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Malignant Peritoneal Mesothelioma with Rhabdoid Features Masquerading as Idiopathic Necrotizing Pancreatitis with Mucinous Ascites: A Case Report and Literature Review

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Abstract: Background: Malignant peritoneal mesothelioma (PeM) is a rare form of malignant mesothelioma that accounts for 10-15% of all cases of mesothelioma. Of these rare tumors, the rhabdoid subtype is exceptionally infrequent with only three cases reported in the current literature. PeM most commonly presents with non-specific symptoms such as abdominal distention, anorexia and weight loss that are difficult to diagnose until the disease is advanced. In this case report, we present a case of malignant peritoneal rhabdoid mesothelioma and review the literature. Case presentation: The patient is a 76-year-old woman who originally presented with necrotizing pancreatitis one year prior to diagnosis. The patient continued to complain of abdominal pain, nausea, vomiting, and weight loss. She experienced recurrent deep vein thrombosis (DVT), recurrent chylous, and mucinous ascites. Diagnostic work-up including MRI, repeat CT, EUS, and MRCP were inconclusive. Additionally, cytology from multiple paracenteses were negative for malignancy. Diagnostic laparoscopy revealed diffuse carcinomatosis, abdominal wall and peritoneal implants and a large epigastric mass. Biopsies of lesions taken during the procedure were identified as peritoneal mesothelioma. Conclusions: To our knowledge, we have presented the first case of PeM with rhabdoid features present in the peritoneum and in gastric polyps. The large amount of histopathogical variation of these tumors requires surgical biopsy, as cytology alone is non-diagnostic.

Keywords: Malignant Peritoneal Mesothelioma (PeM), Malignant Mesothelioma (MM), Rhabdoid Mesothelioma

1. Introduction

Malignant Mesothelioma (MM) is a great masquerader, presenting as an infection, metastasis, another type of cancer or as a paraneoplastic syndrome [1-5]. It arises from mesothelial cells lining the serosal cavities, including the pleura, peritoneum, and pericardium. The most common subtype is pleural mesothelioma, which typically presents in older men (>60 years) [5]. Malignant peritoneal mesothelioma (PeM) accounts for about 10-15% of all cases of mesothelioma, presenting at a slightly younger age, with a median age of 51-59 years [6, 7]. Unlike pleural mesothelioma, it has an even distribution in men and women [8-10]. The most common of its presentations are non-specific symptoms such as abdominal distention (present in 30-80% of patients), anorexia, and weight loss [11, 12]. Due to the non-specific nature of symptoms, patients with PeM frequently have advanced disease at the time of diagnosis. On average, patients are diagnosed 4-6 months after initial presentation [6, 13, 14].

Histologically, diffuse malignant mesothelioma is divided

into 3 types: epithelioid, sarcomatoid, and biphasic. The epithelioid type is extremely heterogeneous, presenting as solid, tubulopapillary, trabecular, clear cell, and small cell, among others [15, 16]. Of the subtypes, PeM with rhabdoid features is exceedingly rare, with only three cases reported in the literature [14, 17, 18]. MM presenting with malignant gastrointestinal (GI) polyps is also rare, with only 16 reported cases of mesothelioma presenting as metastatic disease in the GI tract [19]. To our knowledge, there are no case reports of PeM with rhabdoid features also presenting with malignant GI polyps.

Here, we present the clinicopathologic findings of a case of a malignant peritoneal rhabdoid mesothelioma presenting as a necrotizing pancreatic mass, recurrent deep vein thrombosis (DVT), chylous, mucinous ascites, and late onset weight loss.

2. Case Report

2.1. Patient Presentation

This is a 76-year-old female who initially presented with idiopathic necrotizing pancreatitis one year prior to presentation (Figure 1). Repeated biliopancreatic ultrasounds and triglyceride levels were normal. The patient denied use of alcohol. She continued to return every few months with intermittent, vague abdominal pain, nausea, weight loss, and thick, chylous appearing ascites (on paracentesis). Six months after her initial admission, she developed a DVT and pulmonary embolism (PE) requiring thrombectomy and anticoagulation. Ultrasound of the left lower extremity identified a DVT of the left common femoral, superficial femoral, popliteal, peroneal, and posterior tibial veins. On examination, the patient's left lower extremity was tender to palpation with a palpable cord in the popliteal fossa without pitting edema.

