the splenic and portal veins were associated with the previous diagnosis of polycythemia vera. The uneventful resection of the right atrial myxoma resulted in improvement in clinical symptoms and provided a better quality of life after surgery.

The differential diagnosis of a mass in the right atrium includes myxoma, thrombus, metastasis, and a primary malignant cardiac tumor.[9] Atrial myxoma is the most common primary tumor of the heart. Right atrial myxoma may present as fever, weight loss, right heart failure, and pulmonary embolism. Although atrial myxoma is a slowly progressive disease, our case emphasizes that its diagnosis should be included in the differential diagnosis of tumors that show a progressive growth in cardiac chambers. Echocardiography is a very useful diagnostic imaging modality that may show valvular stenosis or prolapse of the tumor through the atrioventricular valve. Magnetic resonance and CT imaging are also useful, both allowing evaluation of the mediastinum and intrathoracic extension of disease processes. Myxomas usually have a gelatinous nature and therefore may manifest as a low-attenuation mass on CT. In some cases, calcification may be present.[10] Cardiac thrombi are the most frequent cardiac masses, and mainly result from impaired wall motion abnormalities or arrhythmias. Thrombus in the right atrium is very rare and risk factors include central venous lines, enlarged cardiac chambers, and arrhythmias. Coexistence of thrombus in the right atrium and IVC is rare and usually occurs in malignancy such as renal cell carcinoma. Magnetic resonance imaging is particularly sensitive for detecting thrombi, which appear as dark structures surrounded by contrast-enhanced blood. On contrast MR imaging, atrial myxoma is usually a heterogeneous structure with necrotic areas within the tumor. In our case, we did not perform MR imaging.

In conclusion, although a right atrial mass developing in the presence of BCS may suggest atrial thrombus formation, right atrial tumors such as myxoma should also be considered in the differential diagnosis among the causes of BCS even in the presence of a primary pathology. Echocardiography may be useful in the diagnostic work-up of patients with BCS.

Conflict-of-interest issues regarding the authorship or article: None declared

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Key words: Budd-Chiari syndrome/etiology; echocardiography; heart atria/pathology; myxoma/complications/surgery; venous thrombosis/etiology.

Anahtar sözcükler: Budd-Chiari sendromu/etyoloji; ekokardiyografi; kalp atriyumu/patoloji; miksoma/komplikasyon/cerrahi; ven trombozu.