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Case Report

Ovarian Dysgerminoma Associated With Fibrosarcoma: A Case Report

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Summary: A 35-year-old woman presented with abdominal pain and weight loss. Pelvic computed tomography showed a 15 cm mass in the left ovary. Grossly, the removed ovary was completely replaced by a solid tumor mass. On histological analysis (100 sections), the lesion showed the typical morphological features of dysgerminoma (20%) admixed with a major (80%) fibrosarcoma component. Tumors did not have well-demarcated boundaries with a close intermingling of both cell types. Despite surgery and combination chemotherapy, the disease progressed rapidly and the patient died of disease 18 months after diagnosis. Review of the literature showed that soft tissue sarcomas of several types may occasionally be associated with gonadal and extragonadal mixed germ-cell tumors or with spermatocytic seminoma of the testis. However, no previously published report of an ovarian fibrosarcoma associated with a pure dysgerminoma was found in the literature. **Key Words:** Ovary—Dysgerminoma—Fibrosarcoma.

Dysgerminomas account for nearly 50% of primitive germ-cell tumors (GCTs), for 1% of all ovarian cancers, and for 5% to 10% of ovarian cancers in the first 3 decades (1). They may occur in a pure form or mixed with other types of GCTs including embryonal carcinoma, yolk sac tumor, choriocarcinoma, and teratoma (2).

Development of somatic malignancies in GCTs is a rare but well-known phenomenon that can occur in neoplasms of gonadal, mediastinal, and intracranial origin (3,4). The somatic neoplasia may be a carcinoma of any type, or, more frequently, a sarcoma (5). Only very few cases of GCTs with sarcomatous components (SCs) have been reported in the ovary (6).

Herein, we present the clinicopathologic features and immunohistochemical findings of the first documented case of an ovarian dysgerminoma associated with a fibrosarcoma.

CASE REPORT

A 35-year-old, gravida 1, para 1 woman was admitted to the hospital because of lower abdominal and pelvic mass associated with abdominal pain and weight loss. A computerized tomography scan of the abdomen showed a 15×13 cm solid mass in the left adnexa. A serologic test showed an elevated CA125 at 165.7 U/mL (normal range: <35 U/mL); furthermore, the lactate dehydrogenase level was markedly elevated at 950 IU/L at presentation (normal range: 105–333 IU/L) and reduced after surgery; the α-fetoprotein and human chorionic gonadotropin levels were within normal limits.

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The patient underwent exploratory laparotomy. Findings included a normal-sized uterus, a 17 × 14 cm left adnexal mass, a normal-appearing right ovary, and 2 to 3 cm nodules within the omentum. A left oophorectomy, omentectomy, and pelvic wash were performed.

Pathological Findings

The ovarian mass measured 17 × 14 cm and weighed 760 g. Grossly, the tumor was lobulated and solid with a capsular disruption. The cut surface was fleshy, yellow, with multiple areas of necrosis (Fig. 1).

Numerous sections (up to 100 sections) were submitted for microscopic examination. For immunohistochemical examination, tissue sections were stained with the following antibodies: KIT (CD117; 1:100; Dako, Carpinteria, CA), placental alkaline phosphatase (clone 8A9; 1:40; Dako), S100 protein (polyclonal; 1:800; Dako), CD34 (cloneQBEnd10; 1:40; Dako), CD10 (clone c56C6; 1:50; Dako), myogenin (clone F5D; 1:25; Dako), smooth muscle actin (clone 1A4; 1:100; Dako), vimentin (clone V9; 1:2000; Dako), and cytokeratins (clone AE1/AE3; 1:100; Dako).

