

Received: 2017.03.22  
Accepted: 2017.05.31  
Published: 2017.08.24

# Spontaneously Ruptured Paraovarian Tumor of Borderline Malignancy with Extremely Elevated Serum Carbohydrate Antigen 125 (CA125) Levels: A Comparison of the Imaging and Pathological Features

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Statistical Analysis C  
Data Interpretation D  
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**Conflict of interest:** None declared

**Patient:** Female, 22  
**Final Diagnosis:** Paraovarian tumor of borderline malignancy  
**Symptoms:** Hypochondriac pain  
**Medication:** —  
**Clinical Procedure:** —  
**Specialty:** Radiology





**Objective:** Rare co-existence of disease or pathology  
**Background:** Paraovarian cysts are common and are generally benign; however, they are frequently misdiagnosed as being of ovarian origin. Conversely, paraovarian tumors of borderline malignancy are extremely rare. Especially, no cases of spontaneous rupture have been reported, and all previous reports had normal serum carbohydrate antigen (CA) 125 level. As for imaging findings, the presence of papillary projections in the lumen of paraovarian tumors does not always indicate malignancy or benignancy, which makes the preoperative distinction difficult.

**Case Report:** We report a case involving a 22-year-old Asian woman with a spontaneously ruptured paraovarian tumor of borderline malignancy. The serum CA125 concentration was extremely elevated, at 28,831 U/mL. Computed tomography (CT) and magnetic resonance imaging (MRI) demonstrated a collapsed unilocular cystic lesion with multiple papillary projections. On MRI, the papillary projections showed two different patterns, which corresponded to the pathological findings.

**Conclusions:** Ruptured paraovarian tumors of borderline malignancy may show extremely high serum CA125 values. Furthermore, specific MRI findings may be useful in evaluating the malignancy of paraovarian tumors.

**MeSH Keywords:** CA-125 Antigen • Magnetic Resonance Imaging • Parovarian Cyst • Rupture, Spontaneous

**Full-text PDF:** <https://www.amjcaserep.com/abstract/index/idArt/904462>

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## Background

Paraovarian cysts are common, accounting for 5–20% of all adnexal lesions, and most of them are benign [1]. On the contrary, paraovarian tumors of borderline malignancy are extremely rare, with only about 40 cases reported worldwide.

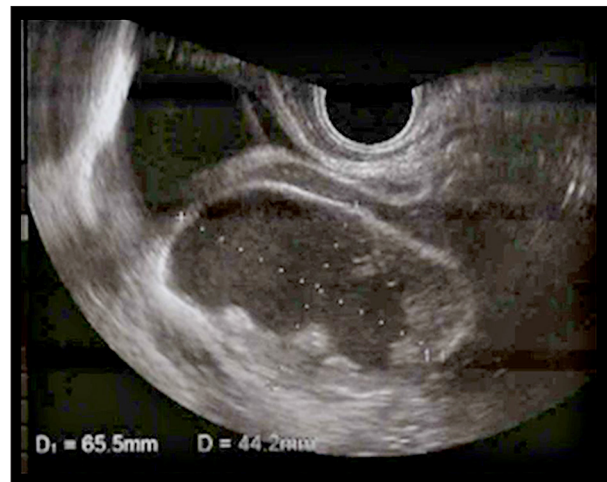
Paraovarian tumors are commonly asymptomatic and incidentally found as a unilocular cyst in the broad ligament of the uterus; thus, they are frequently misdiagnosed as other cystic pelvic masses such as ovarian cysts, lymphoceles, or peritoneal inclusion cysts [2]. Symptoms occasionally appear when they grow excessively or when hemorrhage, rupture, or torsion occurs [3]. Papillary projections are occasionally seen in the lumen of paraovarian tumors. In general, the presence of solid components and mural nodules in the cyst lumen of ovarian tumors indicates their malignant potential [4]. Conversely, 86% of paraovarian tumors with papillary projections in the lumen were found to be benign in one previous report [1]. Therefore, detecting the nodular components of a paraovarian tumor does not always help establish its malignancy, which makes the preoperative distinction of benign and malignant tumors difficult.

