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Pseudomixoma Peritonei, a Rare Entity Difficult to Diagnose and Treat - Case Report

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Rezumat

Pseudomixoma peritonei, entitate rară dificil de diagnosticat şi tratat - prezentare de caz

Autorii prezintă cazul unei paciente de 56 de ani diagnosticată cu pseudomixoma peritonei la 4 ani de la practicarea unei anexectomii stângi pentru chistadenom ovarian. Însămânțarea intraparietală a celulelor mucinoase a facilitat dezvoltarea la acest nivel a unei mase gelatinoase ce a ridicat probleme de diagnostic diferențial cu hernia incizională complicată, încarcerată. Semnele de diagnostic clinic şi paraclinic preoperatorii le considerăm insidioase şi nespecifice pentru stabilirea diagnosticului de certitudine. Tomografia computerizată abdominală a evidențiat prezența unei colecții intraperitoneale masive, însă, cunoscută fiind raritatea acestei patologii stabilirea diagnosticului inițial s-a făcut în cursul laparatomiei exploratorii. Intraoperator s-a efectuat, omentectomie, histerectomie totală cu anexectomie dreapta şi apendicectomie. Examenul histopatologic a confirmat diagnosticul de mucinoză peritoneală. Utilizarea cisplatinului, asociat cu citoreductia chirurgicală agresivă în acest caz de pseudomixoma, a avut o evoluție bună pe termen lung. Stabilirea diagnosticului a reprezentat o provocare, evoluția lentă şi nespecifică a afecțiunii conducând la un diagnostic diferențial dificil.

Cuvinte cheie: pseudomyxoma peritonei, diseminare mucinoasă, laparotomie exploratorie, hernie incizională

Abstract

The authors present the case of a 56 year-old patient diagnosed with pseudomyxoma peritonei, 4 years after being subjected to a left adnexectomy for ovarian cystadenoma. The intra-parietal insemination of the mucinous cells enabled the development, at this level, of a gelatinous mass that raised problems of differential diagnosis with irreducible incisional hernia. In what regards the preoperative signs of clinical and paraclinical diagnosis we consider them obscure and nonspecific. The abdominal computed tomography revealed the presence of a massive intraperitoneal collection, but given the rarity of this pathology the initial diagnosis was made in the course of the exploratory laparotomy. Intraoperatively it became necessary to perform the omentectomy and total hysterectomy with contralateral adnexectomy and appendectomy. The histopathological examination confirmed the diagnosis. Using cisplatin associated with aggressive surgical cytoreduction this case of pseudomixoma had a good long-term evolution. The diagnosis was a challenge, and the nonspecific slow evolution of the disease led to difficult differential diagnostic.

Key words: pseudomyxoma peritonei, mucinous dissemination, exploratory laparotomy, incisional hernia

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Introduction

Pseudomyxoma peritonei is a rare pathology described by Cruveilhie in 1848, with an incidence of 1 in 1,000,000 cases / year (1,2,3). Pseudomyxoma peritonei is characterized by mucinous ascites in the intraperitoneal area and in the peritoneal surface and omentum, secondary to the secondary metastasis from a primary tumour. Recent studies have concluded that most patients with pseudomyxoma peritonei are women (64-75%), usually in the sixth decade of life (2,3,4,5). It is frequently associated with appendicular mucocele or mucinous ovarian tumours(6,8) .

This pathology can have different clinical symptoms. Often the signs and symptoms are nonspecific even in advanced stages of disease. The most common symptoms are pain, abdominal distension, anorexia and nausea. Often, the patients have significant abdominal distension and parietal palpable tumour mass. The most common complication is intestinal obstruction that is accompanied by high morbidity and mortality (1,2,7,8).

Ultrasonography and computed tomography are useful for preoperative diagnosis. The bowel movement, extrinsec compression and intraperitoneal septated cystic images can be observed. However, in most cases, the diagnosis is established intraoperatively during a laparotomy for another pretended pathology and confirmed by the histopathological examination (2,9,10,11).

