

Simultaneous Huge Splenomegaly and Large Liver Hydatid Cyst: A Rare Case

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Abstract

Background: Splenectomy and hydatid cyst resection using either laparoscopy or laparotomy methods are frequently performed procedures. However, cases of patients suffering from both massive splenomegaly and hydatid cyst are rare.

Case Presentation: Patient was a 62-year-old man who complained of upper abdominal pain and was diagnosed with massive splenomegaly. In a Computed Tomography (CT) scan performed prior to surgery, a hypodense lesion with the measurements of 45 × 43 mm was found in the sixth segment of the liver. Therefore we performed an open splenectomy and hydatid cyst resection through one incision.

Discussion: Splenomegaly is a rare condition with the prevalence rate of around 2% in the US. However, tropical splenomegaly is common in Asia and Africa. Splenomegaly may occur due to various reasons, including ET. Symptomatic splenomegaly is the most common cause of splenectomy. In myeloproliferative complications, splenectomy is performed to relieve the patient's pain, early satiety and other splenomegaly related symptoms.

Conclusion: Simultaneous surgical procedures on both massive splenomegaly and hydatid cysts are rare. We suggest a CT scan be performed and the best type of incision be chosen based on the pathology and site of lesion, before performing a midline incision.

Keywords: Massive splenomegaly • Mercedes benz incision • Hydatid cyst

Introduction

The term 'splenomegaly' is used to describe a spleen with a larger than 12 centimeters diameter, a weight of more than 400 g, and a length greater than 20 cm. A spleen weighing more than 1000 g is definitive of a condition known as massive splenomegaly [1]. Splenomegaly may occur as the result of a variety of conditions (Table 1) [2]. The aim of treatment of splenomegaly is treating the underlying disease and preventing the complications of splenomegaly itself.

Table 1. Cause of splenomegaly

Category	Examples
Liver disease	Cirrhosis, Hepatitis
Hematologic malignancies	Lymphomas, Leukemias, Myelo proliferative disorders.
Venous thrombosis	Portal or hepatic vein thrombosis
Splenic congestion	Portal hypertension, Congestive heart failure.
Cytopenia	Immune thrombocytopenic purpura, Autoimmune hemolytic anemia, Immune-mediated neutropenia, Felty syndrome.
Splenic sequestration	Pediatric sickle cell disease, Hemolytic anemia, Thalassemia.
Acute or chronic infection	Bacterial endocarditis, Infectious mononucleosis, HIV, Malaria, Tuberculosis, Histiocytosis, Abscess.
Connective tissue diseases	Systemic lupus erythematosus, Rheumatoid arthritis, Adult-onset still's disease, and some familial autoinflammatory syndromes.
Infiltrative disorders	Sarcoidosis, Amyloidosis, Glycogen storage diseases.
Focal lesions	Hemangiomas, Abscess, Cysts, Metastasis.

Hydatid Cyst Disease (HCD) is a well-known parasitic disease caused by *Echinococcus granulosus*. Hydatid cysts may develop in anybody organ, but occur commonly in the liver (75%) and lungs (15%) [3-4]. The symptoms may be related to the location of the cysts. HCD should be treated even if it is clinically asymptomatic, in order to prevent the possible complications [5]. Treatment options for hydatid cysts include surgery, medical treatment, and percutaneous drainage [6]. This paper reports a unique case of concomitant total splenectomy and resection of hydatid cyst using marsupialization technique via a Mercedes incision.

Case Presentation

A 62 year old man with a history of Essential Thrombocythemia (ET), presented to our hospital with complaints of abdominal pain and distension. The patient had a dull pain in his left upper quadrant and epigastric region which did not shift or radiate, and a left hypochondriac non-tender swollen mass. The size of the mass had increased over the past 4 weeks. The patient did not have diarrhea, fever, nausea, vomiting, jaundice or GI bleeding.

Upon examination, his vital signs were within the normal limits. Physical examination revealed a mildly asymmetric abdomen and a palpable spleen of about 20cm below the left costal margin, and splenomegaly was confirmed. There was no tenderness, rebound tenderness or guarding in his abdomen. No evidence of hepatomegaly and lymphadenopathy was found. Examinations of Chest, cardiovascular system, central nervous system, and musculoskeletal system were all normal. Laboratory studies revealed a white blood cell count of 8800/ul, a hemoglobin level of 10 g/dl, and a platelet count of 466000/mm³, partial thromboplastin time of 33,

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prothrombin time of 13.6, and INR.

