


Ovarian Immature Teratoma with Gliomatosis Peritonei and Nodal Gliomatosis Metastasis: A Case Report

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Abstract: Gliomatosis peritonei (GP) is a rare condition of mature glial tissue within the peritoneum. GP has been reported in approximately 100 cases worldwide, and it rarely occurs in young women or in patients with ovarian mature or immature teratomas during childhood. In patients with ovarian immature teratoma (OIT) combined with gliomatosis peritonei, the tumor is typically large and is often detected intraoperatively. Its neuroglial component may not only spread within the peritoneum but also metastasize via the lymphatic vessels. Pathologically confirmed lymph nodes containing mature neuroglial components and positive for glial fibrillary acidic protein (GFAP) are consistent with nodal gliomatosis (NG). In this report, we present a case of gliomatosis associated with OIT combined with GP, which involves lymph nodes.

Keywords: gliomatosis peritonei, ovarian immature teratoma, nodal gliomatosis, ovarian teratoma

Gliomatosis peritonei (GP) is a rare condition characterized by the presence of mature neuroglial components in the peritoneum, which may be associated with ovarian teratomas of varying grades,¹ with ovarian immature teratoma (OIT) being the more common complication. The coexistence of GP with ovarian immature teratomas is a rare occurrence.² Neuroglial peritoneal implants are more frequently found in larger ovarian mature teratomas.³ The involvement of lymph nodes by mature glial tissue is even less common. Lymph node gliomatosis (also known as nodal gliomatosis, NG) is rarely described in the literature and is mostly associated with ovarian immature teratomas, typically in conjunction with peritoneal gliomatosis. The involved lymph nodes are usually from the pelvic or para-aortic regions, but occasionally, lymph nodes in the lower abdomen, neck, and axilla are also affected.⁴ In this paper, we reviewed the clinical data of a case of ovarian immature teratoma combined with GP and lymph node metastasis, admitted to the Department of Obstetrics and Gynecology at the First Affiliated Hospital of Dalian Medical University. We also reviewed the relevant literature to explore its clinical features, treatment and prognosis.

Case Report

A 23-year-old female patient was admitted to the gynecology ward of the First Hospital of Dalian Medical University on April 2, 2024, for treatment of a lower abdominal mass that had been found for more than 3 months, without any complaints of abdominal pain, distension, or malaise. The patient was 14 years old when she had her first menstruation, and her menstrual cycles had previously been regular, with a cycle of 30–40 d, and a menstrual period of 6–7 d. She was diagnosed with tuberculosis in 2018, which has since been treated and cured. Ano-abdominal bimanual examination revealed normal vulvar development, but the uterus and bilateral adnexa were unremarkable on palpation. A bi-lobed solid mass, approximately 20 cm in size, was palpable in the pelvic cavity. The mass had a smooth surface, well-defined

boundaries, poor mobility, and was tender upon palpation. Upon admission, pelvic ultrasound revealed the uterus to be 62x54x57 mm in size. The right anterior superior portion of the uterus showed a disorganized echogenic area, approximately 180x177 mm in size, with scattered small hypoechoic areas. No significant blood flow signals were detected. Pelvic fluid was not detected. Differential diagnosis includes pelvic-abdominal mass (eg, ovarian gonadal interstitial tumor, tumor of other origin). The enhanced abdominal CT scan suggested the presence of a large mass in the abdominopelvic cavity, with a high likelihood of an ovarian tumor, and the presence of fluid accumulation in the abdominopelvic cavity. Cystic lesions were identified in the left adnexal region, with no other significant abnormalities observed (Figure 1). Serum tumor marker analysis revealed CA125 levels of 207.12 U/mL, CA19-9 levels of 294.26 U/mL, and HE4 levels of 84.90 pmol/L. The admission diagnosis indicated a right pelvic mass with a high likelihood of an ovarian malignant tumor.

