TJTES
TURKISH JOURNAL OF TRAUMA
& EMERGENCY SURGERY

Ulusal Travma Dergisi (2002) 8: 253 - 255

A PATIENT WITH LIVER TRAUMA AND INCOMPLETE BEHCET'S DISEASE

İNKOMPLET BEHÇET HASTALIĞI OLAN VE KARACİĞER TRAVMASI İLE GELEN BİR HASTA

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ABSTRACT

Behçet's Disease (BD) is a complex multisystemic disease, which is characterized by recurrent oral and genital aphteous ulcers and iritis in which vasculitis can also be one of the possible clinical manifestations. A thirty-seven year-old female patient with incomplete BD was admitted to emergency service, with intra-abdominal hemorrhage more severe than that would be expected, with the degree of related trauma. We decided to manage the patient conservatively, observing vital signs, haemogram, computed tomography (CT) and angiography instead. Subcapsular hemotomas were detected in the right and left lobes of the liver. No data was encountered in the literature and textbooks referring to liver trauma with BD, except a case of fatal hemobilia. Here in we present a liver trauma case with hemorrhage and hepatic fragility due to vasculitis in Behçet's disease and review the literature.

Key words: *liver trauma, incomplete Behçet's disease*

ÖZET

Rekürran oral ve genital aftöz ülserler ve iritis ile karakterize olan Behçet Hastalığı, karmaşık multisistemik bir hastalıktır. Vaskülit hastalığın diğer bir görünümüdür. Otuz-yedi yaşında bayan inkomplet Behçet hastası, travma derecesinden daha şiddetli klinik bulgularla acil servise kabul edildi. Karaciğerin sağ ve sol lobunda subkapsüler hematom saptandı. Hasta vital bulgular, hemogram, CT, ve anjiografi ile operasyon yapılmaksızın takip edildi. Bir fatal hemobili vakası dışında, Behçet hastalığında karaciğer travmasına dair bir bilgiye literatürde yada klasik kitaplarda rastlanamadı. Biz bu yazıda vaskülite bağlı hepatik frajilite ve kanama ile seyreden ve konservatif olarak tedavi ettiğimiz bir Behçet hastalığı olgusunu ve bu konudaki literatürü sunuyoruz.

Anahtar Kelimeler: *Karaciğer travması, Inkomplet Behçet Hastalığı*

INTRODUCTION

Behçet's Disease (BD) is a complex multisystemic disease, which was first described in 1937 by the Turkish dermatologist Hulusi Behçet. Behçet referred to a symptom complex consisting of recurrent aphteous ulcers, genital apthae, and iritis that could lead to blindness. If this triad is present, the patient may be diagnosed as complete BD. Pathergy is an additional clinical phenomenon which means the induction of a cutaneous pustular neutrophilic vascular reaction after intradermal trauma. Arthritis, thrombophlebitis involved vasculitis, erytema nodosum-like cutaneous lesions are other manifestations of BD. Neurologic signs and symptoms ranging from benign intracranial hypertension to a condition resembling multiple sclerosis can also be detected(1).

In this article, a patient who had intra-abdominal hemorhage, and wide subcapsular hematoma with incomplete BD, was discussed under the view of literature.

CASE REPORT

A thirty-seven year old female patient being followed with Behçet's Disease (BD) for six years, who felt down from a meter height, was admitted to the emergency service. She had oral and genital recurrent aphteous ulcers (incomplete BD). She had abdominal pain and acute abdominal signs were positive following the trauma. Hematocrit was 24% and diagnostic peritoneal lavage was positive.

Subcapsular hematoma and contusion of the right lobe and medial segment of the left lobe of liver, were detected on the computed tomography (CT) (fig I). The patient was monitored with blood pressure and pulse rate determinations every hour; hemoglobin, hematocrit levels and body

Fig 1: A CT view of the patient on the first day of the trauma. Subcapsular hematoma and contusion were detected in the right lobe and the medial segment of the left lobe of liver.

