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Case Report

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Peritoneal mesothelioma in a young woman: Case report of radiopathologic findings and review of the literature

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Abstract

Peritoneal mesothelioma is a rare diagnosis most often seen in middle-aged men and exceedingly rarely in individuals in their teens and twenties. Diagnosis is often delayed secondary to nonspecific presenting symptoms and a misconception that there must be a history of asbestos exposure to garner such a diagnosis. Here, we present the case of a 21 year-old female with a histologically confirmed diagnosis of peritoneal mesothelioma and review the key radiologic and histologic findings of this rare diagnosis.

Case report

A 21 year-old female presented to our Emergency Department with 3-4 months of abdominal swelling and 1-2 months of upper abdominal pain with nausea and intermittent non-bloody, non-bilious vomiting. Her past medical history was noncontributory and she had no known asbestos exposure. Laboratory work-up including complete blood count (CBC), complete metabolic panel (CMP), urinalysis (UA), inhibin B, thyroid function tests, and lipase levels were normal and HIV testing was negative. Tumor markers including alpha fetoprotein (AFP), carcinoembryonic antigen (CEA), cancer antigen 19-9 (CA19-9), cancer antigen 125 (CA-125) were also negative. Physical exam was remarkable only for a distended, non-tender abdomen.

Of note, the patient had undergone contrast-enhanced CT abdomen/pelvis four years earlier, which demonstrated striated nephrograms suggestive of pyelonephritis. There had been significant increase in abdominal circumference as compared to the prior abdominopelvic scout radiograph (Figure 1).

Contrast enhanced CT of the abdomen and pelvis demonstrated a large volume of simple ascites with peritoneal thickening, enhancement, and nodularity involving the greater omentum (Figure 2). Ascites fluid had attenuation values measuring up to 18 Hounsefield Units (HU). Dedicated chest CT was not obtained but the pleura within the lung bases appeared normal and chest x-ray was unremarkable. Ultrasound guided paracentesis yielded 2.55L of straw-colored ascites, with significant symptomatic relief following the procedure. Ascites fluid demonstrated 661 white blood cells (WBC), 622 red-blood cells (RBC) with macrophages and reactive cells present (Figures 3 and 4).

Shortly thereafter, laparoscopy was performed and demonstrated extensive carcinomatosis, multiple small subcentimeter liver masses, peritoneal studding, and cecal and omental masses. Four liters of non-bloody ascites was aspirated.

Pathology

Histologic sections of the cecal mass demonstrated a proliferation of cells with a variable amount of eosinophilic cytoplasm, round nuclei and prominent nucleoli, and overlying fibroadipose tissue. These cells were arranged in papillary structures that coalesced focally

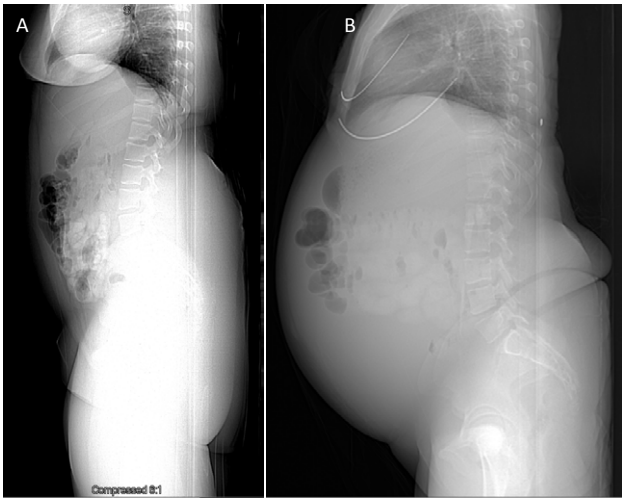

Figure 1. A. Scout lateral abdominopelvic radiograph from CT obtained in 2013. B. Scout lateral radiograph from CT obtained in 2017 demonstrates inreased abdominal girth and increased hazy opacity within the abdomen suggestive of interval development of ascites.

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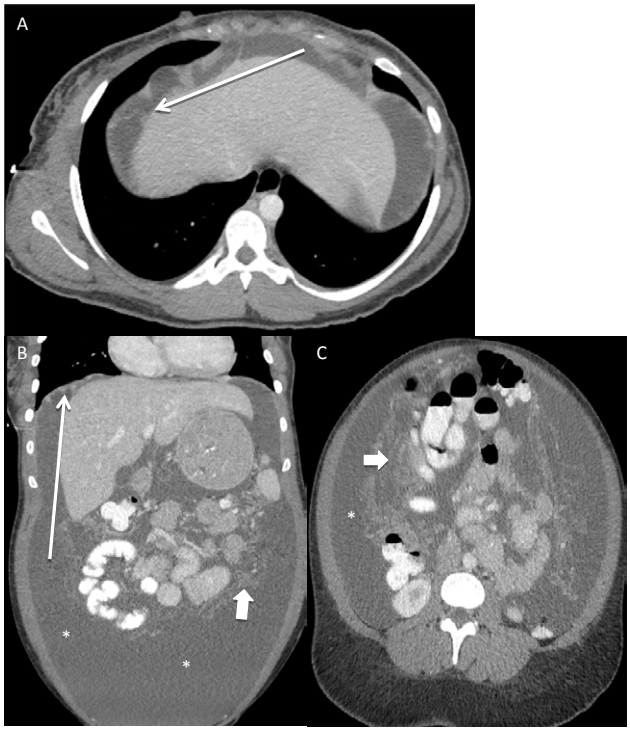

Figure 2. (A) Axial contrast-enhanced CT image at the level of the liver dome demonstrates subtle nodularity along the hepatic dome (long arrow). (B, C) Coronal and axial contrast enhanced CT images demonstrate a large amount of ascites (asterisk), subtle nodularity along the undersurface of the right diaphragm (long arrow), and enhancing omentum (short arrow).