After completing routine preoperative examinations and confirming the absence of contraindications to surgery, a transabdominal right adnexectomy, right pelvic and para-abdominal aortic lymph node dissection, and salpingo-oophorectomy were performed on April 9, 2024. Abdominal lavage fluid was initially retained, and a tumor on the right ovary, measuring approximately 18x16 x 12 cm, was identified. The tumor was grayish-white, predominantly solid, and occupied the entire ovary, making normal ovarian tissue indistinguishable. The peritoneal membrane of the tumor was ruptured at the upper edge, with a fissure approximately 5 cm in length. Tumor tissue extended outward from the fissure, measuring approximately 4.5x4.0 x 3.5 cm. Scattered, localized “yellow-white pus-like” adhesions were

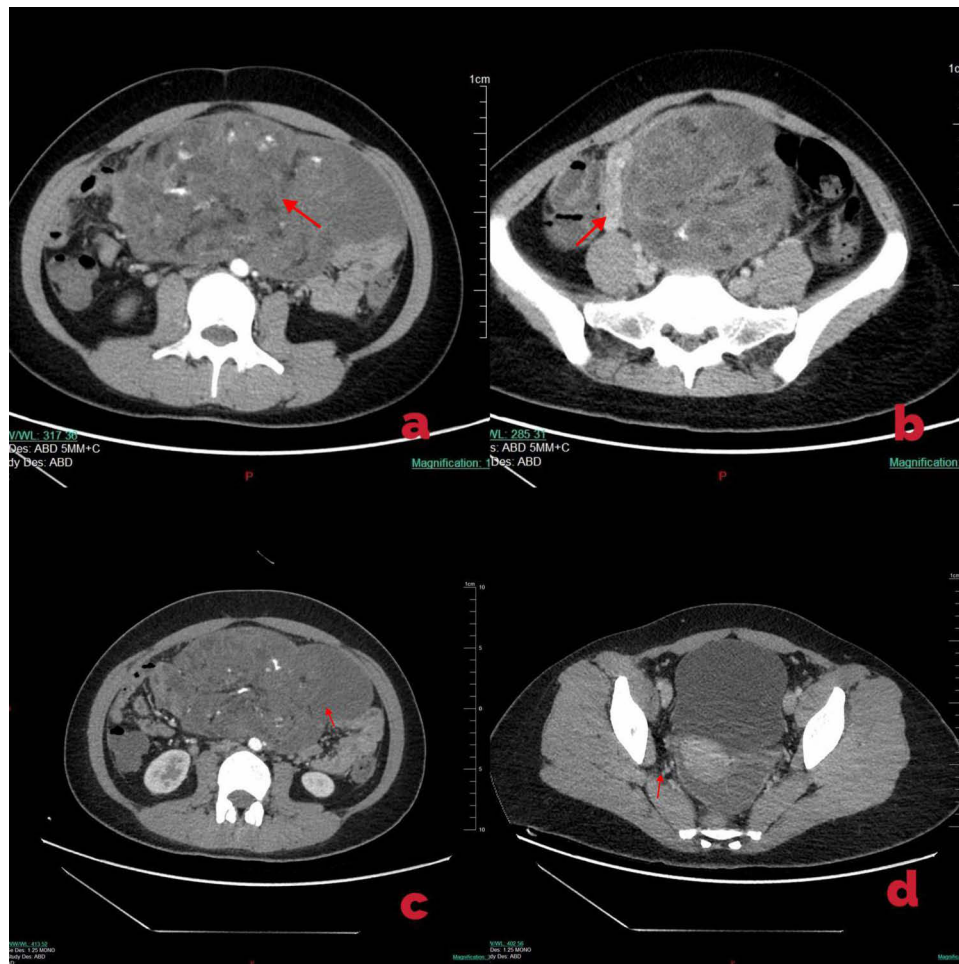


Figure 1 (a) Consistent with immature ovarian teratoma. The red arrow indicates a mass with heterogeneous density, demonstrating prominently enhanced neural tissue components. (b) The presence of dilated ovarian veins indicates a primary ovarian origin of the tumor. The red arrow indicates the dilated ovarian vein. (c) Tumor fissure. The red arrow demonstrates tumor rupture and outward extension. (d) The peritoneal membrane in the rectouterine pouch demonstrated thickening and asymmetrical enhancement. The red arrow indicates asymmetric peritoneal enhancement in the rectouterine pouch.

observed on the peritoneum of the sacral ligaments, rectouterine pouch, right broad ligament, and bladder inversion peritoneum. Enlargement of lymph nodes was noted in the low right parietal abdominal aorta and right intra-iliac region. Intraoperative frozen section pathology revealed a localized immature component in the right ovarian teratoma. Specimen visualization revealed a right adnexal mass measuring 17.5×15 × 9.5 cm with a smooth grayish surface (Figure 2). Upon sectioning, the mass was predominantly solid with cystic structures, and cystic cavities containing mucous fluid were noted. Postoperative pathology confirmed the diagnosis of a grade G1 immature teratoma in the right adnexal ovary (Figure 3). Peritoneal gliomatosis was observed in the greater omentum, uterorectal reflection peritoneum, right colonic lateral sulcus peritoneum, and the peritoneum of the uterovesical pouch. Lymph node gliomatosis was noted in the low right parietal abdominal aorta and right external iliac lymph nodes (2/4, 1/1) (Figure 4). No malignant cells were observed on microscopic examination of the peritoneal lavage fluid. Immunohistochemical analysis was positive for glial fibrillary acidic protein (GFAP) (+) and negative for CD68 (–) and CK (–). The postoperative diagnosis was an immature teratoma of the right ovary with associated peritoneal gliomatosis.