A total bilirubin level of 3.96 mg/dl, direct bilirubin level of 2.01 mg/dl, and amylase level of 325 mg/dl were also reported. Abdominal spiral Computed Tomography (CT) scan showed a hypodense lesion with the measurements of 45 × 43 mm in the sixth segment of the liver, and one with the measurements of 70 × 40 mm in the frontal segment of the spleen, and another with the measurements of 12 × 12 mm located adjacent to the former lesion in the spleen (Figure 1). The spleen measurements were 275 × 106 mm, indicating that the organ was abnormally large. Abdominal 3-phase spiral CT scan showed a cystic lesion with the measurements of 50 × 53 mm in the sixth segment of the liver with no clear enhancement in different phases, and considering the observations on the simultaneously acquired sonogram, the first probable diagnosis was a hydatid cyst. A hypodense lesion with the measurements of 47 × 52 mm was observed, primarily in the frontal segment of the spleen. With the aid of the radiological findings, the diagnosis of hydatid cyst was confirmed and the patient was set to undergo a surgical operation to remove both the spleen and the hydatid cyst. Preoperative preparations were done. A Mercedes Benz incision was performed which revealed a massive spleen. Splenectomy was performed (Figure 2). Hepatic exploration revealed a hydatid cyst in the sixth segment of the liver. The cyst was resected with marsupialization technique, entirely intact (Figure 3). The rest of the abdominal organs including the stomach and pancreas were normal. The cavity was irrigated with 0.5% silver nitrate solution. Corrugated drain was placed.

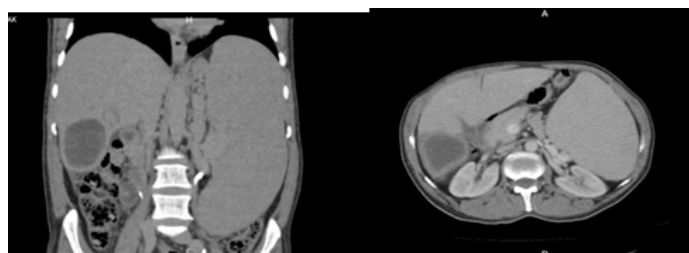


Figure 1. Preoperative computed tomography scan showing large hydatid cysts of the liver and massive splenomegaly.



Figure 2. Intraoperative photo showing massive splenomegaly.



Figure 3. Daughter cyst of liver hydatid cyst.

Histopathological evaluation of the spleen specimen revealed massive, sinusoidal proliferation with extramedullary hematopoiesis. Liver hydatid cyst biopsy report indicated fibrosis and acute inflammation and was compatible with hydatid cyst wall. Two days later, further inspections showed bile leaking along a drain site, which required a Gastroenterologist's consultation.

Following the consultation, Endoscopic Retrograde Cholangiopancreatography (ERCP) was performed within 24h of diagnosis, a procedure which included biliary sphincterotomy and internal stenting. Because of the high volume of bile leak, a plastic double pigtail stent was put in place, which was unsuccessful in stopping the leak. Therefore it was replaced with a simple stent which led to the cessation of leaking. The patient was discharged on a regimen including oral albendazole on the 7th day after surgery. He is now asymptomatic with no recurrence in 4 months.

Discussion

Splenomegaly is a rare condition with the prevalence rate of around 2% in the US. However, tropical splenomegaly is common in Asia and Africa [2]. Splenomegaly may occur due to various reasons, including ET (Table 1) [2]. Symptomatic splenomegaly is the most common cause of splenectomy. In myeloproliferative complications, splenectomy is performed to relieve the patient's pain, early satiety and other splenomegaly related symptoms [7].

Hydatid cyst disease is still considered a communal health issue in Asia, the Mediterranean, South America and Africa. In the recent years, with the increasing number of immigrants, the prevalence of this disease has increased in Europe and North America [6]. Treatment options for hydatid cysts include surgery, medical treatment, and percutaneous drainage.

There is some evidence that show hydatid cysts may cause splenomegaly by increasing the portal vein pressure [8].

In this patient, there were many factors (e.g. hydatid cyst and ET) which could potentially cause splenomegaly. Post splenectomy assessments and pathologic evaluations suggested that splenomegaly was caused in relation to ET.

Although surgery is a method of treating splenomegaly, the simultaneous presence of hydatid cysts in this case required a more complex approach. Ultimately, following the findings on the CT scan, a Mercedes incision was chosen, which enabled the surgeon to have access to both of the surgical sites.

Since a post cyst resection bile leak was predictable, a corrugate drain was placed, with no rush for it to be removed. Two days later, bile was leaking from the site of the drain, therefore a Gastroenterology consultation was performed and the aforementioned therapeutic measures were taken.

Taking this into consideration, approaching such cases is a

multidisciplinary process which requires the co-operation of all medical specialties, including gastroenterologists, oncologists, infectious disease specialists and surgeons. Fortunately, this patient benefited from such co-operation and has not had any issues in the post-operative evaluations so far.

Conclusion

To the best of our knowledge, this report describes the first case of simultaneous surgery for splenomegaly and hydatid cyst of liver. We presented an achievable method for a successful simultaneous splenectomy and hepatic hydatid cyst resection through one incision. It may be considered as one of the treatment options for patients with concurrent splenomegaly and hydatid cyst of liver.

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