Extraovarian mullerian-type cystic tumors of the female. A report of six cases and review of the literature

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Summary

Six cases of extraovarian cystic tumors of mullerian type are reported. Patient age ranged from 27-71 years. Tumor diameters were 5-11 cm and were located in the mesentery (4/6) and retroperitoneum (2/6). Four tumors were of mucinous type (1/4 of borderline malignancy) and two of serous type. These tumors clinically and histopathologically mimic other primary tumors of the peritoneum. The problems of differential diagnosis are discussed.

Key words: Extraovarian neoplasms; Peritoneum; Cystadenoma; Mullerian system.

Introduction

Extraovarian cystic neoplasms that morphologically resemble ovarian tumors are rare, and up to date less than 30 cases have been reported [1, 2].

These tumors arise in the abdominal cavity, the mesentery and the retroperitoneal space, may attain a large size and be misdiagnosed as ovarian or enteric tumors [1]. They may be asymptomatic and found accidentally during a routine examination or they produce symptoms by compression of the abdominal organs causing abdominal discomfort and/or pain.

Their origin is obscure and they are considered as primary tumors of the secondary mullerian system or as tumors arising from ectopic ovarian tissue and mullerian duct remnants [3, 4].

We present five cases examined in our laboratory during the last decade and analyze their clinicopathological features and the problems in the differential diagnosis from other abdominal cystic neoplasms.

Material and Method

Over the last decade 23 abdominal and six retroperitoneal cystic lesions have been examined at the Pathology Department of Areteion University Hospital.

Among these, four abdominal and two retroperitoneal cysts presented morphological features of mullerian type tumors, of mucinous (4/6) and of serous(tubal) type (2/6).

Case 1 (Surgical report no. 47564)

A 54-year-old woman was referred to the 2nd Surgical Clinic of Areteion Hospital with the diagnosis of a tumor of the ascending colon, abdominal pain and discomfort. The diagnosis of a colonic tumor was supported by ultrasonography (US) and computed tomography (CT) studies. The patient underwent right hemicolectomy. During the laparotomy two cystic tumors, one in the mesentery and one in the retroperitoneum, were observed and excised.

Pathological examination showed a stenotic area on the ascending colon 10 cm above the ileocecal valve measuring 4 cm in the greatest diameter. The covering colonic mucosa was normal and the stenotic lesion was confined to the enteric wall. Histology showed development of dense fibrous tissue at the serous surface containing small glands of tubular type, with cuboidal cells, without atypia, consistent with mesothelial or mullerian type inclusions. No evidence of colonic adenocarcinoma was found but a mesenteric cyst measuring 6 cm in diameter was observed, filled with mucinous material. The cystic wall was 0.4-2 cm thick, fibrous and was covered by tall mucinous epithelial lining resembling endocervical mucosa. Acellular mucinous material infiltrated the wall and extended into the mesocolic fatty tissue.

During the surgery another tumor in the retroperitoneum was observed, 11 cm in diameter, which was excised and presented the same histological morphology of a mucinous cystic tumor of mullerian type without epithelial atypia.

Other abdominal organs and the genital system were normal. The ovaries were described as atrophic.

Because of the observed infiltrating tendency of the mucinous material of the mesenteric cyst, the patient was under observation and during a 5-year follow-up no signs of recurrence were observed. After this period, there is no information available on this patient.

Case 2 (Surgical report no. 40563)

A 45-year-old woman was referred to the 2nd Surgical Clinic of Aretaieion Hospital for a mesenteric tumor observed during an abdominal US for gallbladder disease. The tumor was excised and no other lesion was found in the abdomen. The genital system was within normal limits and the ovaries were described as normal in size with small cystic lesions, probably follicular cysts.

Revised manuscript accepted for publication February 12, 2004

Histology showed a cyst, 6 cm in the greatest diameter, with a fibrous wall lined by tall mucinous epithelial cells of endocervical type. Acellular mucinous material infiltrated the wall. The diagnosis of a benign mucinous cystic lesion of the mesentery was made and the patient has been under observation for seven years without any signs of recurrence.

