Pseudomyxoma peritonei due to mucinous adenocarcinoma of the appendix

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A 60-year-old man was referred for evaluation of long-standing abdominal pain. CT scan of the abdomen showed a fluid-filled dilated appendix with mural calcifications (Fig. A, asterisk) and intraperitoneal low-attenuation mass like nodular formations (Fig. B, arrows) causing scalloping of liver contour (Fig. A, C, arrowheads). The patient underwent appendectomy and peritonectomy followed by uneventful postoperative recovery. Histopathological examination confirmed the radiological suspicion of pseudomyxoma peritonei (PMP) due to appendiceal mucinous adenocarcinoma.

Comment

PMP is a rare disease characterized by intraperitoneal spread of mucinous fluid producing neoplasms which originates from the appendix or ovaries. Inflammatory changes associated with peritoneal tumor formations can lead to fistula formations and adhesions which can cause chronic bowel obstruction.

CT is a useful technique in distinguishing simple ascites from PMP since nodular nature of the mucinous material results in a suggestive finding such as hepatic scalloping. However, absence of scalloping does not rule out PMP. Sometimes septae or rim like calcifications could be identified within mucinous nodules. According to redistribution phenomenon, mucin producing cells only seed at peritoneal sites of relative stasis due to their low capability to adhere to the bowel wall that is in constant peristalsis. Therefore, the pouch of Douglas, both subphrenic spaces, and the surface of the liver and spleen are the most common involved sites.

Optimal therapy is considered complete macroscopic tumor removal combined with heated intraperitoneal chemotherapy. The treatment is beneficial in controlling of symptoms, but no absolute cure is common.

Reference