Here, we report an extremely rare case involving a 22-year-old woman with a ruptured paraovarian tumor of borderline malignancy. The tumor itself is extremely rare, and, to the best of our knowledge, none of the previously reported cases spontaneously ruptured or were accompanied by a marked elevation of the serum carbohydrate antigen (CA) 125 level. Further reports of the detailed computed tomography (CT) and magnetic resonance imaging (MRI) findings of these tumors are limited, with ultrasonography being the most commonly reported imaging modality. Hence, we describe and compare

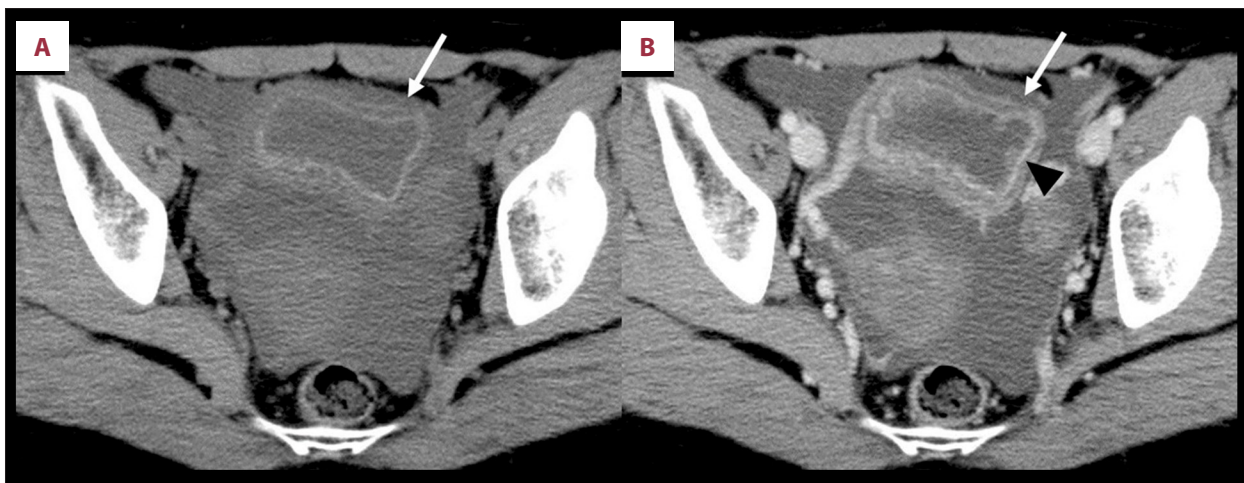
the imaging and pathological findings of a spontaneously ruptured paraovarian tumor of borderline malignancy, with the aim to provide a useful method for estimating the malignancy of this rare condition.

