Case Report

J Korean Soc Radiol 2025;86(1):166-172
https://doi.org/10.3348/jksr.2023.0107
eISSN 2951-0805

JOURNAL of THE KOREAN SOCIETY of RADIOLOGY

MRI Features with Pathologic Correlation of Primary Ovarian Carcinoid: A Case Report

일차성 난소 유암종의 MR 영상 소견과 병리학적 소견: 증례 보고

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Received August 29, 2023
Revised December 10, 2023
Accepted January 7, 2024
Published Online March 8, 2024

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Primary ovarian carcinoid tumors are rare well-differentiated neuroendocrine tumors classified as monodermal teratomas. They usually occur in perimenopausal woman and are accompanied with mature cystic teratoma or mucinous tumors. The diagnosis may be supported by the clinical presentation of carcinoid syndromes such as flushing, diarrhea, or chronic constipation. Here, we report on the case of a 51-year-old female with surgically confirmed primary ovarian carcinoid, describe the MRI features of the tumor, and correlate them with the pathological findings.

Index terms Primary Ovarian Carcinoid; Mature Cystic Teratoma; Perimenopausal; Magnetic Resonance Imaging; Computed Tomography

INTRODUCTION

Primary ovarian carcinoid tumors are well-differentiated neuroendocrine tumors based on the 5th edition of the World Health Organization (WHO) Classification of tumors. These tumors are classified as monodermal teratomas and somatic-type tumors originating from a dermoid cyst. Neuroendocrine neoplasms are categorized independently rather than in individual organ sections for consistent management, in comparison with the 4th WHO classification of cancers of the female reproductive organs. However, primary ovarian carcinoid tumors remain categorized separately from neuroendocrine tumors because of the favorable prognosis of low-grade

neoplasms (1).

Carcinoid tumors are the most prevalent type of neuroendocrine tumors, with most cases detected in the gastrointestinal and bronchopulmonary systems (2). However, primary ovarian carcinoids are extremely uncommon neoplasms, comprising 1% of carcinoid tumors and 0.1% of ovarian neoplasms (2). Consequently, there are only a few reports on specific imaging findings of primary ovarian carcinoids (3). In this report, we describe imaging features of a rare primary mixed trabecular and insular ovarian carcinoid on CT and MRI and correlate them to pathologic findings.

CASE REPORT

A 51-year-old female was referred to our hospital with multiple uterine leiomyomas, detected using ultrasonography at a local clinic. The patient's medical history was unremarkable. She presented with a palpable left abdominal mass, discomfort, and also had complained of chronic constipation for more than 10 years. A physical examination revealed no abnormalities. Laboratory test results, including a complete blood count, serum electrolytes, and CA-125 (5.5 U/mL), were normal. Abdominopelvic CT scan revealed multiple solid pelvic masses with no lymphadenopathy. The patient underwent pelvic MRI with a 3T superconducting unit (Ingenia Elition X, Philips, Best, Netherlands) to evaluate tumor origin and tissue characterization. A comprehensive evaluation of imaging and histopathology were performed after laparoscopic tumor resection.

Serial abdominopelvic CT revealed two, bilateral, adnexal, fat-density mass lesions (Fig. 1A, B). Additionally, a heterogeneous enhancing solid mass measuring 5 cm × 4.2 cm × 5 cm was observed at the anterior portion of the uterus (Fig. 1C). There was no evidence of ascites, lymph node enlargement, or distant metastases.

Pelvic MRI revealed a mass with central high signal intensity and peripheral low signal intensity on T2-weighted images (T2WI) (Fig. 1D). The peripheral solid portion showed homogeneous contrast enhancement similar to that of the uterus (Fig. 1D), demonstrated a high signal intensity on high-b-value (b = 1000) diffusion-weighted imaging, and a low apparent diffusion coefficient (ADC) value on the ADC map, indicating moderate diffusion restriction. Based on CT and MRI findings, the preoperative differential diagnoses included subserosal-type degenerated leiomyoma, ovarian fibroma/fibrothecoma, sclerosing stromal tumor, primary ovarian carcinoid.

