VESICO-APPENDICEAL FISTULA IN A MUCINOUS ADENOCARCINOMA OF THE APPENDIX

Fístula vésico-apendicular em adenocarcinoma mucinoso do apêndice

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ABSTRACT - Background - A rare case of vesicoappendiceal fistula secondary to mucinous adenocarcinoma of the appendix is presented. Case report - A 62-year-old man with a one year history of recurrent urinary tract infections. After two months he developed pneumaturia and fecaluria. An abdominal and pelvic computed tomography demonstrated a trans-mural mass in the posterior wall of the bladder with a vesicoenteric fistula leading to the terminal ileum. Laparotomy revealed a tumor arising from the appendix contiguous with the bladder posterior wall. The bladder was opened and a large fistula and tumor on the posterior bladder wall near the trigone was identified. Frozen pathological analysis showed a mucinous adenocarcinoma. En-bloc right hemicolecetomy and partial cystectomy, preserving bladder trigone was performed. After manipulating the tumor, grossly leakage of mucinous materials occurred into the pelvic cavity. A peritoneal washing with a mytomicin solution at 42º C was then performed, to prevent peritoneal seeding. The patient had a prolonged postoperative ileus and was discharged at the 15th day. Five months after the procedure the patient was receiving chemotherapy with 5-fluoracil and leucovorin and there was no signs of recurrent disease. Conclusion - The presentation with vesico-appendiceal fistula is extremely rare with only a few cases reported in the literature. Knowledge of different types of neoplasm and appropriate treatment allows the surgeon to provide patients optimal care referring to specialized centers whenever appropriate.

vic cavity. Than it was performed a peritoneal washing with a mytomicin solution at 42º C, to prevent peritoneal seeding. Pathological examination demonstrated an exophitic, gelatinous mass with 17 × 12 cm in diameter and vesicoenterical fistula formation (Figure 3). It was classified as a well differentiated mucinous adenocarcinoma of the appendix (Figure 4). Surgical margins were tumor-free and angiolymphatic invasion was not observed. Fifteen dissected lymph nodes were not involved by the tumor. Final pathological diagnosis was a pT4 pN0 pM0 appendiceal mucinous adenocarcinoma.

The patient had a prolonged postoperative ileus and received periferal parenteral nutrition from the 5th to the 11th day when adequate oral intake was achieved. At the 15th day he was discharged. After two months, it was removed double loop catheters and cystoscopy did not reveal any residual lesion. At five months after the procedure, he was receiving chemotherapy with 5-fluoracil and leucovorin, with no image signs of peritoneal and the CEA remains in normal levels.

**DISCUSSION**

Vesicoenteric fistula is usually a complication of an inflammatory or neoplastic process. Common causes include diverticulitis (65-75%), malignant disease (10-15%) and Crohn's disease (5-6%). Presenting symptoms are pneumaturia (52-77%), fecaluria (36-51%) and urinary tract infection symptoms (44-45%). Investigation modalities are usually poor, with abdominal CT being the most useful imaging modality. Although cystoscopy can be performed, it is diagnostic in only 30-40% of cases.

Appendiceal carcinomas are rare with an incidence of 0.12 cases per 1,000,000 people per year. It is estimated that appendiceal cancer is found in 1% of all appendectomy specimens.

Patients with adenocarcinoma of the appendix are usually diagnosed with appendicitis, a right lower quadrant abscess, or a tumour mass. Rarely the tumor can invade the abdominal wall with an enterocutaneous fistula or the urinary bladder with an enterovesical fistula, as shown in this case. International literature report only nine cases of
vesico-appendiceal fistulas derived from mucinous appendiceal adenocarcinomas until present data\textsuperscript{1,2,3,4,5}. This type of tumor usually perforates before diagnosis and may spread to the peritoneal cavity, producing mucinous intraperitoneal ascites, resulting in the pseudomyxoma peritonei\textsuperscript{7}.

The initial assessment of the incidental appendiceal tumor includes assessment of tumor size, involvement of the base of the appendix or the mesoappendix and its perforation\textsuperscript{8}.

If the tumour is confined to the appendix, smaller than 2 cm, without evidence of mesoappendiceal or basal involvement, appendectomy is the appropriate treatment. An emergency cryostat sectioning should be done and if an invasive non-mucinous adenocarcinoma of the appendix is shown a right hemicolectomy can double the survival achieved with routine appendicectomy\textsuperscript{9}. In tumours larger than 2 cm or with invasion of the base or mesoappendix, a right hemicolectomy should be performed\textsuperscript{7}.

If the appendix has ruptured just before removal or during the operation, it is important to remove all free mucin and perform meticulous peritoneal toilet. After resection, and with the abdomen opened, the peritoneal cavity could be washed with warm (41-5ºC) mitomycin solution to avoid peritoneal seeding. Such patients and those who present with a perforated mucinous neoplasm, without evidence of any extra-appendiceal spread, are at risk of developing pseudomyxoma peritonei and should be followed up carefully. CT of the abdomen and pelvis and tumour markers (CEA, CA 125 and CA 19.9), provide baseline measurements\textsuperscript{8}. At the present case the leakage of peritoneal cavity led to perform the warm mitomycin solution wash. Follow up doesn’t show any sign of recurrence, and the patient is been treated with systemic chemotherapy.

Perforated mucinous neoplasm of the appendix with pseudomyxoma peritonei syndrome is best treated by complete removal of tumor (peritoneectomy) and hyperthermic intraperitoneal chemotherapy [mitomycin solution (41-5ºC) during surgery and 5-fluorourcil for four to five days after surgery]\textsuperscript{9}.

Right hemicolectomy, without intraperitoneal chemotherapy or complete removal of the tumor, at an initial procedure for a perforated mucinous adenocarcinoma with peritoneal involvement, results in no survival benefit. In this cases the peritoneal cavity should be thoroughly washed out, and the patient should be referred to an appropriate specialized center for definitive treatment\textsuperscript{9,10,12}.

CONCLUSION

Appendiceal neoplasms are uncommon and consists in an heterogeneous group of pathologies. Many present as appendicitis, but some are encountered as incidental findings at laparoscopy or laparotomy. The presentation with vesico-appendiceal fistula is extremely rare with only a few cases reported in the literature. Knowledge of the different types of neoplasms and appropriate treatment allows the surgeon to provide patients optimal care.