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Case Report

Gliomatosis peritonei arising in setting of immature teratoma of ovary: a case report

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ABSTRACT

A 14 years old girl presented to the gynecology OPD with pain abdomen and huge abdominal lump since 2 months. On clinical examination, a large mass of 20x15 cm size was found extended upto the xiphoid process. Serum studies showed rise of CA-125 up to 406.9U/mL and LDH up to 310U/L. USG shows right ovarian cyst of 14.8x14.1x12.8 cm with internal calcification. MRI revealed a well encapsulated mass of 21x19x17cm with solid and cystic mass and upward peritoneal extension. Exploratory laparotomy was performed with right sided salpingo- ophorectomy with infracolic omentectomy, as the omentum appeared granular. She had an uneventful post-operative recovery. Subsequently HPE showed immature teratoma NORRIS grade 3 with co-existent peritoneal gliomatosis (grade 0). She is under regular follow-up and decided to give six cycles of combination chemotherapy with BEP at regional cancer hospital.

Keywords: Abdominal mass, Chemotherapy, Gliomatosis, HPE, Immature teratoma, Laparotomy

INTRODUCTION

Gliomatosis peritonei (GP) is an infrequent occurrence, exclusively associated with a mature or immature teratoma of the ovary. GP is defined as miliary implantation of glial tissues on the surface of the visceral or parietal peritoneum with secondary maturation into glial nodules of 1-10mm. Robboy and Scully have suggested three possible sources of GP:

- Deposition of immature neural tissue with consequent maturation
- Lymphogenous metastasis; and
- Mature glial cells extruded through a defect in the capsule of the primary tumor.[1]

Surgery is the basic treatment for both mature and immature teratomas as well as for peritoneal gliomatosis.[2] In immature teratoma associated with GP, combined chemotherapy is recommended. Surgery and chemotherapy can give longer survival even in recurrent disease.[3]

CASE REPORT

A 14 years old girl presented to the gynecology outpatient department (OPD) of AGMC and GBP hospital Agartala March 2019 with pain in abdomen and huge abdominal lump since 2 months. Patient had attained menarche one year back and had normal menstrual history. She had poor appetite, weight loss and

backache. Physical examination showed large abdominal mass and clear abdominal veins. A large mass, 20×15cm in size, was found extending up to the xiphisternum mass was firm surface, variegated feel immobile at some places, without tenderness or rebound tenderness. No enlarged lymph nodes were felt. She was moderately pale. (CBC), (LFT), (KFT) were within normal limits. Serum α-fetoprotein (AFP) level was 122.7ng/mL, serum human chorionic gonadotropin (βHCG) was 0.1.0mIU/mL, CA-125 was 406.9U/mL, lactate dehydrogenase (LDH) was 310U/L. Chest radiograph was normal. USG-shows right ovarian cyst 14.8cmx14.1x12.8cm well defined cystic lesion with internal calcification.

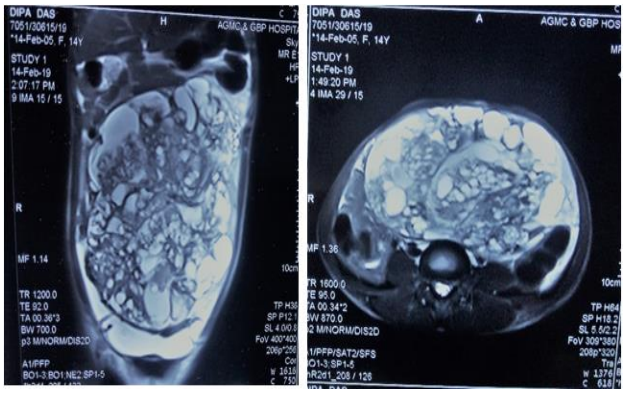

Figure 1: MRI revealed well encapsulated mass with heterogenous, solid as well as cystic mass in pelvic cavity with extension upward to peritoneal cavity left ovary was unremarkable.

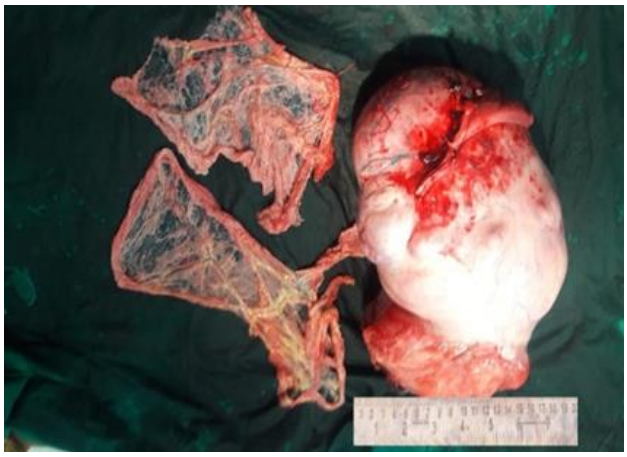

Figure 2: Gross specimen of the huge mass with omentum.

