# Splenic hydatic cyst as a cause of sinistral portal hypertension and isolated gastric variceal bleeding

## Sol portal hipertansiyon ve izole gastrik varis kanamasının bir nedeni: Dalak kist hidatiği

Bülent KANTARÇEKEN[1], Ali ÇETİNKAYA[1], Ertan BÜLBÜLOĞLU[2], Gülen DEMİRPOLAT[3]

*Departments of [1]Gastroenterology, [2]General Surgery, and [3]Radiology, Kahramanmaraş Sütçü İmam University, School of Medicine, Kahramanmaraş, Turkey*

*A 60-year-old male, living in a rural area, presented with recurrent upper gastrointestinal bleeding. Isolated fundal varices were seen on endoscopy. A lesion similar to cyst hydatid was seen in the spleen on abdominal ultrasonography and computerized tomography scanning. Also, sinistral (left-sided) portal hypertension and collaterals were seen due to the compression of the splenic vein by the cyst. Indirect hemagglutination antibody test for Echinococcus granulosus was positive. By screening, no other cyst was found in any other site of the body. The patient underwent open abdominal surgery, and the anterior wall of the cyst was resected partially; within it were multiple daughter cysts and hydatid fluid. After decontamination of the daughter cysts and hydatid fluid, germinative membrane omentoplasty was performed with a part of the omentum. However, the patient suffered from recurrent gastrointestinal bleeding postoperatively and he was re-evaluated. Later, splenectomy was performed in order to relieve left-sided portal hypertension. The patient did not experience further bleeding and gastric varices disappeared following splenectomy.*

**Key words**: Spleen, cyst hydatid, sinistral portal hypertension, bleeding

*Kırsal alanda yaşayan 60 yaşında erkek hasta, tekrarlayan üst gastrointestinal kanama ile başvurdu. Endoskopide izole fundal varisler izlendi. Abdomimal ultrasonografi ve bilgisayarlı tomografide, dalakta kist hidatikle uyumlu lezyon izlendi. Aynı zamanda kist'in splenik vene bası yapması ile sol portal hipertansiyon ve kollateraller izlendi. Kist hidatik için indirekt hemaglütinasyon antikor testi pozitif bulundu. Vücutta kist hidatik için başka bir odak bulunamadı. Hasta tedavi için cerrahiye verildi. Ön duvarı rezeke edildiğinde kız veziküller ve hidatik sıvı izlendi. Germinatif membrane ve hidatik sıvı temizlendikten sonra kist içine omentoplasti uygulandı. Fakat operasyondan sonra hastada tekrarlayan kanamalar devam ettiği görüldü. Bunun üzerine hasta tekrar değerlendirildi ve splenektomi uygulandı. Splenektomiden sonra kanamaların tekrar etmediği ve varislerin kaybolduğu görüldü.*

**Anahtar kelimeler**: Dalak, kist hidatik, sinistral portal hipertansiyon, kanama

## INTRODUCTION

Hydatid cyst, which is caused by *Echinococcus granulosus*, remains a serious health problem in endemic areas, such as in Turkey. Although benign in nature, hydatid disease causes a reasonable rate of morbidity, loss of manpower and resource waste due to its complications. Both primary splenic hydatidosis and invasion of the spleen by hydatid cysts are quite rare (1, 2). Moreover, left-sided (sinistral) portal hypertension and complications due to compression of the splenic vein by splenic hydatidosis are even rarer (3, 4). Sinistral portal hypertension is a rare but important cause of potentially life-threatening upper gastrointestinal hemorrhage. There are many causes of sinistral portal hypertension. The primary pathology usually arises in the pancreas and results in compression of the pancreatic vein. This compression causes back-pressure in the left portal venous system and subsequent gastric varices. Management is usually surgical to treat the underlying pathology and splenectomy to decompress the left portal venous system. Only one case was reported in the literature who presented with upper gastrointestinal bleeding from gastric varices due to compression of the splenic vein by a splenic hydatid cyst (4). We aimed to present our patient with this quite rare and interesting clinical picture of hydatid cyst as the second case.

**Address for correspondence:** Bülent KANTARÇEKEN
Department of Gastroenterology
Kahramanmaras Sütçü İmam University, Faculty of Medicine
46050 Kahramanmaraş, Turkey
Phone: + 90 344 2212337 / 362 • Fax: + 90 3442212371
E-mail: bkantarceken@ksu.edu.tr

