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CASE REPORT

Gliomatosis Peritonei Arising in Setting of Immature Teratoma of Ovary: A Case Report and Review of Literature

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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 1–10 mm. Robboy and Scully have suggested three possible sources of GP: (1) deposition of immature neural tissue with consequent maturation; (2) lymphogenous metastasis; and (3) 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

An 18-year-old female presented with rapidly enlarging lump in right lower abdomen over the last 2 months. Her medical and family history were unremarkable. Local

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examination revealed a huge mass in abdominopelvic region. Rest of the general and systemic examination was within normal limits. Ultrasonography (USG) revealed a 22 × 16 × 9 cm heterogeneously hyperechoic mass in right adnexal region. Magnetic resonance imaging (MRI) revealed a mass with altered signal intensity lesion in midline and pelvis extending to right adnexal region measuring 16.6 × 9.5 × 22.8 cm. Serum tumor markers revealed marked elevation of CA125 [459.8 U/ml (normal <30)]. Other tumor markers were mildly elevated with carcinoembryonic antigen (CEA) levels being 7.97 ng/ml (normal <2.5), alpha-fetoprotein (AFP) levels of 52.27 ng/ml (normal <6) and beta-human chorionic gonadotrophin (HCG) levels were <1 mu/ml (normal <2.5).

Laparotomy revealed a huge right ovarian mass measuring 20 × 15 × 10 cm along with multiple, firm grayish white 0.2–1.0-cm- nodules on peritoneum, omentum, and pouch of Douglas. The patient underwent right salpingo-oophorectomy along with excision of nodules on peritoneal surface, omentum, and pouch of Douglas.

The encapsulated ovarian mass measured 20 × 15 × 10 cm with grayish white glistening external surface. Cut surface was partly solid and partly cystic revealing multiple cystic cavities filled with mucoid material varying in size from 0.3 to 2-cm diameter. Focal areas revealed the presence of hair. (Fig. 1a)

Histological examination revealed features of teratoma comprising skin and adnexal structures (Fig. 1b), foci of mature cartilage and gland formations (Fig. 1c), gut, and respiratory epithelium with a few foci of primitive neuroepithelium (Fig. 1d). The nodules from all the three sites (peritoneal surface, omentum, and pouch of Douglas) contained mature glial tissue (Fig. 2). A diagnosis of immature teratoma (grade 1) with GP was rendered.