MRI (Figure 1) revealed a 21x19x17cm evidence of well encapsulated mass with heterogenous, solid as well as cystic mass in pelvic cavity with extension upward to peritoneal cavity left ovary was unremarkable. Staging laparotomy was subsequently performed, ascitic fluid was sent for cytology. Exploration revealed the same huge mass (Figure 2), with adhesions around omentum appeared granular. No deposits were found elsewhere, neither any enlargement of nodes. Right sided salpingo-oophorectomy with infra colic omentectomy was done.

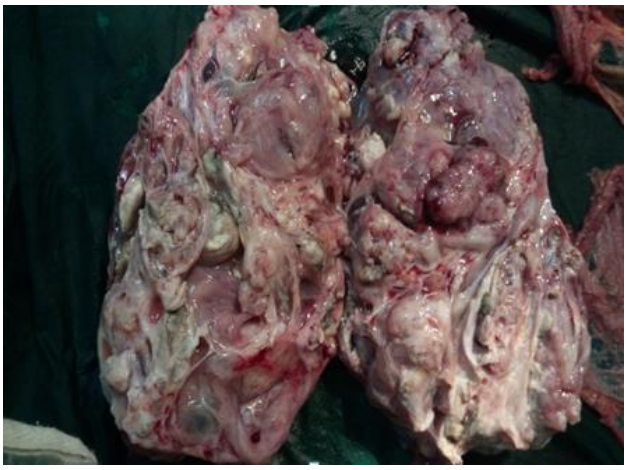

Figure 3: Cut section of huge mass showing solid and cystic areas.

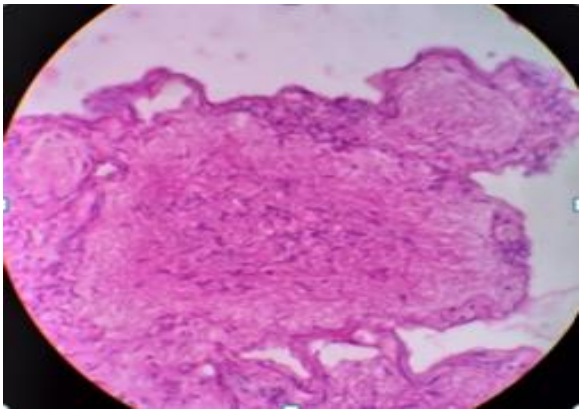

Figure 4: Multiple nodules demonstrating a variety of sizes in the greater omentum, with numerous glial tissues (staining, hematoxylin and eosin; magnification, ×40).

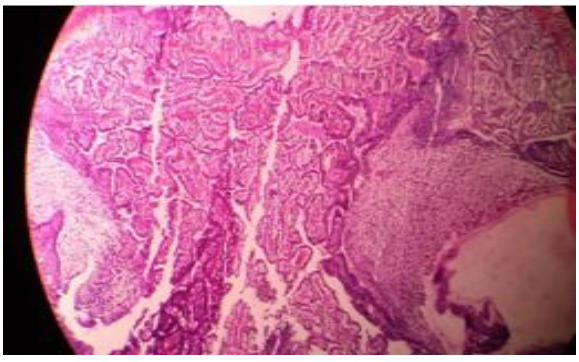

Figure 5: Multiple sections shows mucinous epithelium and cartilage (staining, hematoxylin and eosin; magnification, ×40).

Cut section shows (Figure 3) solid and cystic areas with varigated appearance. Patient had an uneventful post-

operative recovery. Peritoneal fluid was negative for malignant cells on cytological examination. Microscopic examination shows (Figures 4 to 7) large areas of glial tissue, squamous epithelium with keratin, respiratory epithelium, hair follicle, fat, immature cartilage, sebaceous glands and group of mucus glands. Few areas show neuroepithelial tissue occupying <4 low power field. Sections from omentum shows mature glial tissue are surrounded by cuff of lymphocytes representing peritonel implants of mature glial tissue. The diagnosis was immature teratoma NORRIS grade 3 with coexistent peritoneal gliomatosis (grade 0). Immature teratoma component comprised 80% of the tumor. The fallopian tube showed normal histology. She is under regular follow up. In joint oncology meeting, it has been decided to start chemotherapy with multi agent BEP.

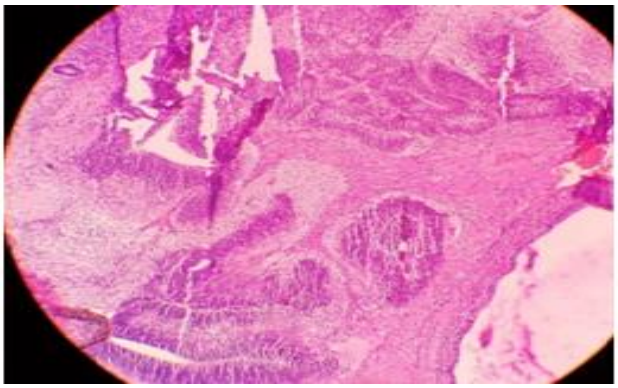

Figure 6: Multiple sections shows neuroepithelium.

