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Letters to the Editor

Mucinous appendiceal neoplasms. Do we all speak the same language?

Key words: Mucinous tumors. Mucocele. Appendix.

Dear Editor,

Appendiceal tumors are unusual entities representing around 1% of the appendicectomies, and 0.5% of the gastrointestinal neoplasms (1). The mucinous adenoma is one of the most prevalent primary appendicular neoplasms, more frequent in middle-aged women. The diagnosis is usually a casual finding in the anatomopathologic study after an appendicectomy performed in a patient with clinical suspect of acute appendicitis (1-3).

Case report

A 53 year-old male without medical past history was admitted to the Emergency Department with a 24 hour low abdominal pain associated to fever (39 °C). In physical examination, pain was focused in the right iliac fossa associated to positive Blumberg's sign. Blood test did not show leucocytosis or neutrophilia (8.8 x 10^9/L white blood cells, 70% neutrophils) and the abdominal radiography showed dilated small bowel. We requested an abdominal computed tomography that evidenced a small fluid collection (12 x 9 mm) in the top of the appendix without free intraabdominal liquid, pneumoperitoneum or retroperitoneal or pelvic adenopathies (Fig. 1).

With acute appendicitis as the first suspected diagnosis, an open appendicectomy was performed, and a gross perforated gangrenous appendicitis with intraabdominal liquid was found. Postoperatory course was uneventful. The anatomopathologic study showed a low-grade appendiceal mucinous tumor with mucin extravasation, but without extra-appendicular epithelium; tumor-free resection margin.

Discussion

The classification of the mucinous appendiceal tumors is controversial and sometimes confusing because of the lack of consensus in diagnostic terms commonly used (2,4,5). The term *mucocele* is a macroscopic concept, which refers to a dilatation of the appendiceal lumen, with or without obstruction, secondary to an abnormal accumulation of mucin. It is usually caused by an appendiceal mucinous adenoma (but it may also be associated to

Fig. 1. Abdominal CT with i.v. contrast (coronal view). It shows appendiceal wall thickening with edema (black arrow) and a small fluid collection at the top (arrow).

ovarian, breast or liver neoplasms, or even non-neoplastic processes) (2-5). *Peritoneal pseudomyxoma* is defined as multiple widespread mucinous implants in the abdominal cavity, usually secondary to a spontaneous or intraoperative rupture of a low-grade malignant mucinous appendiceal neoplasm (4,5).

One of the most widely used classification for these tumors divide them into: mucinous adenoma, low-malignant mucinous neoplasm and mucinous adenocarcinoma (4).

Mucinous adenoma represents 63% of all the cases, and consists in a benign process with microscopic moderate atypia confined to the appendix, without extra-appendicular mucin. The curative treatment is an appendicectomy with free resection margin, without recurrence risk (2,4,6). In cases of affected proximal margin, presence of epithelium in the appendicular wall (without clear signs of infiltration) or doubt about the presence of epithelium in the extravasated mucin, the proper term should be *uncertain malignant potential mucinous neoplasm* (4).

Low-grade appendiceal mucinous neoplasm does not show significant cytological differences compared to mucinous adenoma; it presents wall invasion with or without peritoneal implants (no metastases) (4). Possibility of local recurrence is greater if neoplastic epithelium is present in the extra-appendicular mucin, associated with an increased risk of developing diffuse peritoneal disease and a lower survival (4). It is essential to avoid the appendiceal rupture during surgery to prevent the spread of its content and the subsequent development of peritoneal pseudomyxoma (7,8). The treatment of this condition remains controversial; although appendicectomy was classically considered as the definitive treatment, some authors recommend a more aggressive conduct as a right hemicolectomy and the removal of all mucoid implants associated with cytoreduction (4). Some groups advocated for the need of cequectomy or right hemicolectomy only when the appendiceal base is affected (4). Regarding to the specific treatment for peritoneal implants (peritoneal pseudomyxoma), some groups have experience with the application of intraperitoneal chemotherapy associated with cytoreduction (smaller than 2,5mm implants), with improved prognosis in low-grade cytological atypia tumors (9,10).

Finally, *mucinous adenocarcinoma* presents high-grade atypia with invasion and destruction of the appendiceal wall and risk of metastasis, so the treatment needs to be more aggressive. It is recommended to perform a right hemicolectomy with limphadenectomy; surgical cytoreduction is not recommended because it does not improve prognosis (4,9,10).

The most important factors to consider in a mucinous appendicular lesion are: the presence and/or extension of the extra-appendicular mucin, the presence of epithelium in the mucin or any other peritoneal location, and the cytological atypia, related to the appendix, the mucinous epithelium or the peritoneal implants. All these variables could influence the prognosis (2,5).

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