

CASE REPORT

Giant ovarian mucinous cystadenoma with borderline areas: a case report

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Abstract

Nowadays, giant ovarian mucinous cystadenomas are a rare condition, due to early diagnose and treatment. We report the case of a 44-year-old patient, who was diagnosed and admitted in our department with a giant pelvi-abdominal mass. The patient was operated with very high-risk consent, thus an enormous ovarian cyst of approximately 30 kg was extirpated. Afterwards, an extensive histopathological analysis was performed in order to determine the exact clinicopathological entity of the giant tumor-enteric type of multilocular mucinous ovarian cyst adenoma with borderline areas, with benign proliferation. Although giant ovarian mucinous cyst adenoma with borderline areas is an extremely rare condition, when encountered it is a real challenge for both surgeon and pathologist.

Keywords: giant mucinous cystadenoma, borderline areas, extensive histopathological analysis.

Introduction

Ovarian mucinous cystadenomas are epithelial tumors, which according to histopathological criteria are usually benign, surrounded by a gastrointestinal type epithelium (gastric foveolar-type mucinous epithelium, enteric-type, and far more rarely mucinous cell when they are endocervical type and the cytogenesis is Müllerian cell type) [1–4]. Mucinous borderline tumors and atypical proliferative mucinous tumors also contain gastrointestinal-type mucin and epithelial cells with higher proliferation aspects than those seen in benign mucinous tumors [1–8]. The benign type is the most frequent (80% of the mucinous cystadenomas) and they occur most often in the third to sixth decade of life [6, 7, 9–12]. The tumor is usually unilateral, with a smooth surface, multilocular and can overpass 30 cm in diameter [1, 3, 4]. It can be associated with dermic cysts when the cytogenesis is in germ cell origin or with Brenner tumors when the origin is in surface epithelial origin. The prognosis is very good. Recurrences are very rare (usually when the excision was incomplete or the cystectomy took place without oophorectomy) [1, 3, 4].

Unilateral salpingo-oophorectomy is preferable when the tumor is unilateral and the evolution towards malignancy is unlikely [1–4]. Predictive factors depend on the proliferation factors identified *via* immunohistochemistry, on the hormonal receptors and the positivity of the cytokeratin (CK) CK7 and CK20 [13–19].

However, nowadays, giant ovarian mucinous cystadenomas are a rare condition, due to early diagnose and treatment, even if often they present few and non-specific symptoms [6, 7, 10–12, 20–24]. Even rarer is the situation of a patient with a giant ovarian mucinous cyst adenoma with borderline areas.

Aim

We report the management of a 44-year-old patient with a giant ovarian tumor, diagnosed as mucinous cystadenoma with borderline areas, after an extensive histopathological analysis performed in order to evaluate the precise clinic pathological entity.

Patient, Methods and Results

We report the case of a 44-year-old patient, who was admitted in our department with complaints of diffuse abdominal pain associated with appearance of a large pelvic-abdominal tumor. The woman claimed gradual distension of the abdomen in the last four years, but also constipation and early satiety in the last six months. She denied any previous medical conditions or family history of malignancies.

During the physical examination, we detected a giant abdominal mass, which was palpable throughout the entire abdomen, with smooth surface and elastic consistency (Figure 1). Vaginal examination showed a grossly normal vulva, vagina and cervix, but distended fornices. She weighed 88 kg. By using the forward bend test, a dextro-convex scoliosis was discovered, with a compensatory curvature.

No other abnormalities were noted during physical examination.

Preoperative investigations, including serum tumor markers [CEA, CA-125, CA-19-9 and α -fetoprotein (AFP)] and were within normal ranges.

On ultrasound examination, a large cystic mass with multiple intra-tumoral septations was noted. CT scan revealed a giant pelvic-abdominal cyst, with multiple septations, which extended from the pelvis, towards the

inferior aspect of the diaphragm, measuring 59×48×32 cm. The tumor appeared to occupy the entire abdomen, thus



Figure 1 – Massive abdominal distension.

The patient was operated with very high-risk consent. Under general anesthesia, a pubo-umbilical incision was performed, thus revealing the inferior segment of the giant pelvic-abdominal tumor, which originated in the left ovarian region. There was no free fluid in the abdominal cavity. The cyst was carefully punctured and 29.4 liters of mucinous intra-cystic fluid were slowly drained, in order to prevent hemodynamic and cardiac derangements. The left ovary was included in the large mass. The left fallopian tube was thinned out, adherent, and stretched over the surface of the giant cyst, while the right ovary, right fallopian tube and the uterus were grossly normal (Figure 3).