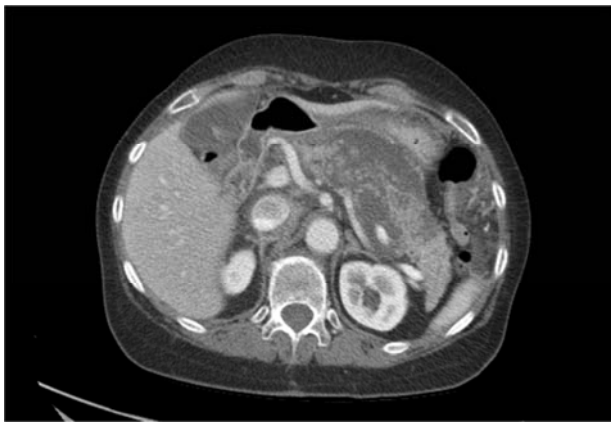

Figure 1. *Initial CT Abdomen/Pelvis (4/2020) with evidence of pancreatic necrosis (note: absence of ascites or carcinomatosis at this point).*

At that time, she was found to have recurrent mucinous ascites and an enlarging peripancreatic fluid collection on MRI (Figure 2). Multiple paracenteses were performed during this time, with the ascites consistently milky white in color and of a gelatinous consistency. Studies were consistently negative for infection and cytology was negative for malignancy; however, most samples could not be analyzed due to fluid viscosity, even with serial dilutions. One month later, the patient was admitted for worsening ascites and persistent abdominal pain. An endoscopic retrograde cholangiopancreatography was performed with pancreatic and biliary stents placed. By this time, the patient was requiring monthly paracentesis for management of symptoms. The fluid remained too viscous for further evaluation. She subsequently developed recurrent DVTs while on anticoagulation and an inferior vena cava (IVC) filter was placed.

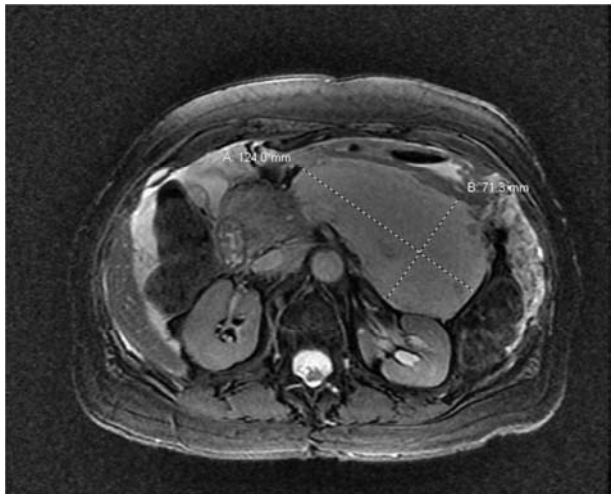

Figure 2. *MRI (8/2020) demonstrating large pancreatic pseudocyst.*

In March of 2021, almost one year after the patient's initial admission, repeat CT scan of the abdomen/ pelvis showed omental caking with enlargement of the proximal pancreas, raising a concern for peritoneal carcinomatosis (Figure 3). At this time, an endoscopic ultrasound with fine needle aspiration was performed for further evaluation of the pancreatic cyst, which was non-diagnostic. The patient was discussed at multidisciplinary tumor board with recommendations to perform lymphangiography to determine the etiology of the ascites, which appeared chylous in nature. This was non-diagnostic as well. The patient then underwent repeat magnetic resonance imaging (MRI) with and without contrast with magnetic resonance cholangiopancreatography. Imaging showed a large fluid volume surrounding the pancreas and extended into the pararenal spaces (Figure 4). There was a mass effect leading to displacement of the surrounding structures. No masses were identified in the pancreas; however, the distal tail demonstrated an abnormal T1 signal with enhancement. Due to inability to diagnose endoscopically and with no specific areas to target by interventional radiology, diagnostic laparoscopy was performed. At the time of laparoscopy, she was found to have diffuse carcinomatosis, with thick, mucinous, abdominal wall and peritoneal implants in addition to a large, firm, and highly vascularized epigastric mass, all of which were biopsied and sent for pathological evaluation. A moderate amount of mucinous ascites was drained during the procedure. An esophagogastroduodenoscopy performed during the operation showed multiple gastric polyps in the lesser curvature, which

were biopsied. Cytology of peritoneal washing was negative for malignancy. Pathology of the implants returned with peritoneal mesothelioma.

