Case Report

J Korean Soc Radiol 2023;84(3):692-697
https://doi.org/10.3348/jksr.2022.0118
eISSN 2951-0805

JOURNAL of THE KOREAN SOCIETY of RADIOLOGY

Clear Cell Borderline Tumor of the Ovary: A Case Report

난소에 발생한 경계성 투명세포종: 증례 보고

Ha Jung Kim, MD[1], Kyeong Ah Kim, MD[1]*, Yikyeong Chun, MD[2], Yang Shin Park, MD[1], Mi Jin Song, MD[1], Jae Woong Choi, MD[1]

Departments of [1]Radiology and [2]Pathology, Korea University Guro Hospital, Korea University College of Medicine, Seoul, Korea

ORCID iDs
Ha Jung Kim http://orcid.org/0000-0002-8279-7361
Kyeong Ah Kim http://orcid.org/0000-0003-4451-9325
Yikyeong Chun http://orcid.org/0000-0002-4235-9690
Yang Shin Park http://orcid.org/0000-0002-1918-2276
Mi Jin Song http://orcid.org/0000-0003-4853-3167
Jae Woong Choi http://orcid.org/0000-0001-5449-1560

Received August 19, 2022
Revised September 12, 2022
Accepted October 2, 2022

***Corresponding author**
Kyeong Ah Kim, MD
Department of Radiology,
Korea University Guro Hospital,
Korea University
College of Medicine,
148 Gurodong-ro, Guro-gu,
Seoul 08308, Korea.

Tel 82-2-2626-1338
Fax 82-2-863-9282
E-mail kahkim@korea.ac.kr

Clear cell borderline ovarian tumor (CCBOT) is quite rare, and only a few cases of CCBOT have been reported. Unlike most borderline ovarian tumors, CCBOTs appear solid because they are almost always pathologically adenofibromatous. Herein, we report the MRI findings of a CCBOT discovered in a 22-year-old female.

Index terms Neoplasm; Ovary; Magnetic Resonance Imaging

INTRODUCTION

Clear cell tumors are a subtype of epithelial ovarian tumors. Most clear cell ovarian tumors are malignant, while benign and borderline clear cell tumors are rare. Clear cell borderline ovarian tumors (CCBOTs) account for less than 1% of all borderline ovarian tumors (1). CCBOT is defined as "a tumor composed of glands or cysts lined by bland cuboidal to flattened cells with clear or eosinophilic cytoplasm and atypia, but without stromal invasion" (2). Due to the rarity of CCBOTs, few cases have been reported in the literature (1, 3, 4). Herein, we present a case of surgically confirmed CCBOT, and discuss the MRI findings and clinicopathological features.

