

## Case Report

# A Rare Case of Immature Ovarian Teratoma with Gliomatosis Peritonei

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### ABSTRACT

Gliomatosis peritonei (GP) is a rare condition that occurs almost exclusively in the setting of ovarian immature teratoma. It is characterized by the occurrence of nodules of mature glial tissues in the peritoneum, omentum and bowel wall. The glial tissue in such cases is usually low grade although there have been cases of malignant evolution described. In general, the prognosis for GP is good. It depends chiefly on the degree of maturity of the implants. In mature GP, usually no additional chemotherapy is necessary. In immature GP, chemotherapy can induce maturation of the implants. We present a case of immature ovarian teratoma associated with low grade GP.

**KEY WORDS:** Glial tissue, gliomatosis peritonei, immature teratoma, India, ovary

## INTRODUCTION

Immature teratoma is a preferred term for the malignant ovarian teratoma usually seen in children and adolescents and is composed of a mixture of embryonal and adult tissues derived from all three germ layers regardless of its gross appearance.<sup>[1]</sup> It spreads on pelvic, abdominal peritoneum and omentum.

In patients with extra ovarian spread, the microscopic appearance of the metastasis is of prognostic importance. Some peritoneal implants or lymph node metastasis contain only mature tissues and do not adversely affect the prognosis. These implants are usually composed partly or completely of mature glial tissue (gliomatosis peritonei (GP)).<sup>[2]</sup>

Despite often widespread involvement of peritoneal surface, GP is reported to impart an improved prognosis even in high grade teratomas. However, a long-term follow up even in the face of mature glial implants is highly recommended because of established cases of malignant transformation of glial components long after initial surgery.<sup>[3]</sup>

## CASE REPORT

A 22-year-old female came to out-patient department of Obstetrics and Gynecology of NKPSIMS and Lata Mangeshkar Hospital, Nagpur, with a history of abdominal pain and increase in abdominal volume of 5-6 months duration. Her family history was unremarkable. Physical examination revealed a huge abdominopelvic mass. Ultrasound and CT scan revealed an abdominopelvic mass of 35 cm × 30 cm diameter arising from the left ovary without lymphadenopathy or liver metastasis.

Preoperative tumor markers were high with an alpha fetoprotein (AFP) levels of 3100 ng/ml and Ca 125 level of 3600 U/ml.

Patients presented with this complaint to outpatient department (OPD) of NKPSIMS.

Laparotomy revealed huge left-sided ovarian mass. There was parametrial thickening on both the sides. Peritoneum, omentum and peritoneal surfaces were grossly normal. No nodules were seen.

### Gross

Left ovarian mass weighed 8 kg and measured 35 cm × 30 cm × 15 cm. The external surface was bosselated [Figure 1] and a cut section showed multiple nodular solid areas and cystic spaces with mucoid

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material [Figure 2]. Gritty areas were also present. Uterus, cervix, right ovary and fallopian tube were unremarkable. Parametrium was thickened on both the sides. A specimen of omentum was received measuring 30 × 10 cm. Grossly no nodules or deposits were visible.

### Microscopy

Sections from the ovarian mass revealed a tumor composed of mixture of mature and immature elements from all the three germ layers. Mature adipose tissue, cartilage, stratified squamous epithelium and glandular spaces lined by columnar to flattened cells were seen [Figure 3]. Foci of fibrillary neural tissue, glial tissue, ganglion cells, melanin pigment, areas of calcification, skin and adnexal structure were also seen. Focal areas showed primitive mesenchyme. Immature neural tissue was seen in the form of primitive neural tubes and rosettes [Figure 4]. Areas suggesting yolk sac tumor or embryonal carcinoma were not seen. These histological features were suggestive of immature teratoma (Grade 1).



