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

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Rezumat

Pseudomixoma peritonei, entitate rară dificil de diagnosticat și 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. Însăminarea intraparietală a celulelor mucinoase a facilitat dezvoltarea la acest nivel a unei mase gelatinoase ce a ridicat probleme de diagnostic diferențial cu hernia incizională, încercată. Semnele de diagnostic clinic și paraclinic preoperatorii le considerăm insidioase și nespecifice pentru stabilirea diagnosticului de certitudine. Tomografia computerizată abdominală a evidențiat prezența unei colecții intraperitoneale massive, 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 dreaptă și appendicectomie. 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 și nespecifică a afecțiunii conduciind la un diagnostic diferențial dificil.

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

Abstract

The authors present the case of a 56 year-old patient diagnosed with pseudomixoma 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: pseudomixoma peritonei, mucinous dissemination, exploratory laparotomy, incisional hernia
The histopathological result establishes the diagnosis of borderline mucinous appendicular cystadenoma, pseudomyxoma peritonei with the pedunculated hypodermis and epiploic mature adipose tissue invaded by mucin. 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 subdiaphragmatic area.
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)
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 presentations 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.

References

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