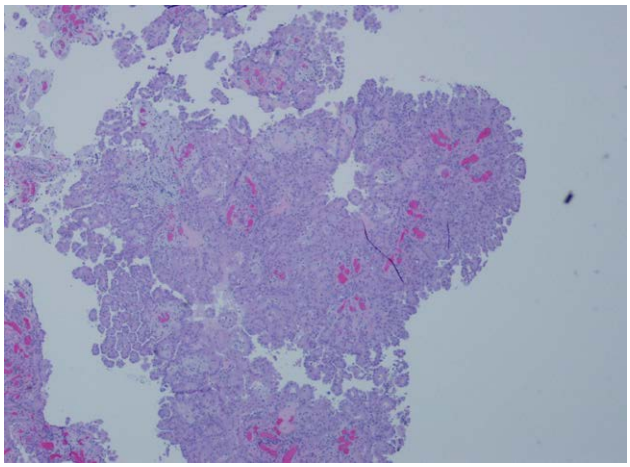

Figure 3. Low power view (4x) of mesothelial proliferation with complex papillary architecture and coalescence.

(Figures 3 and 4). There was also focal invasion in the surrounding soft tissue. Immunostains were performed to characterize the tumor cells: they were positive for calretinin, CK5/6 and WT1 supporting a mesothelial origin. MOC31, BerEP4 and p53 were negative, ruling out adenocarcinoma and serous carcinoma.

Overall the histologic features (presence of focal invasion) and immunochemical profile supported the diagnosis of malignant mesothelioma, epithelioid type.

Discussion

Peritoneal mesothelioma is a rare diagnosis, which presents with

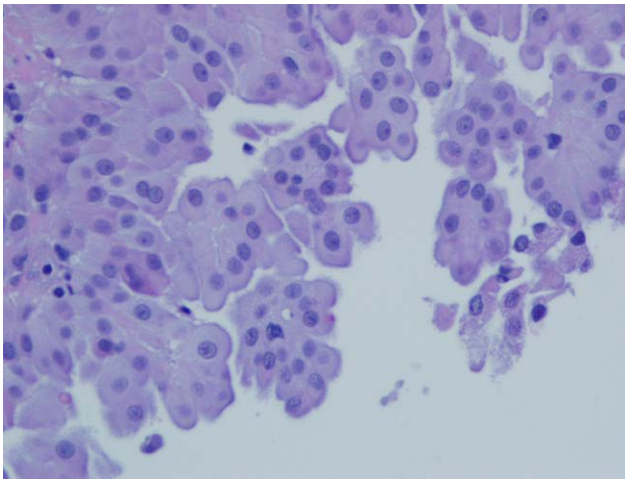

Figure 4. High Power view (20x) of tumor cells with eosinophilic cytoplasm, and round nuclei with prominent nucleoli.

an array of nonspecific symptoms including abdominal distension, abdominal discomfort, and malaise [1]. The incidence is approximately 1 in 1,000,000, most of these middle-aged men [1]. This devastating diagnosis has an average survival of 5 to 12 months [1]. Survival has been shown to increase to 50-60 months with combined cytoreductive surgery and intra-peritoneal chemotherapy [2].

To the best of our knowledge, there have only been only a few reports in the literature of patients in their teens and 20s diagnosed with this entity [3-8], with this population accounting for less than 5% of the 300-400 cases of peritoneal mesothelioma reported worldwide per year [8]. Up to 50% of individuals with this diagnosis have no known history of asbestos exposure, and this number may be higher in children and young adults with such a diagnosis [1,8]. Other entities such as SMV virus 40, radiation exposure, genetics, heavy metals, chronic inflammation, and non-asbestos fibers have been implicated as possible etiologic factors [1,8].

There are two distinct histological types of peritoneal mesothelioma including epithelial, which most often presents with diffuse thickening of the mesentery and peritoneum and/or multiple small nodules [5]. The sarcomatoid type most often presents with a mass, while the biphasic type demonstrates a combination of epitheliod and sarcomatoid features (5). CT findings are variable and it can be difficult to distinguish peritoneal mesothelioma from peritoneal carcinomatosis initially [4]. Findings of peritoneal mesothelioma often include ascites with Hounsefield units between 6 and 25, nodular or thickened peritoneum, thickening of the falciform ligament, mesenteric nodularity, and small or large bowel nodules or masses [9-11]. Small or large bowel, biliary tract, and ureteral obstruction can also be seen. [11]. Fewer months of symptoms prior to diagnosis has been found to be associated with statistically significant prolonged survival [10]. Additionally, CT has been found to be effective as a prognostic tool for patient selection as well as to monitor response to cytoreductive therapy and intra-peritoneal chemotherapy [11]. Thus, it is critical that clinicians, radiologists, and pathologists be aware of this entity and its imaging findings.

Peritoneal mesothelioma rarely occurs in young individuals. However, this entity should remain in the differential diagnosis for patient with unexplained ascites, even in young individuals and those with no known history of asbestos exposure. Earlier consideration of this diagnosis would result in a faster diagnosis and initiation of treatment.

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