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Case Report

Deceiving Case of Spindle Cell Tumor Initially Presented as Endometriosis

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Abstract

Endometriotic cyst is common in radiology practice. Moreover, we tend to overlook as soon we find hemorrhagic cyst on ultrasound or T2 shading on magnetic resonance imaging (MRI). However, although the risk of endometriosis converting into malignancy is very rare, radiologist should keep an open eye to raise a possibility of neoplastic transformation. In this report, we describe a case of a 31-year-old female who presented with lower abdominal pain, abnormal uterine bleeding, and dysmenorrhea, initially diagnosed as endometriotic cyst, which turned out to be a malignancy on MRI.

Key words: Endometriosis, Magnetic resonance imaging, Spindle cell tumor, USG

BACKGROUND

Endometriosis rarely may show a histologic finding of malignancies for which imaging plays an important role for initial diagnosis. The risk of endometriosis converting into malignancy is very rare.[1] The most common variants are endometrioid carcinoma and clear-cell adenocarcinoma. The estimated lifetime risk for surgery for a suspected ovarian neoplasm is 5–10%. For an adnexal mass, the purpose of the evaluation is to diagnose it early, which is crucial to optimize patient care. We managed a challenging case of a premenopausal woman with an initial diagnosis of endometriotic cyst, which was thought to be benign showed features of malignancy in magnetic resonance imaging (MRI) further confirmed on histopathology testing.[2-4] Clinicians should be aware of the utility of MRI especially DWI sequence for initially distinguishing a malignant from benign ovarian pathology which is crucial for patient management.

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CASE PRESENTATION

Thirty-one years old, P2L2, presented to gynecology OPD with complaints of lower abdominal pain, abnormal uterine bleeding, dysmenorrhea, and generalized weakness for 4 years. The patient had multiple surgeries in the past: Open myomectomy in 2013, laparoscopic endometriotic drainage in 2016, lower segment caesarean section (LSCS) with myomectomy in 2017, cholecystectomy with endometriotic cystectomy in 2018, and LSCS in 2019. She denied a family history of any gynecological malignancy. Laboratory investigation for CA125, beta-human chorionic gonadotrophin, and alpha-fetoprotein was normal. The patient was referred to radiodiagnosis outpatient door for USG abdomen and pelvis which showed intramural fibroid and bulky uterus with bilateral complex adnexal cysts [Figure 1a]. The patient was advised MRI abdomen and pelvis which revealed a fairly defined solid lesion measuring approximately 4.8 (AP) × 6.2 (TR) × 5.5 (CC) noted in the right adnexa superior to the fundus of the uterus with indentation of uterine fundus; however, the bilateral ovaries were visualized separately from the lesion. It showed T1 hypointensity and T2/short tau inversion recovery (STIR) hyperintensity whereas the cystic part shows T1/T2/STIR hyperintensity [Figure 1b and c] and diffusion restriction on DWI sequence [Figure 1d and e]. Intense post-contrast enhancement was observed. There were three fairly defined T1 hyperintense lesions showing diffusion restriction on DWI sequence and heterogeneous

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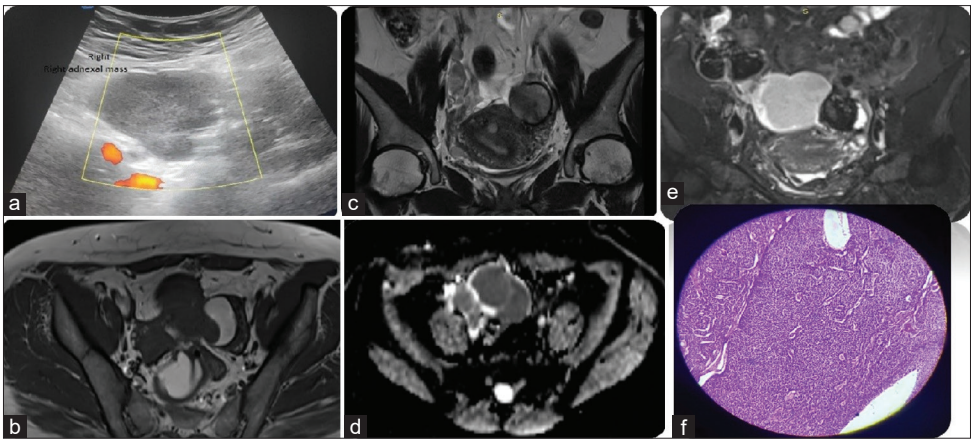