The patient was followed up 6 months postoperatively. Abdominal CT revealed an irregular mass shadow in the right lobe of the liver, measuring approximately 4×2 cm, with uneven density, calcification, and inhomogeneous enhancement on contrast-enhanced imaging. No abnormalities were found in serum tumor marker testing. Due to contraindications to open surgery, laparoscopic hepatic mass resection was performed on October 21, 2024. During the procedure, a yellowish-white mass, measuring approximately 3×5 cm, was identified beneath the diaphragm. The mass exhibited hardness, intact peritoneum, and caused compression of the hepatic tissues, with partial invasion of the liver. Partial invasion of the liver was noted. The postoperative pathological diagnosis confirmed an immature teratoma of the abdominal wall, classified as grade G1. The most recent follow-up was on March 10, 2024. The patient is in stable condition.

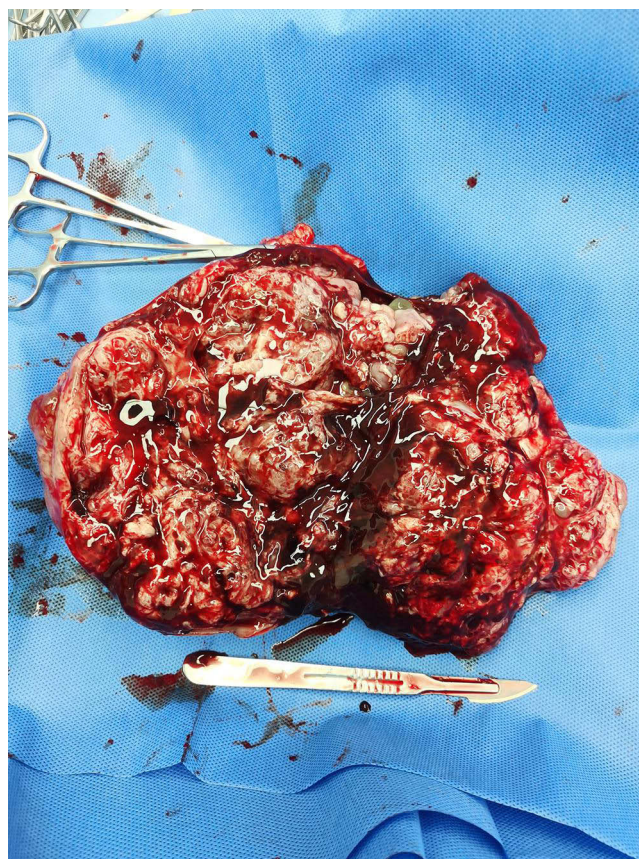


Figure 2 Gross appearance and cut surface of the specimen.

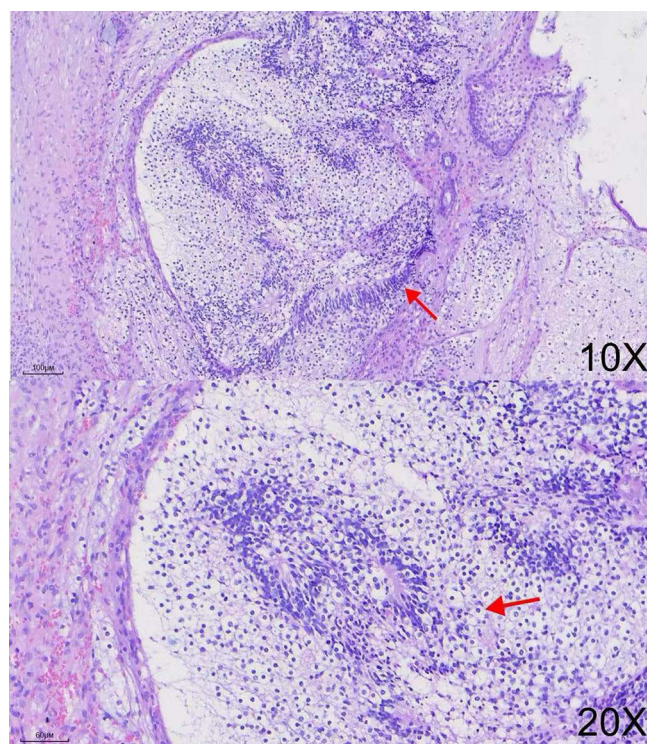


Figure 3 The tumor tissue contained immature neuroepithelial elements and was classified as Grade I (G1). Red arrows indicate the immature glial components observed microscopically.