During surgery, the mass was found in the right adnexa. Subsequently, the patient underwent a hysterectomy and right salpingo-oophorectomy. Gross examination revealed a central cystic component and a peripheral yellowish-grey soft and fibrotic tumor (Fig. 1E), coexisting with one mature cystic teratoma. Microscopically, the tumor was unencapsulated but relatively well circumscribed. Tumor cell nests were surrounded by abundant fibrotic stroma and blood vessels (Fig. 1F). It showed mixed growth patterns, mostly of the trabecular type (not shown), but also of focally insular and solid types. The tumor cells were monotonous with moderate eosinophilic cytoplasm, round to oval nuclei, and granular salt and pepper chromatin. No mitotic figures or necrotic foci were observed. Immunohistochemical staining revealed that the tumor cells were positive for pan-cytokeratin and synaptophysin; less than

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2% Ki-67; and negative for WT-1, estrogen receptors, and progesterone receptors. A unilocular cyst was observed in the remaining right ovary (Fig. 1F). The cyst wall had no epithelial lining cells but contained hair. The final pathological diagnosis was primary ovarian carcinoid tumor with a mature cystic teratoma.

This retrospective case report was prepared according to the ethical principles in the Declaration of Helsinki.

Fig. 1. A 51-year-old female with a primary ovarian carcinoid and two bilateral mature cystic teratomas.
A-C. Serial axial contrast-enhanced CT images show two fat-density mass lesions in both adnexa uteri (arrowheads, A, B), a 5 cm × 4.2 cm × 5 cm heterogeneous enhancing solid mass (arrow, C) at the anterior portion of the uterus (U), and two mild homogeneous enhancing mass lesions, representing leiomyomas (L).
D. Axial turbo spin-echo T2-weighted image (left upper) shows a mass (arrows) with central high signal intensity and peripherally low intensity. Axial T1-weighted (middle upper) and axial contrast-enhanced T1-weighted DIXON images (right upper) show peripheral solid portion with homogeneous contrast enhancement similar to that of the uterus (U). The diffusion-weighted image (b-value = 1000 s/mm^2) (left lower) shows a high signal intensity, and the ADC map (right lower) shows a low ADC value in the peripheral solid portion.
ADC = apparent diffusion coefficient, DIXON = water-selective fat saturation method

