Case report

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Contrast-Enhanced Ultrasound in the diagnosis of Pseudomyxoma peritonei: a case report

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Abstract

Pseudomyxoma peritonei (PMP) is a rare malignancy marked by mucinous ascites from neoplastic cell dissemination. We report a 50-year-old male with abdominal pain, weight loss, and ascites, initially suspected as hepatic metastases. Imaging revealed peritoneal implants and characteristic liver scalloping, but the percutaneous biopsy was inconclusive. Notably, contrast-enhanced ultrasound (CEUS) reproduced the hepatic scalloping typically seen on CT, helping distinguish mucinous peritoneal deposits from metastatic lesions. The patient's condition worsened and passed away. This case illustrates diagnostic difficulties, the limitations of biopsy, and the potential utility of CEUS in PMP.

Keywords: Pseudomyxoma peritonei; contrast-enhanced ultrasound; hepatic scalloping

Introduction

Pseudomyxoma peritonei (PMP) is an uncommon condition usually originating from mucinous tumors of the appendix. The disease is marked by peritoneal mucin, ascites, and organ dysfunction. PMP has a variable prognosis depending on histological classification and the feasibility of cytoreductive surgery with hyperthermic intraperitoneal chemotherapy (CRS/HIPEC) [1].

We present a case of pseudomyxoma peritonei in which contrast-enhanced ultrasound (CEUS) reproduced hepatic scalloping, aiding in the differential diagnosis and illustrating its potential utility in this rare malignancy.

Case report

A 50-year-old male patient presented with progressive abdominal pain, asthenia, and significant weight loss of 16 kg over two months. Physical exam showed cachexia, pallor, abdominal distension, palpable upper abdominal firm masses, and ascites on percussion. Cardiovascular exam showed tachycardia (118 bpm), blood pressure of 143/107 mmHg, and no murmurs. The patient was classified as ECOG (Eastern Cooperative Oncology Group) 3, with poor performance status. Labs revealed leukocytosis (19,170/mm³) with neutrophilia, mild hypoalbuminemia, and mild electrolyte imbalances. Tumor markers (CA 19-9, AFP, CEA) were normal or borderline. Ultrasound (LOGIQ™ S7 Ultrasound/GE Healthcare, United States, C1-5 MHz convex transducer) showed multiple hypoechoic subcapsular liver nodules (10–16 mm) (fig 1, Video 1 on the journal site), hyperechoic thickened omentum and minimal ascites.

CEUS – following the injection of 2.4 ml Sonovue®, contrast reached liver in 13 s – demonstrated non-enhancing hepatic nodules (fig 2, Video 2 on the journal site). Liver parenchyma was indented around the nodules, exhibiting a 'moth-eaten' or 'grape leaf' appearance. In the portal phase, the hepatic parenchyma showed homogeneous enhancement, without washout.

The computed tomography (CT) showed typical liver and spleen scalloping (fig 3), multiple peritoneal implants, and ascitic fluid collections along the splenic and gastric contours, mesentery (with interileal and inter-

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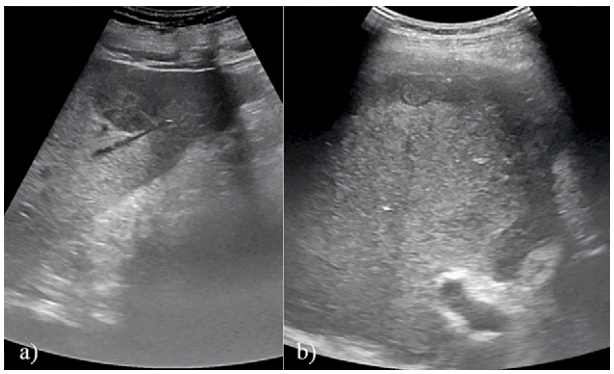

Fig 1. B-mode ultrasound showing multiple hypoechoic subcapsular hepatic lesions in the a) left and b) right hepatic lobes.

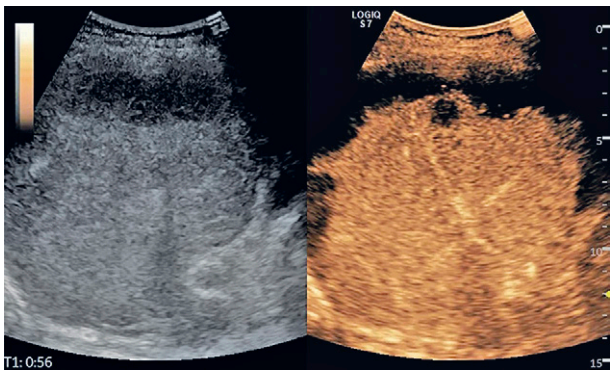

Fig 2. CEUS image (portal phase, 56 s post-injection) illustrating scalloped liver margins without contrast uptake in nodules.

jejunal distribution), omentum, and parietal peritoneum. They also extend into the paracolic gutters bilaterally and the pelvic region, suggestive of PMP.

PMP of unknown origin was diagnosed, as the CT scan did not identify a primary tumor. Gastroscopy showed no masses, while colonoscopy was incomplete due to colon fixation. Translucent membranes were seen in the colonic lumen but deemed histologically irrelevant.

The patient rapidly worsened, with altered coagulation parameters requiring plasma administration. Only a minimally invasive diagnosis approach was feasible. Ascitic fluid analysis indicated an exudate (protein: 2.81 g/dL, elevated LDH: 560 mg/dL, normal glucose: 63 mg/dL) and contained rare, atypical large cells with a hyperchromatic nucleus and irregular nuclear membrane, findings suggestive – but inconclusive – of malignancy.

