Gliomatosis Peritonei Combined with Mature Cystic Teratoma: A Case Report

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Abstract

Co- existent Gliomatosis peritonei (GP) along with mature ovarian teratoma is exquisitely rare. Here, we present a case of mature cystic teratoma with mature gliomatosis peritonei. A 23-year-old woman had been suffering from left sided lower abdominal pain for 2 months. Sonography revealed a heterogeneous complex mass in the pelvis with calcification associated with omental thickening and ascitis. Her relevant tumor markers were normal. A right sided ovarian tumor was found at laparotomy. Whitish nodular deposits were also noted on the omentum and peritoneal surfaces, warrants partial omentectomy. The patient was followed-up for 1 year, with no evidence of recurrence up to the time of writing. Treatment for primary tumor and long- term follow- up is vital for these cases.

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Keywords: Gliomatosis peritonei, Mature cystic teratoma

Introduction

Gliomatosis peritonei (GP) is characterized by the presence of benign glial implants in the peritoneum, omentum, and lymph nodes. It is often associated with immature ovarian teratoma and rarely with mature teratoma. However, it is important to recognize the benign nature of GP due to association with mature ovarian teratoma.[1,2]

Clinical data demonstrated its recurrences, malignant transformation, metastasis and even spontaneus regression. The primary ovarian teratoma may be mature or immature, but the glial tissue is predominantly mature tissue and spreads over the omentum, peritoneum and pelvis. Despite widespread involvement of the peritoneal surface, gliomatosis peritonei does not have a poor prognosis even when associated with immature teratoma.[3]

Four theories are suggested for the development of GP. They are rupture of ovarian capsule and implantation of neural components in the peritoneal wall or greater omentum, angiolymphatic spread, pluripotent stem cells in peritoneum or subjacent mesenchyme undergo metaplasia. The last theories stated occurrence of GP after ventriculoperitoneal shunt operations when glial tissue is transported from cerebrospinal fluid into peritoneal cavity via shunt further support this theory.[4,5] Here, we present a case of mature ovarian teratoma associated with gliomatosis peritonei and a review of relevant literature.

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Case Report

A 23-year unmarried lady having regular menstrual cycle presented to our institute with the complaint of vague lower abdominal pain and a lump in the left lower abdomen for two months. The patients vitals were stable. Abdominal examination revealed a non-tender, palpable mass, left side of lower abdomen, measuring about 16 x18 cm, soft to solid in consistency, and mobile from side to side. Examination of other systems was unremarkable.

Transabdominal ultrasound reveals a solid cystic 10 x12cm left adnexal mass with calcification associated with nodular omental thickening suggestive of infiltration. Mild ascites was also noted. Her tumor markers (Alpha-fetoprotein, CA-125, CA-19.9, CEA) were within normal limit. With the suspicion of a malignant ovarian mass, after all the detailed workup, she was planned for surgery.

During laparoscopic procedure a left ovarian tumor, measuring 12 cm in maximum diameter, was found occupying the entire lower abdominal cavity.The capsule of ovary was intact but there were adhesions to the greater omentum. Numerous whitish nodules are noted in the omental surface, warrants a partial omentectomy. Straw-colored 200 ml ascitic fluid is also aspirated during the procedure.

Gross examination of the ovarian specimen reveals a solid-cystic ovarian mass containing hair, teeth, and dirty material. In addition, numerous whitish nodules with a maximum size of about 1.2 cm were present in the greater omentum.