**Figure 1:** Gross photograph showing a tumor (mass) with multinodular appearance on external surface



**Figure 2:** Gross photograph of the cut surface of the tumor showing multiple nodular and cystic areas

Sections from parametrium showed mature teratoma elements. Foci of adipose tissue, cartilage and glandular spaces were seen. Sections from omentum revealed many foci of mature glial tissue (Grade 0) [Figure 5]. There was no nuclear atypia or mitoses.

A diagnosis of immature teratoma (Grade 1) with multiple glial implants in omentum (Grade 0) was made.

### DISCUSSION

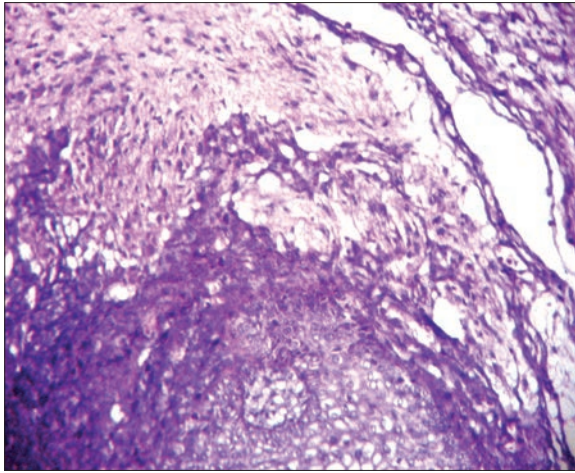
Immature teratoma is a preferred term for the malignant ovarian teratoma usually seen in first and second decade of life. It is composed of mixture of embryonal and adult tissues derived from all three germ layers regardless of its gross appearance.<sup>[1]</sup> Immature teratoma represents 3% of all teratoma, 1% of all ovarian cancer and 20% of malignant ovarian germ cell tumors. In grading of immature teratoma, primitive neural tubes and immature rosettes are counted.<sup>[4]</sup>

Immature teratoma is a predominantly solid, unilateral tumor that averages 18 cm in diameter. The solid component is gray or brown and soft to hard in consistency. Scattered small cysts are typically seen on the cut surface.<sup>[2]</sup> If keratinous debris or hairs are seen it may resemble a dermoid cyst.

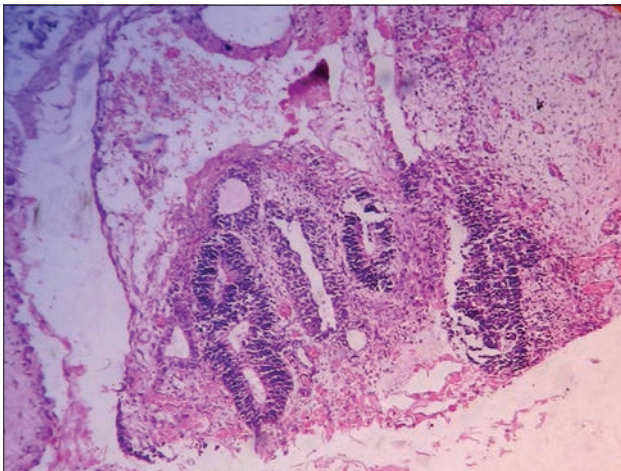
Components of all the three germ layers are present and a mixture of mature or immature elements with haphazard distribution can be seen. The immature elements are mainly mesenchymal and ectodermal in origin. Immature neuroectodermal tissue is the easiest immature tissue to recognize and quantitate. Patients may sometimes present with paraneoplastic syndrome like limbic encephalitis.<sup>[4]</sup>

In patients with extra ovarian spread, the microscopic appearance of the metastasis is of prognostic importance. Some peritoneal implants or lymph node metastases contain only mature tissues and do not adversely affect the prognosis. These grade 0 implants are usually composed partly or completely of mature glial tissue. GP is a rare occurrence and has been found exclusively in females with ovarian teratoma (immature and rarely in mature), though there are stray reports of its association with pregnancy and ventriculoperitoneal shunts performed for hydrocephalus.<sup>[3,5]</sup> The first case of immature teratoma with GP from India was reported by Joshi *et al.* in 1981.<sup>[3]</sup> A review of the literature reported by Chou *et al.* had found 65 cases of GP, which have favorable prognosis after surgical treatment.<sup>[6]</sup>