## Case Report

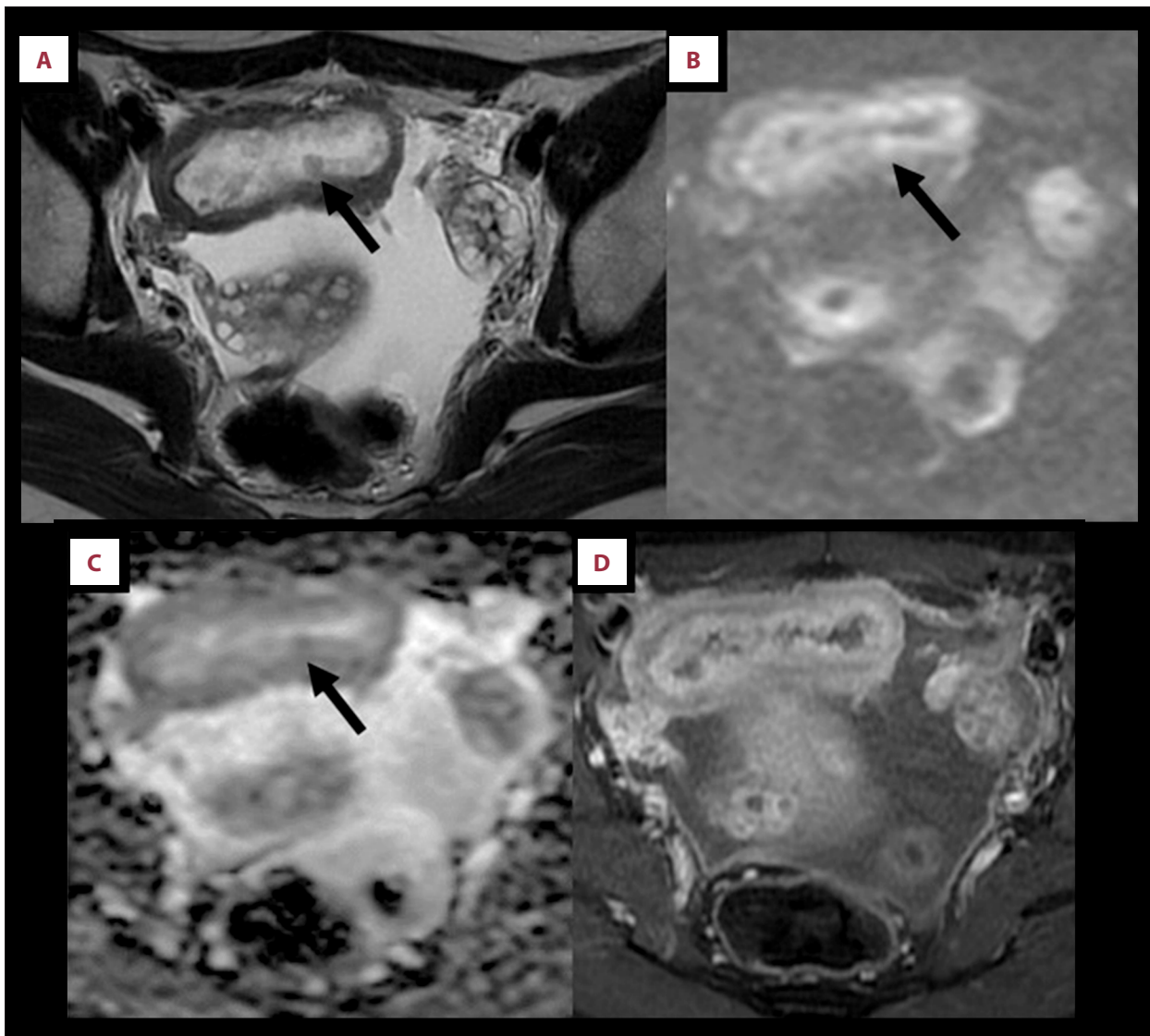
A 22-year-old Asian woman (gravida 0, para 0) presented with right hypochondriac pain.



**Figure 1.** Transvaginal ultrasound imaging showing a unilocular cystic structure that lacks contraction near the right ovary. The structure measures 65.5×44.2 mm, and papillary projections are detected in the lumen. Power Doppler revealed no blood flow within the papillary projections.



**Figure 2.** Computed tomography findings. A unilocular cystic lesion is seen in the median lower abdomen (A, B). The cystic lesion lacks contraction and consists of a thick wall (white arrows). The wall consists of three layers, with the middle layer containing linear calcification (A). The outer and inner layers show contrast enhancement, and papillary projections are seen in the lumen (black arrowhead) (B). Bilateral ovaries are seen apart from the cyst. Obvious ascites is seen in the pelvis.



