Extragenital cystic lesions of peritoneum, mesentery and retroperitoneum of the female. Clinicopathological characteristics of 19 cases

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Summary

The clinicopathological characteristics are presented of 19 extragenital cystic lesions of female patients located at the retroperitoneum (9 cases), the mesentery (6 cases) and the peritoneum (4 cases). Median age was 42.3 years and the main symptom was abdominal pain. The cysts measured 4-27 cm in diameter and were classified as: epithelial (7/19), mesothelial (5/19), vascular (2/19), parasitic (1/19) and developmental in origin (4/19). Of the cases 18/19 were benign lesions and one case was a borderline mucinous cystic tumor. Immunohistochemistry by a streptavidin-biotin method was performed to investigate CEA transcripts (MoAb, Novosan), CA125 (MoAb, CIS Diagnostics), vimentin (MoAb Ve6, Novocastra), secretory component (a polyclonal antibody, DAKO), Factor VIII (MoAb, DAKO), CD34 (MoAb, Scytec), calretinin (a polyclonal antibody, Zymed, San Francisco, CA). Cytokeratins were of low and high molecular weight (Immunon, AE1-MoAb). The results are helpful in the correct classification of various tumors. The treatment of choice is complete surgical resection of the tumors.

Key words: Peritoneum; Custs retroperitoneum; Neoplasm.

Introduction

Peritoneal, mesenteric and retroperitoneal cystic lesions are rare female tumors, with diverse histogenesis.

On Rokitanski presented the first case in 1843 [1] and since then less than 200 cases have been reported with the main emphasis on the clinical features and therapeutic approach [2-4].

These lesions are usually classified into traumatic, infective, degenerative, neoplastic and developmental and most cases presented were considered to be lymphangiomatous malformations [3, 4] .

We present the immunopathological characteristics of 19 cases examined in our laboratory over a 13-year period, with the purpose of investigating the origin and features that permit a correct and differential diagnosis from other cystic lesions of the female abdomen.

Material and Method

The pathological features of 19 cases of cystic lesions that developed in the peritoneum, omentum, mesentery and retroperitoneum of female patients and were examined in our laboratory from 1990-2003 were re-evaluated.

All specimens were fixed in buffered formalin and paraffin-embedded. After routine processing, additional sections were stained by hematoxylin & eosin for immunohistochemistry.

Immunostains were performed with a streptavidin-biotin method to investigate the expression of CEA transcripts (MoAb, Novosan), CA125 (MoAb, CIS Diagnostics), vimentin (MoAb Ve6, Novocastra), secretory component (a polyclonal antibody, DAKO), Factor VIII (MoAb DAKO), CD34 (MoAb Scytec) calretinin (a polyclonal antibody, Zymed) and (Immunon, AE1-MoAb) and low and high molecular weight cytokeratins. Appropriate positive and negative controls were used.

Histochemical investigation of epithelial mucin and proteoglucans in selected cases was performed as well (PAS-diPAS, Mucicarmine).

Clinical information was obtained from the archives of the 2nd Surgical and Gynecological Clinics of our Hospital.

Results

Clinical symptoms. Patients age ranged from 27-71 years, with a median age of 42.5 years. The main symptom was abdominal pain and vague discomfort (11/19 cases), signs of incomplete abdominal obstruction (3/19 cases), a sense of perineal pressure and constipation (3/19 cases).

All cases underwent ultrasound (US) and computed tomography (CT) scanning exploration. The diagnosis of ovarian tumors (11/19 cases) abdominal cysts, probably parasitic (3/19 cases), colonic tumors (3/19 cases) and inconclusive investigation (2/19).

All patients underwent surgical exploration and excision of the cystic lesions. In one case, a right hemicolectomy was performed as well and in another case, partial excision of a retro rectal cyst was performed.

Pathological examination. The lesions were classified according to the histological morphology as follows:

Epithelial cysts of mullerian type (extra genital): Seven cases were found which developed in women 27-71 years