Total hysterectomy with bilateral oophorectomy was performed, because the extemporaneous histopathological analysis displayed the nature of the mass as being a serous ovarian borderline tumor. Postoperative, the patient weighed 57 kg. She recovered without complications and was discharged six days after surgery.

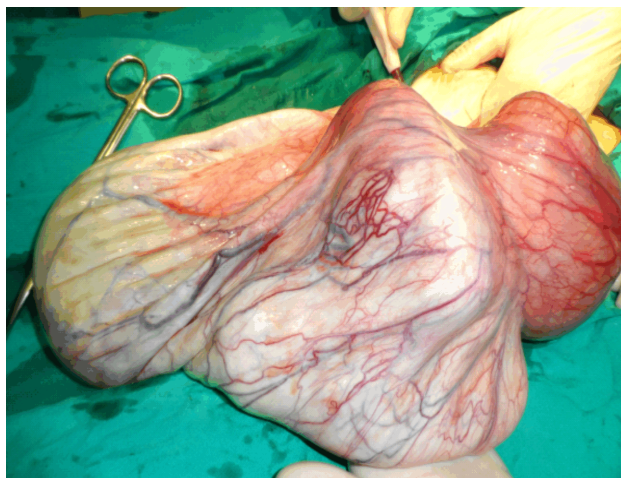


Figure 3 – Intra-operative aspect after the partial depletion of the giant cyst.

compressing adjacent abdominal organs (Figure 2). Bilateral pelvic/iceal system dilatation was also seen.

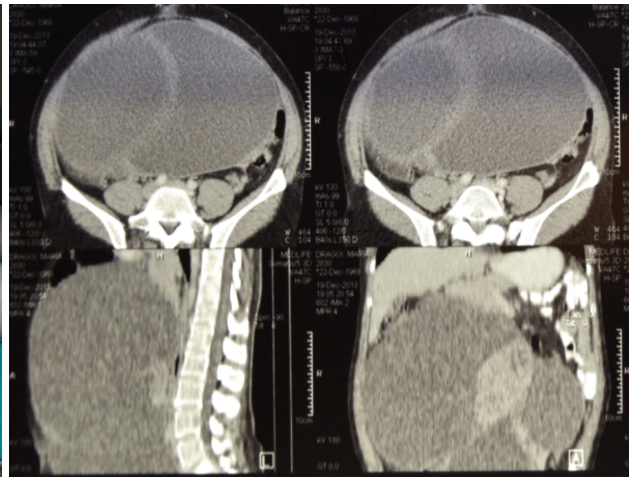


Figure 2 – CT scan showing the giant mass occupying the entire abdomen.

On histopathology, the sections from the uterus, tubes and the right ovary were normal.

An extensive histopathological analysis was performed in order to determine the exact clinicopathological entity of the giant tumor. The initial examination, in Hematoxylin–Eosin (HE) staining, revealed an enteric type of multilocular mucinous ovarian cyst adenoma (Figure 4).

The enteric type of mucinous ovarian cyst adenoma was confirmed using Periodic Acid–Schiff (PAS) method, Alcian Blue staining, Masson's trichrome staining and immunohistochemistry techniques (Figures 5–12). However, because borderline areas were found Ki67 antigen expression was performed, which pointed a benign proliferation (Figure 12).

Therefore, the histopathological diagnosis was enteric type of multilocular mucinous ovarian cyst adenoma with borderline areas, with benign proliferation.

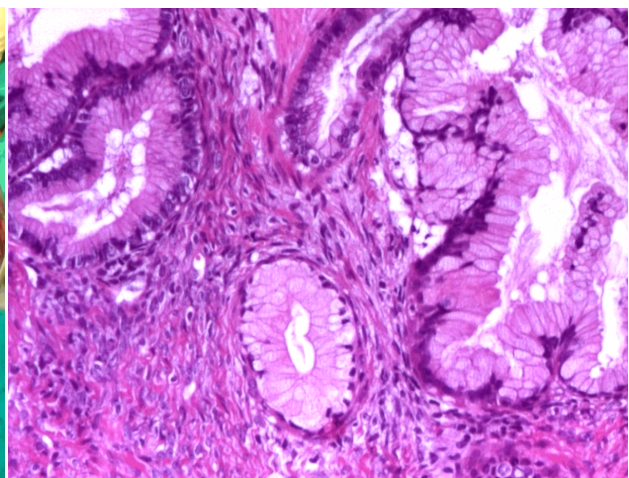


Figure 4 – Enteric type of multilocular mucinous ovarian cyst adenoma: the fibrous stroma contains a benign epithelial neoplastic component that secretes mucus; the nuclei present a discrete pseudostratification (HE staining, ×50).

