



## Immature Ovarian Teratoma (Grade 3) Associated ith Massive Gliomatosis Peritonei

Authors

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### Abstract

*Immature teratoma is defined as a teratoma containing a variable amount of immature, embryonal type tissue. We hereby present a case of immature teratoma in a 12 years old girl presented with a history of abdominal pain and gradual increase in the abdominal size with high preoperative tumor markers like AFP, CA 125 and CEA levels. Histopathology revealed an immature teratoma grade 3 associated with massive gliomatosis peritonei of omentum and pouch of Douglas.*

**Keywords:** Ovarian teratoma, Immature teratoma, Gliomatosis peritonei.

### Introduction

Immature teratoma is the currently preferred term for the malignant ovarian teratoma usually seen in children and adolescents and composed of a mixture of embryonal and adult tissues derived from all three germ layers, regardless of its gross appearance.<sup>[1]</sup> The main immature component is usually neuroepithelial, but mesodermal elements are also common.

In patients with extra ovarian spread, the microscopic appearance of the metastasis if of prognostic importance. These implants are usually composed partly or completely of mature glial tissue (gliomatosis peritonei).<sup>[2]</sup> When the appearance of implants is immature, the prognosis is more guarded than in gliomatosis peritonei.

### Case Report

A 12 years old female came to the outpatient department of Surgery of our hospital, with a history of abdominal pain and gradual increase in the abdominal size for 6-7 months. Her family

histories were all unremarkable. CECT of whole abdomen revealed a large, complex, solid cystic, mildly enhancing pelvic SOL (17 x 9.5cm in trans axis) with an associated right kidney hydronephrosis.

MRI revealed an evidence of a huge, complex, lobulated mildly T2 hyperintense solid and non-enhancing cystic lesion seen to occupy abdomen and adjacent pelvic cavity, extending from infrapancreatic region above up to the false pelvis below.

Multiple foci of calcifications were noted. No intralesional fatty components were seen. The lesion approximately measured about 96mm x 179mm x 225mm.

Preoperative tumor markers were high with an alpha fetoprotein (AFP) levels of 171.48 ng/ml, CA125 level of 688.8 U/ml and CEA level of 18.79 ng/ml.

Laparotomy revealed a huge left sided ovarian mass, and the omentum and peritoneum of Pouch of Douglas were finely granular. Left sided

salphingo oophorectomy was done followed by omentectomy and biopsy of peritoneal implants from Pouch of Douglas and the entire specimen was sent to our Pathology department.

**Gross**

Left salphingo-oophorectomy specimen showed a well circumscribed, encapsulated, large ovarian tumor, 20 cm in diameter. The tumor was partly solid and partly cystic. The cut surface showed greyish white, bony and cartilaginous areas. The specimen of omentum measured 17 cm in greatest dimension and was greyish white in color. The larger deposit from Pouch measured 2 cm in maximum dimension.

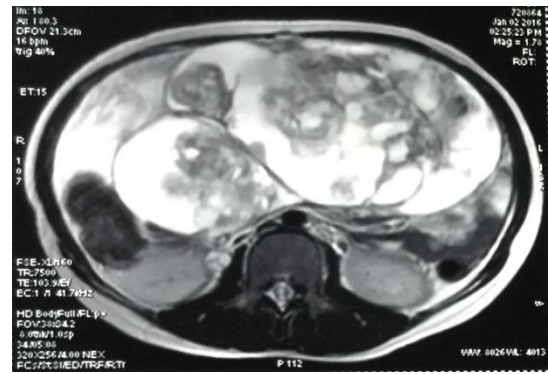
**Microscopy**

Sections from ovarian mass revealed a tumor composed of ectodermal, mesodermal and endodermal elements. Skin and its appendages, cartilage, bone, muscle tissue, choroid plexus, glial tissue, lymphoid tissue and adipose tissue were present. Majority of the elements was mature. Mucin secreting glands were also present. Focal areas of immature mesenchymalelements and neuroepithelial elements were seen. The immatureneuroepithelium in the form of rosettes and tubes were present in four low power field and therefore the teratoma was graded grade 3.

