Abdominal cocoon: a challenging condition

Cocoon abdominal: uma condição desafiadora

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Abstract

Recent articles about congenital peritoneal encapsulation, acquired encapsulating peritonitis and abdominal cocoon are commented. The authors reviewed and discussed these very uncommon etiologies of intestinal obstruction. Additionally, the main features of the two Brazilian case reports of patients with abdominal cocoon and hemorrhagic ascites due to peritoneal involvement by endometriosis are emphasized. Congenital and acquired peritoneal encapsulations are rare conditions. Novel findings about physiology and pathological mechanisms of these conditions are also highlighted.

Key words: Abdominal cocoon; endometriosis, peritoneal encapsulation

Resumo

Artigos recentes sobre encapsulamento peritoneal congênito, peritonite encapsulada adquirida e casulo abdominal são comentados. Os autores revisaram e discutiram as etiologias incomuns como causas de obstrução intestinal. Além disso, destacam-se as principais características dos dois relatos de casos brasileiros de pacientes com casulo abdominal e ascite hemorrágica por envolvimento peritoneal por endometriose. Encapsulamentos peritoneais congênitos e adquiridos são condições raras. Novos achados sobre os mecanismos fisiopatológicos dessas condições também são destacados.

Palavras-chave: Cocoon abdominal; endometriose; encapsulamento peritoneal

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2238-5339 © Rev Med Saúde Brasília 2020; 1(1):133-136

To the Editor,

We read with special interest recent articles about congenital peritoneal encapsulation (CPE) [1], in addition to the acquired encapsulating peritonitis (EP) and abdominal cocoon [2]. The authors reviewed and discussed these very uncommon etiologies of intestinal obstruction. Worthy of note, reports of these conditions have been scarcely described in the whole world; approximately 45 cases of CPE [1] and 5 cases of abdominal cocoon due to endometriosis[,2,3].

Dave *et al*. presented a novel classification for the encapsulated intestinal entities, with the aim of distinguish the exceeding rare congenital condition from other causes of bowel encapsulation as EP and abdominal cocoon, that they call "fibrotic peritoneal encapsulation". The authors summarized the main characteristics of the 45 already described cases of CPE, and in the majority of cases the diagnosis was established by laparotomy with biopsy study [1]. This disorder may origin from a peritoneal adhesion between the linings of the physiological umbilical hernia and the duodenum, further evolving to constitute an extra-peritoneal sac [1]. The CPE has a male predominance of 5:3 and the mean age of diagnosis is 40.8 years [1], and histomorphological patterns allow differentiating it from the clinically similar conditions [1-3]. The diagnosis is usually delayed and may be incidental in surgery for other cause or autopsy. The more specific changes are fixed asymmetrical distention and consistence on palpation [1]. Notwithstanding, the main abdominal manifestations are non-specific: sudden or chronic pain (66%), periumbilical tenderness (58%), distension (40%), and rebound phenomenon (27%) [1]. Imaging studies are useful diagnostic tools and aid in the evaluation of severe complications. The patients with CEP often have very good prognosis after the prompt resection of the sac [1].

Magalhães *et al*. reviewed 42 cases previously published of EP and "frozen" abdomen plus ascites; and described a 28-year-old patient with EP in association with endometriosis [2]. The ages of the women ranged between 20 and 47 years, most of them African descents, with abdominal distention and hemorrhagic ascites (up to 7 L) without malignant cells. In 59.5% of them there was at least one component of the dysmenorrhea, dyspareunia and infertility triad. Other symptoms were abdominal pain, tenderness, palpable mass, dyspnea, and weight loss. Worthy of note were the elevated levels of CA-125, a biomarker for ovarian malignancy [2]. The authors reported

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the second Brazilian and fifth woman in the whole world with confirmed diagnosis of voluminous hemorrhagic ascites coexistent with EP or abdominal cocoon [2,3]. Her clinical manifestations were progressive abdominal pain, cyclic dyspareunia, chest pain and dyspnea with five years of duration. The imaging studies revealed cystic-like ascites with a mass effect, and the fluid cytology showed hemorrhagic effusion negative for malignancy. The diagnosis of EP associated with endometriosis was confirmed by laparoscopic biopsies. The patient was successfully treated with goserelin acetate, and remains asymptomatic [2].

In this scenery, one could comment the first Brazilian report of abdominal cocoon and massive hemorrhagic ascites related to endometriosis in a 40-year-old infertile patient [3]. She had antecedent of ovarian endometriosis treated by goserelin acetate eight months before starting with abdominal pain, vomiting, weight loss and a palpable mass in the mesogastrium. Abdominal images, laparotomy, and the histopathologic studies established the diagnoses. Despite of the intensive care support after surgical intervention, she had peritonitis and died [3]. Cases of EP or abdominal cocoon have morbidity and mortality higher than those of CEP.

Additional comment must be about the peritoneal fluid (PF) promoting myofibroblast differentiation of endometrial mesenchymal stem cells (MSCs), as described by Zhang *et al.* [4] Endometrial MSCs may have increased survival and higher proliferative property in the microenvironment of ectopic lesions; the endometriotic PF can modulate the differentiation of endometrial MSCs and proliferation of endometrial non-MSCs [4]. Endometrial epithelial cells also promote fibrosis in endometriosis by epithelial-mesenchymal transition (EMT), because TGF-β and estrogen of the PF may facilitate the mechanisms of EMT in this disorder [4].

The scarce number of reported cases can favor the lack of consensual approaches related to the herein highlighted conditions. However, case studies might enhance the suspicion index of health workers, contributing to decrease late diagnosis. The authors believe that the commented topics can be useful to surgeons, gynecologists, and general practitioners.

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