Mucinous tumors of the appendix presenting as primary tumors of the ovary. Report of two cases

A. Liapis[1], E. Michailidis[2], P. Bakas[3], A. Kondi-Pafiti[4], G. Creatasas[5]

[1]Assist. Prof. in Obstetrics & Gynecology; [2]Trainee in Obstetrics & Gynecology; [3]Res. Fellow in Obstetrics & Gynecology; [4]Ass. Prof. in Histopathology; [5]Prof. in Obstetrics & Gynecology

2nd Department of Obstetrics and Gynecology, University of Athens "Areataeion" Hospital, Athens (Greece)

Summary

Primary tumors of the appendix are rare and most of them are unrecognized preoperatively, presenting as appendicits, pelvic masses or with no typical abdominal pain. Two cases of mucinous tumors of the appendix presenting as primary ovarian tumors are described. It is important for the gynecologist-oncologist to include mucinous tumors of the appendix into the differential diagnosis of any case of mucinous ovarian tumor and peritoneal pseudomyxoma, especially when these tumors are associated with extraovarian disease.

Key words: Mucinous tumors; Appendix; Adenocarcinoma; Peritoneal pseudomyxoma.

Introduction

Metastatic tumors to the ovary are frequently unsuspected by gynecologists. The metastasis often presents as a primary ovarian tumor and may even be the initial manifestation of the patient's cancer [1, 2]. Because primary ovarian epithelial neoplasms typically are cystic when examined grossly, cystic lesions tend to be routinely regarded as primary tumors. Interestingly, metastases to the ovary are frequently cystic or semicystic, even when the primary tumor is solid [1]. Ovarian metastases may result from genital or extra genital primary tumors. Tumors from the gastrointestinal, pancreatocobiliary and urinary tracts are among the most difficult ovarian metastases to diagnose correctly [3].

Primary tumors of the appendix are rare, comprising less than 1% of the intestinal tumors. While these tumors are not very common, they have a tendency for ovarian spread when they disseminate beyond the appendix and most of them are unrecognized preoperatively, presenting as appendicitis, pelvic masses or with no typical abdominal pain [2-4].

We report two cases of primary appendiceal neoplasms that presented as primary ovarian tumors.

Case Report

Case 1

A 75-year-old woman was admitted to our clinic (2nd Department of Obstetrics and Gynecology, University of Athens, Greece) in September 1998. At admission, she complained of a slight lower abdominal pain, meteorism and she had a temperature of 38.8°C. Clinical examination revealed a descended abdomen because of ascites and pelvic examination revealed a tender mass in the left adnexal region with a quite normal sized uterus, while the right adnexal region was not palpable.

An ultrasound scan demonstrated a 6 cm in diameter solid/cystic mass in the left ovarian region while the uterus and the right ovary were found normal in size and morphology. CT scan confirmed the presence of a 5 x 6 cm pelvic mass that seemed to originate from the left adnexal region and free fluid (ascites) was present but no lymph nodes were revealed. The mass was suspected to be a cystadenocarcinoma of the ovary.

An air-contrast (double-contrast) barium enema and proctosigmoidoscopy revealed diverticular disease of sigmoid, with no other pathology, while I.V. pyelography and an abdomenal X-ray were normal.

The measurement of serum CA 125 levels showed a value of 73.8 U/ml and serum CEA levels showed a value of 175 ng/ml. The other hematological and biochemical tests were normal and a chest X-ray was negative.

Some days later the patient underwent laparotomy. During surgery about 450 ml of ascitic fluid were drained off and then submitted for cytological examination. The omentum was totally infiltrated by the tumor. The last part of the ileum, a great part of the jejunum and the ascending colon were also infiltrated by the tumor which appeared to originate from the appendix.

Intraoperative biopsies of the right adnexal region and the omentum were positive for cancer. Total abdominal hysterectomy with bilateral salpingo-oophorectomy, total omentectomy and right hemicolectomy, which included excision of the terminal ileum and a part of jejunum, with reanastomosis between the ileum and the transverse colon, were performed. The tumor and the other tissues that were removed were sent for pathological examination, which reported a mucinous adenocarcinoma of the appendix (Figure 1). The omentum was infiltrated by mucinous adenocarcinoma and presented the histological features of peritoneal pseudomyxoma (Figure 2).

After an oncology consultation the patient was given some cycles of systemic chemotherapy, but she was admitted again to our clinic in November 2000 with recurrence of the tumor. CT scan revealed a pelvic mass about 10 cm and serum levels of CEA and CA 125 were 831 ng/ml and 79.9 U/ml, respectively. She underwent laparotomy and the pelvic mass was removed. Histological examination reported a recurrence of the previous tumor. The patient was given three cycles of systemic chemotherapy. In January 2001 the serum levels of CEA and CA 125 were 271 ng/ml and 29.3 U/ml, respectively, and she remains under close oncologic observation.