Discussion

Pathogenesis

Currently, three main hypotheses exist regarding the origin of GP:¹ Spillage of neuroglial components due to tumor rupture may lead to peritoneal dissemination, with hematogenous and lymphatic spread as additional possible routes.^{2,5} Glial directed differentiation occurs multicentrally from multipotential stem cells or mesenchymal stromal cells under specific circumstances. Recent molecular genetic studies have found ovarian teratomas to be pure heterozygous and peritoneal glial implantation nodules to be heterozygous, confirming that GP is originated from multipotential stem cells or mesenchymal stromal cells under the epithelium of the peritoneal surface or corpora cavernosa.^{3,6} This may result from the extrusion and dissemination of mature glial tissue, which is closely associated with ventriculoperitoneal shunts.⁷ Similar to GP, the pathogenesis of NG remains unclear. Most reported cases of NG are associated with GP, and it is speculated that NG may result from mesothelial cell chemotaxis due to ovarian tumor secretion,⁴ or it may be secondary to tumor rupture, leading to dissemination via the lymphatics.

Treatment and Regression

There is currently no standardized protocol for the treatment of GP, which primarily depends on the histological grading and staging of the primary tumor. Previous literature suggests that the prognosis of lymph node gliomas is favorable, and no additional treatment is required when the involved lymph nodes are confirmed to be composed of mature tissue without immature components (ie, grade 0), as observed in GP. Therefore, the management of patients with OIT concomitant with GP is guided by the staging and grading of immature teratomas. For all patients desiring fertility preservation, irrespective of tumor stage, fertility-sparing surgery, such as adnexectomy with salpingo-oophorectomy on the affected side, is a viable option. Biopsy should be performed only on the suspected lymph nodes. Following surgery, appropriate combination chemotherapy should be selected based on tumor grade. Chemotherapy is indicated for stage I, G2-3 or II–IV tumors, with the BEP regimen being preferred.

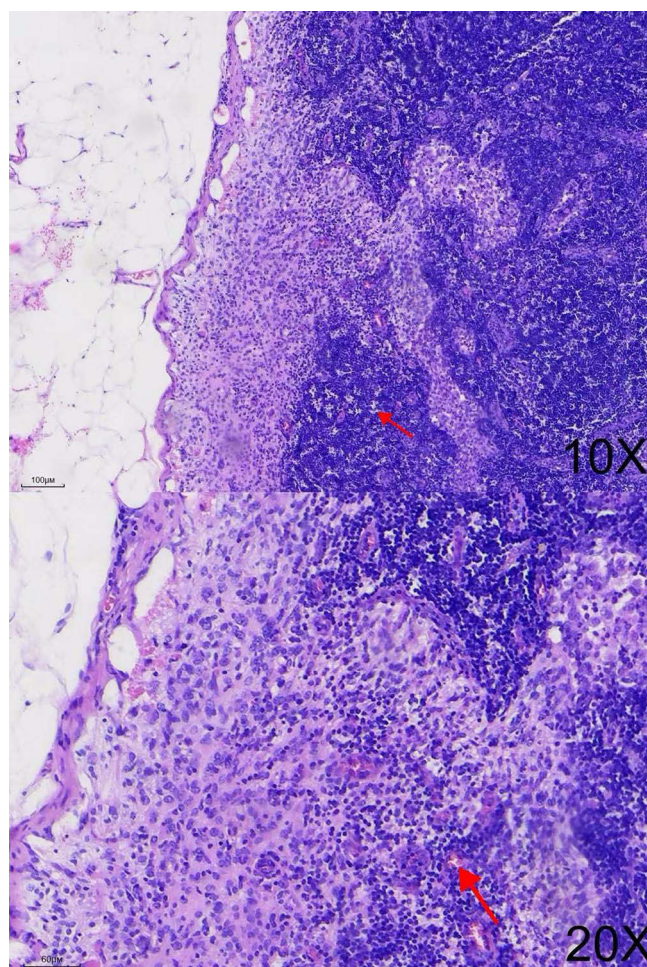


Figure 4 Lymph nodes showed evidence of lymph node gliomatosis. Red arrows identify lymph nodes involved by mature neuroglial tissue.

