Immature teratoma of the ovary is an uncommon tumour constituting one percentage of ovarian teratomas. Gliomatosis peritonei is a condition characterized by implantation of mature glial tissue on the visceral and parietal peritoneum. It is an infrequently reported event in ovarian teratoma. Till date only a few cases have been reported in literature. Here we present a case of Immature teratoma with associated gliomatosis peritonei in a 23 year old female admitted with complaints of abdominal pain.

**Case history**

A 23 year old nulliparous female presented in the gynaecological OPD with complaints of abdominal pain and abdominal distention for 1 month. Her vital signs were normal. Biochemical parameters were normal except for an elevated serum alpha fetoprotein and CA 125 levels. Ultrasonogram suggested a heterogeneous pelvic mass with calcified solid and cystic areas. CT revealed a left adnexal complex cyst with solid components and calcifications. Left salpingo oopherectomy was done. Intra operatively numerous grey white nodules were noted in the pelvic peritoneum.

**Gross features**

Received Unilateral salpingo oopherectomy specimen measuring 12x12x10 cm with an attached fallopian tube measuring 5cm. E/s- Grey white nodular. No Areas of capsular rupture or adhesions made out. No dilated veins seen. C/s –Grey tan, soft, predominantly solid admixed with small cystic spaces of various size ranging from 0.5 cm to 1cm filled with serous and mucinous material. Focal areas of calcification with hair like appendages seen. (Figure:1) Separately received multiple grey white, grey yellow soft tissue bits, largest one measuring 2cm and the smallest one measuring 0.5 cm, with a grey white cut surface. (Figure:2)

**Microscopic:**

H & E sections from ovary revealed mature and immature elements from all three germ layers arranged in a haphazard pattern. (Figure:3,4,5).

**Figure:1** Salpingo oopherectomy specimen measuring 12 x12 x10 cm. C/S - Grey tan, Soft, predominantly solid admixed with cystic spaces of varying sizes filled with serous and mucoid material.

**Figure:2** Peritoneal tissue studded with grey white nodules of varying sizes.

**Figure:3** Tumour is composed of squamous and glandular epithelium along with hair follicles, Pilosebaceous units and areas of melanin pigment. (10x)
Figure: 4 Areas showing glandular epithelium and immature cartilage. (40x)

Figure: 5 Areas showing glandular epithelium and cartilage. (40x)
Ectoderm was represented by predominantly skin elements including pilosebaceous units, sweat glands, hair follicles, neural tissue and choroid plexus. Mesodermal elements included cartilage. Endodermal elements were represented by glandular elements with a columnar lining. Occasional areas showed immature neuroepithelial elements composed of cells with a round nuclei, compact chromatin and scant cytoplasm arranged in the rosettes and tubules(Figure: 6, 7).

Figure: 6 Areas showing choroid plexus and immature neuroepithelial elements. (10x)

Figure: 7 Immature neural tissue forming tubules and rosettes. (40x)
The diagnosis of immature teratoma, grade 2 was given. Sections from the peritoneal implants revealed multiple nodules of mature glial tissue surrounded by peritoneal fat. (Figure: 8, 9, 10) Hence the diagnosis of Immature teratoma grade 2 with Gliomatosis peritonei grade 0 was given.

Figure: 8 Nodules of glial implants in peritoneum surrounded by adipocytes and inflammatory cells. (10x)

Figure: 9 Numerous glial implants in the peritoneum. (10x)

Figure: 10 Nodules of benign appearing mature glial tissue surrounded by the peritoneal pad of fat. (40 x)

Immunohistochemistry:

Figure: 11 Immunohistochemistry – Nodules of glial tissue is positive for GFAP (Gliarial fibrillary acidic protein) (40x)

Discussion:

Ovarian teratoma with gliomatosis peritonei is a rare occurrence. The word teratoma is derived from greek word “Teratos” meaning monstor.1 Immature teratoma is an uncommon tumour comprising 3% of all teratomas and <1% of ovarian teratomas.1, 2, 3, 4, 5 Peritoneal gliomatosis is an uncommon form of dissemination in ovarian teratomas. It was first described by Neuhauser in 1906 as a condition characterized by mature glial implants on the peritoneum and omentum associated with ovarian teratoma. Till date approximately less than 100 cases have been reported in medical literature.2, 6 The first case in India was reported by Joshi et al in 1981.5 Ovarian teratoma arise from the post meiotic, parthenogenetically activated germ cells.16 They contain embryonal and immature elements.7 It occurs predominantly in children and young adults. The average age of occurrence is 20 years. In this case report, it occurred in a 23 year old nulliparous female. It is rare in postmenopausal age group.8 Older patients tend to have a lower grade tumour when compared to younger patients. It is rarely bilateral. The tumour...
remains asymptomatic until it reaches a considerable size. It often manifests as an abdominal mass, increasing abdominal girth, and abdominal heaviness. Few patients experience vaginal bleeding. Rarely it goes for a rupture. Acute rupture produces peritonitis, pyrexia and shock. A slower smaller leakage produces few or no symptoms. Peritoneal implants are composed of tissues derived from all three germ layers. They are homozigous at polymorphic microsatellite loci in contrast to normal tissue which is heterozygous. The other grading systems were proposed by Norris et al in 1976 and Steeper and Mukai in 1986. The two tier grading system classifies Grade 1 ovarian tumours as low grade and Grade 2 or 3 tumours as high grade. Our subject presented with a grade 2 ovarian tumour. The term Gliomatosis peritonei implies the military implants of mature glial tissue in the peritoneum or the omentum. It occurs both in mature and immature teratoma. It is occasionally accompanied by fibrosis and chronic inflammation. It is composed of multiple tiny grey white superficial peritoneal nodules. They are not associated with adhesions and range in size from 0.1 – 1cm with an average of 3cm. In our case the peritoneal nodules ranged in size from 0.5 to 2 cm. It is considered benign as long as it is composed of mature tissue and unaccompanied by other teratomatous elements. Few cases had tumours resembling glioblastoma multiforme arising from these glial implants. In our case the implants were composed of mature glial tissue and hence were of grade 0.

Two theories have been proposed for the occurrence of gliomatosis peritonei.

1. Rupture of capsule and implants in the peritoneum via angiolymphatic spread.

2. Produced from metaplastic transformation of the pluripotent stem cells in response to various intraoperational conditions termed field defects.

65% of the teratomas are derived from a single germ cell after the first mitotic division with subsequent failure of mitosis and endoreduplication of the haploid chromosome. Hence teratoma contains a duplicated set of maternal chromosome. They are homozygous at polymorphic microsatellite loci in contrast to normal or metaplastic tissue that shows heterozygosity in the same loci. It is postulated that ovarian teratoma with abundant glial component secrete substances promoting glial differentiation in the peritoneum. Gliomatosis peritonei has also been reported in children without teratoma. These individuals have had a venticuloperitoneal shunt placed early in infancy. It has been proposed that neural growth factors present in CSF enter the peritoneum via the shunt and cause glial differentiation. One Diagnostic difficulty with Gliomatosis Peritonei is its co existence with a primary tumour implant. Hence Thurlback and Scully proposed the criteria for Gliomatosis Peritonei. It includes:

1. Extensive histological sampling of the peritoneal surface, omentum and diaphragmatic surface.

2. Each of the sampled implants should be composed of grade 0 glial tissue.

Prognosis is closely associated with tumour grade. Grade 1 tumours have excellent prognosis. They require only post operative observation and no adjuvant treatment. Grade 2-3 tumours is an indication for chemotherapy. According to a study by Norris et al Survival for patients with Grade I teratoma is 82%, Grade 2 is 63% and for Grade 3 it is 30%. Paradoxically patients with immature teratoma with mature glial implants have a much better prognosis. Grade 0 glial implants does not appear to have adverse outcome in the tumour. They require no chemotherapy. Hematogenous spread to lung liver and Brain is unusual. Serum AFP levels will be elevated in 50% of the individuals. Modest increase in serum CA125 is noted in some individuals. In our case both these enzymes were elevated. The Serum AFP was 266.4 ng/ml and serum CA 125 was 216.2 U/ml. Rare cases have serious neurological or psychiatric symptoms caused by paraneoplastic syndromes associated with teratoma. Bilaterality is rare. Hence the treatment of choice is unilateral salpingo oopherectomy with collection of samples from the peritoneal implants. No further treatment is required for grade 1 tumours with Grade 0 peritoneal implants. Triple agent chemotherapy using the VAC regimen is advocated for primary tumours of grade 2 or 3 with glial implants of grade 1.

References:


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