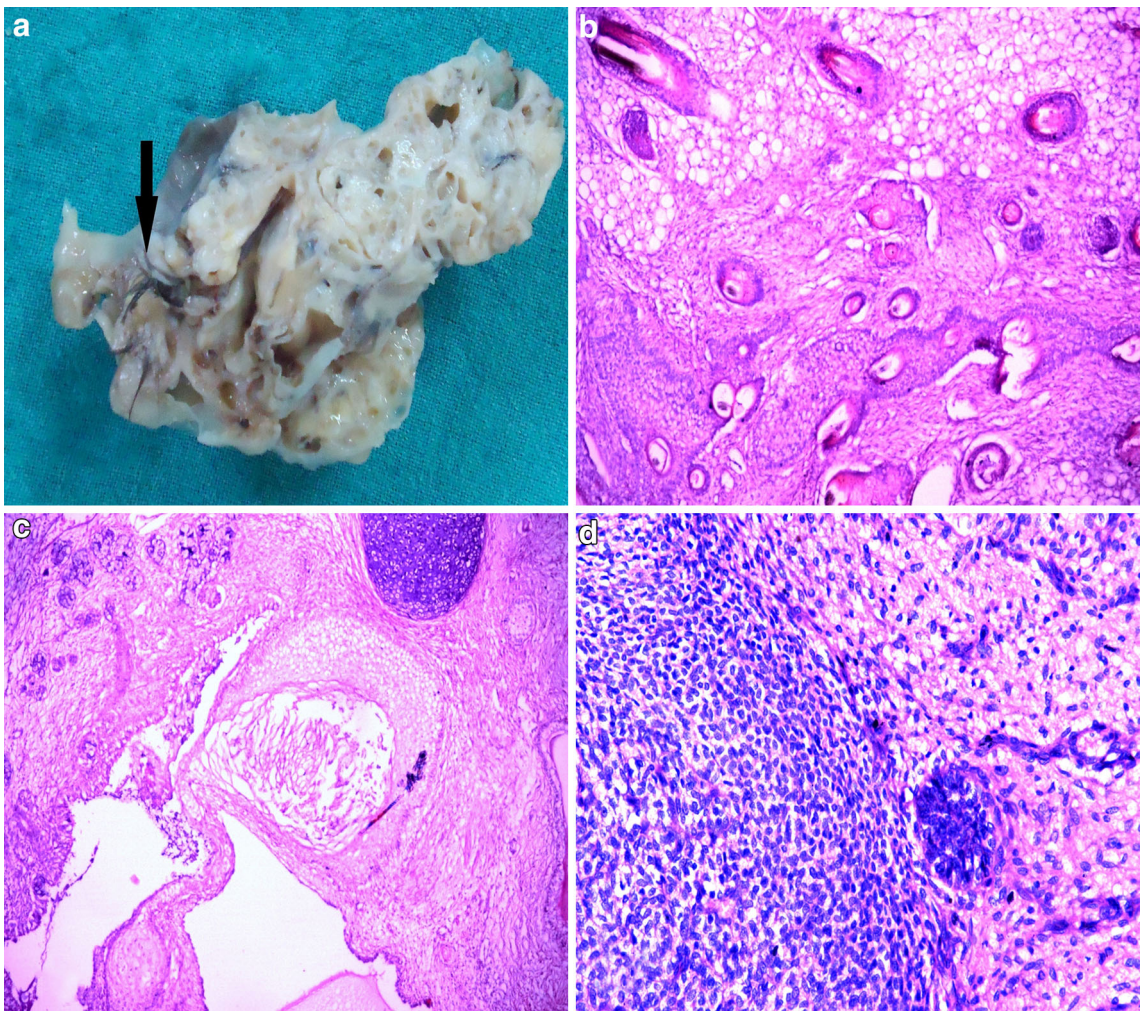

Fig. 1 Cut surface of ovarian mass revealing solid and cystic areas with multiple cystic cavities filled with mucoid material varying in size from 0.3 to 2-cm dia with focal areas revealing the presence of hairs (*highlighted by arrow*). **a** Microphotograph revealing features of teratoma comprising of skin and adnexal structures (**b**; H&E; ×40), foci of mature cartilage, and gland formations (**c**; H&E; ×40), with a few foci of primitive neuroepithelium (**d**; H&E; ×100)