GP is primarily a mature glial nodule, typically a quiescent condition with variable outcomes: i) The lesion persists without change; ii) It may undergo fibrous transformation or degeneration; iii) In some cases, it can be replaced by teratoma or malignant glial tissue. The presence of immature teratoma does not influence the prognosis of patients, and does not impact the overall survival of patients with immature teratoma.³ To date, malignant transformation has not been reported, and cases of NG are exceedingly rare. Therefore, whether it confers an unfavorable prognosis remains uncertain,⁴ although it is believed that all cases are associated with frequent tumor recurrence. Ovarian immature teratomas have a high rate of recurrence and metastasis; however, they exhibit a reversal from malignant to benign behavior. Common sites of metastasis for immature ovarian teratoma include the peritoneum, lymph nodes, and liver. Hepatic metastases can originate either from peritoneal implantation or via hematogenous spread. Most recurrences within the first year of OIT are of the immature type with the same grading as the primary tumor. Previously, a case of ovarian immature teratoma with both gliomatosis peritonei and pleural glial implant in a 4-year-old girl was reported. Glial emboli were present in the pleural implant, suggesting lymphovascular dissemination might be the cause of extra-abdominal glial implantation.⁸ In the present case, the early recurrence of the tumor in the liver raises the possibility of metastatic spread of glial components via pelvic lymph nodes to the hepatic parenchyma. This case highlights the need for enhanced clinical surveillance and suggests that NG may be associated with an adverse prognosis. The site of recurrence, surgical thoroughness, and timing of chemotherapy initiation after the first recurrence are important prognostic factors. Among these, the thoroughness of surgery plays a critical role in the remission of recurrent ovarian immature teratoma. Postoperative administration of effective combination chemotherapy, extending the time to recurrence, and promoting the transformation of the tumor into the mature type can significantly improve prognosis.⁹ Residual

peritoneal lesions in GP can be asymptomatic and may remain dormant or quiescent for extended periods. After ruling out the presence of metastatic immature teratoma, a more conservative surgical approach may be considered for patients with extensive peritoneal dissemination,¹⁰ thereby reducing surgical trauma and minimizing adhesions in young women. In patients with recurrent ovarian immature teratoma, where the initial treatment involved surgery alone, the BEP regimen is recommended due to its excellent response rate.¹¹

Prognosis

The impact of peritoneal gliomatosis or lymph node gliomatosis combined with immature teratomas remains controversial. Some reports suggest that patients with OIT combined with GP may have a better prognosis, whereas other studies indicate that peritoneal glial nodules could have the potential for malignant transformation.¹² Due to the extensive implantation of peritoneal gliomatous nodules, complete excision of the affected tissue is challenging, and patients with residual peritoneal lesions require prolonged and intensive follow-up.

Summary

GP is rare and can be associated with teratomas of any grade, which complicates preoperative diagnosis. Intraoperative attention should be directed towards the rupture of the peritoneal envelope of the immature ovarian teratoma and peritoneal lesions to differentiate between tuberculous peritonitis and peritoneal metastatic carcinomas. The combination of OIT with GP is an unfavorable prognostic factor for recurrence frequency. Therefore, early recognition of this condition is crucial to guide therapeutic regimens and provide patients with OIT sufficient time for the transformation from malignancy to benignity.

Patient Perspective

Ms Q reported experiencing significant anxiety and emotional distress during the preoperative period while awaiting diagnostic clarification regarding potential malignancy. She described heightened worry among family members, particularly her parents, which compounded her own feelings of emotional burden and self-reproach regarding her health status. Following a successful surgical intervention that obviated the need for adjuvant chemotherapy, she expressed a sense of fortunate outcome. This experience underscored her perception of health as a paramount priority.

Abbreviations

GP, Gliomatosis Peritonei; OIT, ovarian immature teratoma; GFAP, glial fibrillary acidic protein; NG, nodal gliomatosis.

Ethics Statement

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. The written informed consent has been stated in the ethical methods and is available for review by the editorial board. It was prepared in accordance with the CARE guidelines. Institutional ethical approval was obtained for this study. The publication of this case report has been approved by the Ethics Committee of The First Affiliated Hospital of Dalian Medical University (First Affiliated Hospital of Dalian Medical University).

Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

Disclosure

The authors have no conflict of interest.

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