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Pathological Case of the Month

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A 12-YEAR-OLD girl presented to the Pediatrics Clinic of Taipei Municipal Chung-Hsiao Hospital, Taipei, Taiwan, with several months' history of abdominal distension and mild dyspnea. Family and medical histories were noncontributory. Abdominal and pelvic examination showed a tender pelvic mass extending into the right costal margin. The other physical findings were insignificant. Ultrasonogram, abdominal x-ray films, and computed

tomographic scan (**Figure 1**) showed a large, partially calcified intraperitoneal mass extending from the pelvis to the xiphoid process. Preoperative serum α -fetoprotein and human chorionic gonadotropin levels were within normal range. During a laparotomy, a large tumor of the right ovary with a capsular tear was found. The omentum and peritoneum were erythematous and finely granular. A right salpingo-oophorectomy and biopsy of the omental implant were performed. The right ovarian tumor measured 23×15×15 cm and weighed 1300 g. The capsule was ruptured and the tumor was solid and cystic. Microscopic sections are shown in **Figure 2** and **Figure 3**. Postoperatively, the patient has been healthy with no recurrent disease for 31 months.

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Figure 1.

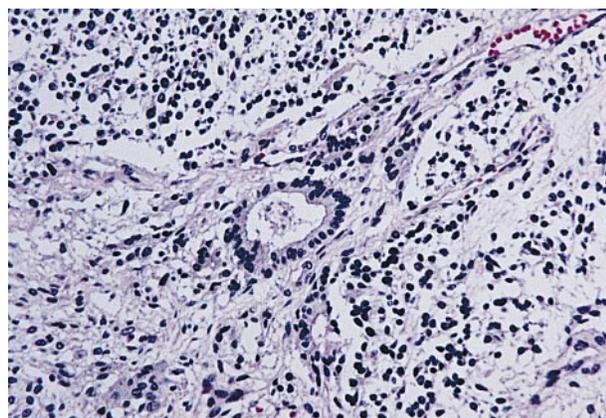


Figure 2.

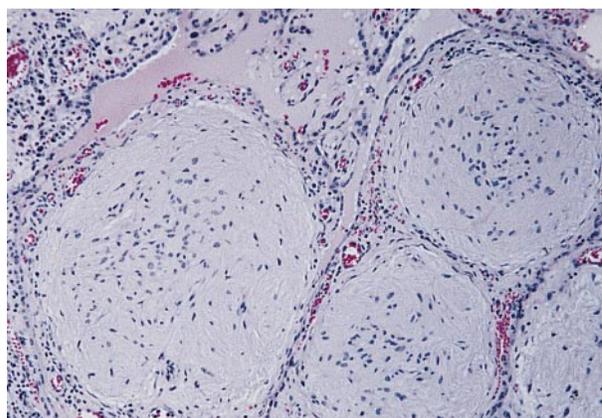


Figure 3.

Diagnosis and Discussion

Immature Ovarian Teratoma With Gliomatosis Peritonei

Figure 1. A large, partially calcified intraperitoneal mass.

Figure 2. Grade 1 immature ovarian teratoma with neuroepithelial rosette formation (hematoxylin-eosin, original magnification $\times 132$).

Figure 3. Grade 0 glial implants in omentum (hematoxylin-eosin, original magnification $\times 66$).

Ovarian teratomas are graded by criteria of Robboy and Scully,¹ which have been modified by Thurlbeck and Scully² and Dehner.³ A teratoma with no immature tissue is grade 0; with fewer than 1 low-power field of immature tissue per slide is grade 1; with more than 1 but fewer than 4 low-power fields per slide of immature neuroepithelial foci is grade 2; and a teratoma with consecutive microscopic fields of immature tissue, either immature neuroepithelial or somatic, is grade 3. All grades 1 and 2 ovarian teratomas in children act in a benign fashion. All grade 3 teratomas that exhibit malignant behavior tend to have foci of yolk sac tumor.

Light microscopy showed a grade 1 immature teratoma of the ovary. Most areas of the tumor were composed of abundant mature glial tissue, choroid plexus, skin and adnexal structures, gut mucosa, bone, cartilage, and teeth. Only scanty amounts of immature mesenchymal elements, cartilage, and primitive neuroepithelium were observed (Figure 2). The omental implants were composed of nodular mature astroglial tissue (Figure 3) and were positive for glial fibrillary acidic protein and S100 protein.

Gliomatosis peritonei (GP) is characterized by milimetric implants of mature glial tissue on the peritoneum or omentum.⁴ Gliomatosis peritonei is usually associated with immature, or rarely with mature, ovarian teratoma (OT). Recognition of this relationship is important because the gross appearance of GP looks like peritoneal carcinomatosis⁵; hence, many cases of low-grade OT associated with GP were treated initially by excision and followed improperly by chemotherapy or radiotherapy.^{1,4,6-10} Despite widespread involvement of peritoneal surfaces, GP does not adversely affect the prognosis of primary OT.

Before making the diagnosis of GP, 2 important prerequisites must be strictly met: (1) peritoneal surface, omentum, and diaphragmatic surfaces must be extensively sampled during the exploratory operation and (2) each of the sampled implants should be composed exclusively, or almost exclusively, of grade 0 glial tissue according to the criteria proposed by Thurlbeck and Scully.²

A review of the English-language literature showed only about 65 isolated cases of glial implants on the peritoneum associated with OT reported before 1994. The largest series of cases are from Robboy and Scully,¹ 5 cases; Norris et al,⁶ 7; Nogales et al,⁷ 4; Nielsen et al,¹¹ 4; and Harms and Janig,⁸ 13. Others were isolated cases.^{2,4,5,9,10,12} Of the reported cases, some immature glial implants (grades 1, 2, or 3) or other teratomatous implants, or both, were found, either at the time of the first operation or within a short period thereafter. However, according to the 2 previously men-

tioned diagnostic criteria, such cases, instead of being diagnosed as GP, should have been diagnosed as metastatic teratoma that would require further aggressive therapy.¹² With these considerations, only 45 of the cases are acceptable as GP. Of these 45 cases, the average age at the time of diagnosis was 15 years (range, 2-50 years); the average time of follow-up was 46 months (range, 6-120 months).

The study by Norris and associates⁶ showed that when only OT was present, the 10-year survival rate for patients with grade 3 OT was 30%; grade 2 OT, 62%; and grade 1 OT, 82%. However, we found that only 1 of the 45 instances of OT with GP underwent malignant transformation 5 years after the initial operation, resulting in the eventual death of the patient. These results suggest that GP does not adversely affect the prognosis of OT and may paradoxically improve the prognosis of primary OT. For the remaining 44 cases, the benign peritoneal glial implants persisted without change, fibrosis, or disappearance. Because most of the patients with GP associated with OT survived, a conservative surgical approach without adjuvant radiotherapy or chemotherapy is recommended for GP.

Extensive sampling of all peritoneal implants is important in the diagnosis of GP. If no other teratomatous elements or malignant glial tissue can be found in the implants, the mature glial implants can be ignored and the methods of therapy should be judged only by the stage and grade of the primary OT. However, if immature glial tissue or other teratomatous components or both are present in the peritoneum or omentum, the treatment should be the same as for metastatic OT.

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