CASE REPORT

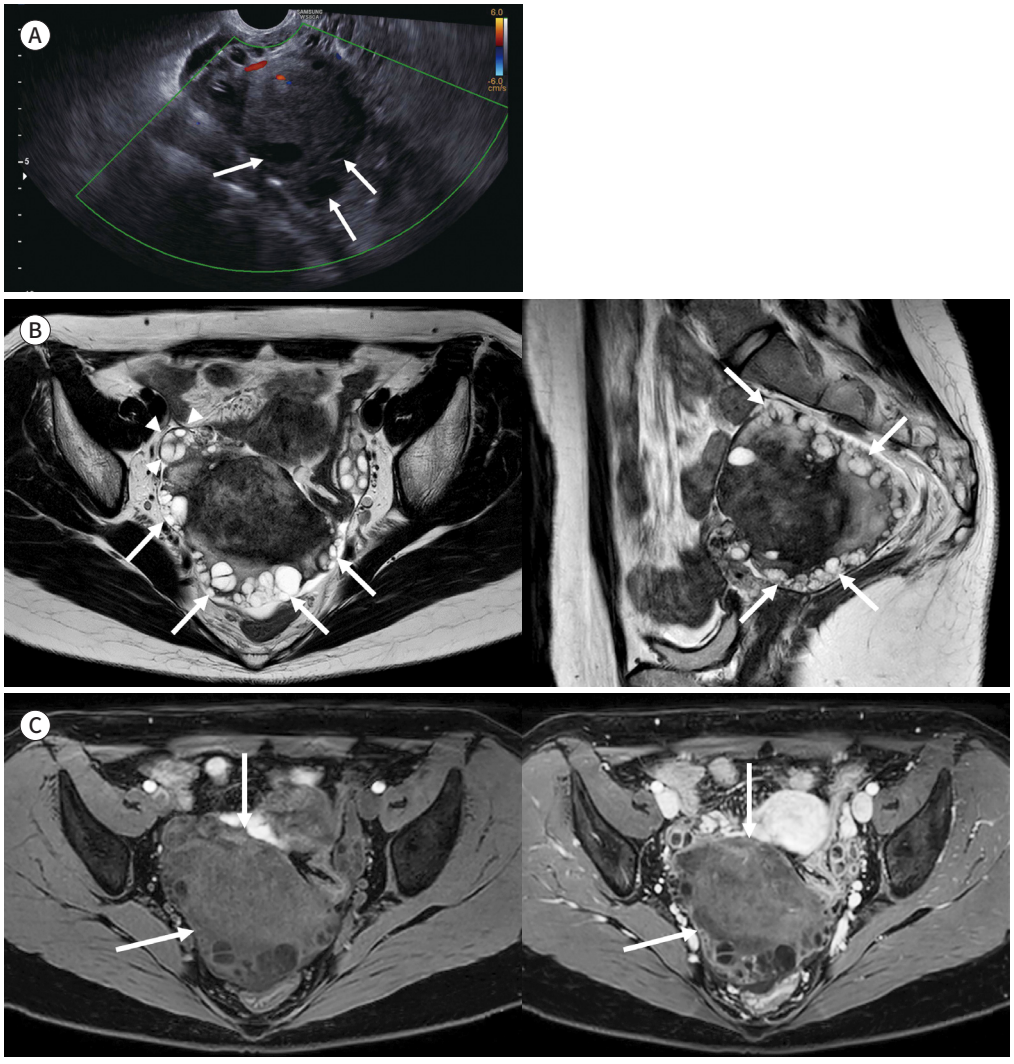
A 22-year-old female visited the gynecology department with a history of menstrual irregu-

JOURNAL of
THE KOREAN SOCIETY of
RADIOLOGY

larity for 2 months. The patient exhibited no additional symptoms and had no recorded medical history. Physical examination revealed a soft abdomen with no palpable masses. Initial tumor markers, such as cancer antigen (CA) 125 and CA 19-9 were within the normal range.

Transabdominal pelvic ultrasonography revealed a 10-cm mass in the pelvic cavity. The mass was mainly echogenic with multiple small peripheral cystic portions (Fig. 1A). Pelvic MRI revealed a solid mass with small peripheral cystic areas in the right pelvic cavity. The mass abutted the right ovary anteriorly; the uterus and left ovary appeared to be normal. On T2-weighted image (T2WI), the tumor showed mostly low signal intensity, similar to that of muscles, with

