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CASE REPORT

Primary Carcinoid Tumor of the Ovary: MR Imaging Characteristics with Pathologic Correlation

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Ovarian carcinoid tumor is a rare neoplasm that may appear as a solid mass or often combined with teratomas or mucinous tumors. We report 2 cases associated with mucinous cystadenomas and describe their magnetic resonance imaging characteristics. On T_2-weighted images, the tumors appeared as multilocular cystic masses with hypointense solid components as a result of abundant fibrous stroma induced by serotonin. Demonstration of prominent hypervascularity of the tumors following contrast administration on dynamic study may be the clue to differential diagnosis.

Keywords: *carcinoid tumor, MRI, ovary*

Introduction

Primary ovarian carcinoid tumors are rare neoplasms that account for 0.3% of all carcinoid tumors and 0.1% of all malignant ovarian tumors.[1] Tumors are usually unilateral and affect post- or perimenopausal women.[1–4] Carcinoid tumor of the ovary may appear as a solid mass but is often combined with mature cystic teratomas or mucinous tumors.[2–4] On computed tomography (CT), about 60 to 80% of ovarian carcinoid tumors may appear as a solid enhancing nodule in the wall of a mature cystic teratoma.[4] When the tumor is a solid mass, imaging features may be indistinguishable from those of solid malignant ovarian tumors.[4] We believe that the specific imaging findings of ovarian carcinoid tumors have not been reported and therefore describe 2 cases of primary ovarian carcinoid tumor with regard to their characteristics on CT and magnetic resonance (MR) imaging with pathologic correlation.

Case 1

A 72-year-old woman, gravida 2, para 2, referred to our hospital for a left adnexal tumor identified at medical checkup was asymptomatic, except for constipation. She did not complain of characteristic symptoms of carcinoid syndrome, such as flushing or diarrhea. Serum tumor markers were not elevated. The patient underwent pelvic MR examination with a 1.5-tesla superconducting unit (Signa Advantage 1.5T, General Electric, USA) that demonstrated a multilocular cystic mass with a solid component of low intensity on both T_2- and T_1-weighted images (Fig. 1A) and a small amount of ascites in the pelvic cavity.

Dynamic CT examination with intravenous administration of contrast medium demonstrated marked early enhancement of the tumor's solid component that suggested its hypervascularity (Fig. 1B, C). Punctate and curvilinear calcifications were observed in the septa of the multilocular cystic components (Fig. 1B, C). We suspected malignant ovarian tumor from the hypervascular solid component. Surgery revealed a left adnexal multilocular cystic mass with solid component measuring 15 cm. Histologic section of the solid component showed cuboidal tumor cells with cellular atypia that resembled small tubules and merged with thyroid follicles containing colloid set in dense fibrous tissue stroma (Fig. 1D). The tumor cells were positive for Grimelius silver staining and chromogranin A and demonstrated slight mitotic activity. The pathologic diagnosis of the solid component was strumal carcinoid of the ovary. The multilocular cystic components were diagnosed as coexisting mucinous cystadenoma of borderline malignancy.

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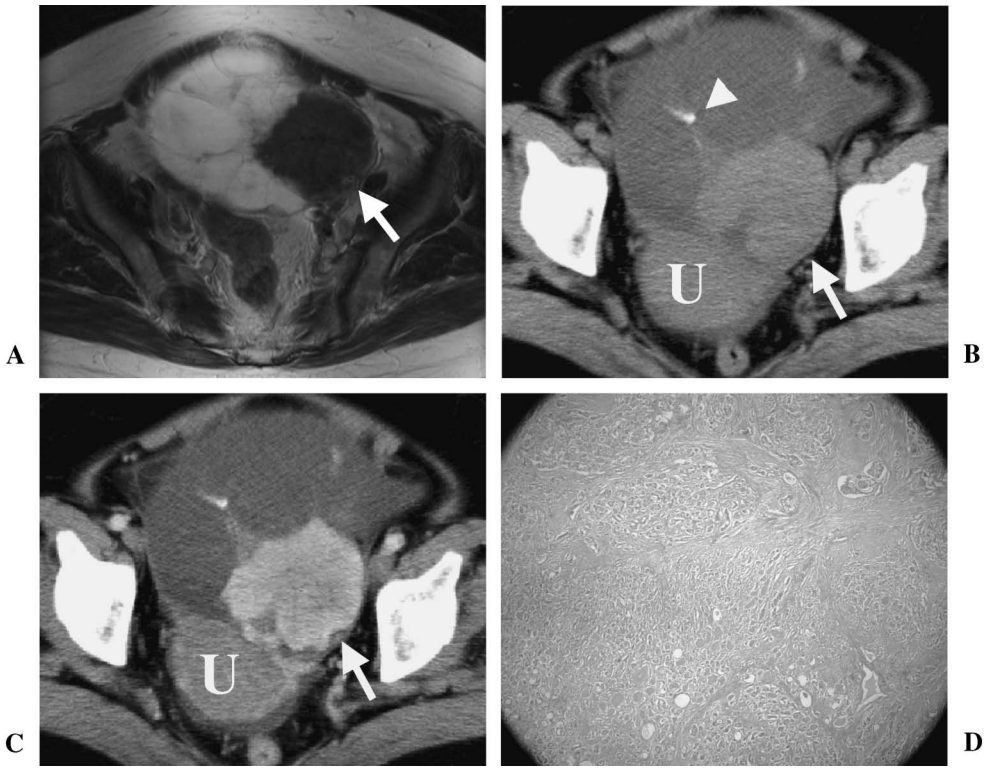

