CASE REPORTS

IMMATURE OVARIAN TERATOMA WITH GLIOMATOSIS PERITONEI- AN UNUSUAL FINDING

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Abstract

Immature ovarian teratoma is a rarely seen germ cell tumor and Gliomatosis peritonei (GP) is a rare condition that occurs almost exclusively in the setting of ovarian immature teratoma. It is characterisized 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, ovary*

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Introduction

Immature teratomas are made of tissues that resemble those found in an embryo, and are the malignant cousins of the very common mature cystic teratomas or dermoid cysts. They may occur in combination with other germ cell tumors and are then called "mixed germ cell tumors." Pure immature teratoma is extremely rare and represents approximately 1% of all ovarian cancers, but within the germ cell tumor group, it is the second most common malignancy.[10]

Case report

A 21 year old lady came to a local physician with complaints of heaviness in whole abdomen with nausea, vomiting for 5 months. Her menstrual cycle was regular with average flow and duration. Her husband was in abroad. She had one child of 3 years. She was not well nourished. On abdominal examination revealed a hugely enlarged mass about 20×17 cm size, mobile, non tender, located in mid abdomen. On per vaginal examination, uterus was normal size, antiverted, fornices were free and mass was not continuous with uterus.

Her physician had taken USG guided FNAC from the mass. Biopsy report showed suggestive of malignant stromal cell tumor of ovary. So she was referred to gynae oncologist for further management. Her oncologist reviewed FNAC report and found that was germ cell tumor. Oncologist did some serum marker like á feto protein, CA- 19-9, CA- 125. All were raised. á feto protein - 4401 u/ml, CA-19-9- 173u/ml, CA- 125- 351u/ml. CT scan was done which showed lobulated mass (20×10.7×17.1) cm arising from right ovary, moderate ascitis, no evidence of pre aortic or pelvic lympha-denopathy.

Patient was admitted in hospital for laparotomy on 27th november'2015. Laparotomy was done on 28th november,2015 with frozen section facility. On opening abdomen, there was moderate amount (800ml) of ascitic fluid, straw colour. A huge rt sided ovarian tumour about 20×18 cm in size which was partly cystic, partly solid, ugly looking, capsule was intact. Two friable nodule (2×3 cm) in pouch of douglas on both sided uterosacral ligament. No nodule in omentum, small and large gut, undersurface

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of diaphragm, liver, gall bladder and parietal peritoneum. Opposite ovary and tube was healthy. Uterus was normal size. Whole ovarian tumour was removed intact and send for frozen biopsy. Biopsy report was mature teratoma. So right salpingectomy, removal of pelvic deposit, total omentectomy was done.

Macroscopic feature

An ovarian mass measuring about 20×9cm. The cut surface showed a solid and cystic variegated tumour. The solid area was roughly about 15×10cm. The cystic area contains haemorrhagic fluid. The cut surface of solid area showed calcified. There are multiple grey brown solid area of omental tissue. There were also irregular grey browm piece of nodule in peritoneum. The largest one measures about 3×2×1cm. The cut surface is grey brown and friable.

Microscopic feature

Section showed ovarian tissue. It presents a tumour made of adnexal structures and squamous epithelium, mucous glands, respiratory epithelium, intestinal glands, bones and mature cartilage. It also showed brain tissue and immature fetal tissue.

Sections of omental tissue and peritoneal nodule showed fibrofatty tissue. It presents a tumour made of abundant mature tissue with loose mesenchymal tissue and glial tissue. These also showed less than 10% immature components.

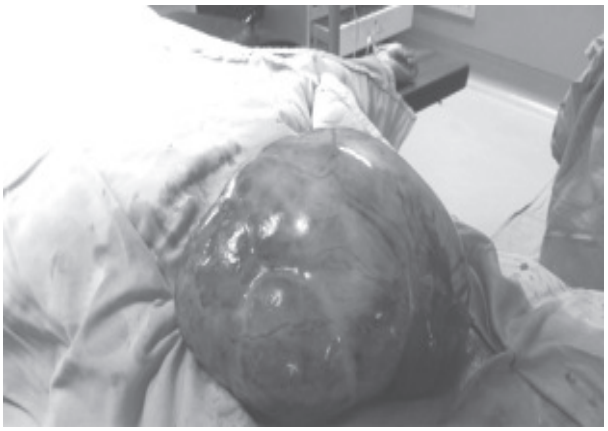

Fig.-1: *Gross appearance of immature teratoma*

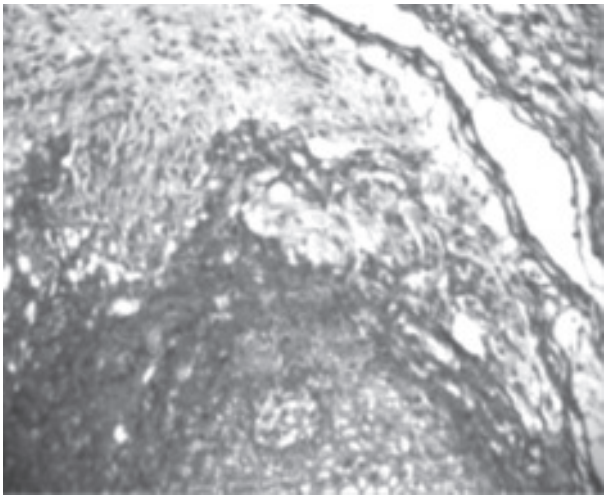

Fig.-2: P*hotomicrograph showing mature elements cartilage, glandular Epithelium*