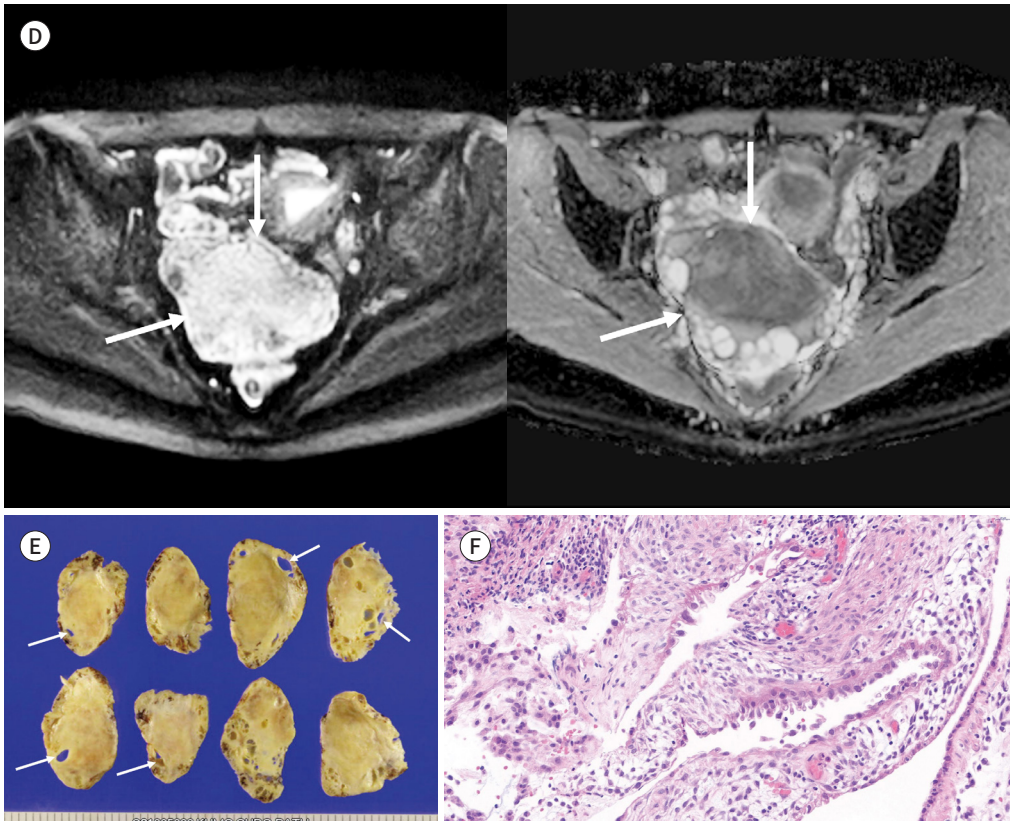
Fig. 1. A 22-year-old female with a clear cell borderline tumor of the ovary.
A. Transabdominal pelvic ultrasonography shows approximately 10-cm sized heterogeneous echoic mass with peripheral anechoic portions (arrows), without increased vascularity.
B. Axial and sagittal T2-weighted image demonstrate a mass with very low signal intensity and peripheral multiple cysts (arrows). The right ovary (arrowheads), which appears normal, is closely attached to the anterior of the mass.
C. Axial T1FS imaging (left) and axial CE T1FS (right) demonstrate mild contrast enhancement of the mass (arrows).
CE = contrast-enhanced, T1FS = T1-weighted fat suppression



https://doi.org/10.3348/jksr.2022.0118

JOURNAL of
THE KOREAN SOCIETY of
RADIOLOGY

Fig. 1. A 22-year-old female with a clear cell borderline tumor of the ovary.
D. High b-value (b = 1000) DWI (left), demonstrates mild increased SI (arrows) with mild decreased SI (arrows) in the ADC map (right).
E. Gross specimen shows a well-circumscribed, whitish fibrotic mass with peripheral multilocular cysts (arrows).
F. Microscopic image of the resected tumor with hematoxylin and eosin stains (× 400) shows glands lined by cuboidal cells with a clear cytoplasm, and atypia without stromal invasion.
ADC = apparent diffusion coefficient, DWI = diffusion-weighted image, SI = signal intensity



multiple variably sized cysts at the periphery (Fig. 1B). The tumor showed iso-signal intensity on the T1-weighted image (T1WI) compared to that of the muscle. The mass displayed mild enhancement on contrast-enhanced T1WI (Fig. 1C). It showed a mildly high signal intensity on diffusion-weighted imaging and low signal intensity on the apparent diffusion coefficient map (Fig. 1D). Based on the MRI findings and considering the hypointense nature of the mass on T2WI, the preoperative differential diagnoses included fibrotic ovarian tumors, such as fibroma, fibrothecoma, and Brenner tumor with cystic changes.