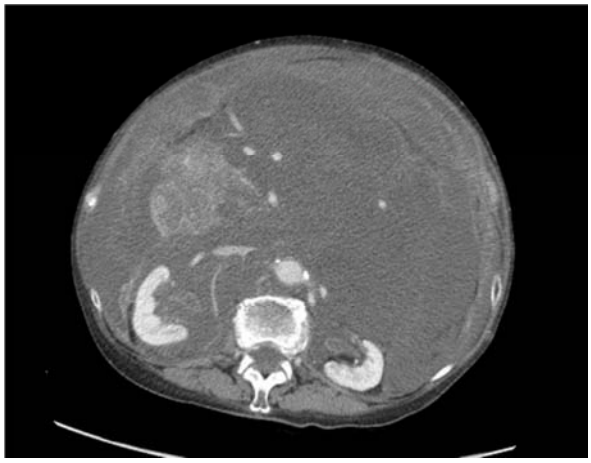

Figure 3. *Repeat CT Abdomen/Pelvis (3/2021) with evidence of ill-defined soft tissue with nodularity, concern for carcinomatosis, and large volume ascites.*

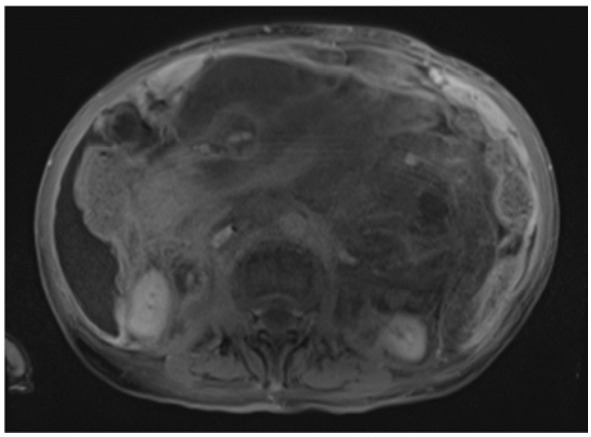

Figure 4. *Repeat MRI (4/2021) demonstrating large volume fluid surrounding pancreas.*

2.2. Past Social History

The patient's past social history was significant for heavy secondhand exposure to asbestos. Notably, her husband died of pleural mesothelioma.

2.3. Lab Values

During hospital admission, patient's lab values were significant for a low albumin of 3.5 (3/2/2021), normal lipase of 101 (3/26/2021), mildly elevated aspartate aminotransferase at 52 (3/2/2021), and normocytic anemia with a hemoglobin of 10.6 (3/2/2021).

2.4. Pathological Findings

The pathological examination of the peritoneal mass biopsy showed tumor cells arranged in an alveolar pattern and consisting of areas of discohesive cells in a background of myxoid stroma. The tumor cells showed eccentrically located nuclei and an abundant amount of glassy, eosinophilic cytoplasm with cytoplasmic inclusions, consistent with rhabdoid cells (Figures 5A-D). Tests for immunohistochemical markers performed on sections of the peritoneal tumor showed diffuse and strong positivity for calretinin, Wilms tumor-1, and CK 5/6, which was supportive of a mesothelial phenotype (Figures 6A-C). CEA, Ber-EP4, and desmin were all negative, excluding a poorly differentiated carcinoma or metastatic tumor. INI nuclear staining was retained in the rhabdoid areas. Given this constellation of findings, a diagnosis of PeM with rhabdoid features was made. Histological findings of the gastric polyp biopsy showed discohesive tumor cells infiltrating into the lamina propria of gastric mucosa, morphologically similar to the peritoneal mass (Figure 7A). Immunohistochemistry studies performed on the gastric polyp demonstrated the same immunostaining profile as in the tumor sample. Tumor cells of the gastric polyp also showed immunoreactivity to mesothelial cell markers (calretinin, WT-1, CK 5/6 (Figures 7B-D). In summary, the gastric tumor was considered as a MM invading gastric mucosa.

