Case Report

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Bilateral Ovarian Fibromatosis in a Postmenopausal Female: A Case Report with Emphasis on MRI Findings and Differential Diagnosis

폐경 후 여성의 양측성 난소 섬유종증: MRI 소견과 감별진단 중점의 증례 보고

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Ovarian fibromatosis is a rare non-neoplastic condition that causes ovarian enlargement in women, typically around the age of 25. This enlargement is due to the proliferation of the collagen-producing ovarian stroma. On T2-weighted MRI, a key diagnostic feature of ovarian fibromatosis is the 'black garland sign,' characterized by multilobulated very low signal intensity along the ovarian surface. This condition also features the preservation of normal ovarian stroma or follicles internally. We present a case involving a 65-year-old postmenopausal female who was pathologically misdiagnosed with ovarian fibroma. However, the diagnosis was later revised to ovarian fibromatosis based on characteristic MRI findings. The case report discusses the differential diagnosis and pathologic findings associated with ovarian fibromatosis.

Index terms Case Report; Magnetic Resonance Image; Ovarian Neoplasms

INTRODUCTION

Ovarian fibromatosis was first described by Young and Scully in 1984 as a rare, non-neoplastic condition that leads to the enlargement of one or both ovaries. This results from the proliferation of collagen-producing ovarian stroma and can be mistaken for ovarian neoplasms (1). Ovarian fibromatosis predominantly occurs in pre-

menopausal female, with the average age of onset being 25 years, and can cause menstrual irregularities, abdominal pain, and hirsutism (2).

In this case report, we discuss an incident involving a 65-year-old postmenopausal female who had accidentally discovered bilateral ovarian masses. After a total hysterectomy and bilateral salpingo-oophorectomy, the masses were initially misdiagnosed as ovarian fibroma by the pathologist. However, after a detailed discussion and analysis of characteristic MRI findings, the final diagnosis was revised to ovarian fibromatosis.

CASE REPORT

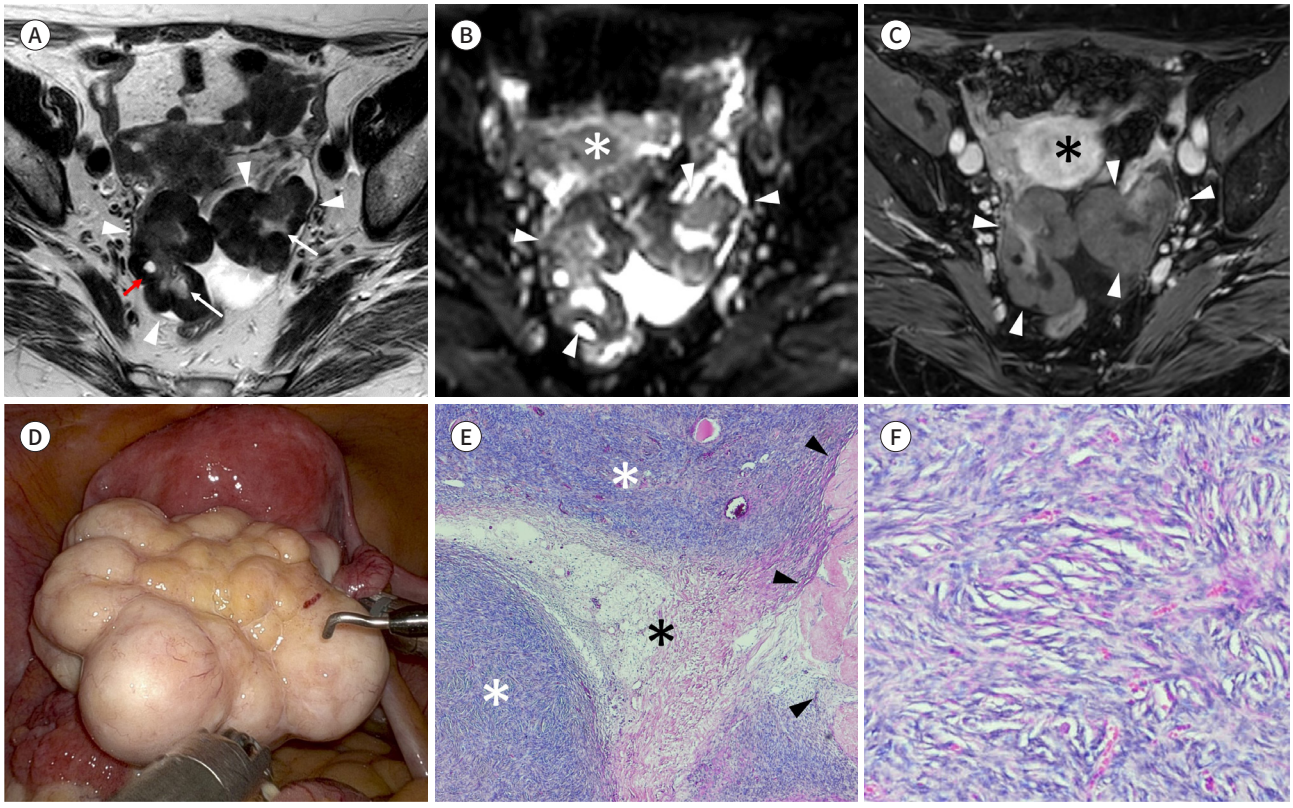
A 65-year-old postmenopausal female presented with bilateral ovarian masses, initially identified by transvaginal ultrasound (TVUS) at an outside hospital. Her vital signs were stable, and she reported no abdominal pain or vaginal bleeding. Physical examination revealed no palpable abdominal mass. Blood tests indicated elevated cancer antigen 125 (CA-125) levels at 50.90 U/mL. The patient's medical history included four vaginal deliveries, breast cancer 12 years ago, and tongue cancer 4 years ago.