During surgery, the mass appeared to be abutting the right ovary. Subsequently, the patient underwent resection of the right pelvic mass, without requiring oophorectomy or pelvic lymphadenectomy. The cut section showed a whitish and fibrotic tumor, with peripheral multilocular cysts (Fig. 1E). Microscopically, the tumor contained glands lined by cuboidal cells with atypia and clear cytoplasm, without evidence of stromal invasion (Fig. 1F). The final pathological diagnosis was confirmed as CCBOT with an adenofibromatous background associated with endometriosis in the right ovary. Regular follow-up was performed after surgery, without chemotherapy or radiation therapy. No recurrence was observed in the post-operative

one year.

This study was approved by the Institutional Review Board of our institution (IRB No. 2022GR0330). Informed consent was waived due to the retrospective nature of the study.

DISCUSSION

Borderline ovarian tumors are noninvasive epithelial tumors, which present with higher levels of cytologic atypia and epithelial proliferation compared to benign tumors. Most borderline tumors of the ovary are serous and mucinous tumors (3). CCBOT is a rare subtype of borderline ovarian tumors. CCBOT is pathologically defined as "an adenofibromatous clear cell tumor with glandular crowding and low-grade nuclear atypia without stromal invasion" in the 2020 World Health Organization classification (1).

Majority of patients with CCBOT are postmenopausal. In the literature, the mean age of patients was reported as 61 years (range, 30–86 years). Only four cases of CCBOT in premenopausal female have been reported so far (1). In our case, the patient was a premenopausal 22-year-old female, making her the youngest patient with CCBOT reported, to our knowledge. Patients with CCBOT have a nonspecific clinical presentation; most are asymptomatic, although potential symptoms include abdominal pain, distention, and abnormal uterine bleeding (4). In our case, the patient's only complaint was an irregular menstrual cycle.

Due to its rarity, the etiology of CCBOT is not yet fully understood. A previous report hypothesized that two possible pathways exist for all types of clear cell ovarian tumors derived from endometriosis. In the first possible pathway, cystic clear cell carcinoma evolves from an endometriotic cyst with epithelial atypia. Second, clear cell adenofibroma develops from a fibromatous reaction in endometriotic cysts, which then progresses to CCBOT and ultimately to adenofibromatous clear cell carcinoma (2). This hypothesis was consistent with our case, as histopathologic examination revealed an endometriotic background associated with the tumor.

Typical MRI findings for most borderline ovarian tumors are purely or predominantly cystic, with papillary projections. This is because over 95% of all borderline ovarian tumors are serous or mucinous (5). In two published reports by Kleebkaow et al. (3) and Lee et al. (6), these tumors appeared as solid or mixed solid and cystic masses. This is consistent with the pathological finding of CCBOT, which is almost always adenofibromatous. In the present case, the tumor appeared as a solid mass with peripheral cysts. The solid portion of the tumor showed low signal intensity on T2WI, which may correlate to fibrotic nature of the tumor.

The differential diagnosis for CCBOT includes fibroma, fibrothecoma, and Brenner tumor. These neoplasms have solid components with low T2 signal intensity due to the fibrous stromal tissue (7). According to Kato et al. (8), ovarian fibromas are observed in cysts in approximately 53% of the cases. These intratumoral cysts are located at the periphery of the masses, and exophytic cyst formation may be observed in ovarian fibromas. In our case, the cystic portions of the mass were smaller in size, more numerous, and distributed circumferentially along the border of the tumor compared to that of fibromas. Brenner tumors are primarily solid tumors, but several small cysts are often observed in the solid components. The cystic portions are often multilocular, as Brenner tumors are frequently associated with mucinous cystic tumors. Amorphous calcification also occurs more frequently in Brenner tumors than in other

fibrous tumors (9).

In the present case, it was difficult to distinguish the origin of the mass in the right pelvic cavity due to the detection of a normal-looking crescent-shaped ovary along the anterior border of the tumor. Oh et al. (10) previously reported that normal-appearing ipsilateral ovaries were noted in 50% of the ovarian fibromas. The shape of the ovary may be preserved in a fibrotic mass growing outward from the ovary. Thus, we believed that the tumor was not an extraovarian mass, but an exophytic ovarian mass.