In the patients with a history of open surgeries or other pre-existing parietal defects, adenomatous epithelial cells can accumulate in the hernial sac, leading to the presence of gelatinous fluid in the intraparietal area, situation involving differential diagnosis with incisional hernia (12).

Nevertheless, there is no consensus on the treatment, most authors considering as first intention surgery, in order to reduce tumour size, accompanied by intraperitoneal chemotherapy (2,4,10,20. Other options of adjuvant treatment are systemic chemotherapy, radiation therapy and intraoperative photodynamic therapy. Disease progression, regardless of the treatment applied, is fatal, the disease being considered a malignant pathology, having a recurrence of 76% at 12 years, recurrence that leads to intestinal obstruction or intestinal fistulas that may cause the patient's death (19,20,21,22).

The results of the surgery are not promising but remain the only option for a patient with a low quality of life.

We present the case of a female patient, aged 56 years, hospitalized in the Surgery III Clinic of the Emergency University Hospital of Bucharest, for an incisional hernia, aspect secondary to intraparietal dissemination of pseudomyxoma peritonei.

Case report

We present the case of a 56 year-old female patient, subjected to cholecystectomy, who came to our department because of the presence of a tumor-like mass in the supra- and sub-umbilical median area, having an irreducible volume, accompanied by significant abdominal pain and altered bowel movement, suggestive for an incarcerated incisional hernia. The history of the patient indicates that she has undergone surgery in 2007, during which left adnexectomy for left ovarian cystadenoma was performed. The usual laboratory investigations did not reveal other pathological aspects.

Under general anaesthesia with endotracheal intubation the surgery is performed, detecting a parietal tumour mass having gelatinous, mucinous content, relatively well delimited from the adjacent adipose tissue, limited in the posterior area by the aponeurotic plane, no parietal defect being identified at this level.

The entry into the peritoneal cavity highlights the same mucilaginous appearance, spread throughout the cavity, the macroscopic appearance raising the suspicion of pseudomyxoma peritonei. The extemporaneous histopathological result after the peritoneal biopsy confirms the diagnosis of pseudomyxoma. The patient is subjected to appendectomy, total hysterectomy with right adnexectomy, omentectomy, accompanied by abundant peritoneal lavage.

The postoperative evolution was simple, without complications, the patient being discharged on the tenth day after the surgery, being evaluated, afterwards, in the oncology department.

The histopathological result establishes the diagnosis of borderline mucinous appendicular cystadenoma, pseudomyxoma peritonei with the tegument hypodermis and epiploic mature adipose tissue invaded by mucus. The parietal metastasis probably resulted following the parietal contamination, secondary to the adnexectomy in 2007. (Figs. 1, 2, 3)

The patient returns because of the recurrence of the disease and subocclusive phenomena, after a period of about two years, during which she did not follow any treatment, but had an acceptable quality of life. The abdominal CT revealed expansive intraperitoneal processes with fluid content, which occupies the entire left flank, displacing the parenchymal organs and the bowel, starting from the subdiaphragmal area

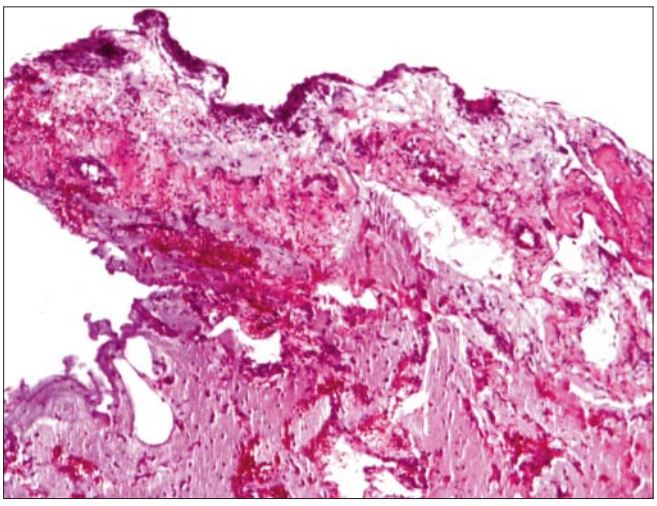

Figure 1. *Peritoneum (the mesothelial limit is up, from left to right) with underlying loose connective tissue infiltrated by mucinous structures with lacunar aspect (on the left side predominantly) (Col. HE, ob. X40)*