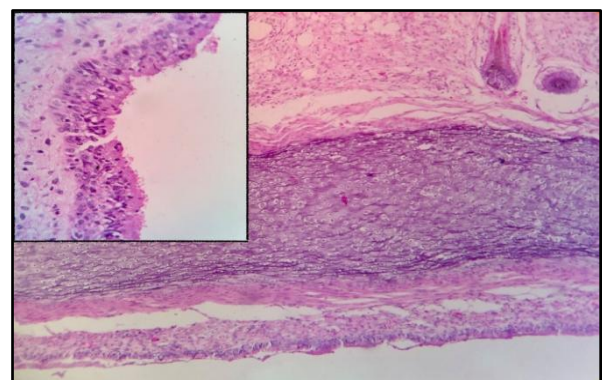
The Fallopian tube was free, however, the capsule of ovary was seen to be involved with presence of teratomatous elements just beneath the inked margin.

The omental tissue and the tissue from Pouch of Douglas showed loose textured fibroadipose tissue with sheets and nodules of glial cells along with focal areas of proliferation of mesothelial cells.

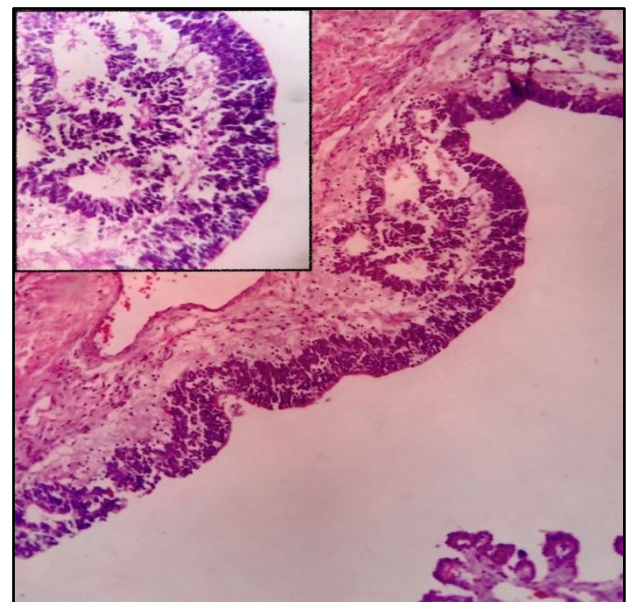
A diagnosis of immature teratoma (Grade-III) ovary with gliomatosis of omentum and Pouch of Douglas was made.



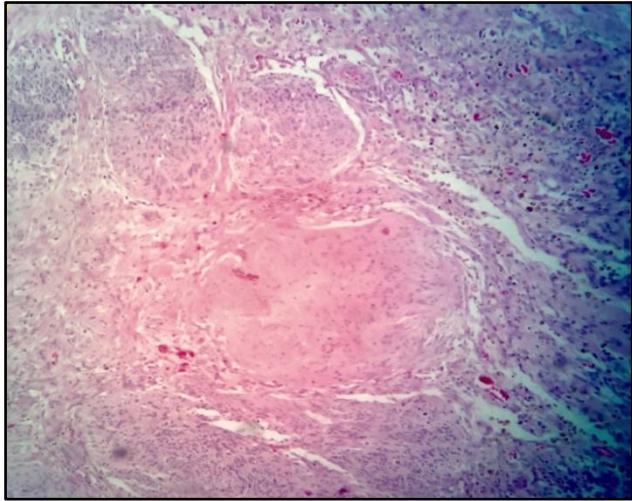
**Figure 1** MRI of whole abdomen showing complex, lobulated, solid and non-enhancing cystic lesion measuring 96mm x 179mm x 225mm.



**Figure 2** Section showing cartilage and hair follicle, inset shows respiratory epithelium. (Hematoxylin- Eosin section)



**Figure 3** Section showing primitive neuroepithelium and choroid plexus, inset shows the primitive neuroepithelium (Hematoxylin-Eosin section).



**Section 4** Section from Pouch of Douglas (Hematoxylin- Eosin section).

### Discussion

According to World Health Organisation, immature teratoma is defined as a teratoma containing a variable amount of immature, embryonal type (generally immature neuroectodermal) tissue.<sup>[3]</sup>

The great majority of teratomas are composed of tissues representing at least two, but usually all three embryonic layers. If the neoplastic tissue is uniformly mature, the tumor is termed mature teratoma (almost always a dermoid cyst); the presence of any immature tissue with an embryonal appearance warrants a designation of immature teratoma.<sup>[4]</sup>

Immature teratomas are the third most common of the primitive germ cell tumors, accounting for almost 20% of all cases and 10% to 20% of cases encountered in the first two decades of life.<sup>[5][6][7][8][9]</sup>

Patients usually present with abdominal pain and abdominal mass over a period. In the present study, the patient presented with the same set of complaints.

It occurs essentially during the first two decades of life as in our case, which presented at 12 years of age.