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Fig. 1

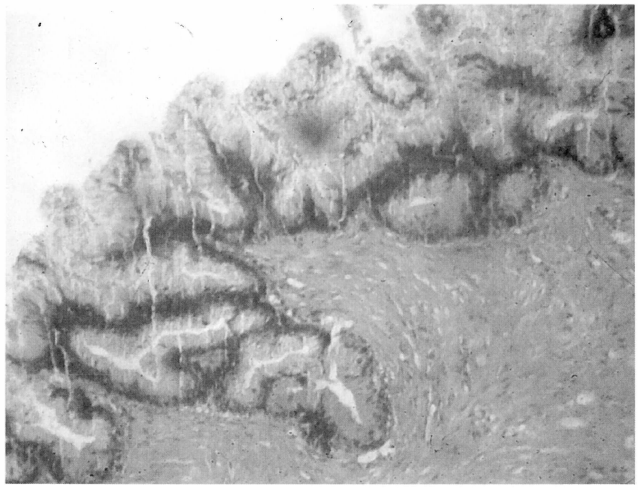

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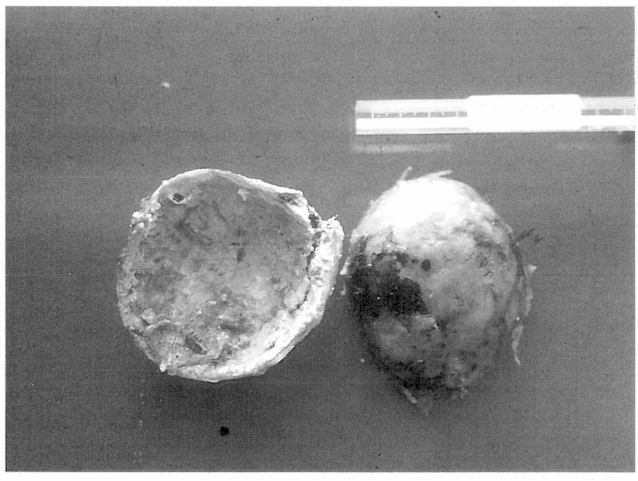
Fig. 3



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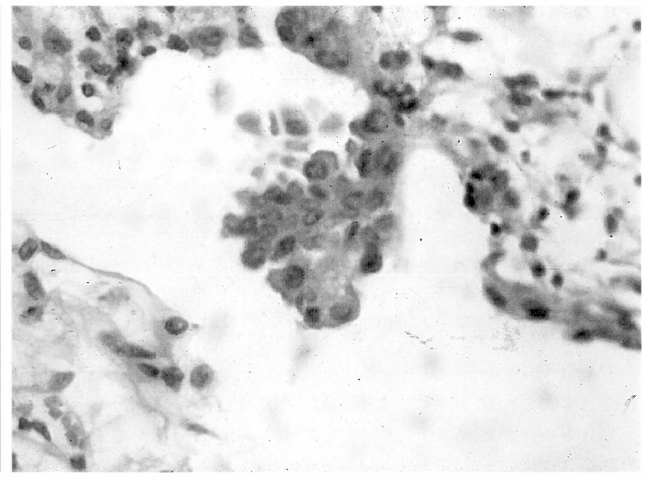

Fig. 5

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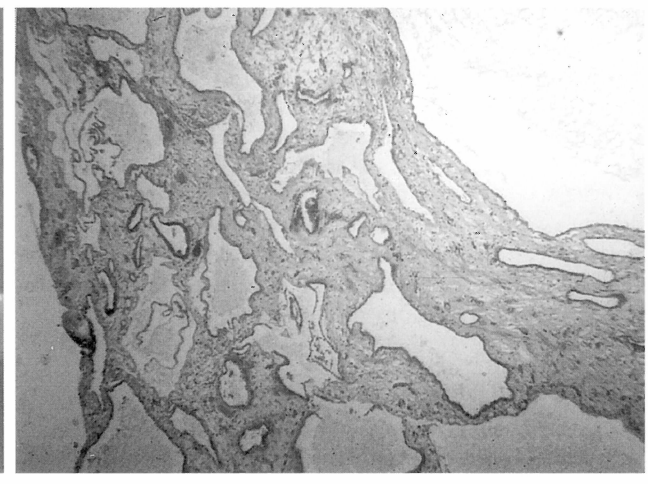

Figure 1. — Histological section of a mucinous cystic neoplasm of borderline malignancy showing hyperplasia and atypia of the lining epithelium (hematoxylin & eosin x 240).
Figure 2. — Histological section of an epithelial cyst showing a thick fibrous wall lined by simple cuboidal serous epithelium (hematoxylin & eosin x 25).
Figure 3. — A single mesothelial cyst measuring 9 cm in diameter (gross picture).
Figure 4. — Histological section of a mesothelial cyst showing a hyperplastic tuft of mesothelial cells (hematoxylin & eosin x 240).
Figure 5. — A multicystic lymphangioma of the retroperitoneum (gross picture).
Figure 6. — Histological section of a lymphangioma showing cystic spaces filled with lymph and separated by fibrous walls (hematoxylin & eosin x 25).

of age. Histology showed five mucinous and two serous cystic neoplasms located in the mesentery (4 cases) and retroperitoneum (3 cases). They measured 5-9 cm in the greatest diameter and consisted of fibrous walls of variable thickness lined by columnar epithelium (endocervical type), mucin-producing epithelium (4 cases) which was atypical and hyperplastic in one case of borderline malignancy (Figure 1) or serous type (3 cases) (Figure 2). The epithelial cells were immuno-positive to cytokeratins of low molecular weight and to secretory component which was located in the apical border of the serous cells and in secretions of mucinous cells, and were negative to the other markers.

