Synchronous pleural involvement
in a patient with pseudomyxoma peritonei

Clinical History:
An 81-year-old female patient was admitted because of fatigue, abdominal distention and breathlessness. At physical examination, there was massive ascites and a palpable omental cake. Evaluation of the respiratory system revealed decreased breath sounds in the right lung field.
Routine laboratory investigations were within normal limits.

Imaging Findings:
Abdominal ultrasonography showed massive, loculated ascites.

We performed a thoracic contrast-enhanced computed tomography (CT) at arterial phase and an abdominal contrast-enhanced CT at portal phase.

Thoracic CT revealed thickened pleura with lobulated masses of water attenuation in the right hemithorax, causing a mediastinal shift (Fig. 1). Neither parenchymatous mass nor mediastinal lymphadenopathy were observed.

Abdominal CT showed a large-volume ascites with scalloping of the liver surface (Fig. 2), curvilinear calcification in the right paracolic gutter (Fig. 3), central displacement of the small bowel (Fig. 4), and a voluminous appendiceal tumour with calcification (Fig. 5). Neither liver metastasis nor lymphadenopathy were present.

Peritoneal and pleural aspiration showed abundant mucinous material. Cytological examination of the fluid showed mucus with cohesive aggregates of mucoproducing epithelial cells (Fig. 6). The immunohistochemical profile revealed epithelial membranous positivity to CK20 and CEA (Fig. 7), while negativity was registered to calretinin.

Discussion:
Pseudomyxoma peritonei is characterized by diffuse collections of gelatinous fluid and mucinous implants on the peritoneal surface and omentum. The primary tumour is an adenoma or a low-grade adenocarcinoma of the appendix that perforates and spreads mucus-producing cells throughout the abdomen [1, 2].

Although this tumour is only superficially invasive, death is mainly due to intestinal obstruction by tumour masses. The mucin-producing cells in PP are poorly adherent, explaining the wide distribution of large tumour deposits. They mainly seed at sites of relative stasis, circulating with peritoneal fluid and dislodged from the surface of the bowel by peristalsis [3]. The pelvis, the subphrenic spaces, the right paracolic gutter and the surface of the liver and spleen are therefore the commonest involved sites [4].

Intrathoracic spread of PP is a rare condition, occurring in 5.4% of cases of pseudomyxoma peritonei, according to
an American study published in 2000 [5]. The authors reported that pleural involvement was due to direct extension through the diaphragm in all 23 patients [5]. Iatrogenic perforation of the diaphragm, occurring during subdiaphragmatic peritonectomy, is the most frequent mechanism. For those patients, pleural extension is detected during follow-up after the initial abdominal cytoreductive surgery [5, 6]. Congenital pleuroperitoneal communications, direct invasion of the diaphragm, haematogenous dissemination can also lead to pleural extension [5]. In our case, the synchronous pleural involvement of pseudomyxoma peritonei was presumed to be the result of the invasion of the central tendon of the diaphragm by the voluminous mucinous tumour, leading to the extension into the pleural space. The patient had an extensive peritoneal involvement with a thick layer of tumour on the undersurface of the right hemidiaphragm.

Imaging plays a major role in the diagnosis of PP. Ultrasonography shows non-mobile echogenic ascites with scalloping of the hepatic and splenic margins [7].

On CT, PP appears of low attenuation, with the presence of the following features: visceral scalloping, septae or calcification in the mucinous material, central displacement of the small bowel, and appendiceal tumour [4, 7]. Magnetic resonance imaging may provide similar information with ascites, peritoneal implants and scalloping [8]. From a histological point of view, different entities should be distinguished: disseminated peritoneal adenomucinosis, peritoneal mucinous carcinomatosis, with a different microscopic appearance [9]. Current treatment of PP consists of complete cytoreduction, using peritonectomy procedures, in combination with heated intraoperative intraperitoneal chemotherapy [5, 10].

**Differential Diagnosis List:** Pleuro-peritoneal pseudomyxoma, Colon cancer peritoneal carcinomatosis, Other malignancies peritoneal carcinomatosis, Cystic lymphangioma, Mesenteric cyst, Enteric duplication cyst, Meckel diverticulum, OVarian cystic lesion, Acute appendicitis

**Final Diagnosis:** Pleuro-peritoneal pseudomyxoma

**References:**


Description: Axial thoracic contrast-enhanced CT at arterial phase: thickened pleura with multiple lobulated masses in the right hemithorax. Origin: David A, Department of Radiology, Centre Hospitalier de Vendée, La Roche-sur-Yon, France
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Figure 2

Description: Axial abdominal contrast-enhanced CT at portal phase: ascites with scalloping of the liver surface. Origin: David A, Department of Radiology, Centre Hospitalier de Vendée, La Roche-sur-Yon, France
Description: Axial abdominal contrast-enhanced CT at portal phase: curvilinear calcification in the right paracolic gutter (arrow). Origin: David A, Department of Radiology, Centre Hospitalier de Vendée, La Roche Sur Yon, France
Description: Axial abdominal contrast-enhanced CT at portal phase central displacement of the small bowel. Origin: David A, Department of Radiology, Centre Hospitalier de Vendée, La Roche-sur-Yon, France
Description: Coronal abdominal contrast-enhanced CT at portal phase: voluminous appendiceal tumour with calcifications (arrow). Origin: David A, Department of Radiology, Centre Hospitalier de Vendée, La Roche-sur-Yon, France
Figure 6

Description: Cytological examination of the pleural and peritoneal aspiration. Origin: Pavageau AH, Department of Pathology, Centre Hospitalier de Vendée, La Roche-sur-Yon, France
Description: Immunohistochemical profile shows positivity of epithelial cells to CK20 (a, X 10) and CAE (b, X 20). Origin: Pavageau AH, Department of Pathology, Centre Hospitalier de Vendée, La Roche-sur-Yon, France