Bizarre big belly ball: intraabdominal abscess mimicking Stauffer syndrome secondary to uterine leiomyosarcoma

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Summary

Background: Stauffer syndrome, a very rare paraneoplastic syndrome, refers to reversible intrahepatic cholestasis in the setting of an abdominal malignancy. *Case:* A 60-year-old female with a past medical history of uterine leiomyosarcoma status post radical hysterectomy, presented three months later with right upper quadrant abdominal pain. Laboratory evaluation revealed intrabdominal cholestasis and ultrasound of the abdomen showed an echogenic solid mass consistent with a metastatic leiomyosarcoma, and it was felt that her hyperbilirubinemia was due to Stauffer syndrome. However, three days later, blood culture grew gram negative bacilli, and CT scan of the abdomen revealed multiple mesenteric masses with air bubbles consistent with an abdominal abscess. The abscess was drained under CT-scan guidance and her cholestasis gradually came back to nearly normal. *Conclusion:* The case highlights the importance of considering infectious etiologies and Stauffer syndrome in the differential diagnosis of liver dysfunction in patients with intraabdominal malignancies.

Key words: Vulvar cancer; Brain metastasis; Meningeal carcinomatosis.

Case Report

A 60-year-old female presented to the emergency room with upper abdominal pain. Her medical history was significant for uterine leiomyosarcoma, status post radical abdominal hysterectomy with bilateral salpingo-oophorectomy (TAH/BSO), omentectomy and periaortic pelvic lymphadenectomy, three months prior. Postoperatively, there was no evidence of residual/metastatic disease (Figure 1). For the past month she had been experiencing progressive anorexia, shaking chills, and constant right upper quadrant abdominal pain (4-5/10 in severity). The pain did not radiate and there was no identifiable aggravating or reliving factors. She was admitted to the oncology inpatient service for further management and work up.

Laboratory evaluation revealed that her total bilirubin had increased from 3.8 to 10.2, with direct bilirubin increasing from 3.4 to 8.7 in the span of seven days (normal 0.1-1.0). Alkaline phosphatase was also found to be slightly elevated (ALP = 182, normal range 41-108). Transaminases were normal and the hepatitis panel was negative, other than evidence of past hepatitis A infection. Ultrasound of the abdomen (including Doppler) was negative for biliary dilatation/obstruction, liver lesion, portal vein occlusion or thrombus, but did show an echogenic solid mass in the left flank that was suggestive of a solid tumor, consistent with a metastatic leiomyosarcoma in this case. Because there was no obvious source of liver dysfunction, it was felt that the hyperbilirubinemia was due to Stauffer syndrome, a paraneoplastic cholestasis process.

Considering the diagnosis of Stauffer syndrome, we contemplated the use of chemotherapy for the sarcoma. However, her blood culture, drawn at the time of admission (because she was complaining of chills), came back positive for gram-negative bacilli *(Escherichia coli and Pseudomonas).* A follow-up CT scan of the abdomen and pelvis revealed multiple new heterogeneous-enhancing mesenteric masses with air bubbles throughout the abdomen, the largest measuring 8.5 x 6.5 cm in size (Figure 2a and 2b). It was aspirated and found positive for polymicrobial flora consistent with an abdominal abscess (inside the metastatic tumor). The patient was started on Piperacillin/Tazobactam (Zosyn), and the abscess was drained under CT scan guidance (Figure 3). Bilirubin and other laboratory parameters started improving within 24 hours, and gradually returned to near normal. The patient was subsequently discharged. However, due to the incurable nature of the cancer, she did not wish to pursue any chemotherapy, and was subsequently admitted to a home hospice program.