**Manuscript received:** 10.02.2009 **Accepted:** 05.11.2009

doi: 10.4318/tjg.2010.0109

## CASE REPORT

A 60-year-old male, living in a rural area, presented with hematemesis and melena. The first signs of upper gastrointestinal bleeding were realized nearly six months before. He went to the doctor with these complaints and was given some medications such as proton pump inhibitors and antacids without endoscopic examination. The patient was admitted with hematemesis and melena to our clinic during his second upper gastrointestinal bleeding. In his physical examination, the mucosa was pale; blood pressure was 110/70 mmHg and pulse was 84/minute, and melena was present in rectal examination. Intravenous fluid replacement was started. Laboratory tests on admission were as follows: hemoglobin: 6.7 g/dl, hematocrit (Htc): 20.9%, thrombocyte: 122000/mm$^3$, leukocyte: 4200/mm$^3$, prothrombin time: 15 sec, and international normalized ratio (INR): 1.17. Bleeding from isolated fundal varices was seen in upper endoscopy (Figure 1). No other bleeding source was observed. Two units of erythrocyte suspension were given to the patient since he had orthostatic hypotension. After those treatments, his clinical condition improved. Abdominal ultrasonography revealed a cystic lesion nearly 17 cm in diameter, with well-bordered wall, at the hilus of the spleen. The liver was mildly enlarged with its normal texture. The spleen was moderately enlarged (long axis was nearly 15 cm). The portal vein was in normal caliber. Abdominal computerized tomography (CT) revealed a well-bordered cystic mass lesion (11x15x17 cm in diameter) consistent with hydatic cyst, with low-attenuated, round-shaped fluid collections (daughter vesicles), in the anteromedial side of the spleen. Moreover, CT revealed a mildly tortuous collateral vascular structure between the splenic vein and esophagogastric junction (Figure 2). Liver enzymes, albumin and bilirubin levels were normal, and hepatitis serology was negative. Iron level, iron saturation and ferritin levels were low. Peripheral blood smear revealed hypochromic microcytic anemia. Indirect hemagglutination antibody (IHA) test for hydatic cyst was positive in a titer of 1/256. The patient was operated and the cyst anterior wall, which was filled with multiple daughter vesicles and cystic fluid, was resected. After decontamination of daughter cysts, hydatid fluid and germinative membrane, a part of the omentum was replaced into the cyst remnant (omentoplasty). Nevertheless, the patient suffered from recurrent gastrointestinal bleeding after the operation and he was re-evaluated. Later, splenectomy was performed in order to decompress the splenic vein. After splenectomy, the patient had no further bleeding and gastric varices disappeared (Figure 3). At the follow-up one year after his second operation, the patient was well and his hemoglobin level was stable.

## DISCUSSION

*Echinococcus granulosus* can affect any organ in the body, and a high suspicion of this disease is justified in endemic regions. The clinical presentation of *E. granulosus* infection depends upon the site of the cysts and their size. Small and/or calcified cysts may remain asymptomatic indefinitely. However, symptoms due to mass effect within or-

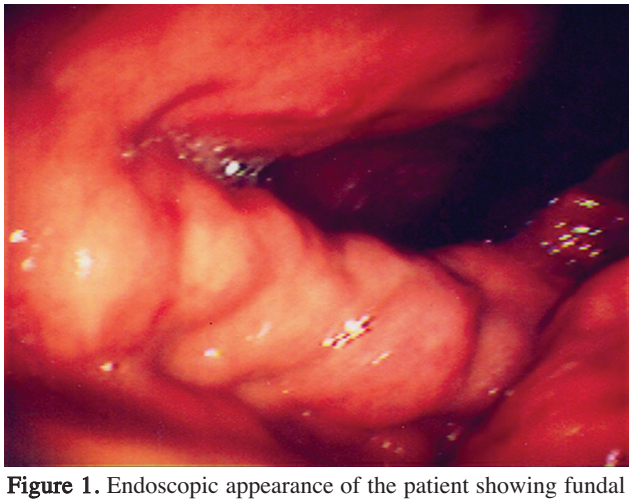

**Figure 1.** Endoscopic appearance of the patient showing fundal varices.

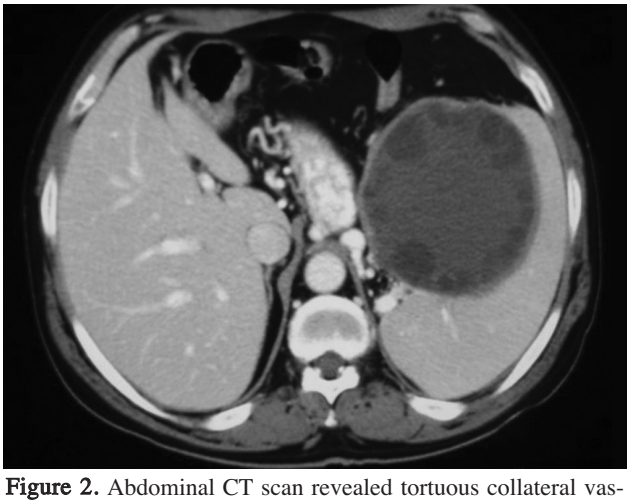

**Figure 2.** Abdominal CT scan revealed tortuous collateral vascular structure in gastric fundus region caused by hydatid cyst located in the splenic hilus.