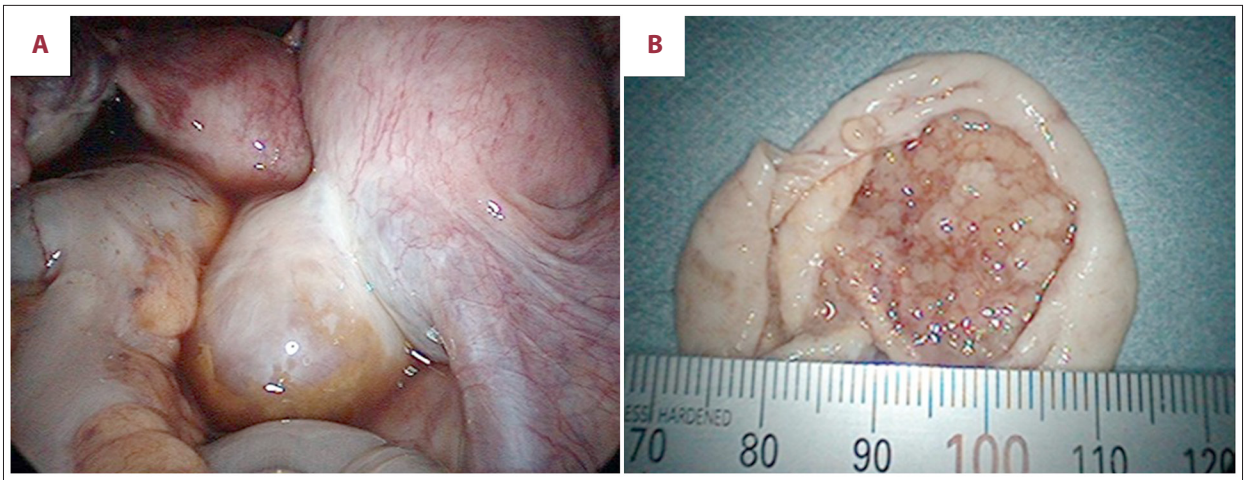
**Figure 3.** Magnetic resonance imaging findings. A collapsed unilocular cystic lesion is seen at the ventral side of the pelvis. T2-weighted imaging (T2WI) shows that the cystic lesion contacts the right fallopian tube (A). Multiple papillary projections are seen in the lumen of the cystic lesion, which is composed of a thick wall. Most of the papillary projections show high signals on T2WI, but some show slightly reduced signals. The papillary projections with low signals on T2WI show high signals on diffusion-weighted images (B) and a slight decline in signal intensity on apparent diffusion coefficient mapping (black arrows) (C). Contrast enhancement is seen in the wall and in the papillary projections (D). The bilateral ovaries are seen aside from the cystic lesion. Prominent ascites is seen in the pelvis, and the peritoneum is thick, with contrast enhancement observed.

Her medical history included epilepsy, which was treated with sodium valproate. Her family history was unremarkable. The patient denied any history of smoking, heavy alcohol consumption, or substance abuse.

Based on her symptoms, acute cholecystitis was suspected and an abdominal ultrasound examination was performed. The images showed normal gallbladder findings; however, a unilocular cystic structure was found in the median lower abdomen (Figure 1).

The values of the serum tumor markers were as follows: CA125, 28,831 U/mL (normal, <35 U/mL); CA19-9, 0.8 U/mL (normal, <37 U/mL); and carcinoembryonic antigen, 2.7 ng/mL (normal, <5.0 ng/mL).

Transvaginal ultrasound imaging showed a collapsed unilocular cystic structure near the right ovary with multiple papillary projections. Power Doppler revealed no blood flow within the papillary projections.



**Figure 4.** Gross findings of the laparoscopic resection specimen. The tumor was detected in the broad ligament of the uterus, adherent to the right fallopian tube. There is no connection between the tumor and the right ovary (A). Multiple papillary projections are seen inside the tumor (B).

CT images demonstrated a unilocular cystic lesion with a thick wall. The wall consisted of three layers, with the middle layer containing linear calcification. The outer and inner layers showed contrast enhancement, and papillary projections were seen in the lumen (Figure 2).

T2-weighted MRI (T2WI) of the cystic lesion demonstrated that most of the papillary projections showed high signal intensity, while some showed slightly reduced intensity. The papillary projections that showed low signal intensity showed high signal intensity on diffusion-weighted images and a slight decline in intensity on apparent diffusion coefficient mapping (Figure 3).

