APPENDICEAL MUCOCELE
IN A PATIENT WITH RECURRENT BREAST CANCER
- A CASE REPORT AND LITERATURE REVIEW

ILIJA GUTEŠA, MLADEN STANEC, DANKO V. VRDOLJAK,
IVAN MILAS and IVAN PENAVIĆ

Department of Surgical Oncology, University Hospital for Tumors, Zagreb, Croatia

Summary

Mucocele or cystic distention of the appendix is a rare entity found in only 0.25% of all appendectomies and 8% of all appendicular tumors. We report a case of a 68-year-old female patient in whom asymptomatic mucocele was found at abdominal CT imaging a month after excision of recurrent invasive ductal carcinoma to the right pectoral region, and 26 years after modified radical mastectomy. After adequate preoperative treatment, a right hemicolectomy was performed with the final pathology of mucocele of the appendix.

KEYWORDS: appendiceal mucocele, recurrent breast cancer

INTRODUCTION AND DISCUSSION

Mucocele of the appendix develops from obstruction of the appendiceal lumen, which leads to accumulation of mucinous substance and abnormal luminal dilatation. The incidence of mucocele ranges from 0.2 to 0.3% (1). It occurs more frequently in women than in men (4:1) and in persons over the age of 50 (2).

Classification. Based on the characteristics of their epithelial lining, 4 different entities of mucocele may be distinguished (2, 3, 6, 7). Simple or retention mucoceles result from obstruction of the appendiceal lumen, in the majority of cases by fecoliths. They are characterized by normal epithelium and luminal dilatation up to 1 cm. Mucoceles with hyperplastic epithelium also have mild luminal dilatation. The most common form of mucocele is a mucinous adenoma/cystoadenoma (accounting for 63-84% of the cases) with epithelial villous adenomatous changes and some degree of atypia, showing a luminal distension of up to 6
cm. Malignant mucinous cystoadenomarcinoma occurs in 11 to 20% of the cases. It is characterized by stromal invasion and/or epithelial cells in peritoneal implants, often with severe luminal distension. In case of a spontaneous or iatrogenic rupture of the mucocele, mucous material extravasates into the peritoneal cavity. The material can be acellular, but it can also contain cells showing some degree of dysplasia. Unlike in colorectal carcinoma, lymph node involvement and liver metastases are rarely reported (8). Instead, these cells are disseminated over the peritoneum, often avoiding the surface area of the small bowel. Dissemination and implantation of malignant cells result in a syndrome called pseudomyxoma peritonei (1, 9, 10).

Clinical picture. Mucocele is symptomatic only in 50% of the cases, and the remaining 50% are discovered incidentally during surgery (2, 5, 6, 11). Symptoms may include: abdominal pain, a sensation of an intra-abdominal mass, weight loss, nausea, vomiting and acute appendicitis (11-13). Rupture of a neoplastic mucocele may clinically mimic appendicitis (14). The clinical picture of pseudomyxoma peritonei is even more non-specific. The most common sign is a gradual abdominal enlargement. In men it may present as a newly developed hernia, and in women as an ovarian mass (15).

Diagnostics. Mucocele can be diagnosed by ultrasonography or computed tomography of the abdomen. A characteristic CT finding shows a round, encapsulated cystic mass. Calcifications can be seen in 50% of the patients (12, 16). Enlarged nodules in the cyst wall increase the probability of malignancy, or cystadenocarcinoma (17). On the other hand, mucoceles of less than 2 cm in diameter are rarely malignant (11). Mucoceles larger than 6 cm in diameter are associated with the presence of often perforating cystadenomas and cystadenocarcinomas (in 20% of the cases) (18, 19). Ultrasonography of a mucocele shows an encapsulated cystic lesion, firmly attached to the cecum. The echogenicity varies with the mucous content density. The finding of pathognomonic multiple, onion skin-like circles along the dilated appendix is also possible (20, 21). Ascites is a non-specific CT sign of pseudomyxoma. As pseudomyxoma cells are of poor adherence ability, and often rearranged by peristaltics, cell accumulations are more likely to be found at sites of relative stasis in the abdominal cavity. The most frequent sites are therefore the pouch of Douglas, subphrenic regions, and the liver and splenic surface (22).

Treatment. Recent studies show that an intact mucocele is a benign process usually not resulting in disease progression (10). Rupture of a mucocele and spillage of its contents into the abdominal cavity are associated with a poorer prognosis. Surgical removal of the mucocele therefore requires extra caution. In case epithelial cells are found in the abdominal cavity a diagnosis of pseudomyxoma peritonei is established (24). If a mucocele is visualized during laparoscopy, the use of a bag (19, 4, 25), or conversion to an open surgery is advisable (26). Until recently, right hemicolectomy was a standard procedure in the management of mucinous and other appendiceal neoplasms (23). However, the study by Gonzalez-Moreno et al. showed that a right hemicolectomy does not confer any survival advantage (14). The authors suggest the following indications for right hemicolectomy: (1) necessity to remove the primary tumor or achieve complete cytoreduction, (2) lymph node involvement demonstrated by histopathological examination of the appendiceal or ileocolic lymph nodes, (3) a non-mucinous neoplasm identified by histopathological examination.

The management of pseudomyxoma peritonei requires a combination of a combination of complete surgical cytoreduction and perioperative intraperitoneal chemotherapy. The recommended chemotherapy regime includes mitomycin followed by five days of post-operative fluorouracil (8). Systemic chemotherapy often has a transient response and is recommended only for patients with extensive peritoneal disease and high grade cystadenocarcinoma (8).

CASE REPORT

A 68-year-old female patient was admitted to the Department of Surgical Oncology, University Hospital for Tumors, Zagreb, Croatia due to an exulcerated, cytologically positive tumor in the surgical scar of the right pectoral region 26 years after modified radical mastectomy.

After adequate preoperative workup (including a gynecological examination with transvaginal sonography to detect status post hysterectomy and bilateral adnexectomy, and an unechogenic cystic mass measuring 4 cm in diameter) radical
tumor excision with partial resection of the large right pectoral muscle was performed. Histopathological analysis showed the diagnosis of a partially hormone-dependant, HER-2 negative recurrent invasive ductal carcinoma. The patient was referred for medical oncology consultation that indicated hormonotherapy with Arimidex, 1 tablet once a day to be taken continuously for 5 years. Postoperative CT of the pelvis was performed to detect a relatively sharply defined hypodense zone of 45 mm in the lower right hemiabdomen of the appendiceal region, visibly connected to the large intestine in its cecal region. After application of intravenous contrast, the lesion was not imbibed which led us to consider a cystic mucinous change of the appendix. As this lesion over 2 cm in diameter might be malignant, explorative laparotomy was indicated and performed. Status post cholecystectomy, hysterectomy and bilateral adnexectomy was shown along with a tumor of about 5 cm in diameter in the appendiceal region. Therefore, a right hemicolectomy was performed. The postoperative course was uneventful. Histopathology confirmed the diagnosis of appendiceal mucocele.

CONCLUSION

Although the histopathological diagnosis of mucocele is most commonly benign, in 11 to 20% of the cases the mucocele is actually a malignant mucinous cystadenocarcina that may rupture and develop into pseudomyxoma peritonei associated with a poorer prognosis. Therefore, the treatment of mucocele requires a detailed workup and extreme care while handling mucocele during surgery.

REFERENCES


Author’s address: Ilija Guteša, M.D., Department of Surgical Oncology, University Hospital for Tumors, Illica 197, 10000 Zagreb, Croatia