Pathologic Quiz Case

A Right Adnexal Mass in a Postmenopausal Patient

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A 63-year-old woman with no significant prior medical history presented to her primary care physician with complaints of abdominal swelling and discomfort. An abdominal ultrasound revealed a large right adnexal mass. Subsequent laparotomy showed a 20 × 9.5 × 5.5-cm, friable, red-tan mass, which appeared to obliterate the right fallopian tube. Shaggy, friable tumor implants were noted on the surface of the right ovary, the serosal surface of the uterus, and on the left adnexa. Apparent tumor implants were also seen on the omentum and diaphragm.

An abdominal hysterectomy with bilateral salpingo-oophorectomy was performed, and biopsies of the omentum and diaphragm were taken. Histologic sections of the tumor showed a neoplastic proliferation of cells distending the right fallopian tube. Residual nonneoplastic fallopian tube plicae were seen adjacent to the neoplasm (Figure 1), which contained a mixture of carcinomatous and sarcomatous elements. The carcinomatous portion of the tumor (Figure 2) assumed endometrioid, papillary serous, and squamous patterns. The sarcomatous portion of the tumor showed undifferentiated areas, as well as areas of cartilaginous differentiation. Frank chondrosarcoma was seen in multiple sections (Figure 3). In focal areas, spindle-shaped cells with bright red cytoplasmic granularity were seen (Figure 4). Carcinoma in a predominantly papillary serous pattern was seen in the biopsies of the omentum and diaphragm. Numerous psammoma bodies were present in the disseminated foci of carcinoma. Carcinoma in a predominantly papillary serous pattern was also seen on the surfaces of the left and right ovaries, the left fallopian tube, and the serosal surface of the uterus. Carcinoma was not identified in the endometrial cavity or in the parenchyma of either ovary.

Cytologic examination of peritoneal fluid sampled at the time of laparotomy showed numerous papillary clusters of tumor cells. Cytokeratin staining was strong in all epithelial components and weak in the sarcomatous portions of the tumor. Stains for actin and desmin were strongly positive in the spindle-shaped cells, with bright red cytoplasmic granularity. Despite aggressive chemotherapy, the patient died 3 months after presentation.

What is your diagnosis?

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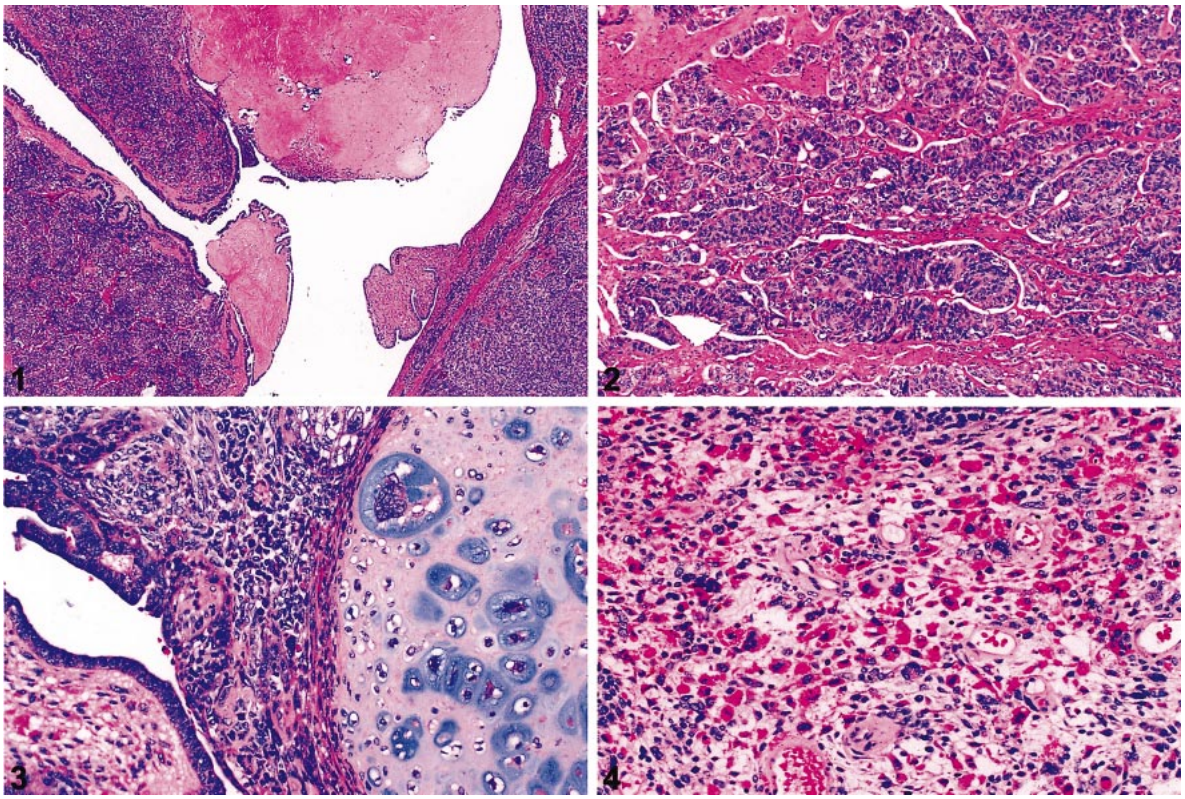

