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PHOTO QUIZ

A 41-year-old man with an increased abdominal girth

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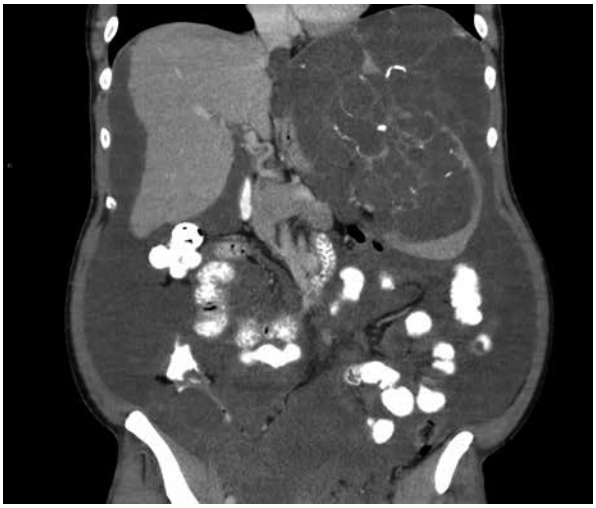
CASE REPORT

A 41-year-old man with no notable medical history was referred to the internal medicine ward because of an increasing abdominal girth and a weight loss of 21 kg. He denied stomach ache, a change in bowel movements, anorexia or night sweats. Physical examination revealed an abdominal distension with a protruded umbilicus and diffuse hyporesonant percussion without evident shifting dullness. A contrast-enhanced computed tomography (CT) scan was performed.

Figure 1. *A contrast-enhanced computed tomography of the abdomen*



WHAT IS YOUR DIAGNOSIS?

See page 436 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (PAGE 432)

A 41-YEAR-OLD MAN WITH AN INCREASED ABDOMINAL GIRTH

DIAGNOSIS

The computed tomography scan revealed a massive covering of the peritoneal cavity with accumulation of mucinous material, most likely to be consistent with the diagnosis of pseudomyxoma peritonei. The mesenterium and the omentum were extensively involved, with scalloping of the liver and spleen. At the lower pool of the caecum there was a mass effect that potentially could be a mucinous tumour of the appendix. A puncture was taken and pathology showed material with a remarkable load of mucus, supporting the diagnosis of pseudomyxoma peritonei, most likely a low-grade disseminated peritoneal adenomucosis. Our patient was referred to a specialised centre for treatment of pseudomyxoma peritonei where he had cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC).

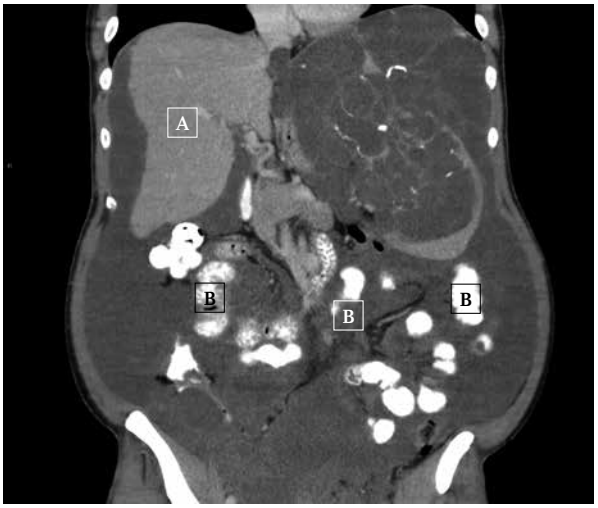
Pseudomyxoma peritonei is a rare condition that originates from a ruptured mucocele of the appendix in the majority of the cases, allowing epithelial cells to redistribute through the peritoneal cavity. In time they will proliferate and produce an excessive amount of mucinous ascites. Histopathologically, pseudomyxoma peritonei can be classified into a low-grade disseminated peritoneal adenomucinosis (DPAM), a peritoneal mucinous carcinomatosis (PMCA), which is the more malignant form, and an intermediate subtype. DPAM is associated with a better prognosis than PMCA. At an advanced stage, patients present with the characteristic distension of the abdomen, the so-called 'jelly belly', eventually leading to intestinal obstruction.[1,2]

Computed tomography of pseudomyxoma peritonei has characteristic features and demonstrates a typical distribution pattern of the mucinous ascites. This can be differentiated from normal watery ascites by density properties. In its end stage, impression of the liver surface ('thumb printing') and compression of the intestines by the mucus can be visualised. Although there are several CT features that are more common in DPAM than in PMCA, and vice versa, there is considerable overlap, which makes radiological differentiation difficult.[3]

Despite differences in biological behaviour and prognosis the current primary treatment is the same for both diseases: cytoreductive surgery followed by an HIPEC procedure. It has an overall ten-year survival rate of 63%. One of the most significant prognostic factors is the degree of extensive and complete cytoreduction. HIPEC contributes predominantly to disease control with a median progression-free survival rate of 8.2 years.[4]

Our patient unfortunately showed early radiological progression, four months after his HIPEC procedure. Therefore he is referred to a gastro-enterologist for further evaluation and potentially additional systemic chemotherapy.

Figure 2. *Abdominal computed tomography with scalloping of the liver (arrow A) and compression of the intestines (arrow B) by the mucinous material*



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