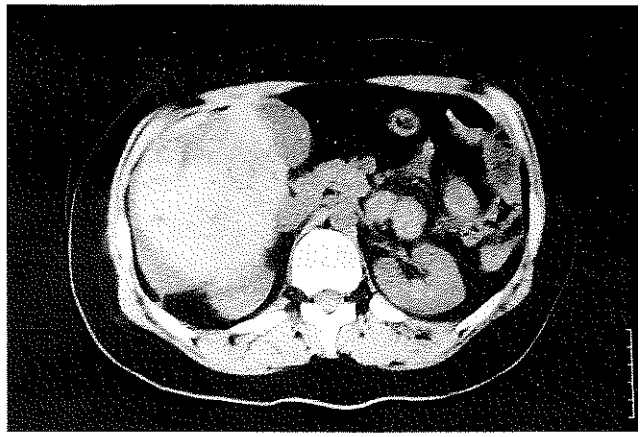


Fig 2: The control CT view of patient on 20th day of trauma. No change was detected in the size of hematoma.

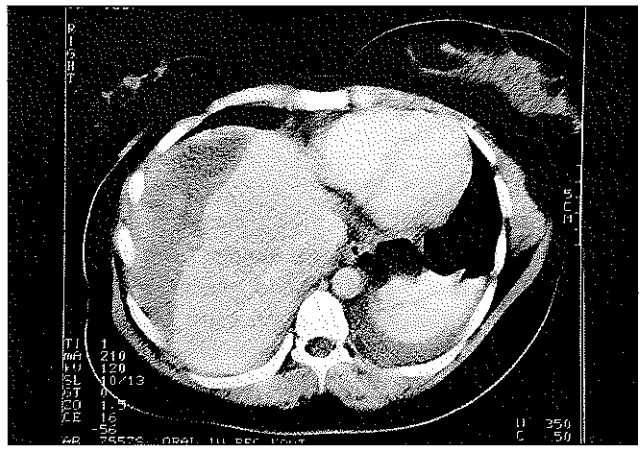
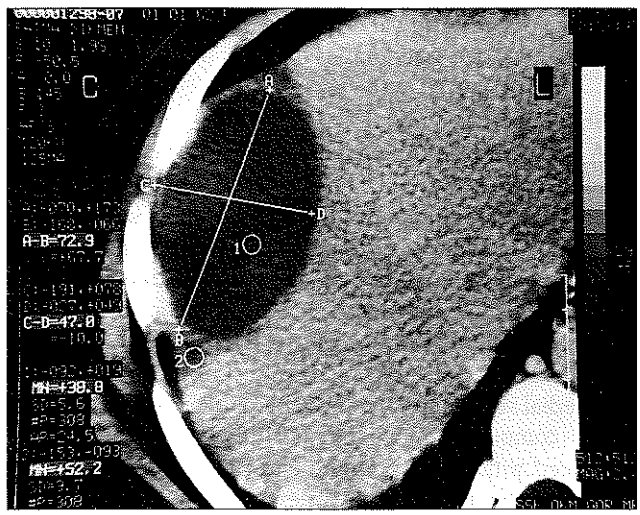


Fig 3: The control CT view of patient, 3 months after trauma. An organized hematoma in the right lobe of the liver could be seen.



temperature every four hours for 15 hours. She was hemodynamically stable despite 6% decrease in the hematocrit level. Two units of whole blood were transfused.

One day later, she was transferred to the hepatopancreatobiliary surgery unit of another hospital where she was followed conservatively. No expansion of the liver hematoma was observed.

Three days later, hepatic arteriography and portography was performed, branches of hepatic artery were found to be normal and no extravasation was detected.

There was no change in the size of the hematoma on the control CT, 20 days after the trauma (fig 2). Three months later, control CT revealed that the hematoma in the right lobe of the liver had been organized (fig 3).