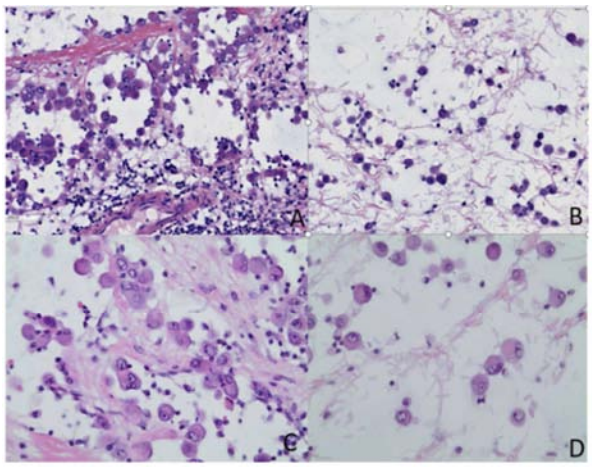

Figure 5. *Histological findings of excision biopsy of peritoneum mass. A: H&E staining shows rhabdoid cells arranged in an alveolar pattern (magnification x200). B: Discohesive tumor cells are seen in the background of myxoid stroma (magnification x200). C: Higher magnification (x400) showing rhabdoid cells with cytoplasmic inclusions. D: Higher magnification (x400) of discohesive cells exhibiting rhabdoid morphology.*

Figure 6. *Immunohistochemical stains of the peritoneal mass. A: Calretinin immunostaining showing nuclear and cytoplasmic positivity in the tumor cells. B: WT-1 immunostaining showing nuclear positivity in the tumor cells. C: Tumor cells exhibits strong membranous expression for CK5/6.*

3. Discussion

PeMs are rare tumors according to current reported rates in the literature [14]. Even more rare are cases of PeM with rhabdoid features. Reported cases have been discovered to involve the liver and the periumbilical peritoneum [17, 18].

We have presented a case of PeM with rhabdoid features present in the peritoneum and in gastric polyps. To our knowledge, this is the first case with these features. Given our patient's rare variant of PeM, we seek to provide the clinicopathological features of this case along with a brief review of the literature.

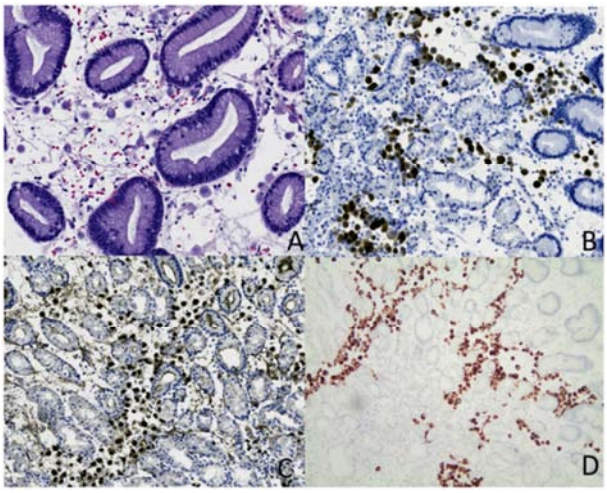

Figure 7. *Histopathology findings of the gastric polyp. A: H&E staining shows discohesive tumor cells infiltration in the lamina propria of gastric mucosa, morphologically similar to peritoneal mass (magnification x200). B: Calretinin immunostaining showing nuclear and cytoplasmic positivity in the tumor cells. C: The immunostaining of WT-1 shows nuclear positivity in the rhabdoid tumor cells. D: Tumor cells exhibits strong membranous expression for CK5/6.*

The clinical presentation of PeM is highly variable and non-specific. Common presenting features include abdominal distention (30-80%), diffuse abdominal pain (27-58%), early satiety, weight loss, and nausea [13]. The patient discussed in this review had all of these features (although the abdominal pain was not particularly prominent). PeM is also known to produce several different paraneoplastic phenomena, including malignancy-related thrombosis [20]. Consistent with this, our patient presented with a hypercoagulable state which required treatment with rivaroxaban and the placement of an IVC filter. The most commonly utilized imaging methods for PeMs are computed tomography (CT) scans; however, the findings are non-specific [13]. As in the case of our patient, imaging was insufficient to assist in a diagnosis. Paracentesis also has a low diagnostic yield for these tumors, as the number of malignant cells present in the ascites is low and they have a wide range of cytologic features. The reactive mesothelial cells and omental induration on CT are indicative of a malignancy. As a result, solid tissue specimens are usually required to make a diagnosis, which was also true for our patient [1, 10, 12-14, 21].

