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Pseudomyxoma Peritonei: A Case Report

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Abstract **Case Report**

Pseudomyxoma peritonei (PMP) is a rare condition, consisting of mucinous ascites and multifocal mucinous epithelial implants. This disease mainly affects women. It may remain asymptomatic, discovered during laparotomy. The most frequent symptom is abdominal distension associated with diffuse abdominal pain. Mucinous tumours of the appendix are the most frequent cause of pseudomyxoma peritoneum, accounting for 90% of cases, and ovarian origin remains very rare. There are essentially two types of treatment for PMP: firstly multiple surgical debulking and secondly cytoreduction surgery with peri-operative intraperitoneal chemotherapy consisting of hyperthermic intraperitoneal chemotherapy with or without immediate post-operative intraperitoneal chemotherapy. We report a case of peritoneal pseudomyxoma secondary to a mucinous tumour of the right ovary.

Keywords: Pseudomyxoma Peritonei tumour.

INTRODUCTION

Peritoneal pseudomyxoma, also called gelatinous disease of the peritoneum is a rare entity that corresponds to diffuse peritoneal infiltration, consisting of mucinous ascites and multifocal mucinous epithelial implants. Its prevalence is 2 cases per million population [1] Peritoneal pseudomyxoma may remain asymptomatic, discovered during laparotomy. CT scans are the most specific diagnostic tool. The appendicular origin of gelatinous ascites is the most common and represents 90% of causes. Mucinous tumours of the ovary are a very rare cause. We report the case of a 79 year old woman with a peritoneal pseudomyxoma of ovarian origin [2].

CASE REPORT

We report the case of a 79 year old woman, without any particular pathological history, never had an appendectomy, who presented an abdominal distension of gradual onset for 12 months with moderate hypogastric pain, without any associated digestive signs, in particular no digestive haemorrhage or transit disorder, or obvious gynaecological signs, in particular metrorrhagia and leucorrhoea. The clinical evolution was marked by the worsening of the abdominal distension with alteration of the general state of the patient, which motivated her consultation at the emergency department of the Er-razi Hospital. The general physical examination revealed signs of malnutrition, such as melting of the adipose panicle, but there was no fever or paleness of the skin and conjunctiva. Abdominal examination showed homogeneous abdominal distension and diffuse dullness of the abdomen, without collateral venous circulation or palpable mass. The rest of the clinical examination was unremarkable.

Biological tests were normal

Abdominal ultrasound revealed abundant heterogeneous ascites with septal defects and fine mucoid echoes. Abdominal and pelvic computed tomography (CT) showed abundant ascites with suspicious-looking nodular infiltration of the peritoneum, with no distinguishable hepatic, splenic or digestive lesions. Tumour markers, namely carcinoembryonic antigen (CEA), carbohydrate antigen (CA) 19-9, and CA 125 were normal. As part of the extension work-up, a pelvic MRI was performed which showed peritoneal carcinosis with irregular and nodular thickening of the peritoneum, giving a pseudo-mass appearance at the level of the cul-de-sac of Douglas, with no clearly distinguishable ovarian mass. The thoracic CT scan did not reveal any pulmonary lesions apart from micro-nodules in the middle and left lobe of the left lung of infectious origin. Due to doubtful diagnosis, the decision was made to perform an exploratory laparoscopy. The surgical exploration

revealed a gelatinous ascites of great abundance with whitish nodules disseminated on the parietal peritoneum, as well as a mass depending on the right ovary. The procedure consisted of evacuation of the ascites with right adnexectomy taking away the tumour. The anatomo-pathological study of the peritoneal biopsies showed an aspect in favour of a peritoneal pseudo-myxoma and a cytology with atypical cells of low grade, that of the ovarian mass was in favour of a borderline mucinous tumour of the ovary. The immunohistochemical complement showed positive anti CK7, anti CK 20 confirming the ovarian origin.

DISCUSSION

Peritoneal pseudomyxoma (PMP) is an unusual condition characterised by mucinous ascites, classically arising from rupture of a low-grade mucinous neoplasm of the appendix, or more rarely of an ovarian neoplasm [3].

The natural history of PMP is centred on the "redistribution phenomenon", through which the mucinous tumour cells accumulate in specific sites. The peritoneal tumour accumulates at sites of peritoneal fluid uptake, namely the greater and lesser omentum and the underside of the diaphragm, particularly on the right. The second main distribution mechanism is gravity, with tumour deposits commonly aggregating in the recto-vesical pouch or pouch of Douglas, the paracolic gutters and the retrohepatic space [3].

PMP is usually diagnosed in the 40-55 age range, and is often discovered incidentally in patients who have undergone laparotomy, laparoscopy or imaging for other medical conditions.(4) Clinically, the symptomatology is non-specific. There are many revealing signs, dominated by progressive and isolated increase in abdominal volume and pain. The other functional signs are mainly related to the impact of the gelatinous peritoneal disease on the digestive tract and/or the urinary tract [2]. Due to its indolent nature and non-specific symptoms, most patients are unfortunately diagnosed at an advanced stage of the disease.

On biological grounds, the tumour markers ACE, CA 19-9 and CA 125 are increased [5] on radiological examination, the unprepared abdominal film (UAP) may show an opacity predominating on one side of the abdomen which is rarely significant. On ultrasound and CT, the diagnosis of pseudomyxoma is based on the identification of three lesions: The mucinous ascites and its characteristics, the nodular peritoneal implants if they are visible, and the primary tumour, which is only exceptionally visualised. The mucinous ascites of pseudomyxoma is heterogeneous and low density. It may be septated and may contain fine calcifications. In our patient, the CT scan showed the radiological features described in the literature, without being able to determine the location of the primary tumour. The CT scan is also of interest in monitoring the evolution of the disease, and in detecting a potential recurrence or complication. (Occlusion, abscess, ureteral compression with dilatation of excretory cavities) [2]. Magnetic resonance imaging (MRI) allows better characterisation of gelatinous masses, hepatic and splenic scalloping, and intra-ascitic septa [6].

Laparoscopy allows direct visualisation of gelatinous ascites and peritoneal implants. However, it is dangerous because it exposes the risk of gelatinous fistula and infection [6]. The spontaneous evolution is slow and mainly local, marked by the frequency of recurrences.

Ultraradical surgery is the gold standard for the treatment of PMP. It is currently based on cytoreductive or debulking surgery, which involves removing any visible tumour formation by performing a series of peritonectomy procedures. The addition of immediate postoperative intraperitoneal chemotherapy (IPC) or intraoperative intraperitoneal chemo hyperthermia (IPCH) improves the prognosis of the disease by acting on the microscopic residual disease [2, 7]

CONCLUSION

Pseudomyxoma peritonei is a rare neoplasm, most often of appendicular origin, but an ovarian origin remains possible. Without early treatment, the prognosis of this disease remains poor [8]. The prognosis can be improved by early diagnosis of mucinous tumours even before the ascites stage. Cytoreductive surgery combined with intraperitoneal chemotherapy remains the gold standard in the management of this disease.

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