Pathologic Diagnosis: Malignant Mixed Müllerian Tumor With Heterologous Elements Arising in the Fallopian Tube

Primary malignant neoplasms of the fallopian tube are exceedingly rare. The most common of these tumors, carcinoma of the fallopian tube, accounts for less than 1% of all primary neoplasms of the female reproductive tract.[1] Tumors with both malignant epithelial and mesenchymal elements, malignant mixed müllerian tumors (MMMTs), are exceptionally rare in the fallopian tube, having been reported in fewer than 100 cases in the medical literature. Much like the more common MMMTs of endometrial or ovarian origin, MMMTs of the fallopian tube show carcinomatous elements with predominantly glandular differentiation, which may assume endometrioid, clear cell, and papillary serous patterns; less commonly, squamous differentiation or undifferentiated carcinomas may be seen. The sarcomatous components of these tumors may consist of generally undifferentiated malignant mesenchymal elements or any combination of skeletal muscle, smooth muscle, cartilaginous, or bony differentiation. If the sarcomatous portions of the tumor show differentiation toward mesenchymal tissues native to the fallopian tube, such as smooth muscle, the tumor is designated as having homologous elements; if differentiation toward mesenchymal elements not normally seen in the fallopian tube (such as cartilage or bone) is present, the tumor is designated as having heterologous elements.[2]

The tumor in the current case contained distinct heterologous elements. While overt chondrosarcoma was easily recognized, rhabdomyoblastic differentiation indicative of rhabdomyosarcoma was suspected on routine stains when clusters of polygonal cells with bright red cytoplasmic granularity were seen. Immunohistochemical stains for actin and desmin highlighted these cells, and electron microscopic studies of the tumor showed ultrastructural features of skeletal muscle differentiation. Cytokeratin stains performed on representative sections of the tumor showed strong staining in the epithelial components and focal weak staining in the sarcomatous portions, a common finding in malignant mixed mullerian tumors.

The true origin of these tumors is somewhat unclear; cases have been described in the ovaries, uterus, vagina, fallopian tube, and even as a primary lesion of the peritoneum with no demonstrable tumor elsewhere in the genitourinary tract.[3,4] Since a high percentage of cases show cytokeratin and epithelial membrane antigen positivity in the sarcomatous foci, some debate exists as to whether these tumors represent true mixed carcinomatous/sarcomatous lesions or whether they represent metaplastic carcinomas.[5] The main differential diagnosis in cases of MMMTs is immature teratoma. This distinction can be somewhat difficult, as both tumors may contain elements derived from all 3 germ cell layers; neuroectodermal tissue, however, has only rarely been reported in MMMTs and should constitute only a minor component of the tumor. In addition, the carcinomatous component of an immature teratoma is classically embryonal in appearance, a pattern not characteristic of the epithelial component of MMMTs. These 2 tumors also occur in different age groups; immature teratomas show a peak in childhood and in young adulthood, while MMMTs occur in the postmenopausal setting.

Malignant mixed mullerian tumors have an extremely poor prognosis regardless of their site of origin. Even with surgical intervention and combination chemotherapy, survival is usually measured in months, with occasional reports of cases in which patients survived 5 years or longer.[6,7] A major factor contributing to this high mortality rate is the presence of widely disseminated disease in the pelvic and abdominal cavities in 75% to 90% of cases at the time of presentation.[5,8] With a highly aggressive nature and limited therapeutic options, MMMTs represent one of the most lethal neoplasms of the female genitourinary tract.

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