

CASE REPORT

Mature ovarian teratoma with gliomatosis peritonei: A rare case report

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Abstract

Gliomatosis peritonei (GP) is rarely observed along with mature ovarian teratoma. However, it is important to recognize the benign nature of GP when associated with mature ovarian teratoma. Treatment for primary tumor and long-term follow-up is vital.

KEYWORDS

gliomatosis peritonei, ovarian mature teratoma

1 | INTRODUCTION

Gliomatosis peritonei (GP) is a rare condition 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.¹ As only about 100 cases of GP have been reported to date, there is no consensus on treatment and follow-up for this condition so far; however, some clinical data had demonstrated its recurrence, malignant transformation, metastasis, and even spontaneous regression.²⁻⁵

Here, we present a unique case of mature ovarian teratoma associated with gliomatosis peritonei and a review of relevant literature. This case is of interest as no cases have been published so far and is the first case to be reported from Nepal.

2 | CASE REPORT

A 21-year unmarried lady having a regular menstrual cycle presented to our institute with the complaint of vague abdominal pain and a lump in the lower abdomen for one and half months. On physical examination, all the vitals were stable. Abdominal examination revealed a non-tender, palpable mass of about 16-18 cm, soft to solid in consistency, and mobile from side to side. Examination of other systems was grossly normal.

Ultrasonography of the abdomen showed a large multilocular cystic mass measuring 12cm × 13cm with internal solid component and multiple septae in an abdominopelvic region. A contrast-enhanced CT (CECT) scan revealed a large abdominopelvic adnexal heterogeneous complex cystic solid mass measuring 27cm × 17cm × 11cm arising from the right ovary, with scattered foci of fat and

calcific component and exerting mass effects in the form of displacement of bowel loops, minimal mass effect in body, and fundus of the uterus. (Figure 1) Additionally, a nodular omental thickening suggestive of infiltration was also noted. Her tumor markers were as follows: Alpha-fetoprotein: 52.3 ng/mL (0–8), CA-125: 253.5 U/mL (<35), CA-19.9:687.16 U/mL (<37), and CEA: 6.4 ng/mL (0–5). With the suspicion of a malignant ovarian mass, after all the detailed workup, she was planned for surgery.

At laparotomy, there was straw-colored ascitic fluid of about 200ml and a huge multiloculated cystic hemorrhagic smooth glistening mass about 25cm x 20cm x 15cm with an intact capsule arising from the left ovary. (Figure 2) Cut section of the mass showed hair, teeth, brain matter, and sebaceous material. In addition, multiple subcentimeter reddish nodules with a maximum size of about 4 cm were present in the greater omentum with no such lesions in the peritoneum. (Figure 3) There were no seedlings in the diaphragm, liver, gallbladder, spleen, and kidneys. The right ovary was bulky, and thus, a wedge biopsy was done. However, bilateral tubes and uterus were normal. Few deposits were present in the rectum, which was sent for histopathology. Delicate adhesions were present between the tumor, lateral pelvic wall, and the large intestine. The frozen section of the mass was suggestive of ovarian teratoma with gliomatosis, and an immature component could not be ruled out. So, a fertility-sparing comprehensive surgery, that is, left salpingo-oophorectomy, bilateral pelvic lymph node dissection, para-aortic lymph

node sampling, multiple peritoneal biopsies, total omentectomy, and appendectomy was carried out.

Her final histopathological report revealed mature tissue derived from all three germ layers comprising of stratified squamous epithelium, skin appendages, glial tissue, adipose tissue, mucous glands, bony tissue, cartilage, choroid plexus, columnar epithelium, and pigmented cells at places. An immature component was not observed in several sections studied. (Figure 4) Sections from the omentum nodule and deposits from the rectum show mature glial tissue. (Figure 5) The peritoneal fluid was negative for malignancy. All these features were suggestive of mature teratoma of the ovary with gliomatosis peritonei.

Her postoperative period was uneventful. No adjuvant therapy was needed. The patient is on regular follow-up, and there is no evidence of recurrence at 22 months of surgery.