Figure 1. Ovary with omental fatty tissue

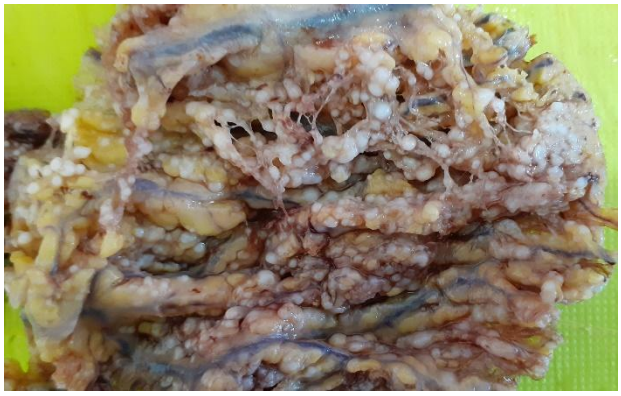
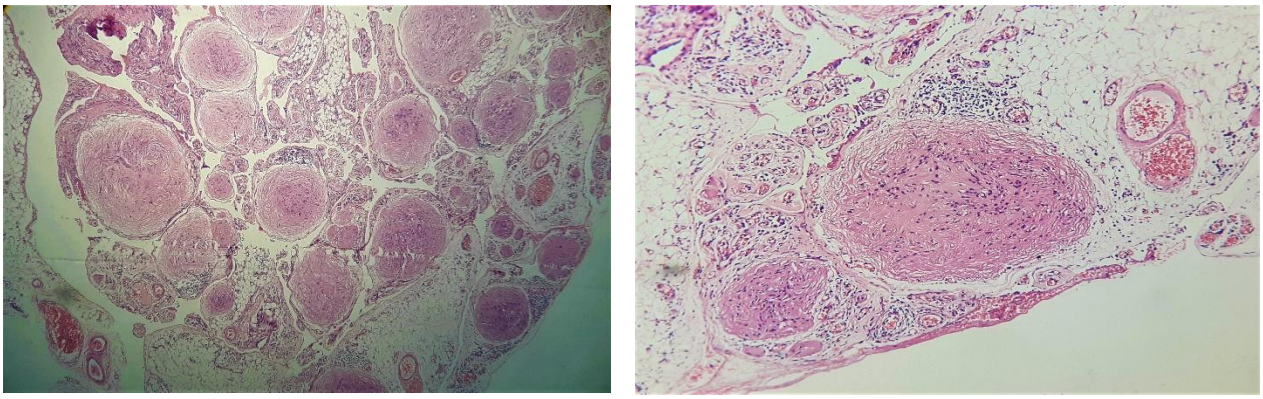


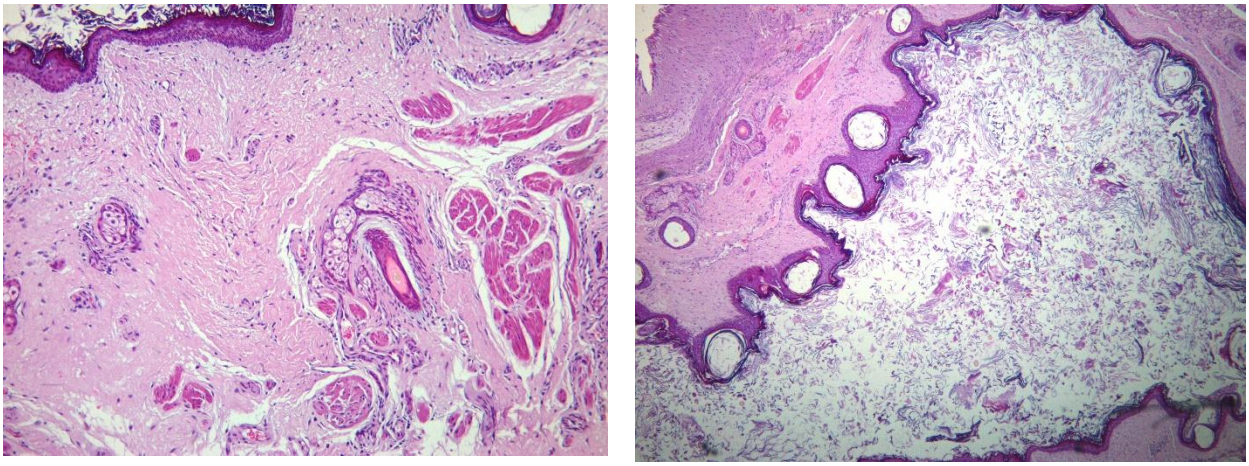
Figure 2. Omental fatty tissue with multiple tiny deposits mimicking metastases

Histopathology reveals mature cystic teratoma with solid part containing hair shafts, tooth, skin adnexal structures,mature glial tissue and mesenchymal tissue including smooth muscle cells, fatty tissue and mature glial tissue. The capsule is intact. Microscopic findings of greater omental nodules reveal mature glial tissue. Fallopian tube does not show any significant change.