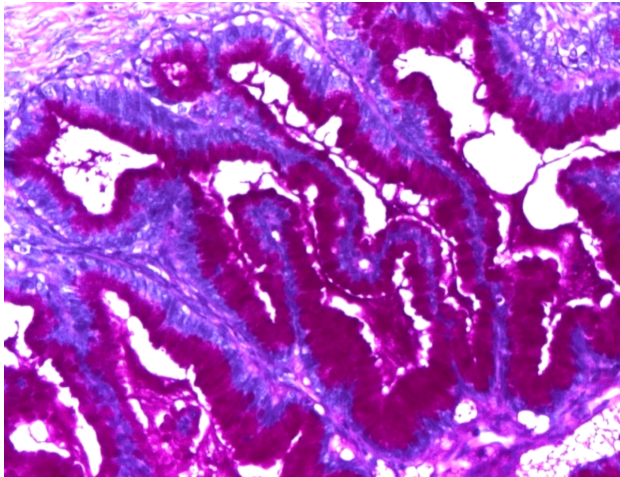


Figure 5 – Enteric type of multilocular mucinous ovarian cyst adenoma: the PAS method was used to highlight the abundance of mucopolysaccharides inside the mucus secreting cells, which is specific for the enteric type of cyst adenoma (PAS staining, $\times 100$).

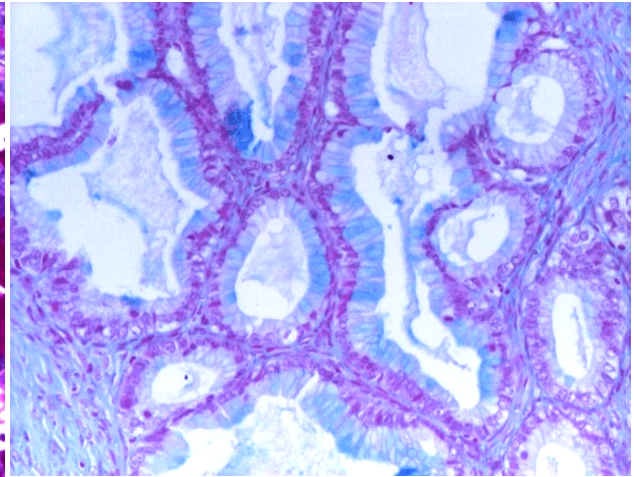


Figure 6 – Enteric type of multilocular mucinous ovarian cyst adenoma: the Alcian Blue staining was used to detect sialomucins, which is specific for the enteric type of cyst adenoma (Alcian Blue staining, $\times 100$).

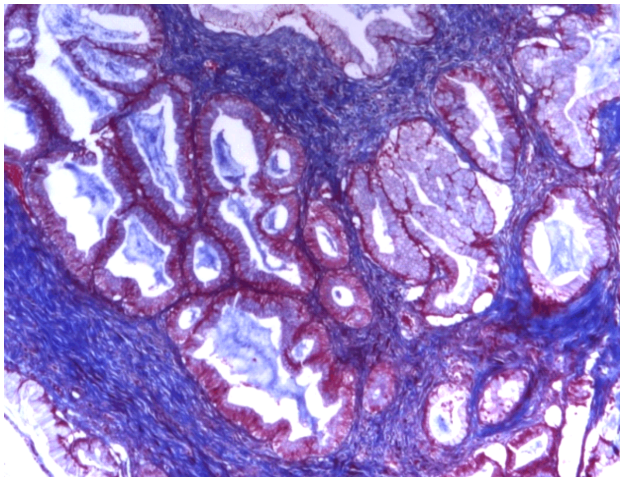


Figure 7 – Enteric type of multilocular mucinous ovarian cyst adenoma: the Masson's trichrome staining was used to differentiate fibrous component (located around the neoplastic secreting epithelium) from the neat muscular fibers (which is specific for the neoplastic proliferation) (Masson's trichrome staining, $\times 50$).