Fig. 1. (**A**) Axial fast spin-echo T_2-weighted image (repetition time [TR]/effective echo time [TE], 4200/105 ms) shows left adnexal multilocular cystic mass containing homogeneous solid component of low intensity (arrow). (**B**) On axial plain computed tomographic (CT) scan, the solid component (arrow) shows homogeneous soft tissue attenuation. Punctuate and curvilinear calcifications (arrowhead) are observed in the septa of the multilocular cystic mass. U, uterus. (**C**) On axial contrast-enhanced CT scan (early phase), intense contrast enhancement of the solid component (arrow) stronger than that of the uterine body indicates hypervascularity. U, uterus. (**D**) Photomicrograph of the mass with low power magnification (hematoxylin and eosin) shows nests of tumor cells separated by abundant fibromatous stroma. Intimate admixture of trabecular carcinoid with microfollicles containing colloid is observed.

Case 2

A 77-year-old woman, gravida 2, para 2, was referred to our hospital for genital bleeding. She did not complain of constipation or characteristic symptoms of carcinoid syndrome. Serum estradiol was slightly elevated (44 pg/mL); serum tumor markers were not. The patient underwent pelvic MR examination with a 1.5-T superconducting unit (Signa Excite HD 1.5T, General Electric) that revealed a right adnexal mass that appeared as a multilocular cystic mass with sponge-like solid portion and peripherally situated solid component of low intensity on both T_2- and T_1-weighted images (Fig. 2A). We observed hemorrhagic loculi of high intensity on fat saturated T_1-weighted images (Fig. 2B). Solid components showed high signal intensity on diffusion-weighted images with relatively low apparent diffusion coefficient (ADC) values (0.76×10^{-3} mm^2/s in the peripherally situated component of low intensity; 1.43×10^{-3} mm^2/s in the sponge-like portion) (Fig. 2C). Dynamic MR examination with intravenous contrast administration demonstrated marked early enhancement of the tumor's solid components (both the sponge-like portion and peripherally situated solid component of low intensity) that suggested its hypervascularity (Fig. 2D). T_2-weighted images revealed uterine enlargement with endometrial thickening. Therefore, we suspected an estrogen-producing tumor, such as granulosa cell tumor. Surgery revealed a right adnexal mass measuring 10 cm. The pathologic diagnosis of the solid components (both sponge-like portion and peripherally situated solid component of low intensity) was insular carcinoid of the ovary (Fig. 2E). The tumor cells were positive for chro-