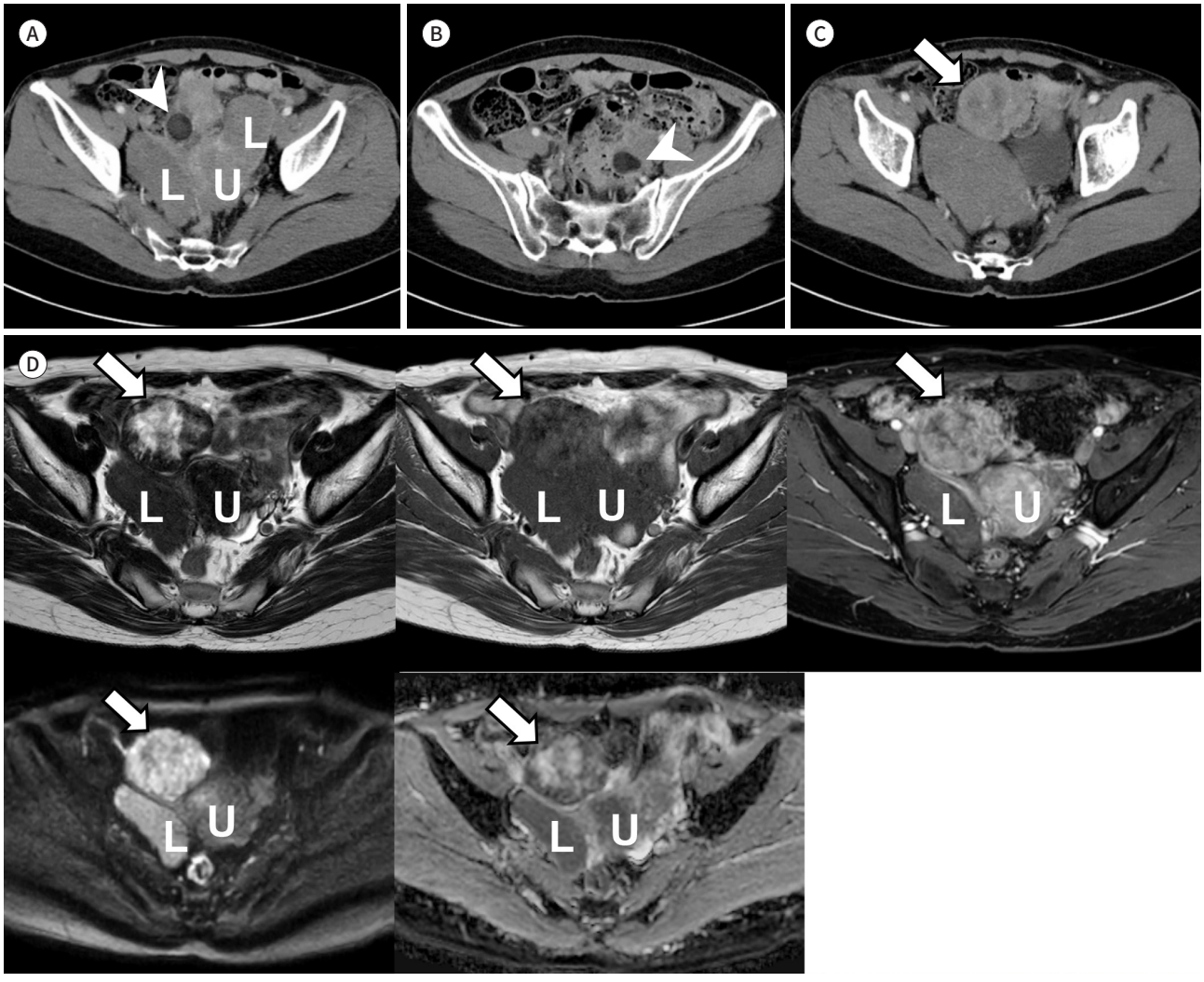
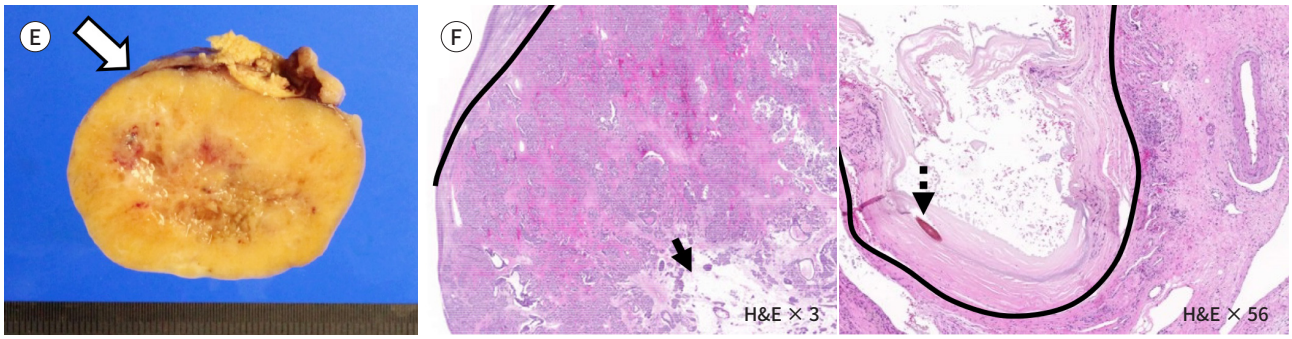