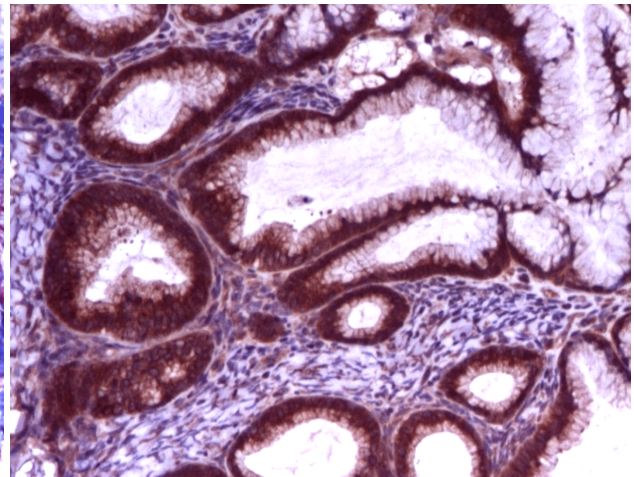


Figure 8 – Enteric type of multilocular mucinous ovarian cyst adenoma: CK20+ immunostaining (a small molecular weight protein specific for the enteric type of cyst adenoma) ($\times 100$).

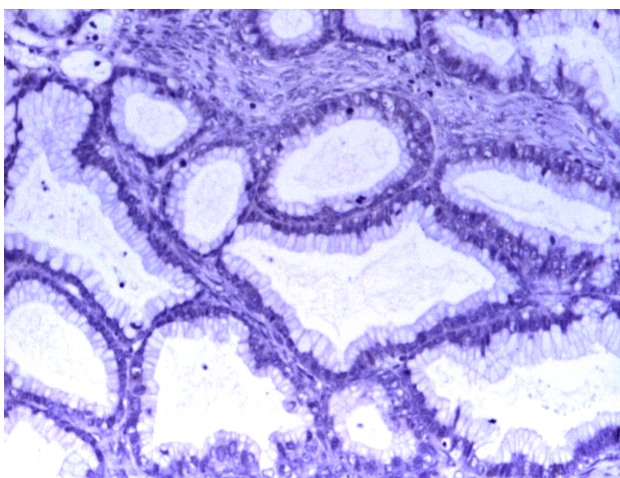


Figure 9 – Enteric type of multilocular mucinous ovarian cyst adenoma: CK7- immunostaining (a small molecular weight protein found in the mammary and pulmonary secretory cells) ($\times 100$).

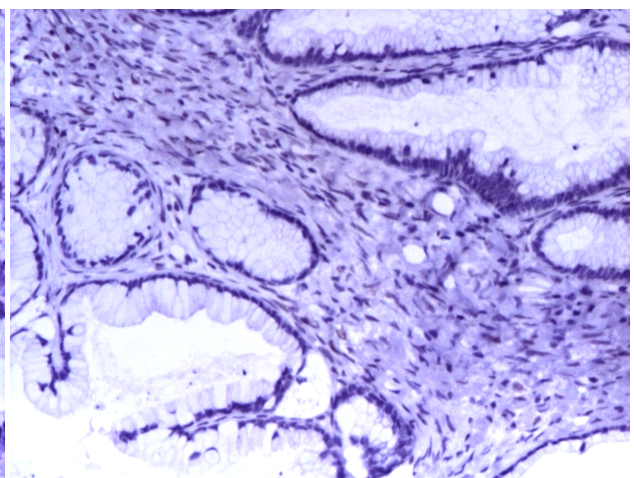


Figure 10 – Enteric type of multilocular mucinous ovarian cyst adenoma: estrogen receptor immunostaining (pledging for the enteric type of cyst adenoma) ($\times 50$).

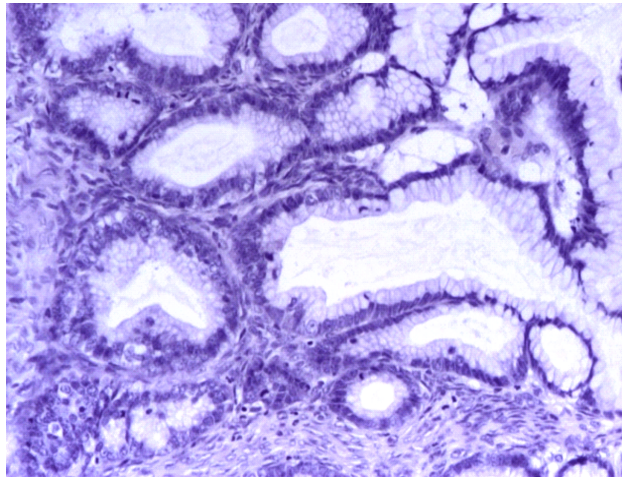


Figure 11 – Enteric type of multilocular mucinous ovarian cyst adenoma: progesterone receptor immunostaining (pledging for the enteric type of cyst adenoma) ($\times 100$).