TVUS performed at our hospital revealed bilaterally enlarged ovaries measuring 4.66 × 3.16 cm on the right and 6.85 × 3.30 cm on the left. These ovaries demonstrate a multilobulated hypoechoic thick surface (0.5 to 1.0 cm) and internally relatively higher echogenicity. Additionally, a hyperechoic nodular lesion was detected in the endometrial cavity, but an endometrial biopsy showed no significant findings, only some segmented endometrial tissue.

On pelvic MRI, the T2-weighted images (T2WI) revealed bilaterally enlarged ovaries, measuring 5.5 cm on the right and 4.5 cm on the left. These ovaries exhibited a thickened and multilobulated surface along the cortex (thickness ranging from 0.7 to 1.6 cm) with very low signal intensity compared to the myometrium (Fig. 1A). The spared internal stroma of the ovaries exhibited intermediate high signal intensity. A small unilocular cystic lesion (5 mm) in the right ovary, considered a normal follicle, and a small amount of ascitic fluid in the pelvic cavity were also noted. The T1WI showed homogeneous iso-signal intensity compared to the myometrium, with no significant diffusion restriction on the diffusion-weighted images (Fig. 1B). The fat-suppressed gadolinium-enhanced T1WI (contrast-enhanced T1WI) exhibited contrast enhancement of the ovarian cortex similar to the surrounding skeletal muscle but weaker than the myometrium (Fig. 1C). No enlarged pelvic lymph nodes were observed. These findings strongly suggested bilateral ovarian fibromatosis. However, the possibility of bilateral ovarian fibroma or Krukenberg tumor could not be completely ruled out due to the patient's medical history of breast cancer.

Two days after the MRI scan, the patient underwent a laparoscopic total hysterectomy and bilateral salpingo-oophorectomy. During the procedure, the ovaries were observed to be enlarged, displaying a tan-colored, multilobulated contour (Fig. 1D). The pathological examination revealed extensive nodular fibrosis throughout the ovarian cortex, accompanied by ovarian edema and an entrapped and atrophied luteal cyst, indicative of normally preserved ovarian stroma (Fig. 1E). High-power microscopic examination revealed the proliferation of spindle cells with collagen without a discrete mass formation (Fig. 1F). Initially, even a pathologist with over 30 years of experience did not recognize the disease entity of ovarian fi-

Fig. 1. A 65-year-old female with bilateral ovarian fibromatosis.
A. Axial T2-weighted MRI image of the pelvis shows enlarged ovaries (right ovary: 5.5 cm, left ovary: 4.5 cm). The image reveals very low signal intensity along the multilobulated cortex of both ovaries (arrowheads, 'black garland sign'), intermediate signal intensity in the internal stroma (white arrows), and a normal unilocular follicle in the right ovary (red arrow). A small amount of ascitic fluid is noted in the pelvic cavity.
B. Axial diffusion-weighted image with a b-value of 400 sec/mm^2 shows iso- to low signal intensity along the ovarian cortex (arrowheads) compared with the myometrium (*), indicating no significant diffusion restriction.
C. Axial fat-suppressed gadolinium-enhanced T1-weighted image shows weaker contrast enhancement (arrowheads) in the cortex of both ovaries compared with the myometrium of the uterus (*), similar to the surrounding skeletal muscle. No enlarged pelvic lymph nodes observed.
D. A photograph taken during laparoscopic surgery shows a tan-colored, multilobulated, and enlarged left ovary.
E. Histologic specimen (H&E stain, ×40) from both ovaries shows multifocal nodular fibrosis (white asterisks) with surrounding ovarian edema (black asterisk) and an entrapped and atrophied luteal cyst (arrowheads).
F. High power-view of the specimen (H&E stain, ×100) shows the fascicular growth of proliferated spindle cells with collagen.
H&E = hematoxylin and eosin



bromatosis due to its rarity, and the proliferation of spindle cells in specimens led to a misdiagnosis of ovarian fibroma. However, after a comprehensive review and re-discussion with the pathologist, considering the characteristic MRI findings, the diagnosis was revised to ovarian fibromatosis. Following surgery, the patient was regularly monitored in the outpatient department without significant symptoms.

