Radiological Appearance of Primary Ovarian Angiosarcoma in a 79-Year-Old Woman: a case report and literature review

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ABSTRACT

Primary ovarian angiosarcoma is an extremely rare disease. Most reports concern young premenopausal patients, and relatively few case reports regarding elderly patients exist [1]. In this report, we present a case of ovarian angiosarcoma in a 79-year-old woman. The patient’s initial chief complaint was abdominal distention. Sonography, computed tomography (CT), and magnetic resonance imaging (MRI) demonstrated a large heterogeneous mass with hemorrhaging, solid compartmentalization, and an abundant vascular network. The diagnosis of primary ovarian angiosarcoma was verified using immunohistochemistry staining with CD34, CD31, and Fli-1. In this report, we describe the sonographic, CT, and MRI features of this case of angiosarcoma.

CASE REPORT

The 79-year-old woman, G3P3, had a history of diabetes mellitus which was controlled using oral medicine for 7-8 years. She was admitted to the hospital because of poor appetite, abdominal distention, and constipation for 2 months. Laboratory data revealed an elevated serum level of CA-125 (183.2 U/mL), but a normal CEA level. Transvaginal ultrasound showed a large mixed hyper- and hypoechoic mass lesion in the pelvic region, which measured 19.7 × 15.0 × 18.3 cm and exhibited low echogenicity in the center (Fig. 1). A pre-contrast CT revealed a large lobulated mass lesion in the lower abdominal cavity with mixed low-density components. An amorphous high-density and post-contrast CT showed marked serpiginous contrast enhancement, which corresponded to the vascular structure (Fig. 2). T1-weighted fast spin-echo (FSE) MR images showed a large solitary mass lesion containing certain hemorrhagic components. Sagittal T2-weighted FS-FSE MR images...
showed a heterogeneous mass lesion with a multi-loculated cystic hyperintense component at the superior dorsal region. A post-contrast T1-weighted FS-FSE MR image showed a strong curvilinear contrast enhancement of the lesion (Fig. 3).

Based on the images of a large pelvic mass lesion, laparotomy, abdominal total hysterectomy, bilateral salpingo-oophorectomy, and pelvic peritoneal tumor resection were performed to remove a large left ovarian tumor with a gray-white to dark red solid, fibrous to fleshy surface, which was abundant with vasculature and large areas of hemorrhaging (Fig. 4a). A transverse section of the specimen showed a hypercellular left ovarian tumor and fibrous hypervascular tissue (Fig. 4b). Histologic features manifested as a proliferated, irregular, slit-like structure lined with atypical endothelial neoplasms undergoing frequent mitosis (Fig. 4c), which were immunopositive for CD31, CD34 (strongly), and Fli-1 (focally) and immunonegative for D2-40 (Fig. 4d). Based on these findings, the patient was confirmed to have angiosarcoma arising from the focal background of the left ovary.

DISCUSSION

Angiosarcoma is a rare neoplasm that accounts for less than 1% of all sarcomas. Angiosarcoma involving the female genital tract accounts for approximately 4% of uterine and 1% of ovarian malignancies. Furthermore, it is especially rare in postmenopausal women [1, 10]. An angiosarcoma is usually large, hemorrhagic, soft, and friable. It may be confined to the ovary, but is often associated with the invasion of surrounding structures [6]. Abdominal and retroperitoneal angiosarcomas usually present as asymptomatic masses. Patients may present with neurologic symptoms from the compression of lumbar or pelvic nerves.
Most previously reported ovarian angiosarcomas have nonspecific or gastrointestinal symptoms, in which the most common symptom is abdominal pain. Our patient presented with only abdominal distention, constipation, and poor appetite.

The definite diagnosis of angiosarcoma mainly depends on the histopathological result. Ovarian angiosarcomas are composed of vascular spaces of varying size and appearance, and are associated with certain sex-cord stromal and germ cell tumors, which are lined with endothelial cells that are usually large and show an atypical appearance, bizarre nuclei, and frequent mitotic activity. Angiosarcomas usually express the typical vascular antigens including von Willebrand factor, CD31, CD34, and Fli-1. Immunopositivity to actin and laminin can also be found [2]. Staining for epithelial markers and HMB, melan A, and S- protein may help in differentiation to confirm the sex-cord stromal or germ cell origin of the tumor cell markers [12]. It may also enable us to confirm this diagnosis.