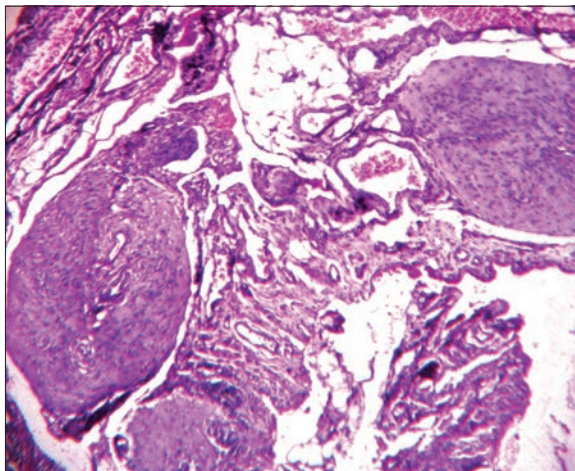
The mechanism of implantation is unknown and two theories to explain the origin of GP have been proposed. In one glial implant arise from the teratoma and in



**Figure 3:** Photomicrograph showing mature elements cartilage, glandular epithelium (H and E, x100)



**Figure 4:** Photomicrograph showing immature neural tissue (H and E, x100)



**Figure 5:** Photomicrograph showing glial implants in peritoneum (H and E, x100)

the other, pluripotent stem cells in the peritoneum or adjacent mesenchyme undergo glial metaplasia.<sup>[6]</sup> Recent molecular studies of glial implants have shown that they

are genetically different from an ovarian tumor but have same genetic patterns as patient.<sup>[2]</sup> Substances produced by a tumor result in metaplastic transformation of peritoneal or subperitoneal tissue into glial tissue.<sup>[2]</sup>

Patients with recurrent teratoma may be affected with the tumor-prone syndrome, where one or more peritoneal cell types or population are predisposed to neoplastic conversion and formation of tumors as a result of an endogenous or exogenous neoplastic stimuli.<sup>[7]</sup>

Glial fibrillary acidic protein (GFAP) is an intermediate filament protein that expresses with the development of astrocytes in the fetal nerve tissue. GFAP immunostain confirms the glial nature of the tissue. A strong expression of GFAP often suggests that tumor cells are mature and well differentiated.<sup>[8]</sup>

Surgery and multi-dose chemotherapy is the main line of treatment. In general, the prognosis of GP is good. It depends chiefly on the degree of maturity of the implants. In mature GP, usually no additional chemotherapy is necessary. In immature GP chemotherapy can induce maturation of the implants.<sup>[9]</sup>

The growing teratoma syndrome (GTS) is defined as conversion of a metastatic immature teratoma into a metastatic mature teratoma as a result of chemotherapy. GTS rarely occurs in association with ovarian germ cell tumors.<sup>[10]</sup> The prognosis of this benign entity however remains favorable, with the survival of up to 9 years in completely resected cases.

Recurrence is influenced by several factors, such as tumor grade, growth pattern, capsular rupture and vascular invasion.<sup>[8]</sup> It is also important to separate immature teratomas that have a yolk sac or embryonal tumor pattern, since the prognosis substantially decreases under these circumstances.<sup>[8]</sup>

The risk of metastasis correlates with the amount of immature neuroepithelial tissue present and this is taken into account for grading according to the system of Robby and Scully modified by Norris *et al.*<sup>[10]</sup>

When associated with mature glial implants within the peritoneum the prognosis is usually much better, Irrespective of the original tumor grade.<sup>[10]</sup> There have been rare descriptions of cases of mature GP which have evolved into malignant tumors long after initial surgery. Hence, long-term follow up even in the face of mature glial implants is highly recommended.

On follow up our patient was found to be disease free 2 years after the surgery.

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