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Pathology

GLIOMATOSIS PERITONEI ASSOCIATED WITH IMMATURE OVARIAN TERATOMA

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ABSTRACT Gliomatosis peritonei (GP) can be defined as the metastatic implantation of glial tissue on the surfaces of visceral or parietal peritoneum. The implants of gliomatosis peritonei resemble benign mature glial tissue with delicate fibrillar processes and scattered supporting cells.GP is characterized by the recurrence of peritoneal implants after the surgical treatment of ovarian teratoma. Malignant transformation is exceedingly rare It has been found to be associated exclusively in females with ovarian teratomas . Immature teratoma (IT) is the term used for malignant ovarian teratoma composed of mixture of embryonal and adult tissues derived from all the three germ layers regardless of its gross appearance. According to WHO, IT is defined as a teratoma containing variable amount of immature embryonal type neuroectodermal tissue. Here, we report a case of immature ovarian teratoma associated with gliomatosis peritonei in a 45 year old female.

KEYWORDS : Gliomatosis pertonei, teratoma, immature neuroepithelial tissue.

INTRODUCTION

Gliomatosis peritonei is metastatic implantation of glial tissue on the surfaces of visceral or parietal peritoneum. It is associated with immature ovarian teratomas. The term itself of "teratoma" was derived from the Greek root "*teratos*" which means Monster. The mechanism of implantation of glial tissue is unknown. Histologically, the implants of gliomatosis peritonei resemble benign mature glial tissue. Malignant transformation is very rare.In rare circumstances, they can undergo transformation to malignant tissue (glial or teratomatous).

Clinical History

A 45-year-old woman presented with abdominal distension and pain. Computed tomography (CT) showed a cystic- solid pelvic mass. Her AFP (1100 ng/ml) and CA 125 level were high but total hCG level was in normal range. She underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy with peritoneal and omental biopsies. All the tumor implants in the peritoneum were removed. Uterus and left adnexa were normal. Right ovarian mass measured 25x20x10 cm. Cut section showed solid areas and cystic areas along with keratinous and necrotic debris. Numerous small nodules were isolated from the peritoneal/ omental tissue. On histologic examination, the tumor showed a large amount of immature neuroepithelial tissue(figure 1). In addition, mature glial tissue was defined in the peritoneal samples (figure 2). So, the final histopathological diagnosis was given as immature teratoma associated with gliomatosis peritonei.

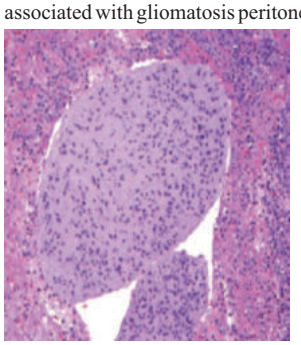

Figure 1: High power view of section from the ovarian mass revealing immature neuroepithelial tissue (H&E, 400X)

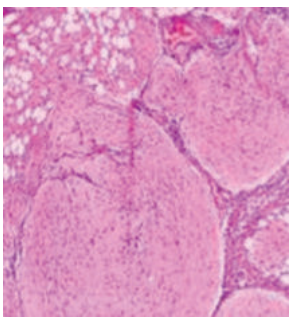

Figure 2 : High power view of sections from omentum showing nodules of matue glial tissue (H&E,400X)

DISCUSSION

Teratomas are most frequently found in the gonads (ovary and testes). Extragonadal teratomas are rare and arise from midline structures (thyroid, retroperitoneum, mediastinum, pericardium and brain). Very rarely, teratomas are found in other solid (e.g. breast, parotid gland, liver) and hollow (e.g. oesophagus, stomach, bladder, uterine cervix) organs [1]. Teratomas may be benign, malignant or a component of a mixed germ cell tumor (GCT) [2].

The first description of teratoma was made in 1960 by Thürlbeck and Scully [3].The ovarian teratomas are represented by mature, immature, and monodermal (as *struma ovarii*, carcinoid tumors, neural tumors) types. They are considered the most common germ cell neoplasm. Teratomas comprise a number of histologic types of tumors, all of which contain mature or immature tissues of germ cell (pluripotential) origin. Mature teratomas are benign tumors, which are most often composed of derivatives of two or three germ cell layers. In contrast, immature teratomas are malignant ovarian tumors, as the present case [4].IT represents 3% of all teratomas, 1% of all ovarian cancers and 20% of malignant ovarian germ cell tumors [5].