Primary mesotheliomas may resemble various primary or metastatic cancers that have directly invaded the serosal membranes. Metastatic malignancies, particularly carcinomas and sarcomas of the pleura, pericardium, and peritoneum, may mimic mesothelioma. Lung cancer with rhabdoid features can be confused with MM. Immunohistochemical staining patterns and specific genetic markers in tumor or somatic tissues differentiate most carcinomas, lymphomas, and metastatic sarcomas from asbestos-related mesothelioma [22]. In women with PeM, most frequent markers are calretinin and WT-1 [23].

Mesotheliomas are known to present with a wide variety of histologic patterns which include epithelioid, sarcomatoid, mixed epithelioid, biphasic (mixed sarcomatoid and epithelioid), desmoplastic, and rhabdoid types [18]. Thus, histologic patterns alone are also insufficient to make a diagnosis, due to the cytologic variation that is typical of these tumors. Immunohistochemical markers are often employed to assist in determination of a definitive diagnosis. A review of the literature identified only three cases of PeM with rhabdoid features. Ordóñez provides an in-depth description of the immunohistochemical features of 10 cases of mesothelioma with rhabdoid features. Of these cases, only one was of peritoneal origin, which had epithelioid features and 70% rhabdoid cells [18]. All of the specimens included in the study were found to stain positively for vimentin and pan-keratin, which was consistent with the non-rhabdoid areas of the specimens [18]. The case reported by Matsukuma et al. was of a biphasic mesothelioma containing sarcomatoid cells with rhabdoid features [17]. The rhabdoid cells of this tumor stained positively for vimentin, keratin, and epithelial membrane antigen expression [17]. Puttagunta et al. described a case of deciduoid mesothelioma with focal rhabdoid change. The cells of this lesion stained positively for keratin, vimentin, and calretinin [21]. The lesions described in Matsukuma et al. and Puttagunta et al. were primary lesions of the liver, while the case described by Ordóñez was a periumbilical peritoneal lesion [17, 18, 21].

Previous studies have shown a wide range in ages for the diagnosis of PeM in women, ranging from 19 to 93 years old, with a mean age at diagnosis of 60 for mesothelioma in general, and a median age of 52 for PeM [24]. It presents frequently with nonspecific abdominal or pelvic pain, paraneoplastic syndrome, or cervical lymphadenopathy. PeM is associated most frequently with high household asbestos exposure [25]. Overall, 70-80% of cases of mesothelioma result from exposure to asbestos, and para-occupational exposure, such as that experienced by our patient, is a known risk factor [26]. Symptoms of mesothelioma may not appear until 30 to 50 years after the initial exposure to asbestos. However, after symptoms become apparent, mesothelioma may rapidly progress to cause life-threatening complications [27].

Ten percent of acute pancreatitis is idiopathic. Mechanical obstruction of the pancreatic duct due to rare etiologies includes structural abnormalities like microlithiasis, pancreas divisium, and pancreatic-biliary tumors. Necrotizing pancreatic ductal adenocarcinoma is responsible for 1.1% of idiopathic necrotizing pancreatitis, with a delay in diagnosis of 5.6 months [28]. Primary pancreatic mesothelioma is extremely rare and metastasis to the pancreas are uncommon. These are found only in a minority (3-12%) of patients with widespread metastatic disease at autopsy. Essentially, any primary cancer may eventually deposit in the pancreas, most commonly renal cell carcinoma.

The incidence of mesothelioma is anticipated to increase worldwide in the next two decades, due to the expected latency between exposure and onset of disease [22]. Since novel potential therapeutic strategies are available, one should be familiar with MM etiology, pathophysiology, clinical appearance, and genetic screening, due to difficulties with diagnosis of primary mesothelioma using only cytology and immunohistochemical staining patterns [29].

4. Conclusion

PeM with rhabdoid features is an exceedingly rare diagnosis with a non-specific clinical presentation. We presented the clinicopathological features of a patient with PeM with rhabdoid features present in the peritoneum and in gastric polyps. Our patient had a vague presentation of abdominal distention, diffuse abdominal pain, early satiety, weight loss, nausea, and hypercoagulable state that was consistent with the literature. Multiple paracenteses and imaging studies were insufficient to establish a diagnosis in our patient. Surgical biopsy was diagnostic; however, these lesions are difficult to diagnose based on the high variability of the histological presentation. Several different metastatic malignancies may appear similar to mesothelioma especially carcinomas and sarcomas of the pleura, pericardium, and peritoneum. For this reason, immunohistochemical stains of samples are often required to make a final diagnosis. Ultimately, due to vague clinical presentation, definitive diagnosis of PeM requires surgically obtained tissue, as was present in this case.

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