All reported cases of CCBOT, including ours, represent stage I disease and have a relatively good prognosis (1). Most CCBOTs are confined to the ovaries at the time of identification (1). Thus, adjuvant therapy and complete surgical staging can be omitted in most CCBOT cases, and fertility-sparing surgery can be a reasonable option in young patients (6). In our case, the patient did not undergo ipsilateral oophorectomy or additional treatment after surgery, and was cancer-free in the post-operative one year.

Here, we present a rare case of CCBOT in a 22-year-old female. The tumor was a fibrotic ovarian mass, exhibiting T2 low signal intensity with multiple peripherally located small cystic areas on MRI. Although CCBOT is very rare, it can be considered as a differential diagnosis upon discovery of an ovarian fibrotic mass accompanying a cystic portion.

Author Contributions

Conceptualization, K.H.J., K.K.A.; data curation, K.H.J., K.K.A., C.Y.; investigation, K.H.J., K.K.A.; writing—original draft, K.H.J., K.K.A.; and writing—review & editing, K.H.J., K.K.A., P.Y.S., S.M.J., C.J.W.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

Funding

None

REFERENCES

1. Ricotta G, Maulard A, Candiani M, Genestie C, Pautier P, Leary A, et al. Clear cell borderline ovarian tumor: clinical characteristics, prognosis, and management. *Ann Surg Oncol* 2022;29:1165-1170
2. Zhao C, Wu LS, Barner R. Pathogenesis of ovarian clear cell adenofibroma, atypical proliferative (borderline) tumor, and carcinoma: clinicopathologic features of tumors with endometriosis or adenofibromatous components support two related pathways of tumor development. *J Cancer* 2011;2:94-106
3. Kleebkaow P, Aue-Aungkul A, Temtanakitpaisan A, Kietpeerakool C. Borderline clear cell adenofibroma of the ovary. *Case Rep Pathol* 2017;2017:3860107
4. Vasenwala SM, Ansari HA, Haider N, Rehman S, Khan T. Atypical proliferating clear cell adenofibroma of the ovary: a case report. *Middle East J Cancer* 2013;4:87-90
5. Alves AS, Félix A, Cunha TM. Clues to the diagnosis of borderline ovarian tumours: an imaging guide. *Eur J Radiol* 2021;143:109904
6. Lee HJ, Kim TJ, Kim JN, Seo EJ, Lee KY. Borderline clear cell adenofibromatous tumors of the ovary: two case reports. *Korean J Pathol* 2007;41:420-423
7. Foti PV, Attinà G, Spadola S, Caltabiano R, Farina R, Palmucci S, et al. MR imaging of ovarian masses: classification and differential diagnosis. *Insights Imaging* 2016;7:21-41
8. Kato H, Kanematsu M, Ono H, Yano R, Furui T, Morishige K, et al. Ovarian fibromas: MR imaging findings with emphasis on intratumoral cyst formation. *Eur J Radiol* 2013;82:e417-e421
9. Montoriol PF, Hordonneau C, Boudinaud C, Molnar I, Abrial C, Kossai M. Benign Brenner tumour of the ovary: CT and MRI features. *Clin Radiol* 2021;76:593-598

10. Oh SN, Rha SE, Byun JY, Lee YJ, Jung SE, Jung CK, et al. MRI features of ovarian fibromas: emphasis on their relationship to the ovary. *Clin Radiol* 2008;63:529-535

난소에 발생한 경계성 투명세포종: 증례 보고

김하정[1] · 김경아[1*] · 전이경[2] · 박양신[1] · 송미진[1] · 최재웅[1]

난소의 경계성 투명 세포 종양은 매우 드문 종양으로 현재까지 몇 개의 증례만 보고되었다. 일반적인 경계성 종양과 달리 경계성 투명 세포 종양은 고형의 종괴로 보이는데 병리학적으로 거의 대부분 선섬유성이기 때문이다. 저자들은 22세 여성의 난소에서 생긴 경계성 투명 세포 종양의 자기공명영상 소견에 대하여 보고하고자 한다.

고려대학교 의과대학 고려대학교 구로병원 [1]영상의학과, [2]병리과