Immature teratoma (IT) is the term used for malignant ovarian teratoma composed of mixture of embryonal and adult tissues derived from all the three germ layers regardless of its gross appearance. According to WHO, IT is defined as a teratoma containing variable amount of immature embryonal type neuroectodermal tissue (usually)[6].Immature elements represent the evolution of a malignant clone, and the prognosis relates to the amount of this component. Tumour grading is based on the amount of immature neuroepithelium [7]. In grading of immature teratoma, primitive neural tubes and immature rosettes are counted. Immature teratomas lack 12p amplification in contrast to its presence in other malignant ovarian germ cell tumours . Thus, its pathogenesis also differs from other malignant germ cell tumours [8].

It is a tumour of children and adolescents, that occurs mainly in first two decades of life and is exceptional in postmenopausal women. It is almost always unilateral [9]. Grossly, immature teratoma is a predominantly solid, unilateral tumor that averages 18 cm in diameter. The solid component is gray or brown in colour and soft to hard in consistency. If keratinous debris or hairs are seen it may resemble a dermoid cyst [10].

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 [11].

Gliomatosis peritonei (GP) can be defined as the metastatic implantation of glial tissue on the surfaces of visceral or parietal peritoneum [12]. It has been found to be associated exclusively in females with ovarian teratomas, although there are reports of its association with pregnancy and ventriculoperitoneal shunts performed for hydrocephalus. All grades of ovarian teratomas have been

described, with immature teratomas being more commonly associated with this gliomatosis peritonei. The first case of immature teratoma with GP from India was reported by Joshi et al. in 1981 [13].

GP is characterized by the recurrence of peritoneal implants after the surgical treatment of ovarian teratoma. Histologically, the implants of gliomatosis peritonei resemble benign mature glial tissue with delicate fibrillar processes and scattered supporting cells. Malignant transformation is exceedingly rare [14]. However, few reports have documented the rapid recurrence of immature peritoneal implants, as implantation is associated with teratomas of all grades [15]. GFAP is an intermediate filament protein that expresses with the development of astrocytes in the fetal nerve tissue. It is used to confirm the glial nature of the tissue. A strong expression of GFAP often suggests that tumor cells are mature and well differentiated [16]. Majority of cases with GP are associated with immature ovarian teratoma and extremely rarely found with mature ovarian teratomas [17].

The mechanism of implantation of glial tissue is unknown and two theories have been proposed to explain the origin of GP. According to one of the theories, glial implants arise from the teratoma through rupture of capsule with subsequent implantation in the peritoneum. Also dissemination through angiolymphatic channels have been documented. In support of lymphatic dissemination, mature glial tissue has been found in mesenteric, para-aortic, and retroperitoneal lymph nodes in association with immature teratomas, in the presence or the absence of GP. The other theory proposes that pluripotent stem cells in the peritoneum or subjacent mesenchyme undergo glial metaplasia in response to unknown neoplastic stimuli. They presumably originate in pluripotent Müllerian stem cells. Molecular studies propose that ovarian teratoma and GP are genetically distinct(multiple independent tumors rather than relapse or metastasis) [18].

The nodules of glial implants are usually 1-10mm in size, localizing in the parietal and visceral peritonei. Intraoperatively, gross appearance of GP can be confused with other multicentric intraabdominal diseases like peritoneal leiomyomatosis, peritoneal carcinomatosis, endometriosis, tuberculosis, melanosis, and ectopic decidua. Hence, the role of microscopy and panel of immunostains is important to arrive at a definite diagnosis [19].

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 [19]. Macroscopically, peritoneal implants are small in size, well circumscribed, and have a grayish color. 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. 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) and Oct 4 is used to rule out malignant transformation, and negative AFP rules out metastasis from an immature germ cell tumor [20].

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). Patients with mature glial implants in a case of immature teratoma have a better prognosis.

CONCLUSION

GP is a rare benign condition associated almost exclusively with immature teratoma of ovary. Despite widespread involvement of peritoneal surfaces, GP does not adversely affect the course and prognosis. Treatment of choice should be judged only by the stage and grade of primary ovarian teratoma. However, long-term follow-up is necessary in view of few cases reporting malignant transformation of glial implants long after initial surgery.

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