In all cases the genital system was normal or without any remarkable ovarian pathology.

Benign cystic mesotheliomas: Five cases developed in women 31-34 years of age, the youngest age group in our study. Three were multicystic peritoneal mesotheliomas composed of cystic spaces of various sizes (7-19 cm in total diameter) and two were large unilocular mesenteric cysts (9 cm and 27 cm) (Figure 3). The mesothelial cells gave a positive immunoreaction only to keratins 5/6 and calretinin (Figure 4). No asbestos exposure of these young women was reported .

Lymphangiomatous lesions: Three cases were found: Two women, 45-54 years of age, presented large multicystic lymphangiomas, 10 and 18 cm in diameter and retroperitoneal (Figure 5). Histology showed cystic spaces with fibrous walls, and lymphocytic infiltration and lymphoid nodules (Figure 6). The cysts were lined by endothelial cells and were immunohistochemically positive only to CD34 and factor VIII. One case was diagnosed as lymphangiomatosis because of the presence of abundant bundles of smooth muscle tissue in the wall of the cystic spaces. This lesion developed in the retroperitoneum of a woman 52 years of age who also presented with a uterine leiomyoma (20 cm in diameter) and angiomyolipoma of the kidney. This triad of tumorous lesions may represent a developmental anomaly described in patients with tuberous sclerosis.

Retrorectal tailgut cysts: Three patients, 29, 33 and 52 years of age presented with these rare lesions. The cysts contained fibrous walls with some smooth muscle fibres and were covered by squamous and cuboidal epithelium. One patient also had a large ruptured endometrioid ovarian cyst and because of extensive fibrous adhesions in the pelvis, the retrorectal cyst was only partially excised.

Parasitic cysts: One patient with a history of hepatic parasitic cysts presented a peritoneal parasitic echinococcus cyst measuring 7 cm in the greatest diameter which was excised with its fibrous capsule.

Discussion

Peritoneal, mesenteric and retroperitoneal cystic lesions are rare entities and up to date less than 200 cases have been reported, mostly as case reports.

The incidence varies from 1/14,000 to 1/250,000 cases of general hospital admissions and they are more common in children with an incidence of 1/200,000 cases of pediatric hospital admissions [4, 5]. Most cysts reported were located in the mesentery and were asymptomatic, incidentally found during surgery [3-8] and are considered as developmental anomalies of lymphatic tissue and rarely of traumatic, infective, degenerative, parasitic or neoplastic origin [4, 5, 8].

In our series the location and histology varied. No traumatic, infective or degenerative cysts were found. Most of our cases (14/19) were considered to be neoplasms, epithelial-mullerian, mesothelioma or vascular tumors with distinctive morphology and immunohistochemical characteristics that permit correct diagnoses. One case only of lymphangiomyomatosis and the tail gut cysts may be considered as genuine developmental defects. Most of our cases [9] developed in the retroperitoneum, six in the mesentery and four were peritoneal.

Most patients presented with various symptoms that brought them to the hospital. It should be noted that in most cases radiological examination revealed the lesions and although most were considered as ovarian or colonic tumors, most of our cases were large solitary or multiple cystic tumors. Despite certain difficulties, all cysts were totally excised with the exception of a retrorectal cyst which was surrounded by fibrotic tissue and adhesions and only partially excised. In one case, during laparoscopic exploration of a large mesenteric cyst, the adhering colonic wall was found hard and stenotic. A right hemicolectomy was performed en bloc with the cystic mass. Permanent sections showed extensive fibrosis of the colonic wall with benign serosal inclusions and a large benign cystic mesothelioma.

There is diagnostic difficulty in recognizing mesothelioma from epithelial cystic tumors and in these cases immunohistochemistry is most helpful.

Vascular tumors and parasitic cysts present a characteristic morphology and the correct diagnosis is easily made.

All our cases presented a benign character with the exception of a retroperitonal epithelial mullerian tumor of borderline malignancy. This patient is well three years after surgery. The possibility of the development of a malignant cystic lesion must always be considered in view of certain reports [4, 7] and the therapy of choice of these unusual and most interesting lesions is complete surgical excision. An alternative method to consider is laparoscopic excision, which presents the advantage of rapid recovery and minimal morbidity [10].

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