Revised manuscript accepted for publication November 5, 2003

Fig. 1

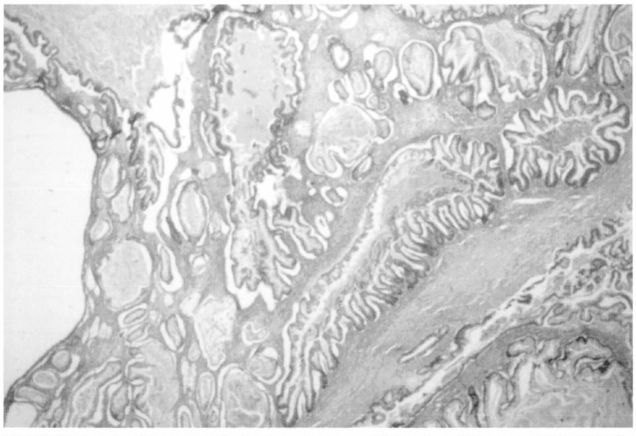
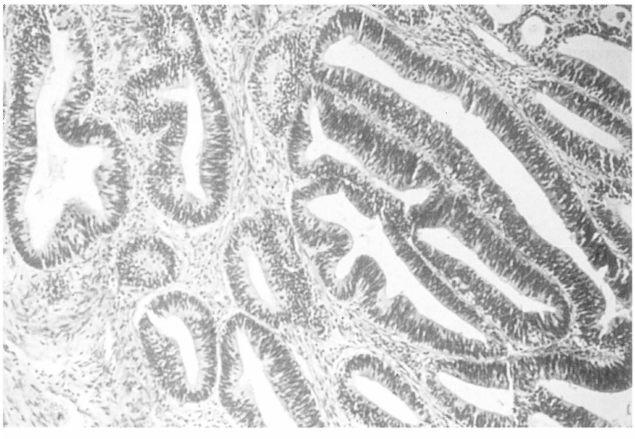


Fig. 3



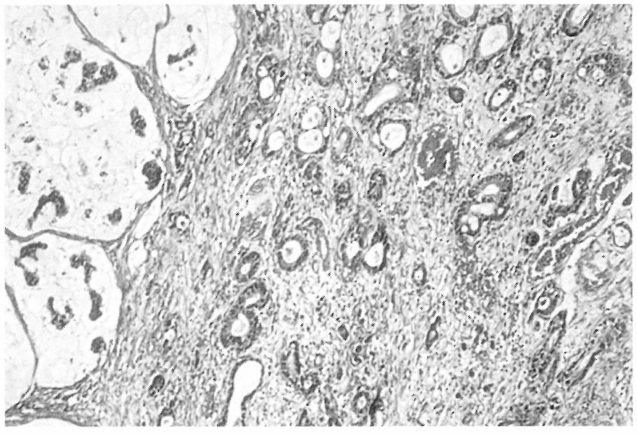


Figure 1. — Histological section of appendiceal wall showing extensive infiltration by a mucinous cystadenocarcinoma (Hematoxylin-eosin, x 25).

Figure 2. — Histological section of the omentum showing infiltration by mucinous adenocarcinoma of the appendix and lakes of loose mucinous material.

Figure 3. — Histological section of appendiceal mucosa showing a mucinous tumor of the appendix of undetermined malignant potential.

Case 2

A 61-year-old woman presented at our gynecologic outpatient department with only the clinical symptom of lower abdominal pain. The pain was deep and had started suddenly, three months previously; it was first localized in the right lower quadrant. The patient had been examined a few days before by a pathologist and an ultrasound scan was performed. The ultrasound demonstrated a 7 x 5 x 5 cm in diameter solid/cystic mass at the anatomic position of the right adnexal region.

Upon her admission to our clinic in May 2002 a vaginal examination confirmed the presence of a semisolid mass at the anatomic position of the right adnexal region and a CT scan was performed. CT revealed a 7 x 4.5 x 7 cm in diameter pelvic mass at the anatomic position of the right adnexal region which probably originated from the right ovary. Assessment of serum CA 125 and other tumor markers were negative except for the serum CEA levels which showed a value of 4.6 ng/ml. The other hematological and biochemical tests were normal and a chest X-ray was negative.

During laparotomy an 8 x 5 x 7 cm in size semisolid cystic mass was found in the last part of the ileum. The mass probably originated from the appendix and morphologically looked like ovarian tissue. Therefore, our first diagnosis was tumor of the appendix or ectopic ovary. Intraoperative histological examination of the tumor showed the morphology of a borderline mucinous tumor, probably ovarian. The tumor was removed while iliac resection and reanastomosis, total abdominal hysterectomy with bilateral salpingo-oophorectomy and total omentectomy were performed.

The final histological diagnosis reported a mucinous tumor of undetermined malignant potential of the appendix (Figure 3). After oncologic consultation the patient remains under close oncologic observation;

Discussion

Neoplasms of the appendix are very uncommon and are usually diagnosed at operation or autopsy. Three histological types of malignant tumors occur: carcinoid, adenocarcinoma and malignant mucocele [5]. While none of these tumors are very common, all have the tendency for ovarian spread when they disseminate beyond the appendix. Many of these tumors mimic primary ovarian neoplasms such as endometrioid adenocarcinomas and intestinal type mucinous cystic tumors [4, 5].

