

Letters to the Editor

Surgical management of appendiceal adenocarcinoid. Case report and literature review

Key words: Adenocarcinoid tumor. Appendiceal neoplasms. Appendicitis.

Dear Editor,

Adenocarcinoid tumor of the appendix is a rare entity characterized by the presence of a double component (neuroendocrine and glandular). It originates in the neuroendocrine cells of the appendicular mucosa. A preoperative diagnosis of a primary appendiceal tumor is uncommon and more so one suggesting an adenocarcinoid pathology.

Case report

A 57-year-old woman consulted for nausea and abdominal pain in the right hemiabdomen. The patient had no fever or any other clinical data. Ultrasonography and CT informed of a perforated acute appendicitis contained by an inflammatory mass. An emergency laparoscopic appendectomy was performed, revealing a gangrenous acute appendicitis. The pathological report informed of infiltration of the appendicular wall by a primary adenocarcinoid tumor (Figs. 1 and 2). Under this diagnosis a second surgery was performed to extend the resection with a right hemicolectomy. There were no tumor data in the anatomopathological specimen.

Discussion

Appendiceal tumor is an extremely rare entity, accounting for 0.5% of gastrointestinal tract tumors. Garin conducted a review (1) of 7,970 cases of acute appendicitis and found 74 appendiceal tumors (0.9%), including metastatic tumors, and only 2% were adenocarcinoid appendiceal tumors (1).

The mean age at presentation of adenocarcinoid is usually around 50 years (unlike typical carcinoid tumors, which appear around 35- years) with predominance in women (2). The most common presentation is an acute appendicitis although it can also appear as abdominal pain with a palpable abdominal mass. Most cases with this type of presentation tend to be wrongly diagnosed as primary ovarian tumors. Adenocarcinoid has not been reported to cause carcinoid syndrome (3,4).

They are characterized by the presence of two well differentiated histological components: neuroendocrine and glandular (5-7).

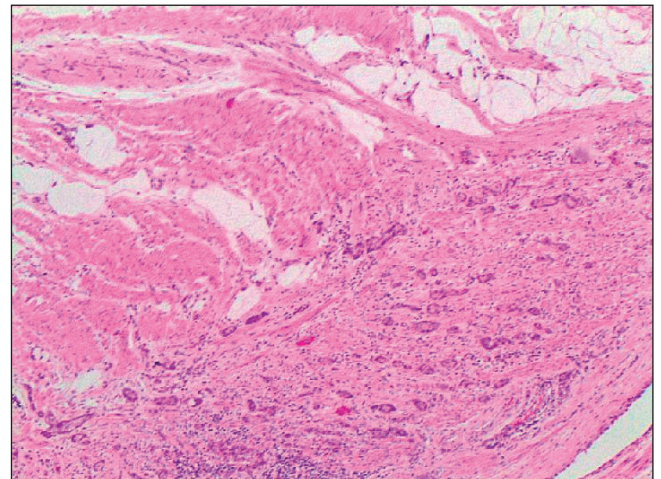


Fig. 1. Histological image (haematoxylin-eosin x20). Infiltration of the appendicular wall by neoplastic glands from the submucosa to the serosa.

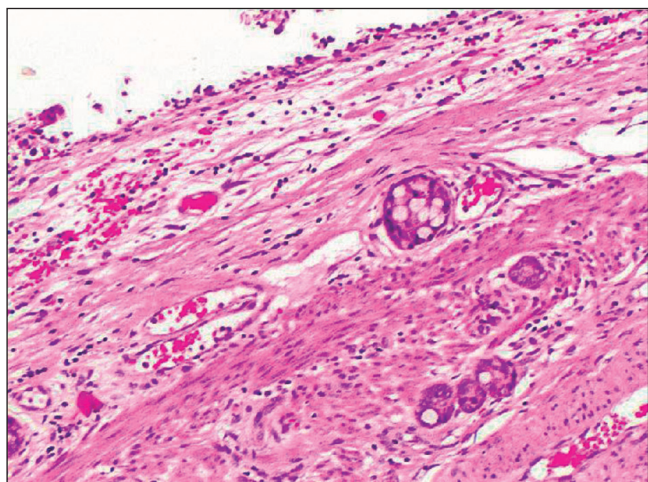


Fig. 2. Histological image (H-E x200). Goblet cell clusters.

They are positive, therefore, for endocrine cell staining such as chromogranin A and synaptophysin, as well as for mucin. Unlike carcinoid tumors, the S100 marker is characteristically negative. Despite arising in mucosal crypt cells, the adenocarcinoid tumor characteristically maintains a normal mucosa with submucosa and muscularis propria extension (8).

Tumors are classified according to histological criteria into three groups: group A, typical adenocarcinoid, with well-defined goblet cells arranged in clusters or cohesive linear pattern, with minimal cytologic atypia; group B, adenocarcinoid with signet ring cells arranged in irregular large clusters, with significant cytologic atypia; and group C, poorly differentiated adenocarcinoid with not otherwise distinguishable from a poorly differentiated adenocarcinoma in which there is minimum focal evidence of goblet cells or a confluence of signet ring cells (2).

Due to the infrequency of this pathology and, therefore, to the lack of experience in its management, there is an ongoing debate regarding the correct treatment option (3,6-8). However, the MSKCC pathological classification recommends an appendectomy for type A (typical adenocarcinoid), a right hemicolectomy for type B (signet ring), and a hemicolectomy, hysterectomy and bilateral ovariectomy for type C (poorly differentiated) (2,9).

Localized adenocarcinoid tumors occurred equally in males and females, while disseminated tumors have been seen in females mainly, mostly in the form of a Krukenberg's tumor or peritoneal carcinomatosis, with a very poor prognosis (9).

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