In 20 sections, the tumor was composed of sheets of cells, with large, round, vesicular nuclei and abundant clear cytoplasm. The stroma consisted of thin-to-broad fibrous bands that contained mature lymphocytes (Fig. 2). These cells were positive for CD117 (c-kit) (Fig. 3) and placental alkaline phosphatase. The histologic and immunohistochemical features were in keeping with the diagnosis of dysgerminoma. Immediately next to this lesion was a second component, which represented approximately 80% of the tumor. It was composed of densely cellular, intersected bundles of spindle cells with moderate-to-severe atypia. Patchy areas with a storiform pattern were also present. The mean mitotic count was 8 mitoses per 10 high-power fields and ranged from 6 to 30/10 high-power fields. Abnormal mitotic figures were seen in all fields. These areas were positive for vimentin and negative for cytokeratins, S100 protein, CD34, smooth muscle actin, CD10, and myogenin, confirming this element of the tumor as

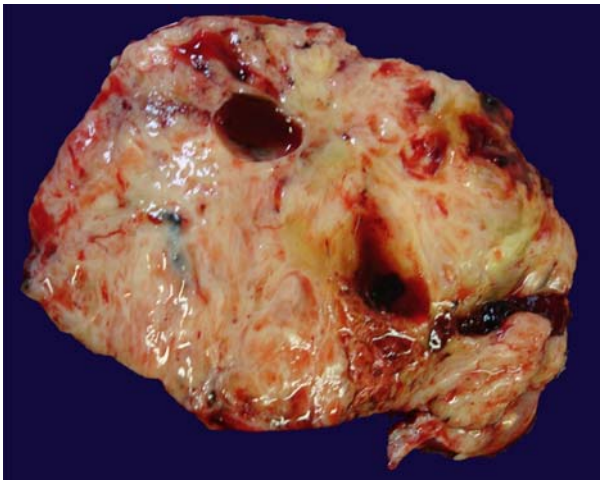

FIG. 1. Ovarian tumor, gross findings. The sectioned surface is fleshy, gray-white and yellow with hemorrhage, necrosis, and a few cystic structures.

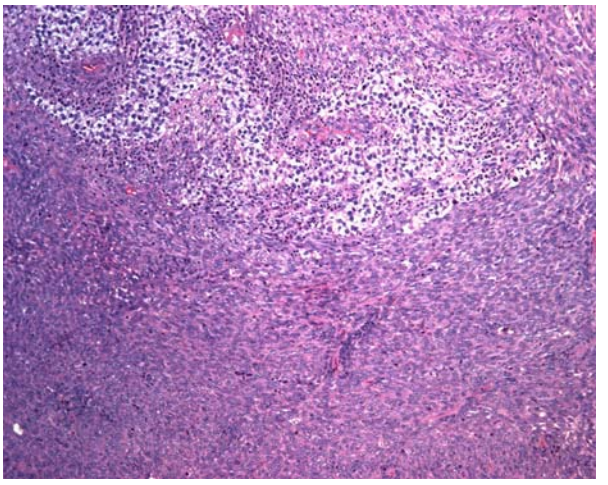

FIG. 2. Low-power view of an area in the ovarian tumor showing intermixing of dysgerminoma with fibrosarcoma (200 ×).

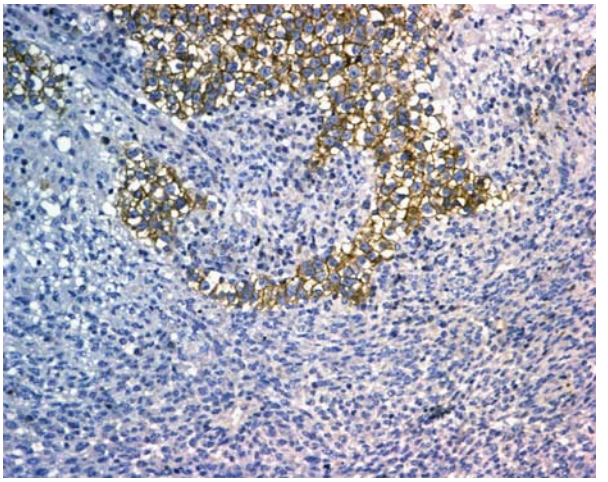

FIG. 3. CD117 stain highlighting the dysgerminoma component (200 ×).