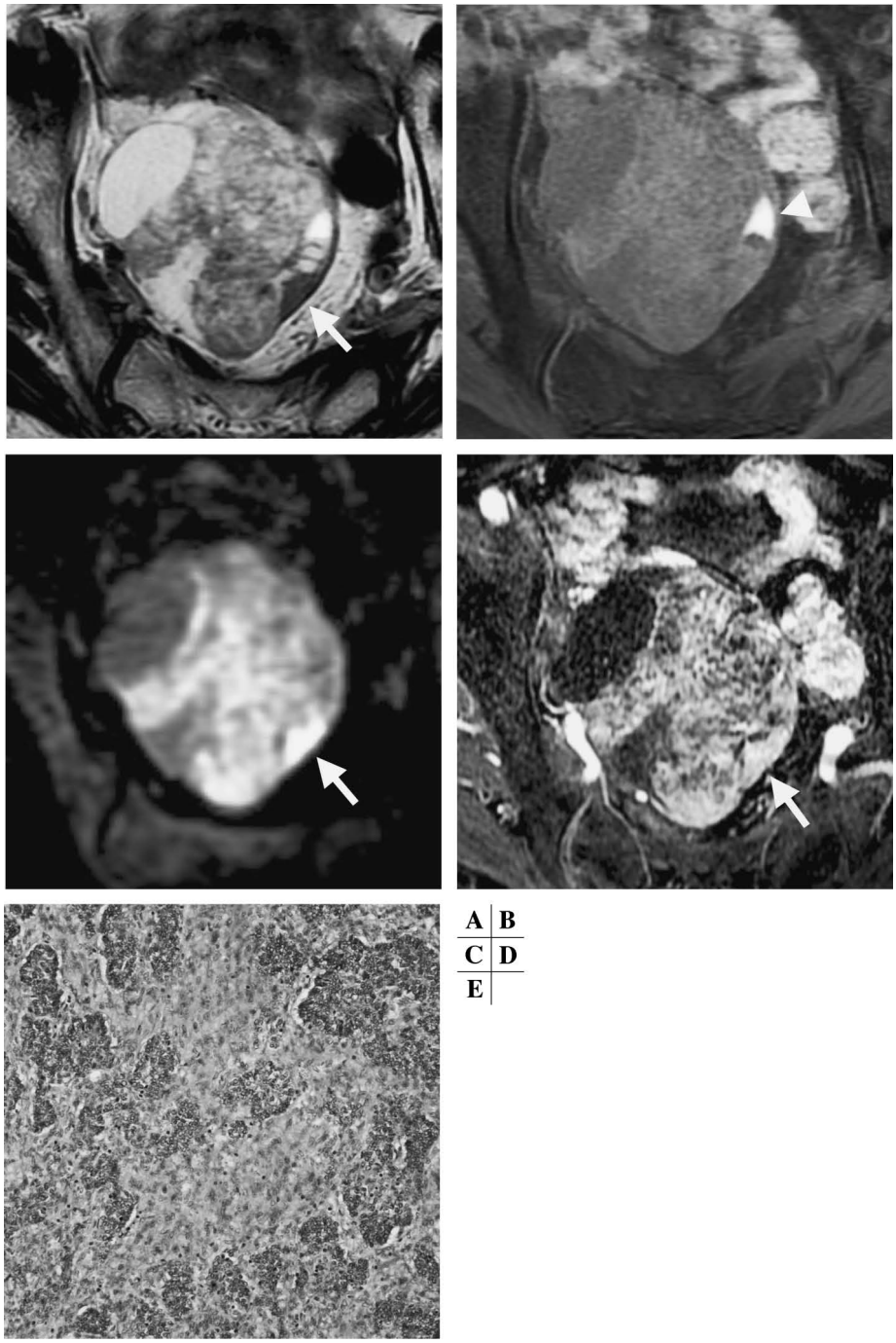

Fig. 2. (**A**) Axial fast spin-echo T_2-weighted image (repetition time [TR]/effective echo time [TE], 4000/99.3 ms) shows right adnexal multilocular cystic mass containing sponge-like large solid portion and peripherally situated solid component of low intensity (arrow). (**B**) On axial fat-saturated spin-echo T_1-weighted image (TR/TE, 516.7/7.6), hemorrhagic high intensity loculus (arrowhead) is observed. (**C**) On axial high b-value ($b = 800\ s/mm^2$), echo-planar, diffusion-weighted image (TR/TE, 6000/61.1), sponge-like large solid portion shows heterogeneous high signal intensity with relatively low apparent diffusion coefficient (ADC) value ($1.43 \times 10^{-3}\ mm^2/s$). The peripherally situated solid component (arrow) shows intensely high signal intensity with low ADC value ($0.76 \times 10^{-3}\ mm^2/s$). (**D**) On axial contrast-enhanced gradient-echo T_1-weighted image with fat suppression (early phase of dynamic study; TR/TE, 4.6/2.2), both the sponge-like portion and peripherally situated solid component (arrow) show intense contrast enhancement, indicating hypervascularity. (**E**) Photomicrograph of the peripherally situated hypointense solid component on T_2-weighted image with high power magnification (hematoxylin and eosin) shows tumor cells of insular carcinoid with fibrous stroma.

mogranin A. The multilocular cystic components were diagnosed as coexisting mucinous cystadenoma.