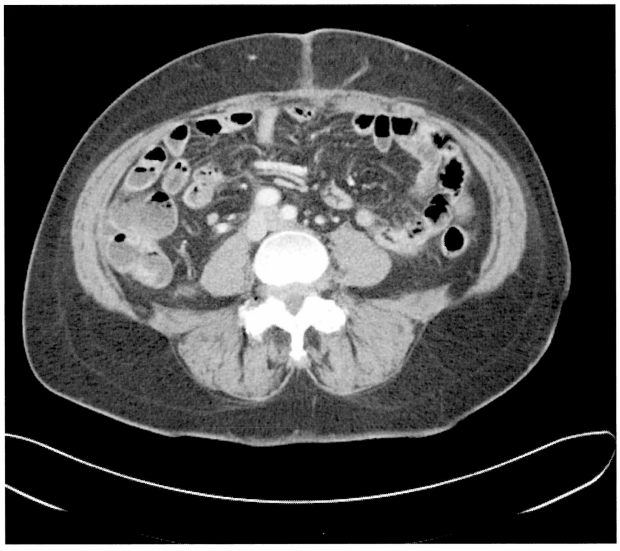

Figure 1. — Normal CT abdomen of the patient status post total abdominal hysterectomy with bilateral salpingo-oophorectomy for uterine leiomyosarcoma.

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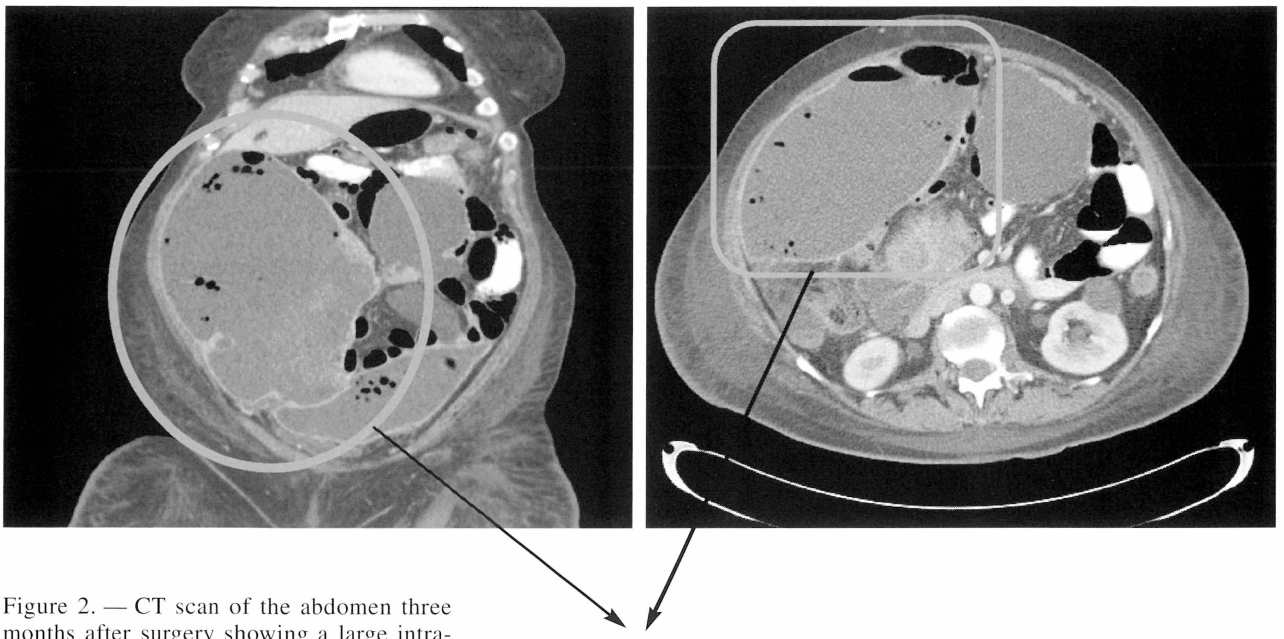

Figure 2. — CT scan of the abdomen three months after surgery showing a large intra-abdominal abscess in the coronal view (a) and saggital view (b).

Solid heterogeneous mass with air bubbles suggesting the presence of a large abdominal abscess.

