Ovarian mixed germ cell tumor composed of endodermal sinus tumor and immature teratoma: Case report

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Introduction

Ovarian germ cell tumors are derived from primitive germ cells of the embryonic gonad and usually affect young women. Mixed germ cell tumors are relatively rare, comprising only 8% of all ovarian germ cell tumors. In ovarian mixed germ cell tumors, the second most common endodermal sinus tumors are presented in 70% and immature teratomas in 53% [1].

We report a case of a 25-year-old female with an ovarian mixed germ cell tumor composed of endodermal sinus tumor and immature teratoma.

Case Report

The patient was a 25-year-old female, gravida 0, para 0. She presented at our hospital with lower abdominal fullness. On pelvic examination a newborn-head sized tumor was palpated and ultrasonography demonstrated a solid tumor measuring 131x87x127 mm with ascites in the cul-de-sac. She was admitted for surgery on February 26, 1997. Chest X-ray, lung CT and upper gastrointestinal endoscopy showed no abnormal findings. Laboratory results were all within normal limits. Tumor markers were 318 IU/ml in CA125, 6316 ng/ml in alpha-fetoprotein (AFP), 11.2 IU/ml in CA19-9 and 0.4 ng/ml in CEA.

The T_2-weighted MR findings revealed a solid tumor with heterogeneous high intense areas (Figure 1), although on T_1-weighted images the tumor emitted a hypointense signal in the majority of parts (Figure 2).

At laparotomy the ruptured tumor arose from the right ovary. No apparent findings of dissemination were observed in the abdominal cavity. Frozen section examination diagnosed an ovarian mixed germ cell tumor in clinical Stage Ic. A total hysterectomy, bilateral salpingo-oophorectomy and omentectomy were performed.

Macroscopically, marked hemorrhage and necrosis were visible in the cut surface of the tumor. The histological findings revealed an ovarian mixed germ cell tumor composed predominantly of endodermal sinus tumor (EST) and rarely of immature teratoma. In the EST component, Schiller-Duval bodies and polyvesicular vitelline patterns were identified (Figure 3) and the grade II immature teratoma was composed of immature neuroepithelial cells forming rosettes (Figure 4). There was no cancerous involvement in the uterus or left ovary.

After surgery the patient received five courses of the PEP regimen as adjuvant chemotherapy. PEP consisted of cisplatin (70 mg/m^2 intraveneously on day 1), etoposide (100 mg/m^2 intraveneously daily from day 1 to 2) and pepleomycin (7 mg/m^2 intramuscularly three times every week). Serum CA125 and AFP decreased to the cut-off values after the second course of chemotherapy and remained below the cut-off values thereafter. She has had no evidence of recurrence for three years since surgery.

Discussion

EST is an uncommon tumor accounting for approximately 10 to 15% of all ovarian germ cell tumors.

Although 71% of patients with EST are stage I at the time of diagnosis, subclinical metastasis is present in 84% of stage I patients [1]. The poor prognosis of EST is characterized by extremely rapid growth and extensive intra-abdominal spread.

Immature teratoma contains immature tissue derived from the ectoderm, mesoderm and endoderm. About 70% of patients are diagnosed at stage I. The survival of patients with stage I was 100% if the tumor was grade I, 70% if grade II and 33% if grade III [1].

Some tumor markers are characteristic of ovarian germ cell tumors. AFP may be positive in EST, immature teratoma, mixed germ cell tumor and embryonal carcinoma [2], and CA125 may be identified in immature teratoma and dysgerminoma [3]. In our case, the elevated levels of serum CA125 and AFP were noted preoperatively but these tumor markers decreased promptly after surgery and the postoperative PEP regimen. Thus, tumor markers are useful in monitoring the response to therapy in germ cell tumors.

The current management is initial surgery with surgical staging and cytoreduction, followed by adjuvant chemotherapy. Generally, ovarian germ cell tumors are much more chemosensitive than epithelial ovarian cancers. Excellent results have been achieved in stage I disease with the PVB regimen (vinblastine, bleomycin and cisplatin). We used the PEP regimen to reduce the risk of pulmonary fibrosis due to bleomycin and confirmed the efficacy of this regimen in this case. Currently, the most effective and popular regimen appears to be the BEP regimen [2] using bleomycin, etoposide and cisplatin. Gershenson *et al.* [4] reported that 25 of 26 patients treated with BEP remained in sustained remission. Williams *et al.* [5] reported that three courses of BEP would