Case 3 (Surgical report no. 85263)

A 71-year-old woman was referred to the 2nd Clinic of Obstetrics and Gynecology with the clinical diagnosis of an ovarian tumor. Serum levels of CA-125 were within normal limits. The diagnosis of an ovarian tumor was supported by the US and CT studies and the patient underwent exploratory laparoscopy during which a mesenteric cyst was found and excised. The ovaries were atrophic and no other remarkable changes of the genital system were reported.

Histology showed that the cyst measured 5.5 cm in the greatest diameter and had a fibrous wall lined by mucinous epithelium of endocervical type. Focally, a prominent stratification of the epithelium was observed with papillary projections into the lumen, rare mitoses and cellular atypia (Figure 1). Among the tall columnar cells, isolated cells of enteric (goblet) type were observed. No infiltration of the wall by the glandular elements was observed and the diagnosis of a mucinous tumor of borderline (low) malignancy was made. The patient is well three years after the surgery without any signs of recurrence.

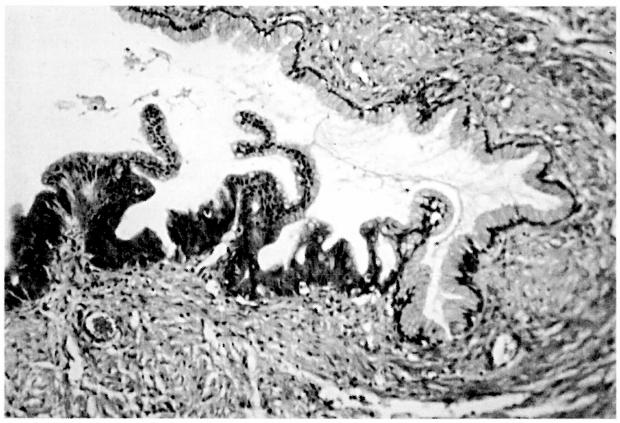

Figure 1. — Histological section of a mesenteric cyst with morphology of a mucinous borderline tumor (H&E x 25).

Case 4 (Surgical report no. 66100)

A young woman of 27 years of age was referred to the 2nd Surgical Clinic of Aretaieion Hospital with the diagnosis of a retroperitoneal tumor with cystic features. No symptoms were described and the tumor was an accidental finding during a CT examination for adrenal evaluation. A cystic tumor was excised measuring 3 cm in diameter with a wall 0.6-1 cm in thickness which consisted of loose connective tissue. The wall was lined by tall epithelial cells of serous (tubal) type without any atypical changes. The diagnosis of retroperitoneal serous cystadenoma was made. The genital system and especially the ovaries were within normal limits.

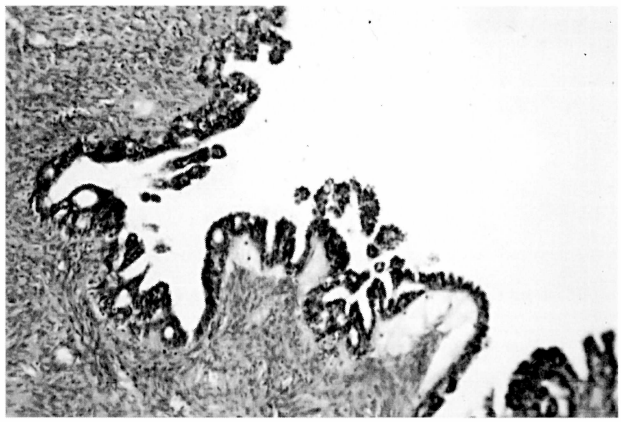

Figure 2. — Histological section of a retroperitoneal cystic tumor with morphology of a serous (mullerian) cyst (H&E x 25).

Case 5 (Surgical report no. 52881)

A young woman of 29 years of age was referred to the 2nd Surgical Clinic of Areteion Hospital with the clinical and radiological diagnosis of a retroperitoneal cyst. The genital system and the ovaries were within normal limits and there was a history of a recent cesarean section. The serum levels of CA-125 were normal.

