Can You Identify This Malignancy?

**History**

Mr. B. is a 39-year-old male with no significant medical history who began experiencing progressive abdominal girth and bloating. Ultrasound and CT of the abdomen and pelvis revealed moderate to large abdominal ascites. Ultrasound-guided paracentesis yielded 7 L of serous fluid, with cytology showing atypical mesothelial cells. Upper endoscopy and chest CT were unremarkable. Exploratory laparoscopy demonstrated a large amount of abdominal ascites and carcinomatosis involving the peritoneal surfaces, omentum, and surfaces of the left and right lobes of the liver. Biopsies of peritoneal surfaces and liver implants confirmed a diagnosis of mesothelioma, and Mr. B. was referred to surgical oncology.

**Chief Complaint**

Mr. B. presents to the surgical oncology clinic with right upper quadrant pain extending to the right flank, 7/10 on the pain scale, exacerbated by lying on his right side. He has not been taking any pain medication. He reports abdominal bloating, progressive abdominal girth since paracentesis, early satiety, excessive gas with large meals, subjective weight gain, and dyspnea on exertion after climbing one flight of stairs. He denies nausea, emesis, melena, hematochezia, diarrhea, constipation, fevers, night sweats, and chills.

Mr. B. has never had a colonoscopy. He is a lifelong nonsmoker and denies any alcohol or drug use or any prior asbestos exposure to his knowledge. He works as a customer service representative at a department store and specifically denies any knowledge of occupational or recreational chemical exposure.

**Physical Examination and Diagnostic Studies**

Upon physical exam, Mr. B. is a healthy-appearing male of stated age, in no apparent distress. His vital signs are normal. Abdominal exam reveals a dome-shaped abdomen with gross ascites. Normal bowel sounds are present with no hepatosplenomegaly. Shifting dullness and fluid wave are noted. There is mild tenderness to deep palpation of the right mid-abdomen. No stigmata of cirrhosis, jaundice, scleral icterus, or lower extremity edema are noted. Cardiac exam is unremarkable. Laboratory analysis reveals Mr. B.’s CA-125 level to be 263.3 U/mL (normal range: 0–35 U/mL). His CEA and CA 19-9 levels are normal, as are his complete blood count, liver function tests, and kidney function.

Cross-sectional imaging of the abdomen and pelvis (see Figure) show a large amount of ascitic fluid extending into the peritoneal cavity, with the omentum displaced and compressed by fluid as well as omental caking. No dominant mass is identified, and intra-abdominal and pelvic organs are unremarkable except for a hepatic cyst. Repeat ultrasound-guided paracentesis is performed for symptomatic relief, yielding 5 L of green fluid, with cytology again revealing atypical mesothelial cells. Mr. B. subsequently undergoes an exploratory laparoscopy revealing a large amount of abdominal ascites and carcinomatosis involving the peritoneal surfaces, surfaces of the left and right lobes of the liver, and the omentum. Representative biopsies of peritoneal and liver implants are consistent with mesothelioma.

**WHAT IS THE CORRECT DIAGNOSIS?**

☐ Malignant peritoneal mesothelioma
☐ Benign multicystic peritoneal mesothelioma
☐ Well-differentiated papillary mesothelioma

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J Adv Pract Oncol 2014;5:231–232
Malignant peritoneal mesothelioma is a rare and fatal malignancy originating from the mesothelial cells that line the peritoneum. Mr. B.’s symptomatology is characteristic of this disease, as patients typically present with a combination of ascites, abdominal pain, swelling, and early satiety (Loggie, 2001). There are approximately 400 new cases diagnosed annually in the United States, with an equal incidence in both males and females (Alexander et al., 2013). Although there has been an association of asbestos exposure with this malignancy, the cause ultimately remains unknown (Yan et al., 2009). On gross examination, there are many tumor nodules of varying size diffusely seen throughout the peritoneal cavity, often leading to massive malignant ascites. The omentum is typically infiltrated by disease; thus, radiographic evidence of omental nodularity and caking is not uncommon.

Malignant peritoneal mesothelioma should be suspected in any individual with a diffuse malignant process in the abdomen. As was the case for Mr. B., it is not uncommon for the cytology on the ascitic fluid to be negative. A definitive diagnosis can be established only through tissue biopsy with appropriate immunohistochemical staining. The major histologic variants for malignant mesothelioma are epithelioid and sarcomatoid. The epithelioid subtype accounts for approximately 75% of patients and is typically associated with better response to therapy and subsequently a better prognosis than the sarcomatoid subtype.

The role of CA-125 as a tumor marker in this malignancy is not well understood. A study assessing 46 patients undergoing cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC) for malignant peritoneal mesothelioma reported a preoperative diagnostic sensitivity of 53.4% for CA-125 (Schaub et al., 2013). In a follow-up report, the authors described an 82% 5-year overall survival for patients with baseline CA-125 ≤ 35 U/mL and a 42% 5-year overall survival for patients with CA-125 > 35 U/mL. Thus it is possible that CA-125 may parallel with disease progression and regression after CRS/HIPEC, implying that the monitoring of serial measurements in follow-up patients may be helpful.

Follow-up

Mr. B. underwent CRS and HIPEC with cisplatin. His tumor was reduced to a CCR = 1 (completeness of cytoreduction), meaning that there was remaining disease that was 2.5 mm in thickness, with the bulk of the disease remaining along the interface of the mesentery to the small bowel. He was discharged from the hospital on postoperative day 8 with no postoperative complications but does suffer from minimal high-frequency hearing loss as a result of the cisplatin. Mr. B. received adjuvant carboplatin and pemetrexed for 4 cycles and is currently on maintenance pemetrexed with radiographically stable peritoneal implants.

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Correct Answer

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