Fig. 1. A 51-year-old female with a primary ovarian carcinoid and two bilateral mature cystic teratomas.
E. The gross specimen shows a central cystic component and a peripheral yellowish-grey soft fibrotic tumor (arrow). Adapted from Choi et al. Medicine (Baltimore) 2023;102:e34391 (10).
F. On microscopic examination (H&E stain, ×3) (left) of the ovarian mass, tumor cell nests are surrounded by thick fibrotic stroma (left, below the black line). The remaining normal ovary and ovarian capsule are shown (left, above the black line). The periphery of the tumor is cellular, and the center is cystic (left, arrow). Microscopic examination (H&E stain, ×56) (right) shows a mature cystic teratoma (right, inside the black circle). The cyst wall has no epithelial lining cells but contains hair (right, dotted arrow).
H&E = hematoxylin and eosin



DISCUSSION

According to the 2020 WHO classification of female genital tumors, primary ovarian carcinoid tumors are rare, well-differentiated neuroendocrine tumors classified as monodermal teratomas (1). Carcinoid tumors of the ovary were first described by Stewart et al. (4) in 1939, who reported two cases of insular carcinoid tumors arising from an ovarian teratoma.

They are known to have the following characteristic clinical features: first, the age of incidence spans a wide range but most of these patients are post- or perimenopausal (2). In our case, the patient was a 51-year-old perimenopausal female. Second, clinical manifestations of primary ovarian carcinoids are usually asymptomatic and occasionally include abdominal pain, vaginal bleeding, dysmenorrhea, heart disease, diarrhea, constipation, hypoglycemia, and hirsutism, based on a systemic review by Zhai et al. (2). Chronic constipation is caused by the tumor-producing gut hormone peptide YY, which inhibits intestinal motility (1). In our case, the patient had chronic constipation, a clinical manifestation of the carcinoid syndrome.

Histologically, the insular type is characterized by the presence of small acini and solid nests of uniformly polygonal cells (5). These cells are rich in basophilic-to-amphophilic cytoplasm and contain a central hyperchromatic nucleus with low mitotic activity. The trabecular variant is composed of ribbons, cords, and parallel trabeculae lined with one or two layers of cells. The cords, ribbons, and trabeculae are surrounded by dense fibrous connective tissue (5). The present case showed a predominantly trabecular histological pattern with a few admixed areas of insular and solid types, indicating a fibrotic component. The stroma was fine, highly vascularized, and hyalinized, suggesting the presence of a cystic component. A unilocular cyst was observed in the remaining right ovary. The cyst wall had no epithelial lining cells but contained hair, suggesting a cystic teratoma.

CT and MRI findings clearly reflected these pathological features. Most primary ovarian carcinoids are unilateral and often combined with mature cystic teratomas or mucinous tumors (3). On CT, approximately 60%–80% of ovarian carcinoid tumors appear as solid-en-

hancing nodules in the walls of mature cystic teratomas (3). MRI shows marked early enhancement of solid masses, and the solid components show hypointensity on T1-and T2WI as a result of the abundant fibrous stroma and diffusion restriction due to hypercellularity (3). The low signal intensity of smooth muscle or fibrosis on T2WI is linked to the T2 shortening effects of intramuscular actin, myosin, and collagen and the lack of extracellular fluid in comparison to the surrounding tissues (6). Takeuchi et al. (3) demonstrated two cases of multilocular cystic masses with hypointense solid fibrotic components. In the present case, the tumor appeared as a peripheral, well-enhanced, solid mass containing a central cystic component, co-existing with two, bilateral, adnexal, mature cystic teratomas. The peripheral solid portion of the tumor showed a low signal intensity on T2WI and moderate diffusion restriction, which may be correlated with its fibrotic nature and hypercellularity. However, the enhancement pattern was limited because dynamic-phase images were not available.