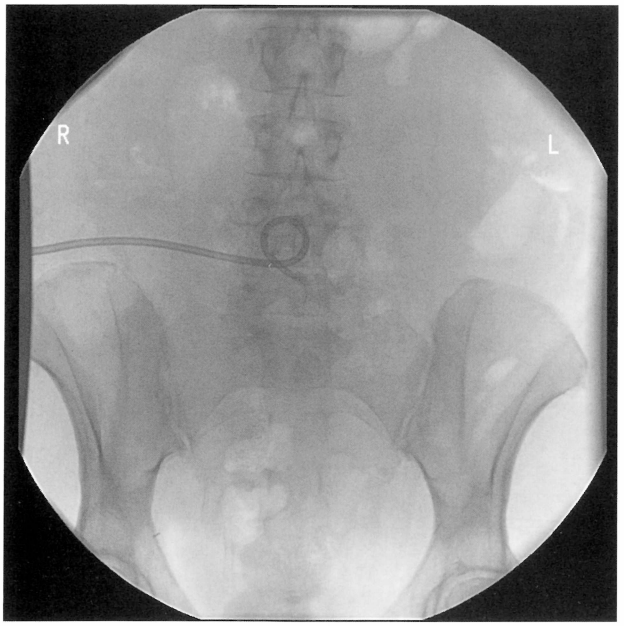

Figure 3. — CT scan guided drainage of the abscess.

Discussion

Stauffer syndrome refers to reversible intrahepatic cholestasis in the setting of an abdominal malignancy and is a diagnosis of exclusion. The pathogenesis is unclear but likely is due to over-secretion of interleukin-6 (IL-6) by the tumor [1]. The syndrome was originally described by MH Stauffer in 1961 [2]. It has been variously called Block-Stauffer-Rothmand's syndrome, Thomson-Rothmand's syndrome, 'nephrogenous hepatosplenomegaly' or 'nephrogenous hepatic dysfunction' [3]. It is usually seen as a paraneoplastic syndrome of renal cell carcinoma [4-9]. Stauffer's syndrome has also been reported in other malignancies including prostate [10], lung [11], and leiomyosarcoma [12].

The clinical features of Stauffer syndrome can include the presence of cholestasis, with or without hepatosplenomegaly [4-12]. Laboratory evidence of cholestasis with elevated ALP is usually present. Other laboratory features include thrombocytosis, hypoalbuminemia, and prolongation of prothombin time. Jaundice is not usually present, however an icteric variant of Stauffer's syndrome with elevated bilirubin has been reported [5, 6]. If a liver biopsy is performed, the histology is usually nondiagnostic and shows non-specific infiltration by leukocytes [13]. The management is directed towards the underlying malignancy. Stauffer's syndrome can be the initial clue towards the presence of an intraabdominal malignancy, and reappearance of the syndrome after surgical resection can herald the recurrence of the tumor [6, 14].

An intraabdominal abscess is an unusual cause of intrahepatic cholestasis. Blood cultures are generally not recommended as work up of hyperbilirubinemia if infection is not suspected [15]. However, in cases of intraabdominal metastasis, the porous bowel mucosa might permit intramural translocation of gut flora (gram-negative bacilli) resulting in development of abdominal abscesses and sepsis which could result in cholestasis. Sepsis is particularly known to result in cholestasis. The causative agent is usually a gram-negative bacilli with *Escherichia coli* being the most common agent [15] (such as in our case). The pathophysiology of cholestasis involves

impairment in hepatocellular and ductal bile transport mechanisms due to proinflammatory cytokines (including IL-6) [16, 17]. An intraabdominal abscess could also result in cholestasis from external biliary compression which can be seen radiologically (either by ultrasound or CT scan) as dilatated extrahepatic bile ducts. Cholestasis due to infection usually responds well to antibiotic therapy (targeted towards the specific organism; and if the organism is unknown, broad spectrum antibiotics targeted particularly against gram-negative bacilli is useful) [18].

This case presented as a diagnostic dilemma, and highlights the importance of considering infectious etiologies and Stauffer syndrome in the differential diagnosis of liver dysfunction in patients with intraabdominal malignancies.

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