Mature ovarian teratoma with gliomatosis peritonei – A case report

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Abstract

Gliomatosis peritonei (GP), a rare condition related to ovarian teratomas, is characterized by miliary implants of mature glial tissues on the peritoneum or omentum. We report herein a case of mature teratoma of the ovary with GP with imaging features and pathological correlation.

Key words: CT scan, Gliomatosis peritonei, Ovarian mature teratoma

Introduction

Gliomatosis peritonei (GP) is the implantation of mature neural glial tissue on surfaces of visceral or parietal peritoneum. This condition is usually seen in patient with immature ovarian teratoma and rarely found with mature teratomas.¹ We present an unique case of mature teratoma of the ovary associated with GP.

Case History

A 20-year-old woman was admitted to gynecology outpatient department with 1-month history of vague abdominal pain and lump in the lower abdomen. Her serum a-fetoprotein (AFP) level was normal. Plain X-ray abdomen was unremarkable. Ultrasonography revealed a poorly defined heterogeneously hyperechoic mass with foci of distal shadowing. Contrast enhanced CT scan [Figure 1] showed complex solid cystic mass in left lower abdomen and pelvis. Fat containing areas and multiple foci of calcification are also seen within the mass. At laparotomy, a right ovarian tumor was found, which measured 8 cm in diameter with solid and cystic component. Delicate adhesions were noted between the tumor and the lateral pelvic wall. In addition, multiple firm, gray-white, 0.3–1.2 cm nodules were present on the anterior peritoneum, uterine serosa, and omentum. The left ovary was unremarkable. Right salpingo-oophorectomy, partial omentectomy, and biopsy of the nodules were performed. The postoperative period was uneventful. No additional therapy was given. The patient is alive and well at follow up 26 months postoperatively.

Pathologic findings

The ovarian tumor weighted 450 g and measured 8 x 5 x 3 cm³. The mass was lobulated, solid, and cystic. The capsule was breached. The solid components were
largely firm and white to pink with interspersed mucoid
regions. Cystic structures contained serous fluid with
yellow sebaceous material and keratinous debris.

Histopathologic examination revealed features of a
mature teratoma comprising skin and adnexal structures,
gut epithelium, respiratory epithelium, foci of mature
cartilage, and mature glial tissue [Figure 2]. Twenty
sections were examined from the ovarian tumor and
none of them revealed any immature element, including
primitive neuroectodermal tissue.

The peritoneal implants and the omental deposits all
revealed Grade 0 mature astroglial tissue. The glial
fibrillary acidic protein (GFAP) immunostain confirmed
the glial nature of the tissue.

Discussion

Gliomatosis peritonei, the miliary implants of mature
glial tissue on the peritoneum, is an infrequently
reported complication of ovarian teratoma. It has been
found to occur almost exclusively in females with
ovarian teratomas, though there are stray reports of its
association with pregnancy, ventriculoperitoneal shunts
performed for hydrocephalus. The mechanism of
implantation is unknown and two theories to explain
the origin of GP have been proposed. In one, glial
implants arise from the teratoma, whereas in the other,
pluripotent stem cells in the peritoneum or subjacent
mesenchyme undergo glial metaplasia.

All grades of ovarian teratomas have been described,
with immature teratomas being more commonly
associated with this condition. The condition is
relatively rare and only about 88 cases of glial implants
on the peritoneum associated with ovarian teratomas
have been reported in the literature. The first case
from India was reported by Joshi et al. in 1981 after
which three more cases had been reported. Despite
the fact that neural tissue may be found in > 30% of
mature teratomas, it is rare to have GP associated with
mature teratomas.

The prognosis of ovarian teratoma is closely associated
with tumor grade as proposed by Thurlback and Scully, which was modified by Norris et al. In the
series by Norris et al. of 58 patients, 5-year survival rate
for patients with Grades 1–3 were 82, 63, and 30%,
respectively. Paradoxically, patients who have
immature ovarian teratomas in association with mature
glial implants appear to have a much improved
prognosis. This statement holds true only if stringent
criteria for diagnosis of GP is adhered to, as proposed
by Thurlback and Scully: (a) peritoneal surface,
omentum, and diaphragmatic surfaces must be
extensively sampled histologically and (b) each of the
sampled implants should be composed exclusively, or
almost exclusively, of Grade 0 glial tissue. If these
two conditions are met, the prognosis of the disease is
excellent.

There are 11 cases reported so far, that had an adverse
outcome. Shefren et al. reported a 16-year-old girl
with Grade 3 teratoma, who developed malignant glial
peritoneal implants and died 5.5 years after her initial
surgery. Dadmanesh et al. reported a patient with
Grade 1 teratoma and Grade 1 immature glial implant
diagnosed 10 months after initial operation, who died
8 years after oophorectomy. These two cases are of
interest in this context as both the cases developed
malignancy after a long symptom-free interval.

Regarding treatment, therapy should be directed by the
grade of the primary tumor and not by the glial
implants, if they are extensively sampled and all are
mature. However, extensive sampling of all peritoneal
implants is important. If no other teratomatous
elements or malignant glial tissue is found in the
implants, the mature glial implants can be ignored and
the method of therapy should be judged only by the
stage and grade of the primary ovarian teratoma.
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. Despite often wide spread
involvement of peritoneal surfaces, GP is reported to
impart an improved prognosis even in high-grade
ovarian teratomas.

In summary, GP is a rare condition that generally
imparts a favorable prognosis to patients with ovarian
teratomas. If patients undergo extensive staging,
peritoneal implants are well sampled, and histologic

Figure 2: Photomicrograph of the ovarian mass showing
keratinized epithelium with underlying appendageal structures,
consistent with mature teratoma (H and E, x200). Inset shows the
glial nodule in the omentum, which is immunoreactive for glial
fibrillary acidic protein (GFAP) (Immunoperoxidase technique
x400)
description shows the implants to be completely mature, a benign clinical course is to be expected. However, long-term follow up, even in the face of mature peritoneal glial implants is highly recommended because of established cases of malignant transformation of the glial components long after initial surgery.

References