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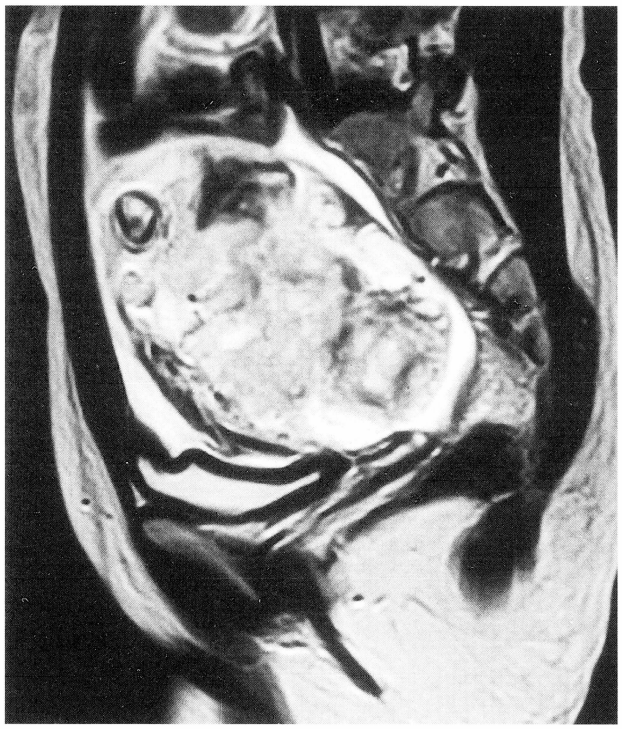

Figure 1. — Sagittal MRI findings of a mixed germ cell tumor on T_2-weighted images.

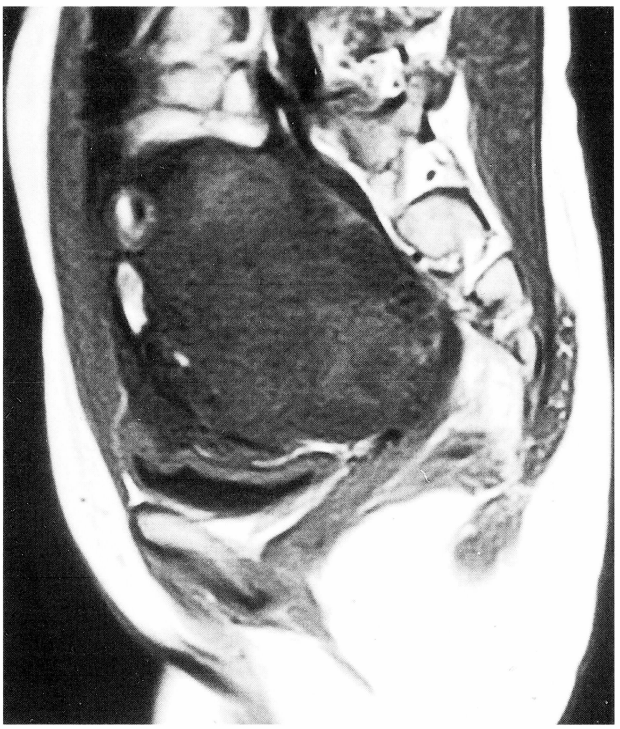

Figure 2. — Sagittal MRI findings of a mixed germ cell tumor on T_1-weighted images.

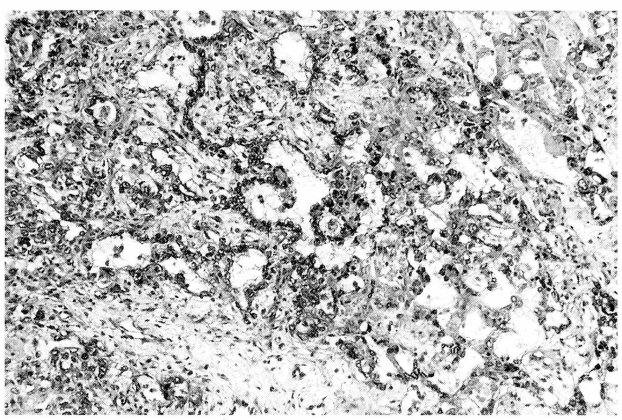

Figure 3. — Histological finding of mixed germ cell tumor (endodermal sinus tumor component).

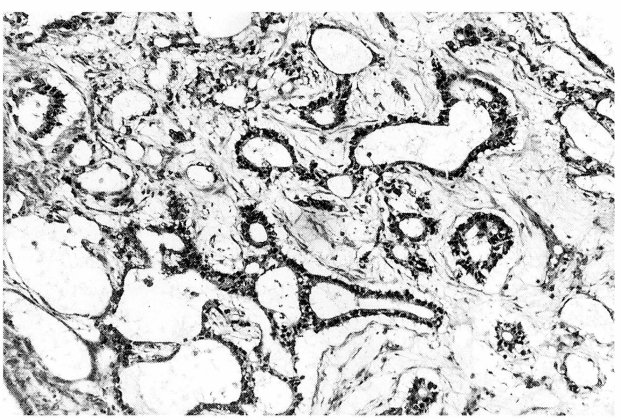

Figure 4. — Histological finding of mixed germ cell tumor (immature teratoma component).

nearly always prevent recurrence in well-staged patients with completely resected ovarian germ cell tumors.

In summary, we have confirmed the effectiveness of the PEP regimen in a patient with an ovarian mixed germ cell tumor composed of EST and immature teratoma who had been initially resected. Further improvement of chemotherapy may follow new surgical approachs to preserve fertility in young patients.

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