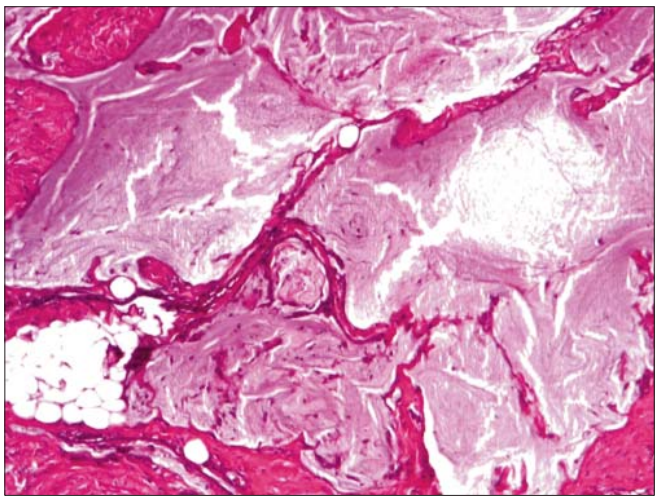

Figure 2. *Peritoneum (top) with mucinous underlying infiltration and reactive, nonspecific aspects: a concentric lymphoid inflammation (right) and diffuse hyperemia (mostly down the center-right), (Col. HE, ob. X 100)*

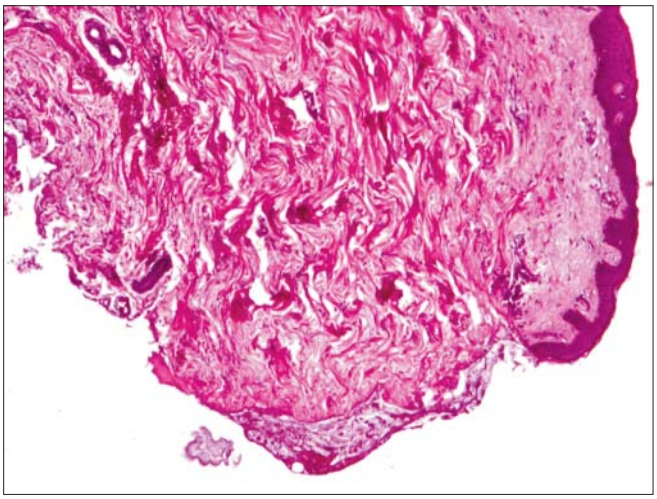

Figure 3. *Peritoneal mucinosis, localized parietal, which exceeded the hypodermis and penetrated into the dermis (top left), (col. HE, ob. X 100)*

up to the pelvis, with a maximum axial diameter in the subdiafragmal area of 15 cm and similar appearance in the abdominal wall along the midline with diameters of 6.5/4 cm representing parietal recurrence. (Fig. 4)

The patient underwent a new surgery during which a massive gelatinous intraperitoneal collection was identified, occupying the entire left flank, starting from the subdiafragmal area up to the pelvis. The content is evacuated, with abundant lavage of the peritoneal cavity. Postoperative evolution with no incidents.

The patient is taken over by the oncology department and starts chemotherapy with Xeloda (Capecitabine) 2500 mg / day in 14-day sessions, repeated every 21 days.