Immature teratomas of ovary are almost always unilateral and occasionally have elevated serum AFP levels.<sup>[10]</sup> In the present study, the tumor was unilateral with an elevated AFP, CA 125 and CEA levels.

Immature teratoma is usually large, with a median diameter of 18 cm, and form encapsulated masses with smooth, glistening outer surfaces. The cut surfaces are predominantly solid in most cases, but small cysts containing mucinous, serous, or bloody fluid, or hair are frequently present.<sup>[4]</sup> In the present case, the ovarian tumor was well circumscribed, encapsulated with a maximum diameter of 20cm. On cut section, it was partly solid and partly cystic.

The immature, embryonic-type tissue varies from small foci to a predominant component and is composed of neuroectodermal elements. These elements consist of neuroepithelial rosettes and tubules, cellular foci of mitotically active glia, and, occasionally, small areas resembling glioblastoma multiforme or neuroblastoma.<sup>[4]</sup>

It is noted that the amount of neuroepithelium correlates with survival and is the basis of grading these tumors. The only type of neural tissue that should be counted in grading a tumor for immaturity is primitive neural tubes and immature rosettes.<sup>[11]</sup>

In a grade 1 immature teratoma, immature neuroepithelium is limited to 1 low power field (lpf). Grade 2 has immature neuroepithelium in 2 or 3 lpf on any slide. A grade 3 contains immature neuroepithelium in 4 or more lpf on any slide. In our case, the immature neuroepithelium in the form of rosettes and tubes were present in 4 lpf and therefore a diagnosis of Grade 3 was given.

Tumor markers serologically evaluate germ cell tumors. In case of immature teratoma, AFP is widely used since occasionally patients with immature teratoma have elevated serum AFP levels. However, many published articles state that the AFP levels in immature teratoma is not correlated to either stage or grade. Diagnosis of recurrence of immature teratoma by tumor markers appear to be more sensitive when combined with detection of CA 125, CA 153 and AFP.<sup>[12]</sup> Gliomatosis peritonei can be defined as the metastatic implantation of glial tissue on surfaces of visceral or parietal

peritoneum. Gliomatosis peritonei 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 ventriculo-peritoneal shunts performed for hydrocephalus.<sup>[13][14]</sup>

The mechanism of implantation is unknown and two hypotheses to explain the origin of gliomatosis peritonei have been proposed. The first hypothesis suggests that glial foci arise from primary teratoma through rupture of capsule with subsequent implantation in peritoneum.<sup>[15]</sup> The second hypothesis suggested that glial foci are genetically unrelated and pluripotent stem cells in the peritoneum or adjacent mesenchyme undergo glial metaplasia.<sup>[16]</sup>

According to two defining criteria of gliomatosis peritonei proposed by Thulrbeck and Scully,<sup>[17]</sup> it should be composed of entirely mature glial tissue.<sup>[18][17]</sup> Other cases where immature glial implants were found should be diagnosed as metastatic teratoma and require further aggressive therapy. Our present case showed massive glial implants both in the omentum and peritoneum of Pouch of Douglas. These implants were composed of mature glial tissues only.

Immature teratomas are treated based on FIGO (International Federation of Gynecologists and Obstetricians) staging and grading of tumor. Since this is a tumor primarily occurring in young patients, preservation of fertility is an important factor in its management. Grade I and FIGO Stage I are usually treated with unilateral oophorectomy.<sup>[19][20]</sup> Grade II and III with advanced stage receive adjuvant chemotherapy in addition to surgery.<sup>[8]</sup> Our present case was diagnosed with Grade III immature teratoma and hence was referred to the Oncology department for adjuvant chemotherapy. She has received 6 cycles of treatment with Bleomycin/Etoposide/Cisplatin. At present the patient is being called for follow up every 6 months and she is doing well without any evidence of any residual disease or recurrence.

## Conclusion

GP is a very rare benign condition associated almost exclusively with immature teratoma. Despite its widespread involvement, it does not affect the prognosis. Ovarian teratomas commonly have a slow course and most cases are diagnosed accidentally or incidentally. Early diagnosis in case of pelvic mass in children helps to formulate appropriate management protocol thus in turn causing minimum impact on life span. Immature teratoma associated with gliomatosis peritonei, surgery with adjuvant chemotherapy is recommended. It can give longer survival even in recurrent disease. Scanning techniques like MRI may have a role in monitoring patients for many years for patients having immature teratoma associated with gliomatosis peritonei.

**Consent:** Written informed consent was obtained from the patient.

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