From the images, a ruptured paraovarian tumor of borderline malignancy or malignancy was suspected, and a laparoscopic resection of the tumor was performed. In the gross findings, the uterus and both ovaries were normal. The tumor was detected in the broad ligament of the uterus, adherent to the right fallopian tube (Figure 4A). Multiple papillary projections were seen in the lumen of the tumor (Figure 4B). There was no obvious exposure to the serous membrane (Figure 5A).

Microscopically, the wall of the tumor consisted of thick collagen fibers, and most of the papillary projections consisted of simple cuboidal and columnar epithelium with small oval nuclei. The stroma was edematous (Figure 5B, 5C). Multiple psammoma bodies were seen in the subepithelial lesion (Figure 5D). Some of the epithelium lining the papillary projections showed stratification and separated small clusters (Figure 5E). There was no obvious stromal invasion or microinvasion.

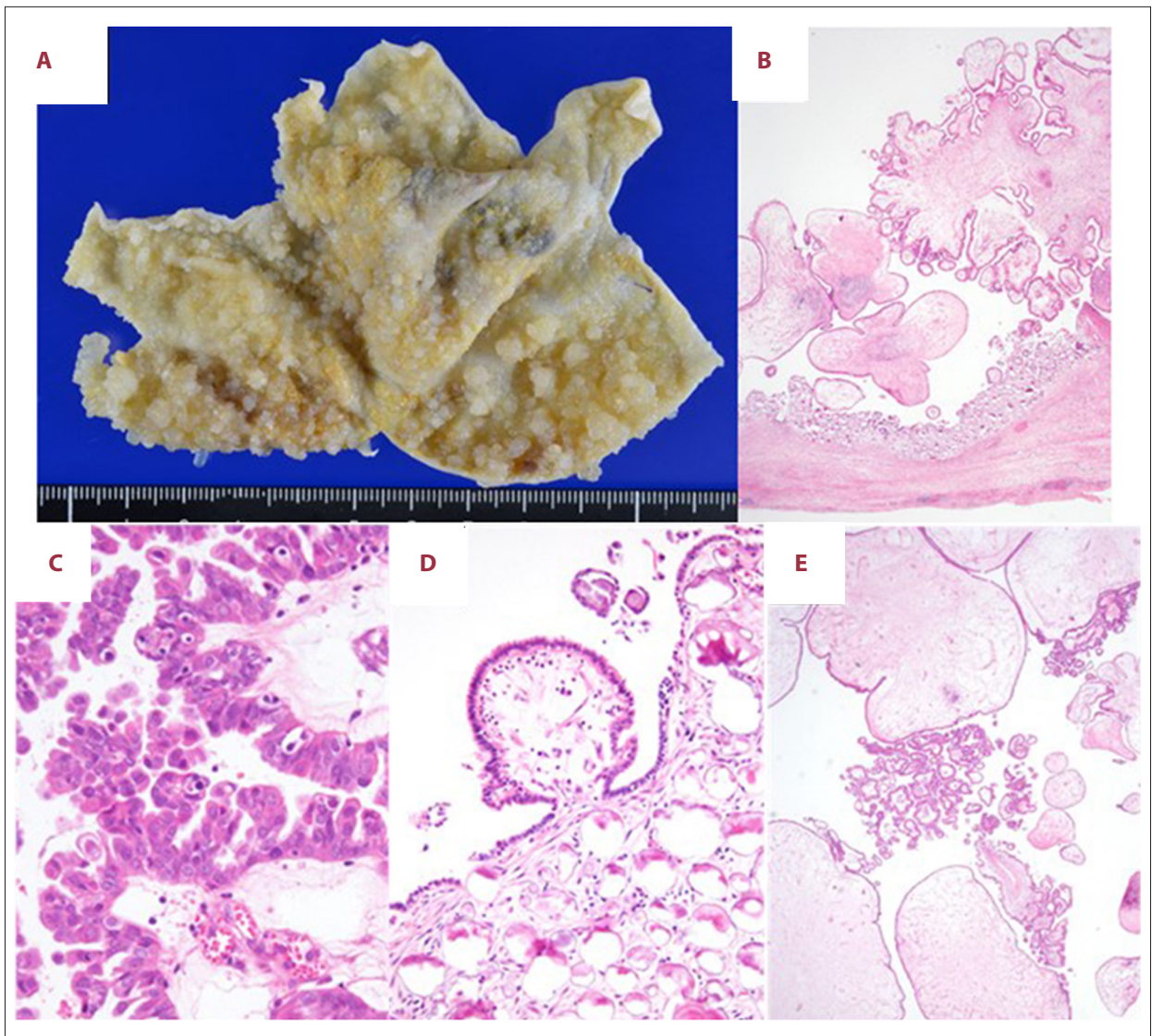
Based on the aforementioned findings, the tumor was diagnosed as a serous paraovarian tumor of borderline malignancy, Stage Ic (2) in the International Federation of Gynecology