Our differential diagnoses for primary ovarian carcinoid included subserosal-type degenerated leiomyoma, ovarian fibroma/fibrothecoma, sclerosing stromal tumor, primary ovarian carcinoid. The anatomical origin of a solid pelvic mass can be ovarian or extraovarian owing to the presence of small peripheral follicles and concurrent contact with the uterine fundus (6, 7). Subserosal uterine leiomyoma is a benign tumor of smooth muscle origin with varying quantities of fibrous connective tissue that often exophytically projects outwards from the subserosal location. Leiomyomas are typically multiple and tend to have an enhancement pattern similar to that of the myometrium and a low signal intensity on T2WI (7). Fibrous-containing tumor such as fibroma and thecoma exhibits low signal intensity on T2WI because of dense fibrous stromal proliferation (6). According to a previous report by Oh et al. (8), it is common to observe the remaining ovary on the same side as the fibroma on MRI, particularly in premenopausal women. Furthermore, the shape of the ovary appeared normal in cases where the fibroma exhibited exophytic growth from the periphery of the ovary (8). A sclerosing stromal tumor is a rare, sex cord-stromal tumor that occurs at a young age (< 30 years) and is commonly accompanied by menstrual irregularities (9). MRI revealed a solid tumor with a central round or cleft-like cyst with peripheral enhancement (9).

Primary ovarian carcinoids have a favorable prognosis in their early stages; however, their malignant potential remains unclear (2). The 10-year survival rate in early stages is as high as 100%, whereas the 5-year survival rate in advanced stages decreases to 33% (2). Therefore, it is crucial to diagnose organ-confined, early stage, ovarian carcinoid tumors before extraovarian progression (3). In our case, the patient underwent robotic hysterectomy and right salpingo-oophorectomy and was cancer-free, two years post-op.

In conclusion, we report a rare case of a primary ovarian carcinoid tumor. Even though this tumor is extremely uncommon, it can be considered as a differential diagnosis when an ovarian fibrotic mass is in conjunction with the mature cystic teratoma or mucinous tumor in perimenopausal woman. This diagnosis may also be supported by clinical presentations such as flushing, diarrhea, or chronic constipation.

Author Contributions

Conceptualization, all authors; data curation, all authors; investigation, K.B.J., Y.S.K.; project administration, Y.S.K.; supervision, Y.S.K., P.M.G.; validation, Y.S.K.; writing—original draft, K.B.J.; and writing—review & editing, Y.S.K.

Conflicts of Interest

Seong Kuk Yoon has been a Section Editor of the Journal of the Korean Society of Radiology since 2017; however, he was not involved in the peer reviewer selection, evaluation, or decision process of this article. Otherwise, no other potential conflicts of interest relevant to this article were reported. All remaining authors have declared no conflicts of interest.

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Funding

This study was supported by research funds from the Dong-A University.

Acknowledgments

This paper has the same patient as the previously published paper (Medicine 2023;102:e34391), but it doesn't include any previously published contents.

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일차성 난소 유암종의 MR 영상 소견과 병리학적 소견: 증례 보고

구본재[1] · 윤성국[1*] · 박민경[2] · 김경아[3]

원발성 난소 유암종 종양은 잘 분화된 신경내분비 종양으로 단모세포 기형종으로 분류되며, 드물게 보고되고 있다. 종양은 보통 폐경전후기 여성에게 발생하며 성숙 낭기형종을 동반한다. 홍조, 설사 또는 만성 변비와 같은 카르시노이드 증후군의 임상적 증상들이 진단하는데 도움이 된다. 저자들은 51세 여성에서 수술로 확인된 원발성 난소 유암종 종양의 자기공명영상 소견을 병리학적 소견과 연관하여 보고하고자 한다.

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