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Letter to the Editor

PERITONEAL PSEUDOMYXOMA: CYTOREDUCTIVE OPERATION AND INTRAPERITONEAL CHEMO-HYPERTHERMIA - REPORT OF TWO CASES

Pseudomixoma peritoneal: operação citorredutora e quimio-hipertermia intraperitoneal - relato de dois casos

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INTRODUCTION

Peritoneal pseudomyxoma (PMP) can be defined as carcinoma of low grade malignancy that is characterized by the phenomenon of tumor redistribution involving large amounts of extracellular mucin[3,5]. Primary tumors are usually in the appendix or ovary, and there may be involvement of other organs[3,10]. Is a slow progressive disease, characterized by accumulation of mucus within the abdomen and pelvis[8].

Although this tumor is not considered biologically aggressive, because they do not metastasize via hematogenous or lymphatic as gastrointestinal adenocarcinomas, it results in a deadly process.The space inside the abdomen and pelvis becomes replaced by mucinous tumor. Therefore, it often results in death unless it is successfully treated[8].

The primary tumor rarely causes symptoms. When originating from the appendix arises within an adenoma, and its progressive growth, produces obstruction and distention caused by mucus production of the mucinous tumor[8]. With the rupture of the appendix happens a slow leak of mucus containing epithelial cells adenoma to the peritoneal cavity, followed by its implantation.

CASE REPORT

Case 1

Woman with 42 years reported that during last eight years is complaining of abdominal obesity. Ascites was found by transvaginal ultrasound. Six years ago, she did laboratory and radiological exams with non-specific results. Laparoscopy was performed with biopsies. It was found ovarian tissue with fibrous thickening without signs of malignancy, chronic nonspecific peritonitis and negative for AFB. Over the years several abdominal ultrasound had very similar reports, with large amount of free fluid with debris in the pelvic region. In 2009, was performed computed tomography of the abdomen and pelvis with oral and intravenous contrast showing blurring of fat of the greater omentum, presence of free fluid and retrocecal cystic mass. She underwent laparotomy for diagnosis. Carcinomatosis was found and the presence of large amounts of mucinous fluid in the cavity. Left oophorectomy was performed with appendectomy and sent for pathology and immunohistochemistry. It was confirmed mucinous cystadenoma and peritoneal pseudomyxoma, undergoing to cytoreductive operation and chemo-hyperthermia.

Case 2

Man of 45 years was admitted to the emergency department with clinically suspected acute abdominal inflammation for acute appendicitis. He underwent appendectomy that revealed the presence of large amounts of mucinous fluid in the cavity. In pathology test and immunohistochemistry was considered adenoma of the appendix with the presence of mucin and peritoneal pseudomyxoma. He underwent cytoreductive operation and chemo-hyperthermia by the same technique presented in case 1, 20 days after diagnosis. Additionally in both cases took place right colectomy, splenectomy, omentectomy, cholecystectomy, hysterectomy, right salpigooforectomy (case 1), bilateral diaphragmatic peritonectomy. After cytoreduction, was performed hyperthermia, chemotherapy perfusion with closed intraabdominal technique with mitomycin C at a dose of 35 mg / m2 associated with dialysis solution heated at 42° C. The total operation time was 11 hours without intraoperative complications. The patient was discharged on 8th day.

DISCUSSION

It was believed that the majority of cases of peritoneal pseudomyxoma were from ovarian tumors. This belief has changed recently after immunohistochemical and molecular genetic studies show that a large proportion of these tumors are secondary tumors of the appendix[11]. Peritoneal pseudomyxoma is the implantation of mucinous epithelium in the peritoneal cavity following rupture of a mucocele of the appendix[6].

Adenomucinous peritoneal cells accumulate in specific abdominal and pelvic sites. The most important factors that determine the location of these cells are the absorption of peritoneal fluid and gravity. Voluminous deposits are often found in the greater omentum, lesser omentum and below the right hemidiaphragm site, as well as rectovaginal fornix, right retrohepatic space and others making these sites filled with mucoid tumor mass[8].

Despite various therapeutic modalities, it remains difficult to cure, with survival rates at five and ten years between 75% and 60% respectively[12]. Recurrent mucinous ascites and intestinal obstruction are associated with higher morbidity[1].

The current treatment remains surgical resection of the lesion. In fact, the current treatment strategy includes cytoreductive operation combined with chemo-hyperthermia. Based on the operation Sugarbacker - the peritonectomy - Deraco et al.[2] demonstrated the cytoreduction associated with chemo-hyperthermia with mitomycin C in 42 °C allowed complete removal of the tumor, providing better local control and better survival[15]. Mitomycin C remains confined in the peritoneal cavity for a long period, being more effective in controlling the disease site[9].

The intraoperative intraperitoneal chemotherapy allows the administration of higher drug concentration with less nephrotoxicity and better distribution of the chemotherapy into the peritoneal cavity. Hyperthermia is also beneficial allowing easier drug penetration into tissues[9].

CONCLUSION

A better understanding of the natural history of this disease has allowed to develop better treatment plans with improved patient survival.

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