and Obstetrics (FIGO) staging. As there is no clear guideline for the treatment of this tumor; the treatment strategy was decided on the basis of the ovarian tumor guidelines, while preserving fertility. Two months later, a right unilateral salpingo-oophorectomy, omentectomy, and peritoneal biopsy were performed. No malignancy was found in the biopsy specimen, and, at the latest follow-up (one year postoperatively), no recurrence was noted.

## Discussion

“Paraovarian cysts” is a general term for cystic lesions that occur between the hilus of the ovary and the ovarian fimbria within the mesosalpinx and broad ligament, and which originate from the Wolffian duct, Müllerian duct, or mesothelium [5]. Paraovarian cysts are common, accounting for 5–20% of all adnexal lesions, and most of them are benign [1]. On the contrary, paraovarian tumors of borderline malignancy are extremely rare, with only about 40 cases reported worldwide.

Paraovarian cysts are generally asymptomatic, although they are occasionally identified by clinical symptoms resulting from excessive growth, hemorrhage, rupture, or torsion [3]. They are usually anechoic unilocular cysts, with a thin and uniform wall observed on ultrasound images. However, the preoperative diagnosis of paraovarian cysts is often difficult, and they are commonly misdiagnosed as other cystic pelvic masses (e.g., ovarian cysts, lymphoceles, and peritoneal inclusion cysts). In one previous report, a preoperative diagnosis of paraovarian cysts by ultrasound images was only possible in 6.7% of cases, and 73.3% of cases were misdiagnosed as tumors of ovarian origin [2]. Another report showed that only 2% of patients with paraovarian cysts had malignant potential [6]. However, to



**Figure 5.** Macroscopically, papillary projections are seen in the lumen of the tumor. There is no obvious exposure to the serous membrane (A). Microscopically, the wall of the tumor consists of thick collagen fibers, and most of the papillary projections consist of simple cuboidal and columnar epithelium with small oval nuclei and edematous stroma (B, C). Multiple psammoma bodies are seen in the subepithelial lesion (D). Some of the epithelium lining the papillary projections shows stratification and separated small clusters (E). There is no obvious stromal invasion or microinvasion.

date, there has been no report describing the detailed imaging characteristics of borderline/malignant paraovarian tumors.

Savelli et al. compared the ultrasound images of 50 cases of paraovarian tumors, and found that 15 cases showed multiple papillary projections growing from the cyst wall into the lumen [1]. Of these 15 cases, 11 showed no blood vessels within the papillary projections, as determined by color Doppler. However, 13 of the 15 cases with multiple papillary projections were benign tumors, including eight cases of cystadenofibromas and five cases of cystadenomas. Only two cases were serous borderline tumors [1].

In general, the presence of solid components and mural nodules in the cyst lumen of ovarian tumors indicates their malignant potential [4]. On the contrary, about 86% of paraovarian tumors with papillary projections were found to be benign in Savelli study [1]. Therefore, detecting nodular components of a paraovarian tumor does not always indicate its malignancy, making the distinction of benign and malignant tumors difficult.

