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Case report
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Cholecystitis caused by hemocholecyst: An unusual complication of Hemophilia A

Oguzhan Karatepe[1*], Mustafa Tukenmez[2], Gökhan Adas[1], Gamze Çitlak[2], Kemal Hunerli[2], Muharrem Battal[1], Osman B. Gülçiçek[1], Servet Karahan[1], Deniz Özcan[3]

[1] *Department of Surgery, Okmeydani Training and Research Hospital, 34380, Istanbul, Turkey*

[2] *Department of Surgery, Istanbul University,Istanbul Medical Faculty, 34390, Istanbul, Turkey*

[3] *Departments of Pathology, Okmeydani Training and Research Hospital,34380, Istanbul, Turkey*

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Abstract: Hemocholecyst is a rare disorder that has been reported in patients with gallstones. Previous reports describe cholecystitis resulting from hemocholecyst after iatrogenic trauma. We report the first case of acute cholecystitis secondary to hemocholecyst in a patient with Hemophilia A.

Keywords: Hemocholecyst, Hemophilia

1 Introduction

A hemocholecyst (HC) is a clot-filled gallbladder caused by bleeding within its lumen most commonly seen after trauma [1, 2]. Distinguishing a HC from hemobilia may be difficult [1–4]. Hemophilia is a genetic bleeding disorder characterized by prolonged or spontaneous bleeding usually manifested as intra-particular and oral hemorrhages as well as bleeding into muscle, retroperitoneum and bowel wall [5].

We report the first patient with acute cholecystitis resulting from hemocholecyst secondary to Hemophilia A.

* E-mail: drkapatepe@yahoo.com

2 Case Report

A 16-year-old-male with history of Hemophilia A was seen in our emergency department with a three day history of right upper quadrant pain, nausea and vomiting. The patient's vital signs were as follows: temperature, 37.8°C; pulse, 112bpm; blood pressure, 110/70. On physical examination there was rebound tenderness in the right upper quadrant of the abdomen and a palpable gallbladder. The peripheral blood count revealed a hemoglobin level of 8.8 g/dL, hematocrit of 25%, platelet count of 110000/mm^3, and white blood count of 14000/mm^3. The liver function tests were within normal range. The chest X-ray was unremarkable. An abdominal ultrasound (US) revealed an enlarged gallbladder with wall edema containing a hematoma. The spiral tomography showed the gallbladder to have a heterogenous appearance, which was suggestive of intraluminal bleeding (Fig. 1). He was given two doses of Factor VII (Novoseven) of 90 μg/kg (3,6 mg) at 4 hour intervals before he was transferred to the operating room for surgery. During exploratory surgery a hematoma and disperse foci of gangrene were found in the gallbladder (Fig. 2). A Cholecystectomy was performed. Postoperatively the patient received 60 μ/kg (2,4 mg) of Factor VII every 3 hours. During the following days he had no complications. Histopathology revealed erosion of the gallbladder mucosa as well as hemorrhagic foci (Fig. 3).

Fig. 1 Computed tomography revealed isodense structure measuring 25 mm inside the gallbladder lumen, consistent with a hematoma. Also, increased wall thickness and hydrops was noted accompanied by an increase in intraluminal density.

The patient was sent home on postoperative day 5.

3 Discussion

The usual presentation of hemophilia is bleeding in childhood. In 30% of cases the first presentation is excessive bleeding after a circumcision and intracranial hemorrhage in $1-2$% of full term neonate. Intramuscular or intra-articular hemorrhage is also a common finding [5]. Spontaneous gastrointestinal bleeding is rare. Bleeding elsewhere is often precipitated by pre-existing pathology or trauma [5].

Fig. 2 Hematoma inside the gallbladder.

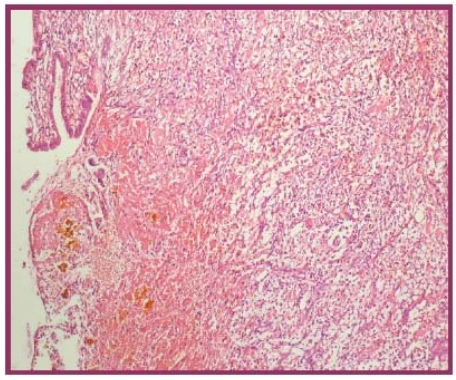

Fig. 3 Histopathology revealed erosion of the gallbladder mucosa with hemorrhagic foci.

The term "hemocholecyst" (HC) was first used in 1961 by Fitzpatrick [6] to refer to a clot-filled gallbladder in which the gallbladder itself is the source of the bleeding. Recent studies have documented that blood accumulation in the gallbladder is seen with iatrogenic trauma, anticoagulant therapy, lupus, liver biopsy, or ruptured aneurysms of either the hepatic or cystic artery [1–4] .

The pathogenesis of hemocholecyst is poorly understood. It has been suggested that both the rate of bleeding and bile flow affect the tendency to form a clot. Despite the fibrinolytic nature of bile, a greater tendency to form a "pure" clot is seen with minor bleeding. These pure clots are more likely to lead to cystic duct obstruction and cholecystitis [2–4]. Direct causes of hemocholecyst are more limited and include malignancy, cholelithiasis, vascular disorders, and the presence of heterotopic gastrointestinal mucosa within the gallbladder. Indirect causes of hemocholecyst appear to come from the liver secondary to trauma. HC can cause hemobilia, and conversely, hemobilia may result in HC due to the reservoir function of the gallbladder [1–4]. We therefore believe that the term "HC" is appropriate in this case, since the blood clots were confined to the gallbladder even though they did not appear to have originated there. We do not believe that the HC was a result of hemobilia because there was no history of trauma. However, it should be noted that it is not always possible to differentiate primary HC from HC secondary to

trauma even in patients with history of trauma. There is a case report in the literature of a HC secondary to hemobilia following a liver biopsy.

Primary hemocholecyst should be distinguished from hemobilia resulting from mucosal erosion in pre-existing cholecystitis. Hemobilia involving the gallbladder can present with signs and symptoms of upper gastrointestinal bleeding, lead to gallbladder rupture and hemoperitoneum, or remain confined to the gallbladder [2–4]. The clinical presentation can mimic that of acute cholecystitis. Obstruction of the cystic duct by a clot can lead to acute abdominal pain, hematemesis, jaundice, melena, hemoperitoneum, and acute cholecystitis progressing to gangrene [2, 3].

The diagnosis of HC may be difficult despite the available imaging methods. CT scan can show hemorrhage while ultrasound reveals the presence of echogenic material within the gallbladder, as occurred with our patient. These findings suggest a differential diagnosis of gallstones, gallbladder carcinoma, polyps, sludge, or blood clot [1, 3]. However, the sonographic demonstration of blood clots is quite variable and can be nonspecific. Therefore, when a clinical suspicion for cholecystitis exists, the ultrasound findings of intraluminal membranes, focal gallbladder wall irregularities or non-shadowing, non-layering intraluminal echoes should suggest hemorrhagic and/or gangrenous cholecystitis [2, 4].

We report a case of hemophilia complicated with HC which resulted in acute cholecystitis. Although HC is rarely seen in patients with hematological diseases, a high index of suspicion is essential for prompt diagnosis and early intervention.

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