3 | DISCUSSION

Gliomatosis peritonei is a rare occurrence characterized by the implantation of glial tissue on the peritoneal surfaces, usually associated with solid ovarian teratomas, and its peak incidence occurs during the second decade of life.² GP is composed of miliary-like grayish 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.⁶

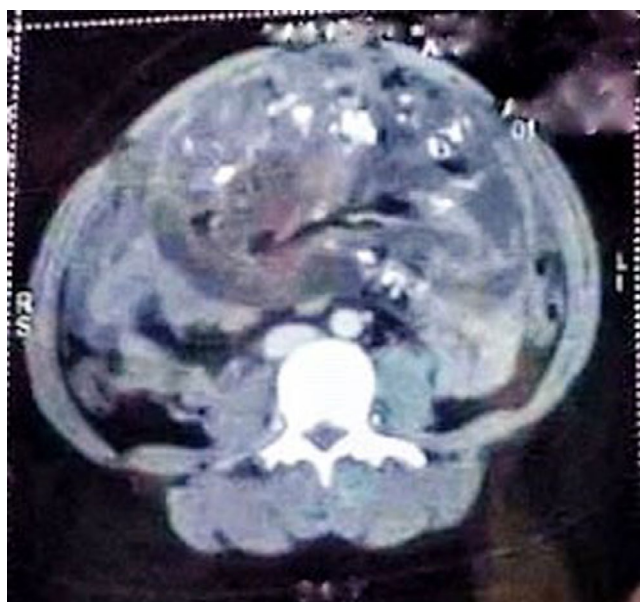


FIGURE 1 Axial image of contrast-enhanced computed tomography (CECT) of lower abdomen shows a mixed solid and cystic mass with scattered areas of fatty components and calcifications suggestive of an ovarian teratoma

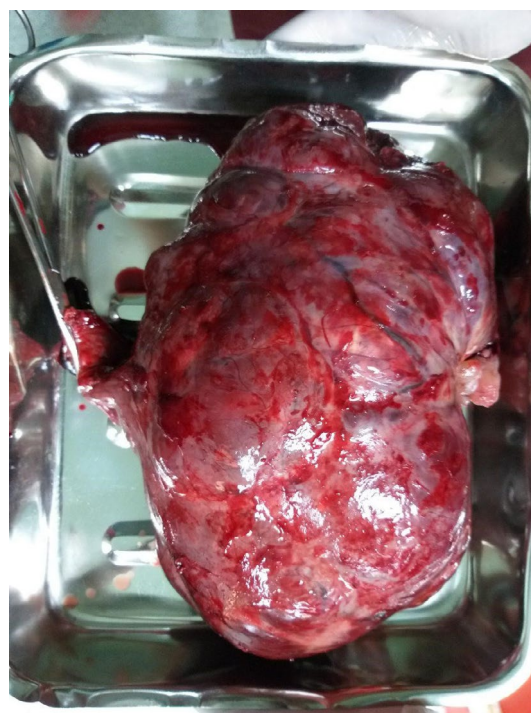


FIGURE 2 Gross specimen of huge left ovarian mass

Though the origin of GP remains uncertain, two theories to explain its etiology have been proposed. In one, implantation of glial tissue in the peritoneum occurs either through rupture of the capsule with subsequent implantation of teratomatous tissue or via lymphatic spread as in metastasis of carcinomas, while in other, pluripotent stem cells in the peritoneum or subjacent mesenchyme undergo glial metaplasia.⁷ The capsule of the ovarian teratoma in our case was intact, and also, the peritoneal fluid was free of malignant cells.

At imaging, mature cystic teratoma appears as pure cystic mass to complex solid cystic mass, and in the majority, identification of intratumoral fat components forms the key diagnostic imaging feature.⁸ GP is seen as the nodular enhancement of the peritoneum in CT, indistinguishable from conventional metastases by imaging alone.⁹ In our case, radiologically, a heterogeneously enhancing



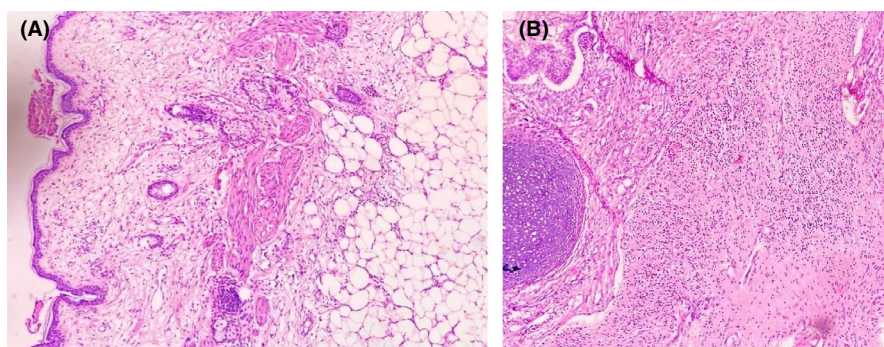
FIGURE 3 Gross specimen of greater omentum with multiple subcentimeter nodules; the maximum size of the nodule is about 3.8 cm