A B

Figure 3. Microscopic features of multiple mature glial tissue in omentum . A. Multiple nodules of glial tissue; 4x). B. High power view shows mature glial tissue; 10x)



A B

Figure 4. Mature cystic teratoma of ovary. A. Photomicromicrograph shows skin, pilo-sebaceous units and sweat glands; 4x). B. Photomicrographs show lamellated keratin; 10x)

The postoperative course was uneventful. No further chemotherapy was administered. The patient was followed-up regularly at our institution, and no evidence of recurrence was found after 1 year

Discussion

Gliomatosis peritonei is a rare condition characterized by the implantation of glial tissue on the peritoneal surfaces, usually associated with solid ovarian teratomas. Its peak incidence occurs during the second decades of life.[2] GP is composed of miliary-like grayish white nodules of mature glial and neuronal tissue without any other teratomatous component on the peritoneal surface and omentum and, thus, is considered a benign process.[5]

Most common hypothesis of disseminated GP is intraperitoneal spread via capsular defect of the primary tumor, either spontaneous or surgical. In our case, the surgical procedure reveals left adnexal tumor with intact capsule

and slightly adhered to the greater omentum. So in this case origin of GP may be subjacent mesenchymal or peritoneal glial metaplasia. Moreover adnexal tumors show mature cystic teratoma with predominantly ectodermal and mesenchymal components. All the glial implants are matue and ascitic fluid cytology is negative for malignant cells fortunately. If immature glial tissue or other teratomatous components or both are present in the omentum, the treatment should be the same as for metastatic ovarian teratoma.[6]

As the patient is unmarried, fertility-preserving surgery is preferred in our case. Role of ancillary techniques like immunohistochemistry is also considered if the GP is found immature. Glial fibrillary acidic protein is usually positive for mature glial tissue and negative for immature glial components. There are other positive staining reactions of the glial tissue, these are GFAP, vimentin and the neuron specific enolase (NSE) and S 100. The negative staining with Ki 67 also indicates mature non-proliferating glial tissue. The staining with the antibody against CD 68 revealed quite a few macrophages scattered within the glial tissue.[9] As our case is clearly benign in nature in histopathology, we didn't go for immunohistochemistry.

Most of the cases in various studies GP is mostly associated with immature teratoma. However immature teratoma with GP showed better prognosis than would be expected based on grading of immature teratoma.[11]

Norris et al reports survival of patients with immature teratoma grades 1, 2, 3, were 82%, 63% and 30% respectively.12 Yoon et al reports 16 patiens with GP and immature teratoma are alive while reporting this review article.[13]

Though the prognosis of our case is excellent, but long term follow up is highly recommended. Because many cases were found as malignant transformation of glial components long after initial surgery.[7, 8]

According to Federation of Gynaecology and Obstetrics (FIGO) for ovarian immature teratoma, the primary tumour and implants should both be graded according to the histologic quantity of neuropithelium components. This system is widely accepted for therapeutic and prognostic implantation.[10]

Conclusion

Gliomatosis peritonei is a rare condition and generally has a favorable prognosis. If patients undergo extensive staging intraoperatively, the peritoneal implants are well sampled, and histologic description shows the implants to be completely mature, a benign clinical course can be expected Peritoneal implants in both cases usually mature glial tissue. GP with mature cystic teratoma requires only long term follow up for recurrence or malignant transformation. Recognition of benign nature of implants is pivotal to avoid unnecessary extensive surgery in young patient. Recommended for intra-operative frozen section.

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