The patient underwent surgical exploration and a retroperitoneal cyst measuring 17 cm was observed located in the left adrenal gland.

Histological examination of the cyst showed a fibrous wall lined by cuboidal or tall cells of serous type with focal hyperplasia but no remarkable atypia or infiltrative tendency (Figure 2). A small part of adrenal tissue was observed at the exterior surface of the cyst.

Additional paraffin sections of the specimens showing a serous type tumor were investigated by immunohistochemistry. Staining for CEA (moab, Monosan CA-125 (moab, CIS Diagnostics) and CK 34bE12, CK AE3 (moab Monosan) secretory component (Poly-Dako) was positive but negative for Vimentin (moab, NOVOCASTRA). The morphology and the immunocharacteristics were consistent with a mullerian serous type tumor and not mesothelioma.

Discussion

The development of extraovarian cystic tumors with similar morphological characteristics as epithelial ovarian tumors is an unusual event of uncertain histogenesis. Three theories are offered that attempt to explain the development of these tumors. The theory of the neoplastic potential of a secondary mullerian system [3] seems the most plausible since the theories that these tumors arise from ectopic mullerian epithelium or from a supernumerary or ectopic ovary are not supported in that no remnants of ovarian tissue were found in the cases reported. In none of our cases, totally examined by serial sections, was ovarian tissue found in the form of stroma or other ovarian elements.

Most of the reported up-to-date 26 extraovarian cystic tumors were mucinous, of benign, malignant and borderline histology [2, 4-6]. They developed in women and

were located in the retroperitoneum with the exception of only one case that developed in the inguinal region [7]. In the first of our cases, two mucinous cystadenomas were present – one located in the retroperitoneum and one in the mesentery. This is a unique event in our experience and its association with the presence of mesothelial inclusions at the colonic serosa is remarkable. The other two mucinous tumors we reported were located in the mesentery as well, and this is an unusual finding [8].

No evidence of teratomatous tumor was observed in contrast with one theory about histogenesis that suggests that mucinous tumors may arise from ectopic teratomas [9].

Two of our cases with tumors measuring 6 and 11 cm were cytologically benign but presented infiltration of the wall by acellular mucinous material which was a worrying picture although the follow-up has not shown recurrence of disease up to now.

Another case was diagnosed as a mucinous cystic tumor of borderline malignancy. The diagnosis was based on the criteria used in the examination of ovarian mucinous tumors. The patient did not receive any other therapy and remains well.

With the exception of one case where goblet cells were observed among the endocervical-type epithelium, no other elements such as gastric or specialized enteric mucosa or ovarian-type stroma were found in the wall or in the epithelial lining of the mucinous cysts we have reported [10].

The two reported serous tumors were large cysts located in the retroperitoneum and were histologically benign. Both these characteristics are rare and in contrast to the reported serous extraovarian tumors which have a peritoneal location and are malignant or of borderline malignancy.

Clinical symptoms were absent in three of five cases and the discovery of the tumors was an accidental finding during an examination for unrelated causes. In one case, the tumor was mimicking a primary colonic tumor and in another case an ovarian tumor, both at the clinical and specialized radiological examination.

The diagnosis of mucinous tumors does not present any diagnostic difficulty. The differential diagnosis of serous extraovarian tumors must be made from peritoneal cystic mesotheliomas. It is based on the morphology of the epithelium and the immunohistochemical characteristics which in our study showed a positive cellular reaction to CEA, CA-125, CK 34bE12, AE3 (h.mw), secretory component, and a negative reaction to Vimentin [12].

In conclusion, extraovarian tumors of mullerian type are unusual cystic lesions of the peritoneum, mesentery and the retroperitoneal space that most probably arise from the specialized cells of the secondary mullerian system. They may attain a great size and produce symptoms that mimic other primary tumors of the colon or ovary. Surgical excision is the treatment of choice and their recognition is important in order to avoid unnecessary and extensive surgical procedures.

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