mass with calcification and fatty component in the pelvic cavity suggested that adnexal mass was a teratoma. As the size of the mass was more than 20cm, immature teratoma was the preferred diagnosis because immature teratomas are typically larger.¹⁰ As most of the glial tissue is small and numerous, they are often difficult to identify by ultrasound and limiting the role of USG in the diagnosing GP, which holds for our case.¹¹ Moreover, the omental deposits of GP were not identified in the CT scan in our case. Grossly, Glial tissue implants are 1–10 mm in size, usually less than 3mm, without any fatty component making them grossly indistinguishable from tuberculous peritonitis and peritoneal carcinomatosis, and therefore, the diagnosis is made by histopathological examination of the implants. Also, omental caking and ascites can be observed in some cases.⁹ In our case, numerous implants were present in the greater omentum and rectosigmoid colon with a maximum size of 4cm along with mild ascites, and the diagnosis of GP was reached after histopathological examination of the frozen section of the deposits.

As the GP is considered a grade 0 teratoma, it usually has a favorable prognosis and is managed conservatively. Further, because the lesion is often extensive, complete resection is usually demanding. Fortunately, residual GP deposits are asymptomatic and inert over a long period or may disappear over time.¹² Thus, the treatment decision is based on the grade of the primary tumor and not the glial tissue implants, provided they are extensively sampled and all are mature.⁸ However, if immature glial tissue or other teratomatous components or both are present in the peritoneum or omentum, the treatment should be the same as for metastatic ovarian teratoma.¹³ Our patient underwent fertility-preserving surgery for the mature ovarian teratoma, omentectomy alongside multiple peritoneal biopsies all of which confirmed the presence of mature glial component only.

Even though the prognosis of patients with GP is excellent, long-term follow-up, even in the face of mature peritoneal glial implants, is highly recommended because of the established cases of malignant transformation of the glial components long after the initial surgery.^{3,14} The diagnostic modality used for assessing residual masses after

FIGURE 4 Section from left ovarian mass shows features of mature cystic teratoma comprising of skin and its appendageal structures (A) ($\times 100$) and nodules of glial tissue along with mature cartilage and columnar cells (B) ($400\times$) [H&E stain]



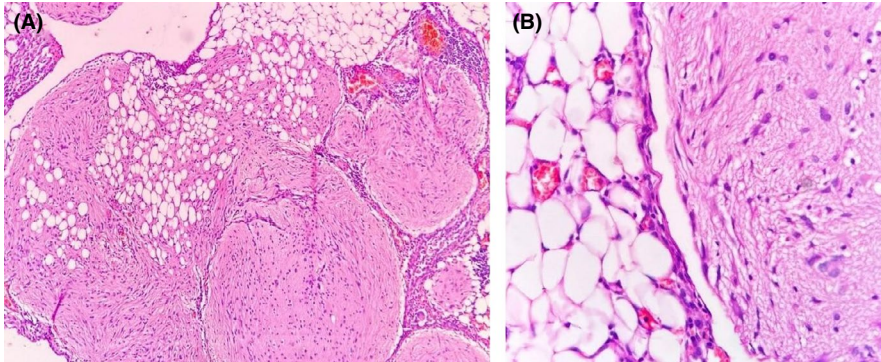


FIGURE 5 Histologic sections (A and B) from the omentum show nodules of mature glial tissue

treatment for germ cell carcinoma, that is, FDG PET/CT is now widely available; this could be useful for the follow-up of GP.¹⁵ However, as this condition is rare there is no widely accepted guidance as to how long and by which means these patients should be followed up. Our patient is on regular follow-up every six months.

4 | CONCLUSIONS

GP in mature cystic teratoma of the ovary is a rare occurrence. Vigilant conservative management with continued clinical evaluation and long-term follow-up is paramount. Moreover, it is pivotal to recognize the benign nature of the seedlings that can avoid unnecessary extensive surgery, especially in young females.

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CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

AB was involved in counseling and treatment for the patient. HPD and MS examined and interpreted the pathology. AB collected all the required case information, images, slides, and reports and contributed to writing manuscripts. AB and SS reviewed the literature and contributed to both writing and editing the manuscript. All authors read and approved the final manuscript.

ETHICAL APPROVAL

Not applicable.

CONSENT

Written informed consent was obtained from the patient before the submission of the report. The signed Institutional Consent Form is on file.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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