Metastatic ovarian tumors account for approximately 17% of ovarian malignant tumors and are of gastrointestinal origin in 24% to 42% of cases. Most of these tumors are derived from the colorectum or stomach and only 1% to 2% originate from the appendix [3]. When an extraovarian tumor is in the genital tract, an independent origin is favored for ovarian tumors in most cases. When they are outside the genital tract, however, ovarian tumors are considered almost metastatic. Additional difficulty in interpretation of cases is caused when the extraovarian primary tumor is mucinous, as the metastatic ovarian tumors in these cases, and may stimulate a primary mucinous carcinoma or occasionally, a mucinous borderline tumor of the ovary.

In a recent study Sehouli *et al.* [2] reported a case of a 58-year-old female patient with the suspected diagnosis of ovarian cancer. Upon surgical exploration, examination of the appendix revealed the histological diagnosis of primary mucinous adenocarcinoma.

McBroom *et al.* [4] reported three cases in which the patients were referred to their institution from 1994-1999 for presumed late stage ovarian cancer but finally after laparotomy were found to have primary appendiceal adenocarcinoma, adenocarcinoid and mucinous cystadenocarcinoma metastatic to the ovaries.

To investigate the relationship between ovarian and appendiceal mucinous tumors Young *et al.* [6] reported 22 cases of concurrent ovarian and appendiceal mucinous neoplasms and favored an appendiceal origin for most of these tumors. The appendiceal and ovarian tumors were synchronous in 21 cases. Laparotomy typically disclosed large cystic ovarian tumors, usually multilocular, an appendix that was usually dilated and covered with mucus, and abundant intra-abdominal mucus.

In contrast Seidman *et al.* [7] reported 25 cases with synchronous ovarian and appendiceal mucinous neoplasms and their findings suggest an independent origin of the ovarian and appendiceal tumors in most cases and do not favor an origin in a single site. Ovarian pseudomyxoma was present in 22 cases.

In another study Ronnett *et al.* [3] analyzed 20 cases of ovarian metastases deriving from appendiceal adenocarcinomas. Appendiceal and ovarian tumors were diagnosed concurrently in 15 cases. In the remaining five, the ovarian tumors were diagnosed before the appendiceal tumor. The ovarian tumors were bilateral in 16 cases and were histologically similar to the associated appendiceal tumor in each case.

Prayson *et al.* [8] analyzed 19 cases of peritoneal pseudomyxoma with emphasis on site of origin and nature of associated ovarian tumors. A primary appendiceal mucinous neoplasm was found in 16 of the 17 patients with an evaluated appendix and five of the nine women had cystic mucinous ovarian neoplasms; each also had an appendiceal mucinous neoplasm. All five ovarian tumors also had histologic features of ovarian pseudomyxoma. Finally the authors concluded that the appendix is the primary site of origin of pseudomyxoma in the vast majority of cases in both men and women, and associated mucinous ovarian tumors are most likely secondary neoplasms resulting from incorporation of implanted mucus and neoplastic mucinous epithelial cells of the peritoneal pseudomyxoma.

Conclusion

It is interesting that in all the studies that were reported and in our two cases reports, patients usually presented with a pelvic mass and abdominal or pelvic pain. Metastatic mucinous tumors of the appendix should be considered in the differential diagnosis of any case of mucinous ovarian tumor and pseudomyxoma peritonei, especially when these tumors are associated with extraovarian disease.

References

[1] Hart W.: "Metastatic tumors to the ovary". International Society of Gynecological Pathologists Symposium, March 15, 1992, 1.
[2] Sehouli J., Kopetsch O. J., Ricke J., Buchmann E., Stengel D., Riess D. *et al.*: "Primary mucinous adenocarcinoma of the appendix: A rare entity in the differential diagnosis of ovarian cancer". *J. Obstet. Gynaecol. Res.*, 2000, *26*, 333.
[3] Ronnett B., Kurmen R., Smookler B., Sugarbaker P., Young R.: "The morphologic spectrum of ovarian metastases of appendiceal adenocarcinomas". *Am. J. Surg. Pathol.*, 1997, *21*, 1144.
[4] McBroom J. W., Parker M. F., Krivak T. C., Rose G. S., Grothers B.: "Primary appendiceal malignancy mimicking advanced stage ovarian carcinoma: A case series". *Gynecol. Oncol.*, 2000, *78*, 388.
[5] Schwartz S., Shires T., Spencer T.: "Appendix". Principles of Surgery, 6th ed., 1994, 2, 1307.
[6] Young R., Blake Gilks C., Scully R.: "Mucinous tumors of the appendix associated with mucinous tumors of the ovary and pseudomyxoma peritonei". *Am. J. Surg. Pathol.*, 1991, *15*, 415.
[7] Seidman J., Elsayed A., Sobin L., Tavassoli F.: "Association of mucinous tumors of the ovary and appendix". *Am. J. Surg. Pathol.*, 1993, *17*, 22.
[8] Prayson R., Hart W., Petras R.: "Pseudomyxoma peritonei". *Am. J. Surg. Pathol.*, 1994, *18*, 591.

Address reprint requests to:
A. LIAPIS, M.D.
N. Paritsi 9A, N. Psychiko
0030-1 Athens (Greece)