Mucocele and intussusception of the appendix are very rare, found respectively in less than 0.2% and 0.01% of all appendectomies. We report a case of a mucinous cystadenoma of the appendix causing appendiceal intussusception in a patient with anomalous right colonic fixation, ascites and a right ovarian cyst.

**Case report**

A 36-year-old woman was admitted to our ward complaining of colicky pain located in the central abdomen, radiating to the flanks and associated with nausea. The pain had been intermittent for 6 days and almost continuous over the previous 24 hours. She...
also referred a 3 kg weight loss in a week. Physical examination revealed a tender, movable mass, measuring 5 cm in diameter, with undefined limits, located in the mesogastrium. Laboratory data were normal.

US scan revealed thickened double intestinal walls like a concentric ring sign suggestive of invagination corresponding to the palpable mass, and a cystic hypoechoic mass, measuring 3 cm in diameter at the tip of the intussuscepting complex, apparently located in the last ileal loop (Fig. 1). A right ovarian hypo-anechoic cyst and a small amount of fluid collection in Morrison’s space and the Douglas’ recess was also found.

On the following day the patient referred absence of pain, the palpable mass was reduced in volume and showed no tenderness. In the following hours she intermittently complained of the same clinical presentation shown at admission. CT scan revealed a concentric ring sign suggestive of right colonic intussusception; at the tip of the appendiceal invagination complex was a cystic, encapsulated mass (Fig. 2).

It also confirmed the suspicion of a cystic neoplasm of the right ovary. Synchronicity of appendiceal mucocele, ascites and the ovarian mass suggested an associated mucinous tumour of the right ovary with pseudomyxoma peritonei. At laparotomy the appendix was inflamed with a palpable, round-shaped mass measuring 3 cm in diameter located at the base. The caecum and ascending colon showed abnormal fixation to the parietal peritoneum.

Appendiceal mucocele is a very rare clinical condition. Associated ascites and an ovarian mass could suggest synchronous ovarian cystadenocarcinoma with pseudomyxoma peritonei. We describe the case of a 36-year-old female with a mucinous cystadenoma of the appendix causing intussusception, diagnosed by CT but not by US scan, since the associated anomalous fixation of the caecum was misleading in defining the precise anatomical site. Although the CT findings were accurate, the synchronous presence of an ovarian cyst and ascites did not allow us to rule out preoperatively a concurrent cystadenocarcinoma of the ovary with pseudomyxoma peritonei. The appropriate surgical treatment was performed on the basis of intraoperative frozen section examination.

Surgical treatment depends on the nature of the mucocoele: retention forms are effectively treated by appendectomy, while neoplastic conditions require a more extended resection. Treatment of associated ovarian cystadenocarcinoma and pseudomyxoma peritonei includes right colectomy, bilateral ovariectomy and omentectomy. Although a precise preoperative diagnosis of mucocele associated with intussusception of the appendix has been reported as possible, concomitant ascites and ovarian masses, as in the present case, could mimic pseudomyxoma peritonei from concurrent ovarian cystadenocarcinoma. Intraoperative histopathology is required in order to perform the most appropriate treatment.

Key words: Mucinous cystadenoma - Appendiceal mucocele - Intestinal intussusception

Chir Ital 2006; 58, 1: 101-104

Discussion

Intussusception of the appendix is a very rare clinical finding3,4, with an incidence of 0.01% of all appendectomies2. It is associated with mucocele, found in 0.20.3% of surgical specimens from appendectomies1, adenocarcinoma, carcinoid tumour, duplication or mesenteric cyst and lymphoma. Mucocele is described as an obstructive dilatation of the appendiceal lumen filled with mucin. According to Zissin5 it can be related to four histological subtypes: retention cyst, mucosal hyperplasia, cystadenocarcinoma and cystadenoma, the latter accounting for 63% of cases5. Ascites is found in 6% of cases, consisting of a mucinous fluid...
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containing epithelial cells and termed pseudomyxoma peritonei. This can also develop from ovarian cystadenocarcinoma, and in this case an appendiceal mucinous tumour is almost always associated.

The clinical presentation of appendiceal cystadenoma is non-specific, and in most cases is found incidentally at surgery.

Symptoms may include a palpable mass, and chronic or intermittent abdominal pain caused by its intussusception.

When the latter is present, US and CT scan findings are typical, and include the cystic mass at the tip of the invagination complex.

Surgical treatment depends on the nature of the mucocele: retention forms are effectively treated by appendectomy, while neoplastic conditions require a more extended resection.

Treatment of associated ovarian cystadenocarcinoma and pseudomyxoma peritonei includes right colectomy, bilateral ovariectomy and omentectomy. In our case, a precise preoperative diagnosis was not possible.

The association with anomalous colonic fixation was misleading in defining the actual site of the tumour by means of US scan.

CT scan findings were accurate, but since ascites and an ovarian mass were coexistent, we could not exclude concurrent ovarian carcinoma.

Surgical treatment was appropriately performed on the basis of the histopathological examination.

Although precise preoperative diagnoses of appendiceal intussusception caused by mucocele have been reported, the differen-
tial diagnosis with associated pseudomyxoma peritonei and ovarian cystadenocarcinoma may be impossible when concurrent findings such as ascites and ovarian cysts are present.

The diagnosis and most appropriate surgical treatment are based on intraoperative histopathological analysis. Inappropriate surgical under- or over-treatment of the condition in an emergency setting could depend on the unavailability of intraoperative histopathology.

Fig. 4. Histological examination (EE 250x): (↑) cystadenoma of the appendix.

References