Furthermore, in one previous report [3], the CT and MRI showed unilocular cystic tumors adjacent to the ovary or fallopian tubes. The shape of the affected-side ovary was normal, without deformation or exclusion, which distinguished paraovarian

tumors from ovarian tumors. However, only a few reports have described the detailed CT and MRI findings of paraovarian tumors of borderline malignancy, with most only mentioning that papillary projections were seen in on imaging.

The present case highlights two important clinical issues: 1) MRI can be useful in the preoperative diagnosis of these tumors, and 2) ruptured paraovarian tumors of borderline malignancy can present extremely high serum CA125 levels.

First, in our case, the ultrasound images showed multiple papillary projections in the lumen, and the color Doppler showed no blood flow. The CT study demonstrated a thick stratified wall of the tumor. The extension of the wall was assumed to have decreased secondary to the rupture. The high attenuated layer coincided with the pathological layer of multiple psammoma bodies, which are frequently found in serous cystadenocarcinomas of the ovary and in malignant tumors; however, this is not a conclusive factor [7].

On MRI, the signals of the papillary projections of the tumor showed two different patterns. On T2WI, most papillary projections demonstrated high signals, and no diffusion restriction was seen. On the other hand, some of the papillary projections demonstrated low signals, and slight diffusion restriction was seen in these. Pathological analysis showed that most papillary projections were composed of a simple layer of epithelial cells and prominent edematous stromal tissue, which corresponded to the nodules with high T2WI signals. Stratification and separated small clusters of papillary projections, which indicate borderline malignancy potential, were only sporadically present. However, the low T2WI intensity and diffusion-restricted lesions may correspond to such epithelial-rich lesions. Therefore, the detection of papillary projections with low T2WI intensity and diffusion restriction may suggest the malignancy of paraovarian tumors.

Second, the serum CA125 value in the present case of spontaneously ruptured paraovarian tumor of borderline malignancy was extremely high (28,831 U/mL). In contrast, in previous reports, the values of the serum tumor markers, including CA125, were normal [3,8–11]. CA125 is a high molecular weight glycoprotein, which is produced by epithelial cells of the peritoneum, pleura, pericardium, and endometrium [12,13]. The production of CA125 is increased when these epithelial cells are stimulated, such as by inflammation or coelomic fluid collection.

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Therefore, the CA125 level is elevated in various benign and malignant conditions, including benign ovarian cysts, tuberculosis, and ovarian cancer [14–17]. However, in these conditions, the highest reported value of CA125, noted in a case of spontaneously ruptured ovarian endometrioma, remained in the four-digit range [18].

There is a possibility that the CA125 value in our patient was modified by the presence of peritonitis caused by the rupture. However, it is also possible that the tumor itself may have been the cause of the high value, as it belongs to the spectrum of serous tumors. The CA125 level showed a prominent decline after the operation, stabilizing around 15 U/mL.

There is currently no established standard treatment for borderline paraovarian tumors, largely owing to their rarity. However, as paraovarian tumors have the same embryological origin as epithelial ovarian tumors [5], the treatment is usually selected according to the guidelines for borderline or malignant ovarian tumors [19]. Fertility is preserved taking the patient's age, characteristics, and preferences into consideration. The 10-year survival rate is close to 100% in patients with FIGO Stage I malignant ovarian tumors [20], and, to the best of our knowledge, no recurrence has been reported in patients with borderline paraovarian tumors. However, since the tumor in our case had ruptured and obvious ascites was seen, we consider that careful follow-up was necessary. Postoperatively, long-term follow-up by vaginal ultrasound and tumor marker (CA125 and CA19-9) examinations is recommended [21].

## Conclusions

In conclusion, ruptured paraovarian tumors of borderline malignancy may show extremely high serum CA125 values. Furthermore, specific MRI findings can be useful in estimating the malignancy of paraovarian tumors.

## Acknowledgements

We would like to thank Editage ([www.editage.jp](http://www.editage.jp)) for English language editing.

## Conflicts of interests

None.

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