Based on our research and experience, this 79-year-old woman is the eldest patient of this disease entity. The imaging features of ovarian angiosarcoma have not been reported in detail in previous literature.

Sonography, CT, and MRI studies of the tumor were performed in our case. A transvaginal ultrasound showed a large mixed-echoic mass lesion exhibiting central hypoechogenicity in the pelvic region, indicating the existence of central necrotic or cystic components. Unenhanced CT images showed a large tumor with an irregular margin and being isodense as adjacent organ. The presence of low-density areas within the tumor indicates the occurrence of necrotic or fibrotic changes. On contrast-enhanced CT images, some areas showed marked serpiginous contrast enhancement, which may correspond to the vascular structure. The peripheral and progressive centripetal enhancement patterns of the angiosarcoma group have been reported to be similar to that of cavernous hemangioma [13]. On T1-weighted FSE MR images, the mass lesions contained irregular areas of high signal intensity, which may represent the focus of the hemorrhage. Some reports have mentioned the high frequency of hemorrhaging on angiosarcomas, which is demonstrated in the T1-weighted images [14]. In addition, the presence of heterogeneous signal intensity on FSE-FS T2-weighted images, with some multi-loculated hyperintensity at the superior aspect of the mass lesion, may represent cystic components. Moreover, hypointensity that leads to the separation of the mass lesion is another finding that reflects the compartmentalization of the fibrous solid portion of ovarian angiosarcomas [14]. This is consistent with pathological results indicating that angiosarcoma may arise from the focal background of a fibroma. Images obtained after contrast agent administration showed a substantial contrast enhancement of a viable peripheral tumor, which is consistent with the abundant vasculature of angiosarcoma. Thus, MRI can be used to demonstrate the cystic, hemorrhagic, and hypervascular features of an ovarian angiosarcoma, and MRI can provide more information on soft-tissue characteristics compared to a sonography and a CT in an imaging evaluation of such a case. However, angiosarcoma arising from the ovary must be distinguished

**Figure 3.** The abdominal MRI with and without contrast showed a large pelvic mass. a. The pre-contrast axial T1-weighted FSE MR image and b. sagittal T2-weighted FS-FSE MR image demonstrate a mass with an intermediate signal intensity and multi-loculated hyperintensity, which indicates the presence of cystic components. (H) c. The post-contrast coronal T1-weighted FS-FSE MR image showed marked peripheral heterogenous contrast enhancement of the tumor after intravenous administration of gadolinium-DTPA, which corresponds to the abundant vasculature (V) of the tumor.
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The prognosis of ovarian angiosarcoma is poor, and a standard therapeutic method that can be applied to angiosarcoma treatment still does not exist. Surgical treatment, radiotherapy, chemotherapy, immune therapy, and other comprehensive therapy treatments are often adopted. Surgical excision is the main treatment of angiosarcoma. Radiotherapy or chemotherapy before or after the operation may reduce the likelihood of local recurrence and metastasis [15]. After a surgical resection of the tumor, our patient received chemotherapy as an adjuvant treatment.

Figure 4. The histologic feature of ovarian angiosarcoma. a. The gross surgical specimen and b. the transverse section of the surgical specimen showed gray-white to dark red, solid, fibrous surface tissue (F) and an abundant vasculature (V) with large areas of hemorrhaging (H). c. A proliferating, irregular, slit-like structure lined with atypical endothelial neoplasms undergoing frequent mitosis d. The neoplasms were immunopositive for CD31, CD34 (strongly), and Fli-1 (focally) and immunonegative for D2-40.
CONCLUSION

Ovarian angiosarcoma is a rare vascular, malignant soft-tissue tumor with highly malignant, invasive, and multifocal biological characteristics [1]. It uncommonly occurs in postmenopausal patients. The imaging features in this case may demonstrate a variation depending on the biological characteristics of the patient. MRI is superior to sonography and CT in demonstrating the complex components of this tumor. We suggest radiologists to consider ovarian angiosarcoma when conducting a differential diagnosis, especially regarding a complex mass with heterogeneous compartments, multi-loculated cysts, an abundant vasculature with peripheral curvilinear enhancement, hemorrhaging, and internal necrosis, as seen in our case.

REFERENCES

6. Evtushenko NT. Angiosarcoma of the ovary in a 7-year old girl. Akush Ginekol (Mosk) 1958; 34: 105