Discussion

Dense fibrous stromal proliferation in benign ovarian fibrous tumors, such as fibromas, thecomas, cystadenofibromas, and Brenner tumors, may cause low signal intensity on T_2-weighted images.[5] Effects of T_2-shortening by abundant collagen contents and decreased extracellular fluid compared with that of surrounding tissues contribute to the low signal intensity of these fibrous tumors on T_2-weighted images.[5] Because these fibrous tumors are usually benign neoplasms, low signal intensity in a mass may suggest benignity.[5] However, for our 2 cases of primary ovarian carcinoid tumor, which is categorized as a neoplasm with low-grade malignant potential, the low signal intensity on T_2-weighted images mimicked that of benign fibrous tumors.[1–3]

Ovarian carcinoid tumors are divided into 4 subtypes, insular (islet), trabecular, mucinous (goblet cell), and strumal carcinoids,[2–4] each characterized by fibromatous stroma from serotonin produced by the tumor that separates nests of tumor cells. Frequently, the stroma is dense and hyalinized, as in Brenner tumors or adenofibromas.[2,3] In our cases, histopathologic examination revealed dense fibrous stroma that we considered the cause of low signal intensity on T_2-weighted images. Kim also demonstrated a case exhibiting low signal intensity on T_2-weighted images,[6] an ovarian strumal carcinoid in a 66-year-old woman. A T_2-weighted image shows a septate cystic mass containing a solid portion with dark signal intensity that enhances on a contrast-enhanced T_1-weighted image.[6] Various amounts of fibrous stroma in ovarian carcinoid tumors may not always yield low signal intensity on T_2-weighted images.[4] In our Case 2, most solid portions in the carcinoid tumor appeared as a sponge-like mass that mimicked granulosa cell tumor, and only the peripherally situated small component showed low signal intensity on T_2-weighted images. Serotonin-induced stromal proliferation is generally a pathological feature of ovarian carcinoids[2,3]; low signal intensity on T_2-weighted images, which is rarely observed in other malignant ovarian tumors, may be a characteristic imaging finding for ovarian carcinoid tumors. In our second case, the peripherally situated, small, hypointense component on T_2-weighted images showed intense high signal on diffusion-weighted image with low apparent diffusion coefficient (ADC) value. Hypercellularity of tumor cells may cause signal increase on diffusion-weighted images, whereas benign fibrous tumors that exhibit low signal intensity on T_2-weighted images usually show low signal intensity on diffusion-weighted images.[7,8] Hypervascularity on dynamic study suggests functioning tumors, such as carcinoid tumors, but is rarely observed in benign fibrous tumors.[9]

Most ovarian carcinoid tumors are asymptomatic. A third of insular ovarian carcinoid tumors may be associated with the flushing and diarrhea of typical carcinoid syndrome, but other subtypes do not usually cause the syndrome.[1–3] Some trabecular or strumal carcinoid tumors may produce peptide YY, which inhibits intestinal mobility and causes chronic constipation.[1–3,10] The tumor in our Case 2 showed estrogenic activity. Various ovarian tumors with estrogenic manifestations attributable to functioning stroma have been reported.[11] Tanaka and associates reported 2 cases, a carcinoid tumor and a mucinous cystadenoma, with functioning stroma that appeared as multilocular cystic masses with varying signal on MR.[12] Any ovarian tumor, including carcinoid tumor, could have hormonal activity and mimic ordinary hormone-producing tumors.[12] It is important to diagnose carcinoid tumors before extraovarian progression because organ-confined, early stage ovarian carcinoid tumors have good prognosis and surgical excision is curative.[1]

In conclusion, clues to the differential diagnosis of primary ovarian carcinoid tumor may be: its appearance on T_2-weighted images as a fibroma-like solid mass with low intensity or as solid components of low intensity within a multilocular cystic mass that mimic Brenner tumor with co-existing mucinous cystadenoma or mucinous cystadenofibroma; high signal intensity on diffusion-weighted imaging with low ADC value; and hypervascularity following contrast administration on dynamic study. Clinical manifestations, such as typical carcinoid syndrome or chronic constipation, may also support such diagnosis.

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