A rare tumor revealed by abdominal trauma: case presentation

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Abstract
Pseudomyxoma peritonei is a rare and poorly understood form of disease characterized by mucin deposits in the peritoneum. It was first described by Werth in 1884 as peritoneal, serosal and ommental accumulation of mucin within the abdomen and pelvis [1]. Pseudomyxoma peritonei includes a broad range of neoplasms with different patterns of evolution, from benign to borderline or even to malignant lesions [2]. The disease may be asymptomatic until advanced stages. We present the case of a 65-year-old patient who presented for pain in the right hemiabdomen, after a trauma by falling from small height. Abdominal imaging studies oriented the diagnosis to a traumatic disease. At laparotomy, a mucinous tumor attached to the right colon was discovered. The main particularity of the case is that the origin of the pseudomyxoma could not be identified.

Keywords: pseudomyxoma peritonei, mucinous cells, hemicolectomy.

Introduction
Pseudomyxoma peritonei is a rare and poorly understood form of disease characterized by mucin deposits in the peritoneum. It was first described by Werth in 1884 as peritoneal, serosal and ommental accumulation of mucin within the abdomen and pelvis [1]. Pseudomyxoma peritonei includes a broad range of neoplasms with different patterns of evolution, from benign to borderline or even to malignant lesions [2]. The diagnosis of pseudomyxoma implies the presence of mucinous neoplastic cells and mucinous ascites. For the positive diagnosis, epithelial glandular cells need to be identified within the mucin deposits at histopathological exam. The disease is more frequent in women [3]. Usually, pseudomyxoma peritonei arises from ruptured ovarian or appendiceal adenomas or adenocarcinomas, but there are also cases with indeterminate site [4, 5]. Other possible primary sites are the colon, rectum, gallbladder, pancreas, urinary bladder, breast, lungs, urachus [6]. Sugarbaker et al. defined pseudomyxoma as an intestinal mucinous adenocarcinoma grade 1, which arises from a primary adenoma [7]. The patients with pseudomyxoma peritonei usually die by intestinal obstruction, because of advancing disease.

Patient, Methods and Results
A 65-year-old woman, with history of chronic constipation, appendectomy 30 years ago, presented for pain in the right hemiabdomen, after a trauma by falling from small height. At physical examination: the abdomen mobile, right flank painful at palpation, without signs of peritoneal irritation. She has no other abdominal, gynecological or urinary symptoms. Laboratory investigations including full blood count, amylase, liver function tests, urea and electrolytes were all within normal limits. Abdominal ultrasonography revealed a transonic mass at right para-renal level, 15/8 cm, with thick wall, hyper-echoic septum, relatively well defined, with weak Doppler color signal, and free fluid in the pouch of Douglas (Figure 1). No other abdominal abnormalities were noted. Due to the suspicion of right perirenal hematoma/urinoma, abdominal MRI with contrast agent was done, which identified an ovoidal mass, measuring 17 cm cranio-caudal, with mass effect on the right kidney, well defined, with septum inside and containing few tissular masses that captures gadolinium. Superior, the mass comes into contact with the liver and right kidney, anterior with the cecum and ascending colon, and medial with psoas muscle; pelvic ascites 2 cm thickness. Conclusions: retroperitoneal expansive lesion in the right flank, with characters of malignancy (sarcoma?), without signs of local invasiveness (Figure 2). Tumoral markers were negative (carcinoembryonic antigen, CA125, CA19-9, alpha-fetoprotein).

Surgical intervention is decided. On opening the peritoneal cavity, a tumor attached to the right colon, containing mucinous fluid, with 2–3 hole punching, was seen, along with mucinous ascites (jelly-like) (Figure 3). On exploration, the uterus and ovaries were found to be normal. A right hemicolectomy and peritoneal washout were performed. Postoperative recovery was uneventful. The microscopic examination disclosed tissular fragments of a cystic formation, having mucinous content, with a fibros-connective tissue wall of variable thickness. Some areas presented a columnar epithelium, uni- or bi-stratified, secreting mucus, having the nuclei mostly basally located and with minimal atypia, disposed as
glandular or papillar structures. Large areas of cystic appearance were filled with mucinous content dissecting the connective fibers (Figure 4). The epithelial lining was positive for CK20 (Figure 5), CDX2 (Figure 6), CEA (Figure 7), and weakly positive for MUC2 (Figure 8).

The proliferation rate was relatively high – MIB-1 labeling index was 20% (Figure 9). The overall picture suggested a tumor proliferation characterized as pseudo-myxoma peritonei, with uncertain origin, most probably appendicular.
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Figure 8 – MUC2 is weakly expressed in rare epithelia, mostly at the apical pole. Immunohistochemistry, ×200.

Figure 9 – Numerous tumor cells are proliferating, as shown by the high Ki67 labeling index. Immunohistochemistry, ×200.

The postsurgery oncological consult did not recommend any chemotherapy or radiotherapy. The patient was followed by abdominal ultrasound every two months, without any recurrence for two years.

Discussion

Pseudomyxoma peritonei is often an asymptomatic disease, which preferentially affects women with an average age of 53 years [3]. For long time, it was considered that pseudomyxoma originate from ovarian tumors. However, there are studies showing that many of these tumors are secondary to appendiceal tumors in both men and women [8].

Pseudomyxoma peritonei may be asymptomatic until advanced disease, making the exact diagnosis in early stages very difficult. Laboratory studies are of little help for diagnosis. In most of the cases, the suspicion is raised at imaging studies of the abdomen. Abdominal ultra-onography shows echogenic masses with multiple septations and ascites. Abdominal computed tomography may identify the “scalloping effect” on the surface of the visceral organs, resulting from compression by the viscous mucinous secretions and the organizing fibrosis. Magnetic resonance imaging may be more useful than computed tomography in assessing the visceral invasion by mucinous tumors. However, in most of the cases, the tumor is an unexpected finding at exploratory laparoscopy or laparotomy, as in our patient, the final diagnosis being confirmed by histopathology.

At histopathology, this neoplasm contains solid growth with conspicuous epithelial cell atypia and stratification, loss of gland architecture and necrosis, similar to colonic cancer in appearance [9]. The well-differentiated columnar epithelial cells producing mucus usually have minimal nuclear features of malignancy [10].

The optimal treatment of pseudomyxoma peritonei is not well known. In all patients, surgical debulking of the tumor should be done. In 1996, Sugarbaker et al. stated that improvements of surgical techniques, to increase total clearance of the tumor, and chemotherapy are required to control the residual disease on the peritoneal surfaces [11]. Repeated cytoreductive surgical debulking procedures are common due to recurrence of the disease. The current treatment strategy consists of cytoreductive surgery associated with drainage of the mucus and intraoperative hyperthermic intraperitoneal chemotherapy, aiming to destroy small residual mucinous tumor nodules. Commonly, at laparotomy a right hemicolectomy is performed.

In the majority of cases, mortality is related to intestinal obstruction caused by advancing abdominal disease. Five and 10-year survival rates are thought to be around 50% and 20% respectively [1].

Conclusions

The peculiarity of our case is the incidental diagnosis in an asymptomatic patient, in the context of abdominal trauma, which initially oriented the diagnosis in the direction of a traumatic pathology. Another particularity of the case is that the origin of the pseudomyxoma could not be identified. The histopathological and immunohistochemical report indicated the appendix as the possible origin of the tumor. However, our patient had appendectomy 30 years ago, and the imaging studies of the abdomen did not visualize any ovarian, gallbladder, pancreatic, and colonic or rectal pathology.

References


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