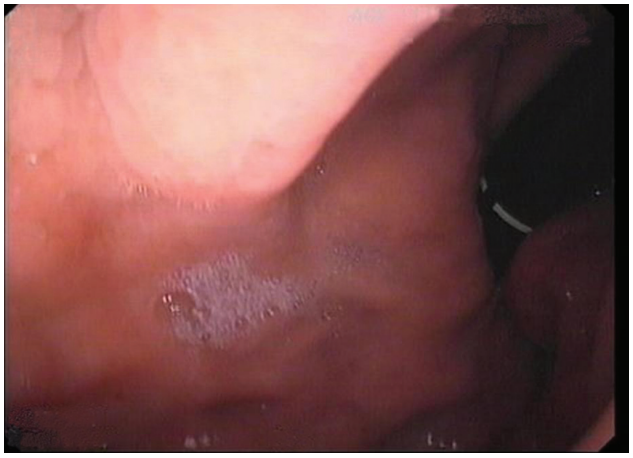

**Figure 3.** Follow-up gastroscopic appearance of the patient with no apparent fundal varices.

gans, obstruction of blood or lymphatic flow, or complications such as rupture or secondary bacterial infections may be encountered. Most patients (as many as 80%) have single-organ involvement and harbor a solitary cyst. The liver is affected in approximately 75% of patients. The other sites in which hydatic cyst locate are: lung (51%), peritoneum, kidney and brain (9%), mediastinum (7% ), heart (4%), bone, spinal cord, spleen (2%), pleura, adrenal gland (1%), and infrequently the bladder, thyroid and other organs. The reported prevalence of splenic involvement varies from 0.9% to 8% (5). Primary splenic hydatidosis is quite rare. In our case, there was no other hydatic cyst involvement site in the body. Sinistral, or left-sided, portal hypertension, is a rare but important cause of potentially life-threatening upper gastrointestinal hemorrhage. There are many causes of sinistral portal hypertension. The primary pathology usually arises in the pancreas and results in compression of the pancreatic vein. This compression causes back-pressure in the left portal venous system and subsequent gastric varices. The left portal system includes the short gastric veins, veins in the upper half of the stomach, coronary vein, and gastroepiploic veins (6). Gastrointestinal bleeding associated with isolated splenic vein obstruction is due to varices that usually develop in the short gastric and left gastroepiploic veins. Management is usually surgical to treat the underlying pathology and splenectomy to decompress the left portal venous system. Sinistral portal hypertension due to Echinococcus cyst is an unusual complication (7). The calcified dead hydatid cyst, located in the splenic tissue close to the hilus, grows very slowly, leading to chronic compression on the hilar vessels, which leads to dilatation and varix formation. Therefore, left-sided portal hypertension and isolated gastric variceal bleeding as a complication due to splenic hydatic cyst is quite a rare entity. Based on the English literature, only two patients were reported with sinistral portal hypertension due to splenic hydatidosis. One of those patients had variceal bleeding (3); the other had only splenic perihilar varices but did not have gastroesophageal varices with bleeding history (4). We present our patient as the second case of upper gastrointestinal bleeding from isolated gastric varices due to left-sided portal hypertension caused by splenic hydatic cyst. It may be difficult to diagnose isolated splenic varices both endoscopically and radiologically. Gastric varices may not be recognized with endoscopy but we were able to see bleeding gastric varices in the first endoscopy. Due to the high mortality risk of variceal bleeding from sinistral portal hypertension, whether caused by thrombosis or obstruction, splenectomy is strongly recommended. In our case, the first therapeutic approach was cystectomy and omentoplasty, but that approach did not relieve portal hypertension and recurrent variceal bleeding, so the second operation was needed. Splenectomy was performed, after which the gastric varices disappeared and no other bleeding was seen. At the follow-up one year after the procedure, the patient's hemoglobin level was stable.

In conclusion, although a very rare entity, it may be beneficial to keep in mind splenic hydatidosis as a cause of sinistral portal hypertension and isolated gastric varices in the differential diagnosis to facilitate more rapid treatment, especially in endemic areas. Furthermore, we give special emphasis to the fact that splenectomy should be performed to cure and prevent severe complications such as variceal bleeding in sinistral portal hypertension due to primary splenic hydatidosis.

## REFERENCES

1. Ammann RW, Eckert J. Cestodes. Echinococcus Gastroenterol Clin North Am 1996; 25: 655-89.
2. Durgun V, Kapan S, Kapan M, et al. Primary splenic hydatidosis. Dig Surg 2003; 20: 38-41.

3. Klompmaker IJ, Haagsma EB, Slooff MJ. Splenic vein obstruction due to a solitary echinococcal splenic cyst, resulting in gastric fundus varices: an unusual cause of variceal bleeding. HPB Surg 1993; 6: 229-32.
4. El Fortia M, Bendaoud M, Taema S, et al. Segmental portal hypertension due to a splenic Echinococcus cyst. Eur J Ultrasound 2000; 11: 21-3.
5. Polat P, Kantarci M, Alper F, et al. Hydatid disease from head to toe. Radiographics 2003; 23: 475-94.
6. Adams DB, Mauterer DJ, Vujic IJ, Anderson MC. Preoperative control of splenic artery inflow in patients with splenic vein occlusion. South Med J 1990; 83: 1021-4.
7. Thompson RJ, Taylor MA, McKie LD, Diamond T. Sinistral portal hypertension. Ulster Med J 2006; 75: 175-7.