This case report was ethically approved and the need for informed consent was waived by the Institutional Review Board of Inha University Hospital (IRB No. 2023-11-054-000).

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DISCUSSION

Ovarian fibromatosis is histologically defined by the proliferation of spindle cells that produce collagen in the cortical layer of the ovarian surface. This results in collagen accumulation in the cortex, leading to fibrosis and ovarian enlargement (3). Radiologically, ovarian fibromatosis presents a unique multilobulated shape and very low signal intensity on T2WI along the thickened ovarian cortex, known as the 'black garland sign' due to its fibrous composition. Moreover, its fibrous composition causes weak contrast enhancement in contrast-enhanced T1WI compared to the myometrium (2). As a non-neoplastic entity, ovarian fibromatosis does not exhibit an invasive or destructive growth into the surrounding follicular structure, thereby preserving the normal ovarian stroma or follicles (4). This characteristic is critical in differentiating ovarian fibromatosis from other fibrous ovarian neoplasms.

In this case, radiological findings such as bilateral ovarian enlargement, the distinctive 'black garland sign', and the preservation of ovarian follicles and stroma strongly suggested ovarian fibromatosis. However, this case was atypical. Most reported cases of ovarian fibromatosis occur in premenopausal women aged between 13 and 39 (2), with the oldest documented case in a 49-year-old female (5), based on our research. The pathogenesis of ovarian fibromatosis is not fully understood, but it is thought to be a 'burned out' stage resulting from reactive fibroblastic proliferation initially triggered by ovarian edema at a young age (6). It is conceivable that in this case, the condition developed in early life, remained asymptomatic, and was only detected at the post-menopause state.

In the differential diagnosis of fibrous ovarian neoplasms, it is crucial to distinguish ovarian fibromatosis from ovarian fibroma. Ovarian fibroma is more commonly found in middle-aged women, typically between 40 and 50 years. Similar to ovarian fibromatosis, they exhibit very low signal intensity on T2WI due to their fibrous composition and show weaker contrast enhancement on contrast-enhanced T1WI compared to the myometrium. However, ovarian fibroma often presents in various oval or round shapes, in addition to a multilobulated shape, and the follicular structures are usually lost, or if preserved, mainly located peripherally around the ovarian surface layer. It is also more commonly found unilaterally (7, 8).

Given the bilaterality of ovarian neoplasms and the patient's medical history of breast cancer, the possibility of Krukenberg tumor cannot be completely ruled out. Krukenberg tumor may exhibit low signal intensity on T2WI due to dense stromal reaction. However, it is typically oval in shape and exhibits strong contrast enhancement (7, 9).

The association between ovarian fibromatosis and CA-125 has not yet been established. In the three reported cases of ovarian fibromatosis where CA-125 levels were presented, they were within the normal range (4, 6, 7). Some studies suggest elevated CA-125 can arise from non-neoplastic cells under conditions such as ascites, increased abdominal pressure from a large mass, or physical irritation of the peritoneum (10). It may provide some explanation for the cause of the patient's elevated CA-125. However, further research is needed to clarify the specific relationship between ovarian fibromatosis and CA-125 levels.

The case is important for several reasons. To our knowledge, there have been no prior radiologic reports of ovarian fibromatosis in Korea. Additionally, among the limited cases reported worldwide, none have documented ovarian fibromatosis in postmenopausal women.

Importantly, the characteristic MRI findings were instrumental in correcting the initial pathological misdiagnosis. These insights are invaluable for accurately diagnosing ovarian fibromatosis, potentially preventing unnecessary surgical interventions.

Author Contributions

Conceptualization, all authors; supervision, K.M.Y.; visualization, H.S.C., K.J.M., H.S.O.; writing—original draft, H.S.C.; and writing—review & editing, H.S.C., K.M.Y.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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폐경 후 여성의 양측성 난소 섬유종증: MRI 소견과 감별진단 중점의 증례 보고

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난소 섬유종증(ovarian fibromatosis)은 콜라젠을 형성하는 난소의 버팀질의 증식으로 인해 난소의 비대를 유발하는 희귀한 비신생물성 상태로 25세 전후의 여성에게서 발생한다. 난소 섬유종증의 주요한 진단적 소견은 MRI의 T2 강조영상에서 난소의 표층을 따라 특징적인 다소엽 형태의 매우 저신호 강도를 보이는 '검은 화환 징후'(black garland sign)이다. 내부에 정상 난소 버팀질과 난포가 보존되는 소견 역시 특징적이다. 저자들은 65세의 폐경 후 여성에게서 병리적으로 난소 섬유종(ovarian fibroma)으로 오인되었으나 특징적인 MRI 소견에 기반하여 난소 섬유종증으로 진단이 수정되었던 증례를 보고한다. 난소 섬유종증과 관련된 감별진단에 관한 논의와 병리적 소견을 함께 기술하였다.

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