Case Report

Pseudomyxoma pleurii: unusual location of a rare disease

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ABSTRACT

Pseudomyxoma peritonei with pleural extension is very rare. Dissemination in the pleural cavity can be trans diaphragmatic or hematogenous. Appendicular and ovarian mucinous tumors are the most often involved. Imaging plays an important role in the diagnosis which is confirmed histologically. We report the case of a pseudomyxoma peritonei and pleurii in a 55-year-old female patient who underwent surgery for mucinous adenocarcinoma of the ovary. This diagnosis must be suggested when there is subpleural nodular thickening associated with pseudomyxoma peritonei.

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Introduction

Pseudomyxoma peritonei is a rare disease. It is an anatomo-clinical entity producing gelatineous peritonitis, invading the entire abdominal cavity, whether or not associated with neo-plastic epithelial cells. Its extension to the pleural cavity is exceptional and until now only few cases had been described in the literature [1].

The diagnosis is anatomo-pathological by transcutaneous or by celioscopic biopsy.

The treatment is surgical, associating hyperthermic intraperitoneal chemotherapy [1,2].

We report the case of a pseudomyxoma peritonei and pleurii in a 55-year-old female patient who underwent surgery for mucinous adenocarcinoma of the ovary.

Case report

A 55-year-old patient, with ovarian tumor treated by total hysterectomy with bilateral adnexectomy a year ago. The anatomopathological examination was in favor of tumor infiltration of the ovary and peritoneum by a mucinous-type adenocarcinoma. She was presented in consultation for abdominal distension with mild dyspnea and weight loss (12kg). The clinical examination found an altered patient, capable of only some personal care, with performances status as the World Health Organization = 3. There was diffuse abdominal dullness without collateral venous circulation or palpable mass. An abdominal ultrasound was performed showing ascites of great abundance. A contrast-enhanced thoracic computed tomography (CT) at arterial phase and

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Fig. 1 – Contrast enhanced chest CT scan in axial (a) and sagittal (b) section at the parenchymal and mediastinal (c) windows: showing pleural thickening with subpleural and scissural masses lobulated with water density, containing curvilinear calcifications (arrow).
an abdominal contrast-enhanced CT at portal phase were performed.

The thoracic CT scans revealed a unilateral pleural thickening of the right hemithorax with water-density lobulated sub pleural and scissural masses. There was no lung parenchyma lesion or mediastinal lymphadenopathy (Fig. 1). The abdominal CT scans showed ascites of great abundance, containing arciform calcifications, with scalloping on the liver and the spleen without other associated lesions, in particular no appendicitis, hepatic nodule or lymphadenopathy (Fig. 2).

An exploratory laparotomy with omentum biopsy was performed and the anatomopathological analysis was in favor of low-grade malignancy mucinous peritoneal carcinoma. The diagnosis of pseudomyxoma peritonei with pleural extension was confirmed. Unfortunately, the patient died few days following the diagnosis.

Discussion

Werth first described the pseudomyxoma peritonei (PMP) in 1884. Pleural extension of PMP is exceptional, rare cases have been reported in the literature. The most common tumors leading to PMP are appendicular and ovarian mucinous tumors. Pleural extension of PMP is due to trans-diaphragmatic, hematogenous or iatrogenic dissemination of a mucinous implant to the pleural cavity [2,3]. The rupture in the peritoneal...
cavity of mucin and epithelial cells, will induce the high production of mucus. Its distribution in the abdominal cavity will depend on three factors: the phenomenon of redistribution, visceral sparing and tumor cell trap [3,4].

The phenomenon of redistribution depends primarily on the sites of absorption of the peritoneal fluid. Tumor cells will therefore tend to accumulate mainly on the surface of the right diaphragmatic dome and the greater and lesser omentum. Once these sites are saturated, the redistribution process can then extend to the left diaphragmatic dome, peri-splenic and the rest of the peritoneal cavity. The phenomenon of redistribution also depends on gravity. The most sloping parts in a standing position and in the supine position will be the most affected: pouch of Douglas, hepatic retro space and para-colic splints [3,5].

Clinical presentation is not specific. It can be revealed by an increase in abdominal volume, abdominal pain, digestive or urinary signs or by a deterioration of the general condition [4].

Imaging plays an important role in the diagnosis, which must be confirmed histologically.

On ultrasound and CT scans, PMP is characterized by the presence of gelatinous ascites, sometimes with peritoneal implants. On CT, it is a heterogeneous hypo dense peritoneal effusion, sometimes partitioned, containing curvilinear calcifications and exerting a scalloping on the liver and the spleen with central displacement of the small bowel as in our patient. It predominates in the diaphragmatic peritoneum and the greater omentum. The primary tumor is rarely individualized.

Magnetic resonance imaging also shows ascites and peritoneal implants with scalloping. The pleural extension appears as thickened pleura with multiple lobulated masses [2,3,5].

The differential diagnosis arises with mesothelium and peritoneal carcinomatosis. The mesothelium gives a diffuse or nodular thickening of the peritoneal serosa with infiltration of the greater omentum, mesenteric masses, and pleural plaques.

Peritoneal carcinomatosis is characterized by an enhancement and irregular thickening of the peritoneal layers with the presence of peritoneal nodules and infiltration of the greater omentum and mesentery. In the PMP, there is no mesenteric fat infiltration [5,6].

The treatment is based on 2 methods: multiple surgical debulking and cytoreduction surgery with perioperative intraperitoneal chemotherapy. The goal of debulking is to remove as much gelatin as possible and tumor formations.

Cyto reduction surgery (CRC) consists of removing the primary lesion as well as all peritoneal nodules and infiltrative masses and any organ affected by contiguity. Lesions of the visceral peritoneum will be removed by segmental or atypical resection. It will also be possible to remove nodules on the Glisson capsule.

Treatment of pleural extension of mucinous tumor from PMP is based on intrapleural chemotherapy combined with pleural cyto reductive surgery [3–5].

Conclusion

Invasion of the pleural cavity with PMP is a rare entity with a poor prognosis. The differential diagnosis arises mainly with the mesothelium and peritoneal carcinomatosis. There are some radiological signs that can guide the diagnosis, which must be confirmed histologically. Treatment is based on cytoreduction with intrapleural and intraperitoneal chemotherapy.

Patient consent statement

Written informed consent for publication was obtained from the patient.

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