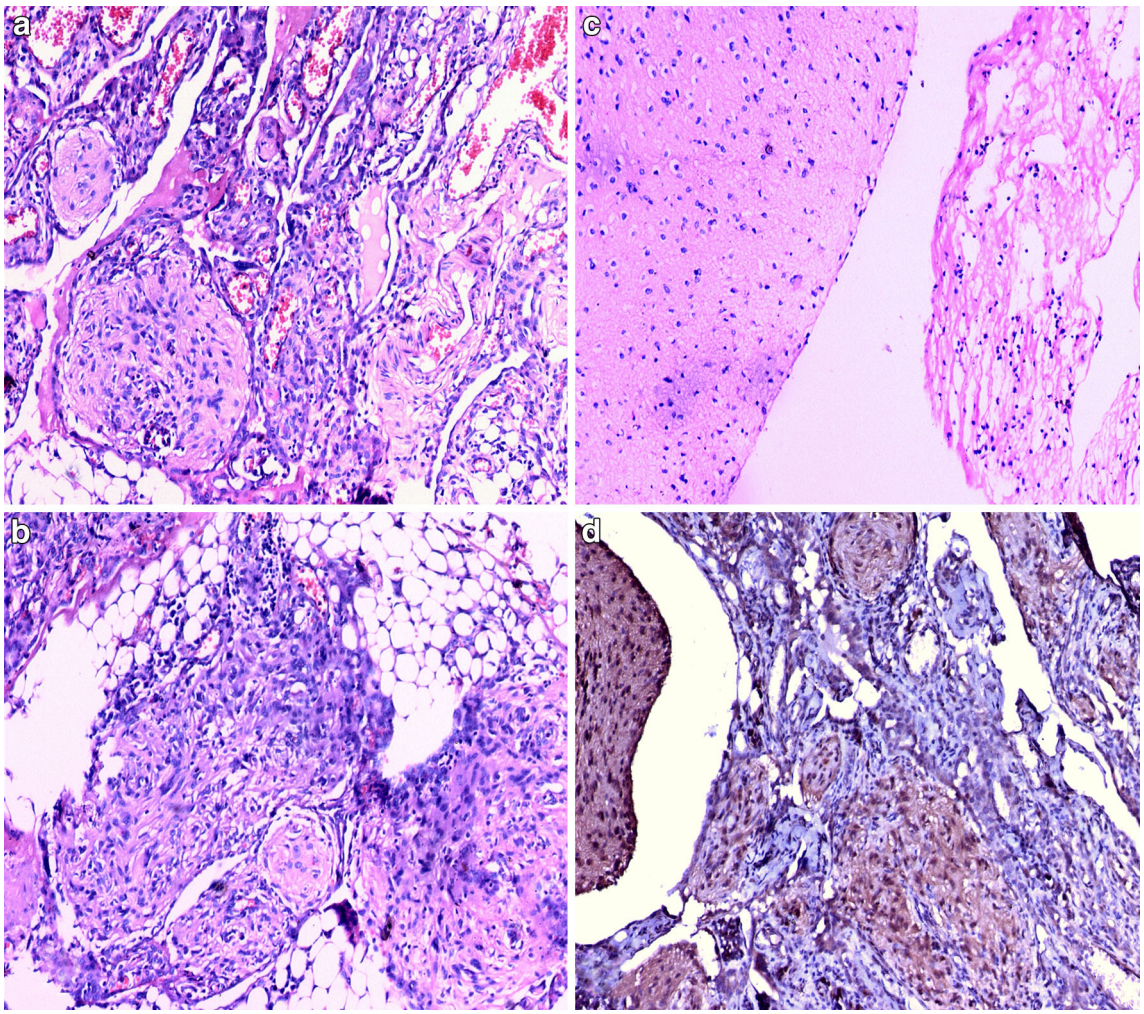

Fig. 2 Microphotograph revealing glial tissue embedded in omental fat (**a** & **b**; H&E; ×40). The glial tissue was mature (**c**; H&E; ×100) and revealed positivity with neuron-specific enolase (**d**; IHC; ×40)

The postoperative period was uneventful, and the patient is still under follow-up for the past 1 year.

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. Tumor grading is based on the amount of immature neuroepithelium presence [3].

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. All grades of ovarian teratomas have been described, with immature teratomas being more commonly associated with this condition [4].

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 [4]. Molecular studies suggest that ovarian teratoma and GP are genetically distinct (multiple independent tumors rather than relapse or metastasis) [3].

The nodules of glial implants are usually 1–10 mm 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].

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 [4]. 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]. Moreover, there is a high incidence of adhesion sequelae in GP, which warrants more careful intervention to avoid tumor rupture, to prevent adhesion, and as a postoperative follow-up [1]. Potential for its recurrence is high, and therefore it requires a careful monitoring of residual lesions using scanning imaging such as computed tomography [3].

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 [2].

A favorable prognosis is determined by the following: (1) histological nature of glial tissue implants that are completely mature regardless of the nature of immature ovarian teratoma; and (2) 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 Thurlback 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 [4].

Conclusion

A mature gliomatosis implant constitutes a harmless situation with a good prognosis, even when associated with an immature teratoma of the ovary. It is important to diagnose these patients carefully because it imparts a favorable prognosis to patients with immature ovarian teratomas, which may be mistaken for intra-abdominal carcinomatosis or tuberculosis, and a close follow-up is required to monitor recurrence and rarely malignant transformation.

Compliance with Ethical Requirements and Conflict of interest All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008 (5). Informed consent was obtained from the patient before being included in the study. Nisha Marwah, Ashima Batra, Sumiti Gupta, Savita Rani Singhal, and Rajeev Sen declare that they have no conflict of interest.

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