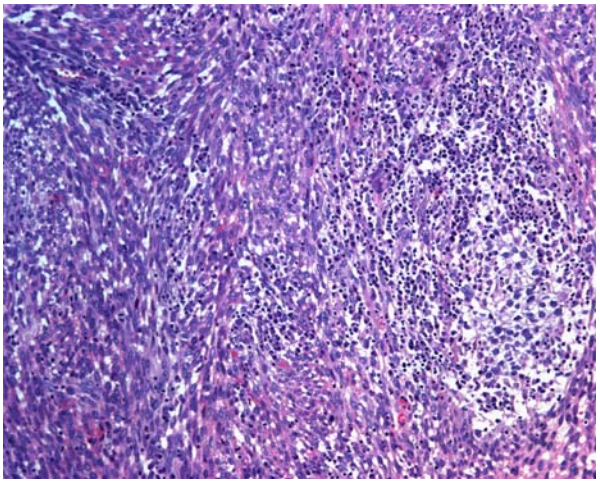

FIG. 4. High-power view of ovarian dysgerminoma associated with fibrosarcoma (400 ×).

being a fibrosarcoma. With these histopathologic findings, the case was diagnosed as dysgerminoma associated with fibrosarcoma (Fig. 4). The omental nodules were composed of only the fibrosarcomatous component and there were no cancer cells in peritoneal lavage. Thus, the patient was placed in the Federation of Gynecologic and Obstetrics stage III category.

Follow-up

After surgery, the patient was treated with 4 cycles of bleomycin plus etoposide and cisplatin, but the abdominal computerized tomography scan 1 year later showed a recurrence of the SC in the pelvis and abdominal cavity. In spite of 5 additional cycles of chemotherapy administered, the disease progressed rapidly and the patient died of disease 18 months after diagnosis.

DISCUSSION

About 8% of all malignant GCTs of the ovary show more than 1 type of neoplasm (7). They also have the capacity to progress to a higher or lower grade of differentiation, change from malignant to benign behavior, or develop mature metastasis from an immature primary (8). In contrast, the development of a secondary somatic (or nongerm cell) malignant component in GCTs is a well-known phenomenon. This phenomenon has been observed in GCTs of various origins, including those of the testis, ovary, mediastinum, and intracranial cavity (6,9). Histologically, the secondary component may resemble a somatic malignancy derived from any of the 3 germinal layers. Sarcomas are the most common somatic malignancies observed in GCTs (10). The SC may appear in the primary tumor or manifest subsequently in the recurrences or in the metastases. Ovarian GCTs with SCs are extremely rare, with very few reported cases in the literature. The most common SCs are leiomyosarcoma and rhabdomyosarcoma (6).

To the best of our knowledge, the association of a dysgerminoma with fibrosarcoma has never been reported. A similar case of sarcoma associated with a pure dysgerminoma was reported by Akhtar et al. (11) in 1989. However, in this case, the SC was that of a rhabdomyosarcoma. The possibility of a mixed GCT in our case was excluded by a careful and thorough sampling from all parts of the tumor.

The SC tends to occur in GCTs that contain a teratomatous mesenchyma. However, SCs have also been reported in GCTs without a teratomatous component. About 6 cases of spermatocytic seminoma with SCs have been reported. In these cases, no teratomatous component was present (12). These results suggest that the development of an SC in a GCT may use other mechanisms, such as aberrant differentiation of primitive germ cells (9,11).

Furthermore, it is important to note that the sarcomatous elements may overshadow the accompanying ovarian tumor, giving the erroneous impression of a pure ovarian sarcoma (13). It is therefore imperative that the tumor containing sarcomatous elements be sampled thoroughly to reach an exact diagnosis.

Primary fibrosarcoma of the ovary is an exceedingly rare tumor that usually occurs in the fifth to eight decade of life, with an average of 58 years. Regarding the histogenetic aspect of ovarian fibrosarcoma, there is currently a general impression that this tumor develops as a result of malignant change within an ovarian fibroma rather than occurring as a malignant tumor *de novo* (14).

A few cases of ovarian fibrosarcomas coexisting with benign cystic teratomas have been described (14). Perhaps some of these cases arose from the teratoma component. In contrast, Ali et al. (13) have recently reported a case of an ovarian yolk sac tumor associated with fibrosarcoma. In this case, the development of fibrosarcoma could be the result of transformation of the blastematous stroma in the yolk sac tumor.

The presence of a SC in GCTs seems to be associated with a poor prognosis, as most patients presented at an advanced stage of disease (6,9).

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