Figure 1: (a) Transabdominal ultrasound, transverse plane illustrating right adnexal lobulated solid hypoechoic mass; (b) MRI, T2 coronal sequence illustrating endometrial cyst showing blooming and shading; (c) MRI STIR image illustrating intensely hyperintense solid right adnexal mass; (d) MRI T1 sequence axial plane illustrating hyperintense thick walled cyst suggestive of hemorrhage; (e) MRI ADC sequence axial plane illustrating hypointense adnexal mass showing diffusion restriction; (f) Histopathology image of spindle cell tumor, granulosa cell type, illustrating call exner bodies

post contrast enhancement noted in the subcutaneous and intramuscular plane of the right lower abdominal wall as well as left rectus abdominis muscle possibly metastatic deposits. There was a well-defined T1 hyperintense, T2/STIR mildly hyperintense cystic lesion noted in the pelvis with shading artifact on GRE pelvic cavity measuring approximately 4(TR) × 3.4(AP) × 3.3(CC) cm likely suggestive of an endometriotic cyst. Wall was diffusely thickened and showed diffusion restriction. Biopsy of the right adnexal mass was planned and revealed suggestive of sex-cord stromal tumor-granulosa cell type [Figure 1f].

The patient was referred to a tertiary care center for further management; she was evaluated thoroughly by computed tomography of the pelvis as well as a biopsy. Computed tomography scan showed bilateral bulky ovaries with few abdominal and pelvic peritoneal deposits along the right paracolic gutter and inframesocolic omentum along with small bowel loops. Solid masses were observed anterior and posterior to the uterus, largest measuring 6.5 × 3.5 cm along anterior location. Repeat right adnexal biopsy was done which revealed a low-grade tumor which composed of spindle cells with minimal cytological atypia.[2] On immunohistochemistry cells were positive for desmin, CD10 (patchy), negative for SMA. Reticulin shows pericellular staining. Mib 1 labeling index is approximately <5%. As the patient was young with raised inhibin, the possibility of a sex-cord stromal tumor was considered.

DISCUSSION

Endometriosis is defined as the presence of endometrial glands and stroma outside the uterine cavity. The main symptoms are pain and infertility. Initially, thought as a benign disease, now considered that endometriosis especially of ovarian origin can have the neoplastic potential for becoming malignancy, although a rare phenomenon.[1] Endometriosis-associated carcinoma is commonly unilateral. The literature showed that 0.3–1.6% of endometriosis may transform into cancer. The most common endometriosis-associated histological types are endometrioid adenocarcinoma and clear-cell adenocarcinoma.[3] Ovarian lesions comprising of spindle cells comprise of different groups; most are neoplastic conditions but several non-neoplastic conditions are also composed of spindle cells.[2] There are two broad classes of spindle cell tumors; one includes the fibromatous group and another sex-cord stromal group which includes granulosa cell, thecoma, Sertoli-Leydig cell tumor.[4] Granulosa cell tumor is the most common sex-cord stromal tumor. Adult granulosa cell tumors are more common than the juvenile type and account for 95% of all granulosa cell tumors, as in our case. The radiologic appearance of sex cord-stromal tumors varies from a small solid mass to a large multi-cystic mass. Up until 2009, no gene expression profile had been identified as a specific marker of granulosa cell tumor. It is found that 97% of granulosa cell tumors had a mutant FOXL2 gene.[4]

This case illustrates the difficulty of establishing a diagnosis of spindle cell tumor and granulosa cell type, arising from an endometrioma.

CONCLUSION

Ovarian cancer is a leading cause of death from a gynecological malignancy. Spindle cell tumors comprise a small portion of ovarian cancer, due to potential long-term implications, diagnosis is critical. Radiologists should have a clear outlook

in evaluating endometriosis, especially for solid cystic components which represent neoplastic transformation. MRI must be on the checklist of such complicated endometriotic cysts. MRI must be on the checklist when evaluating such complicated endometriotic cysts. Biopsy gives the final answer about the histological diagnosis.

TEACHING POINT

Complex solid cystic masses on ultrasound should be evaluated in-depth. MRI is a preferred test with the minimum required sequences including diffusion sequence, which can be a game-changer, due to its inherent property to detect highly cellular masses. In endometriosis, solid-cystic components/ solid masses in relation to an endometriotic cyst can prove helpful on MRI as an indicator of malignant transformation.

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