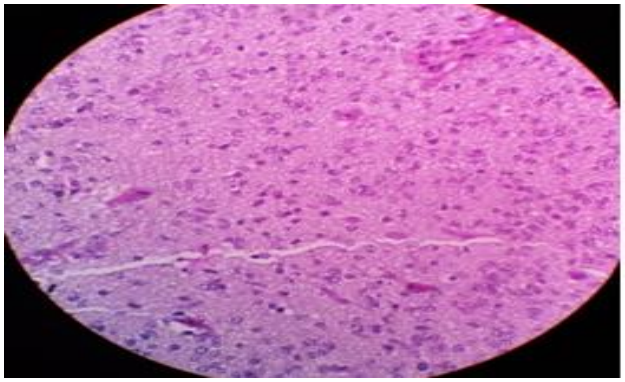

Figure 7: Microscopic view of the omental implants exhibiting mature glial tissues, consistent with gliomatosis peritonei (staining, hematoxylin and eosin; magnification, ×100).

DISCUSSION

Immature teratoma (IT) is the currently preferred term for the malignant ovarian teratoma composed of a mixture of embryonal and adult tissues derived from all three germ layers. According to WHO, IT is defined as a teratoma containing a variable amount of immature embryonal type (generally) neuroectodermal tissue.[3]

The lack of 12p amplification in immature ovarian teratomas, in contrast to it spresence in other types of malignant ovarian germ cell tumor, demonstrated a different pathogenesis compared to other malignant ovarian germ cell tumors.[4] IT of the ovary is almost always unilateral and is a tumor of children and adolescents, that occurs essentially during the first two decades of life.[5]

Gliomatosis peritonei (GP) can be defined as the metastatic implantation of glial tissue on the surfaces of visceral or parietal peritoneum.[3] It has been found to occur almost exclusively in females with ovarian teratomas, although there are stray reports of its association with pregnancy and ventriculoperitoneal shunts performed for hydrocephalus. Immature teratomas being more commonly associated with this condition.[6]

The mechanism of implantation is unknown, and two theories to explain the origin of GP have been proposed. According to one of the theories, glial implants arise from the teratoma, whereas the other proposes that pluripotent stem cells in the peritoneum or subjacent mesenchyme undergo glial metaplasia.[6] Molecular studies suggest that ovarian teratoma and GP are genetically distinct.[3]

The nodules of glial implants are usually 1-10mm in size, localizing in the parietal and visceral peritonei, and are grossly indistinguishable from tuberculosis or carcinomatosis. Microscopically, GPs may consist of mature or immature glial tissues. The mature nature of the implants generally implies a favorable prognosis, even in patients with immature ovarian teratomas.[1]

Macroscopically, peritoneal implants are small in size, well circumscribed, and have a grayish color. Microscopically, implants are composed of mature glial tissue regardless of the nature of the teratoma. When peritoneal implants contain immature glial tissue, one must rule out metastasis of immature ovarian teratoma.[2] The mature nature of glial tissue is reflected by its immunopositivity for vimentin and neural markers like Neuron specific enolase (NSE), glial fibrillary acidic protein (GFAP), and S100. Negativity for Mindbomb E3 Ubiquitin Protein Ligase 1 (MIB1) is used to rule out malignant transformation, and negative AFP rules out metastasis from an immature germ cell tumor.[1,2]

Recent study indicates that Oct4 might serve as a promising biomarker for the diagnosis of highly malignant cases of immature teratoma because Oct4 expression was exclusively detected in immature neuroepithelium of high-grade immature teratomas.[7]

These peritoneal implants may undergo fibrosis and eventually disappear or sometimes persist without any morphological changes. In rare circumstances, they can undergo transformation to malignant tissue (glial or teratomatous).[2]

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.[6] The treatment mode for IT and GP is complete surgical resection, which is also useful for identifying the presence or the absence of malignant lesions and for preventing malignancy transformation of the GP residual fragments. Because the lesions are extensive, complete excision is usually very difficult.[3] Potential for recurrence is high, and there for requires a careful monitoring of residual lesions using scanning imaging such as computed tomography. The prognosis of IT heavily depends on the FIGO stage.[8] It is influenced by several factors, such as tumor grade, growth pattern, capsular rupture and vascular invasion. It is important to separate from this group the teratomas that also have a yolk sac tumor pattern, since the prognosis substantially decreases under this circumstance.[9]

There is no clear guidance as regards how long these patients should be followed up. There is no consensus about the duration of follow-up care for these patients. England et al proposed MRI and tumor markers for the monitoring of patients with immature ovarian teratoma and mature glial tissue implants. CT and ultrasound have also been proposed for monitoring of the disease.[6]

A favorable prognosis is determined by the following:

- Histological nature of glial tissue implants that are completely mature regardless of the nature of immature ovarian teratoma; and
- Loss of proliferative activity of the peritoneal implants.[2]

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 are adhered to, as proposed by Thurl back 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.[6]

CONCLUSION

A mature gliomatosis implant usually constitutes a harmless situation with a good prognosis, even when associated with an immature teratoma of the ovary.

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