The evolution one year following the last surgery is favourable, with a good general condition, no clinical complaints. The tomography reveals an expansive process in the left subphrenic area with axial dimensions of 11.5 cm, in the anterior-posterior area, and 13.5 / 8.5 cm in the lower pole, with no signs of progression in the following three months and a satisfactory clinical evolution. (Fig. 5)

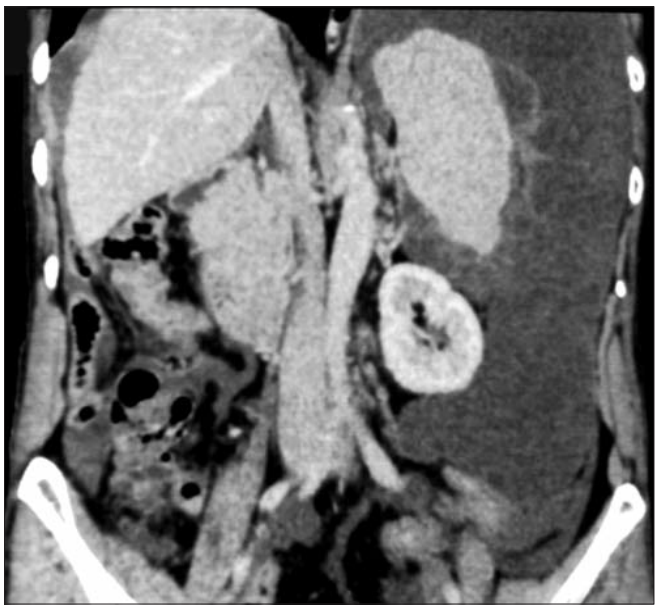

Figure 4. *Expansive process extended to the entire left flank. (tomographic appearance, 2 years after the first surgery)*

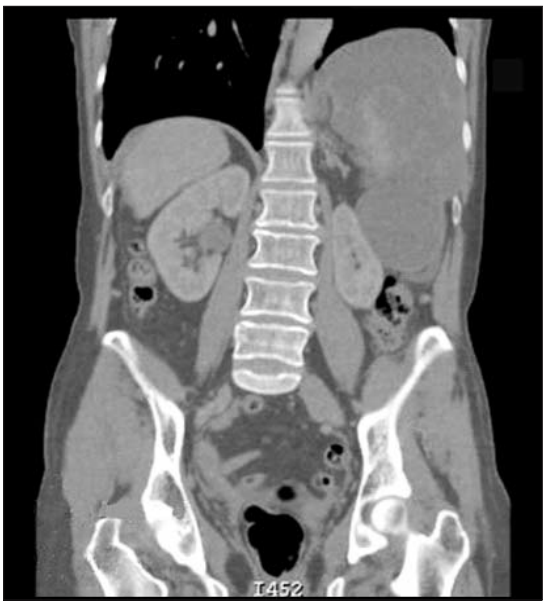

Figure 5. *Appearance tomography 12 months after the last surgery. Left subphrenic expansive process is observed with a maximum diameter of 11.5 cm, polycyclic, poliseptat*

Discussions

Pseudomyxoma peritonei also called "gelatinous disease of the peritoneum" is a rare disease, with no specific clinical manifestations, sometimes even absent for a long time, which affects mostly females with a mean age of approximately 53 years (19).

Recent immunohistochemistry and molecular genetics studies demonstrated both the ovarian and appendiceal etiology involvement in the determinism of this pathology (13,14,15,16).

The clinical manifestation of this disease remains non-specific, establishing a diagnosis which still represents a challenge for the clinician. This feature makes the presentation in a medical department to be late, in advanced stages (18,19). The painless nature of this disease makes the chances of curative treatment to be exceeded, patients generally presenting in advanced stages of disease with massive intraperitoneal dissemination, significant adhesions, partial or complete intestinal occlusions.

Therefore, not having a specific symptomatology, it is difficult to determine a diagnosis of certainty in the early stages, pseudomyxoma peritonei often remaining an incidental intraoperative discovery. Laparotomy thus remains the main way of diagnosis, subsequently confirmed by the histopathological examination of the mass removed.

Conclusions

The use of cisplatin associated with aggressive surgical cytoreduction results in a good long-term evolution. Establishing a diagnosis in this case was a challenge, the slow and nonspecific progress of the disease leading to a difficult differential diagnosis. The patient presented with the signs of an irreducible incarcerated incisional hernia, the diagnosis of certainty being established intraoperatively and subsequently confirmed by the histopathology exam.

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