DISCUSSION

Connective tissue disorders are a protean group of acquired diseases, which have common widespread immunologic and inflammatory alterations of connective tissue. Gastrointestinal and hepatic involvement in connective tissue disorders are not the most important features, although they appear almost regularly(2).

Although, there is no data about liver fragility in patients with BD in textbooks or in the literature, we preferred to follow the patient conservatively without surgical intervention.

Orloff et al. reported that when BD is complicated with Budd-Chiari syndrome (BCS), mortality is increased. In their clinical series, one of the five BD with BCS died, two years after porto-caval shunt surgery because of diffuse vasculitis. Four of the five BD with BCS had no hepatic dysfunction. Serial liver biopsies showed normal architecture in 60% of the patients with BD. However they reported that liver biopsy could be irrational(3).

Four types of vascular lesion are recognized in BD: arterial occlusions, aneurysms, venous occlusions and variceal development. The incidence ranged from 7% to 29% in literature(4). Aneurysms are the main reason for bleeding in other organ systems with BD, but there is no data about bleeding due to hepatic artery aneurysm. Kuzu et al. reported in their 1200 patients' clinical series followed for 8 years, 14% venous and 1.6% arterial manifestations and 0.4% inferior vena cava syndrome, only one patient had hepatic vein thrombosis in that series(4). Bayraktar et al. reported in their clinical series of 66 patients followed for 25 years of period that six of these patients had cavernous transformation of portal vein. Five of these six patients had an additional large vein involvement(5).

Patients with BD can present with recurrent intracerebral, intraretinal, and gastrointestinal hemorrhage, hematuria and fatal hemoptysis. Excessive bleeding from genital ulcers secondary to varicose veins, that may even be postcoital is also possible(6). The prevalence of hemoptysis due to pulmonary vasculitis in patients with BD has been reported to be 5 to 10%. However the prognosis is poor and often fatal, because of ruptured aneurysm combined with

thrombotic angiitis(7). Al Daloon et al. reported hematuria in one case of nine patients with BD having renal involvement. They biopsied four of those nine renal involved patients. Mesengial proliferative changes were observed in three patients, an evidence of immune complex deposition by immunoflorescence. Amyloidosis was present in one patient who had vascular involvement and presented with nephrotic range proteinuria(8). When BD involves digestive tract, the lesions are mainly observed around the ceacum and frequently lead to hemorrhage which requires surgery(9).

Although, any vessels of any organ systems may be affected in BD, there is no data about bleeding caused by thrombosed hepatic, portal, inferior vena caval veins in BD in the literature, except the case of fatal hemobilia with BD(10).

Diagnosis and management of hepatic bleeding with BD by angiography and portograhy is an alternative, but there is no specific angiographic findings for BD(11). In the angiography and portography: we detected only hematoma in the liver. Branches of hepatic artery were normal, branches of portal vein could not be seen in the area of the hematoma (because of external pressure of hematoma), and there was no extravasation both arteriographically and portographically.

Bozkurt et al. presented a case of a 27 year-old man with BD, who had a surgical intervention during the active period of his disease. They observed that severe superficial perivascular dermatitis developed at the incision site, and his Behçet's disease progressed poorly(12). There is an augmented inflammatory response to trauma, particularly of the skin, in-patients with BD but wound healing is not altered(13).

Our case was an incomplete BD and her disease was not aggravated after trauma. We preferred nonoperative approach, afterwards we followed her with CT and we detected an organized liver hematoma three months after trauma (Fig. 3).

In conclusion, although there is no data in the literature about hepatic trauma in BD except a case of fatal hemobilia, we think that due to increased fragility and vasculitis there is a tendency for bleeding in BD patients with even minor liver trauma. Consequently, we preferred to follow our patient with BD, nonoperatively. Despite there is a tendency for extensive bleeding in patients with BD, conservative management could be tried unless the hemodynamic parameters deteriorate.

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