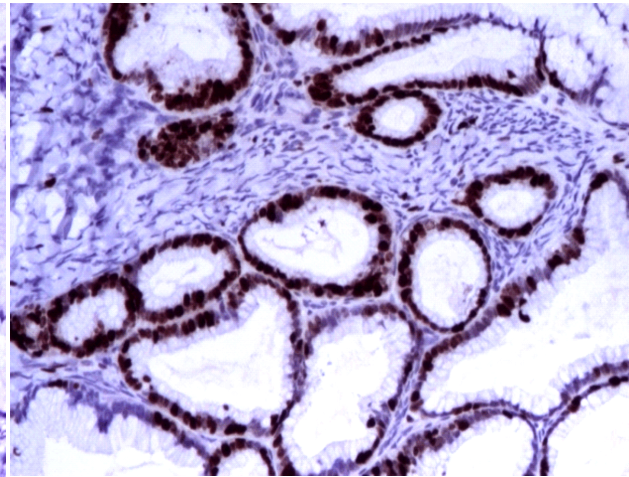


Figure 12 – Enteric type of multilocular mucinous ovarian cyst adenoma: the Ki67 antigen is present only in the dividing cells – approximately 10–15% of the cells have entered the cellular division phase, thus demonstrating that the proliferation is benign (Ki67 immunostaining, $\times 50$).

Discussion

Giant mucinous ovarian cyst adenomas are very rarely observed [6, 7, 10–12, 20–24]. In 10% of cases, these are bilateral [8]. In our case, the tumor was unilateral affecting the left ovary.

The giant mass compressed the adjacent viscera, explaining the symptoms claimed by the patient (constipation, early satiety) and also the bilateral pelvic caliceal system dilatation noted on the CT scan. The giant tumor, also determined long-term traction, therefore causing the dextro-convex scoliosis. In cases of giant ovarian tumors previously reported, the patients described similar symptoms [5, 6, 8–11, 20].

Although some studies claim that laparoscopic surgery has replaced laparotomy, we preferred open surgery through a pubo-umbilical incision, taking into consideration the dimensions of the tumor and the imagistic aspect of the giant cyst – the intra-tumoral septations suggested a malignant proliferation [20, 24–28]. However, because serum tumor markers were within normal ranges and also intra-operative findings we decided to puncture the giant cyst and slowly drain it, in order to prevent the already studied decompression syndrome [8, 29, 30].

Because it was evacuated intraoperative, we do not know the exact weight of the tumor, but we estimate it at about 30 kg (the weight of the patient: preoperative – 88 kg, postoperative – 57 kg). Although larger ovarian tumors have been formerly described, this is the largest ovarian tumor ever reported in our department [6, 31–33].

The diagnosis exam done on a fragment extracted from a proligerous, papillary area resulted in MBT/APTM. After the relevant fragments have been manipulated, the HE staining showed an MBT and a partial APTM. Special stainings were made, and to further strengthen the diagnosis, immunohistochemical tests have been completed. These demonstrated that the enteric type of multilocular mucinous ovarian cyst adenoma does show a fibrous stroma including a benign epithelial neoplastic

component that secretes mucus. In addition, the nuclei present a discrete pseudostratification. When the PAS method was used, it highlighted the abundance of mucopolysaccharides inside the mucus secreting cells. This is specific for the enteric type of cystadenoma [1–4]. Further, sialomucins, which are specific for the enteric type of cystadenoma, were detected *via* the Alcian Blue staining. The Masson's trichrome staining was used to differentiate fibrous component (located around the neoplastic secreting epithelium) from the neat muscular fibers. Lastly, the Ki67 antigen is present only in the dividing cells – approximately 10–15% of the cells have entered the cellular division phase, thus demonstrating that the proliferation is benign [18, 19].

It is important to note that the APMT diagnosis showed a mild segmented proliferation, without microinvasion. The recurrence rate for MGT/APMT with microinvasion is 5% with adverse behavior restricted to FIGO stage IC tumors [17–19].

Conclusions

Although giant ovarian mucinous cyst adenoma with borderline areas is an extremely rare condition, when encountered it is a real challenge for both surgeon and pathologist.

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