Percutaneous peritoneal biopsy showed biphasic malignant proliferation, featuring an atypical epithelial component lining mesenchymal axes with increased cellularity. Unfortunately, immunohistochemical staining was omitted, limiting tumor classification. The patient's rapid clinical deterioration prevented further work-up; he passed away four days later.

Discussion

PMP is an uncommon clinical entity, affecting approximately 1–3 individuals per million annually, and typically presents in the fifth decade of life without significant gender predominance. PMP arises from the rupture of mucinous tumors – most frequently of appendiceal origin – and leads to mucin accumulation in the peritoneal cavity, producing gelatinous ascites or "jelly belly" [2,3]. The disease course is often insidious, with nonspecific symptoms such as abdominal discomfort, weight loss, or increased abdominal circumference, frequently delaying diagnosis or leading to misdiagnosis [2].

In this case, PMP presented with hepatic subcapsular nodules, initially suggesting metastatic liver disease. Notably, CEUS was able to replicate the characteristic "scalloping" of the liver surface typically visualized on CT or Magnetic Resonance Imaging (MRI), a feature caused by mucinous peritoneal implants compressing the hepatic contour. This is, to our knowledge, the first report describing hepatic scalloping identified via CEUS in PMP, representing a novel application of this modality in differentiating mucinous deposits from true hepatic metastases.

CEUS, while not routinely employed in PMP diagnosis, may offer diagnostic value in specific scenarios, particularly when standard imaging raises concern for liver metastases. Previous studies have employed CEUS to characterize appendiceal tumors and metastatic spread [4], but its ability to depict peritoneal mucinous scalloping has not been previously reported.

The diagnostic work-up of PMP typically relies on contrast-enhanced CT, which reveals mucinous ascites, liver and spleen scalloping, and peritoneal implants [5,6].

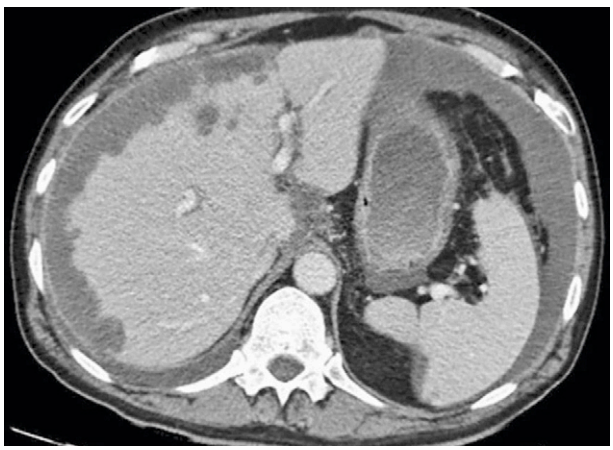

Fig 3. CT showing multiple loculated collections (native densities of up to 30 HU) along liver margins producing scalloping.

The imaging appearance of mucin (around 20 Hounsfield units – HU) is distinguishable from simple ascitic fluid (approximately 5 HU) [7]. MRI can be helpful for detecting the primary tumor and evaluating disease extent in the hepatoduodenal ligament or mesentery. PET/CT may be beneficial in high-grade variants with suspected extra-abdominal spread [2]. Tumor markers (CEA, CA 19-9, CA 125) support prognosis and monitoring [2,8]. Histological confirmation remains the gold standard, yet percutaneous biopsy has limited sensitivity due to the frequent acellularity of mucin pools [8]. In this case, both the cytology and biopsy were inconclusive or incomplete, highlighting the limitations of minimally invasive approaches in PMP and the importance of early surgical biopsy or laparoscopy.

PMP is histologically classified according to the Peritoneal Surface Oncology Group International (PSOGI) 2016 consensus into four categories: acellular mucin, low-grade mucinous carcinoma, high-grade mucinous carcinoma, and high-grade carcinoma with signet-ring cells [9]. These classifications hold significant prognostic value and guide therapeutic decisions.

The current standard of care is cytoreductive surgery (CRS) combined with hyperthermic intraperitoneal chemotherapy (HIPEC), which has shown the best long-term survival rates, especially in low-grade disease [8,10]. The completeness of cytoreduction (CC-score) and the peritoneal cancer index (PCI) are critical prognostic indicators [11]. Systemic chemotherapy may be used in high-grade disease, although its role remains limited, particularly in the preoperative setting [8,11]. Given the complexity of PMP management, referral to high-volume centers with multidisciplinary expertise is strongly recommended [12].

This case emphasizes the importance of considering PMP in the differential diagnosis of hepatic scalloping and peritoneal implants, even in the absence of an identifiable primary tumor. CEUS may serve as a complementary imaging modality in the diagnostic work-up of PMP, especially when CT findings are equivocal, or biopsy results are inconclusive. Its ability to visualize hepatic scalloping – a hallmark typically associated with CT – may support earlier, non-invasive suspicion of PMP and prompt timely surgical referral. The added diagnostic value of CEUS in this context warrants further investigation.

In conclusion, CEUS can replicate CT features of pseudomyxoma peritonei, such as hepatic scalloping, and may help distinguish mucinous implants from metastases. Early surgical biopsy remains crucial for diagnosis and management.

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Video 1. B-mode ultrasound appearance of the liver – multiple hypoechoic subcapsular liver nodules.

Video 2. CEUS of the liver – arterial phase where enhancement of the liver parenchyma begins at 13s and beginning of portal phase, both showing hepatic scalloping and homogeneous enhancement, without washout.