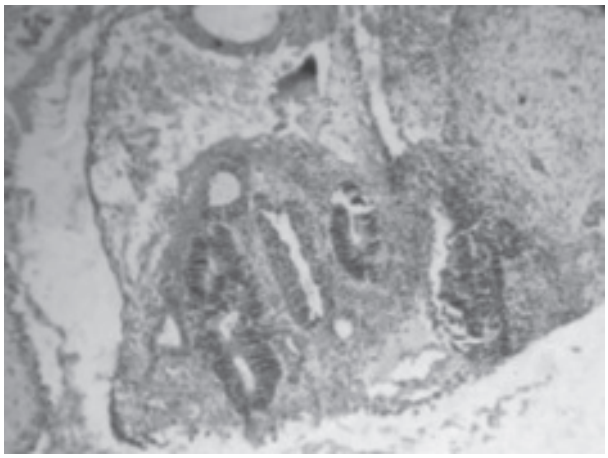

Fig.-3: *Photomicrograph showing immature neural tissue*

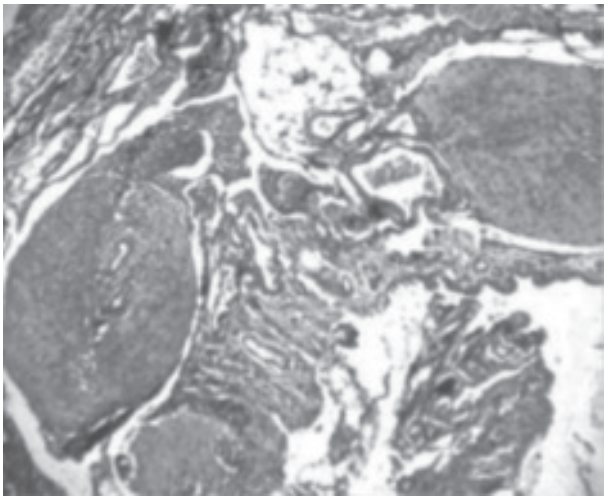

Fig.-3: *Photomicrograph showing glial implants in peritoneum*

A diagnosis of immature teratoma (Grade 1) with multiple glial implants in peritonium and omentum (Grade 0) was made. Her post operative period was uneven full. Ascitic fluid was free of metastasis.

After that she was referred to radio oncologist. He reviewed biopsy report which showed same result. Then she was advised to come for follow up after one month.

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.[1] 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.[4]

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.[2] 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.[4]

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 ventriculoperitonial shunts performed for hydrocephalus.[3,4]

The first case of immature teratoma with GP from India was reported by Joshi *et al.* in 1981.[3] A review of the literature reported by Chou *et al.* had found 65 cases of GP, which have favorable prognosis after surgical treatment.[6]

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 the other, pluripotent stem cells in the peritoneum or adjacent mesenchyme undergo glial metaplasia. [6] Recent molecular studies of glial implants have shown that they are genetically different from an ovarian tumor but have same genetic patterns as patient.[4] Substances produced by a tumor result in metaplastic transformation of peritoneal or subperitoneal tissue into glial tissue.[2]

Treatment for immature teratoma includes both surgery and chemotherapy, Patients with Stage Ia Grade 1 immature teratoma are usually treated with surgery alone, because the prognosis is excellent. When the grade advances to 2 or 3, or the stage goes beyond Ia, chemotherapy is usually recommended.[9]

In reproductive-age women who desire to retain fertility, removal of the involved ovary and surgical staging can be performed, leaving the uterus and the other ovary alone. This can be done because the other ovary is rarely involved, but staging is still required to make sure the cancer has not spread.[9]

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.[7]

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 Robbry and Scully modified by Norris *et al.*[8]

Grade is the single most important prognostic factor in early-stage disease. Even in the advanced stages, however, the grade is very important, assuming all of the visible cancer can be removed surgically. Across all stages, the five-year survival for grade 1 disease is approximately 82% and drops to approximately 30% when grade 3 disease is present. The five-year survival rate for stage 1 disease is 90% to 95%; while advanced stage survival drops to about 50% with Grade 1 to 2 cancer and to 25% or less when the tumors are found to be Grade 3.[9]

When associated with mature glial implants within the peritoneum the prognosis is usually much better, Irrespective of the original tumor grade.[10] 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.

Follow up after treatment is usually based on clinical exams and symptoms with expert use of CAT scans. Routine scans are not recommended, and there are no reliable tumor markers.[9]

Our patient is on follow up and till today she is found disease free.

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