

Body Imaging

Imaging features of unusual lesions and complications associated with ovarian mature cystic teratoma

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ABSTRACT

Mature cystic teratoma (MCT) is a common neoplasm of the ovary that typically contains mature tissues of ectodermal, mesodermal, and endodermal origin. This tumor tends to affect younger women, its presentation ranges from pure cystic mass to complex solid cystic mass, and the detection of intratumoral fat component is the key diagnostic imaging feature. MCT can be associated with various complications and it demonstrates a wide spectrum of imaging findings. Associated complications include rupture, torsion, malignant transformation, and gliomatosis peritonei. MCT may also have unusual imaging features that can lead to misdiagnosis. These features may expand the differential diagnosis to include immature teratoma, monodermal teratoma, mature cystic teratoma with minimal or no fat, and collision tumor. The aim of this article was to highlight and describe the imaging features of unusual ovarian MCT lesions, and the complications associated with ovarian MCT.

1. Introduction

Mature cystic teratoma (MCT) is the most common germ cell tumor of the ovary, and it accounts for approximately 20% of all ovarian neoplasms [1]. It usually affects younger aged women (mean age: 30 years) [2]. At least two of three germ cell layers (ectoderm, mesoderm, and endoderm) are found within this tumor. At imaging, its presentation ranges from pure cystic mass to complex solid cystic mass. In most cases, detection of intratumoral fat component is the key diagnostic imaging feature.

Diagnosis of MCT can be obtained by ultrasound (U/S), but U/S is nonspecific and it has a spectrum of appearance that depends on bones, teeth, hair, and echogenic fat material. The sonographic signs associated with ovarian MCT that have been well-described in the literature include the Rokitansky nodule or dermoid plug (the most common typical finding), diffuse or regional high-amplitude echoes (the second most common finding), iceberg sign, dot-dash sign, and floating balls sign. On computed tomography (CT), fat density and calcification inside a cyst are hallmark. On magnetic resonance imaging (MRI), the fat component is detected via fat saturation techniques (Fig. 1) and chemical shift imaging. However, unusual imaging features and complications may be encountered in routine clinical practice. It is, therefore, necessary to be familiar with the imaging features of unusual ovarian MCT lesions, and the complications associated with ovarian MCT.

2. Unusual lesions of ovarian teratoma (Table 1)

2.1. Immature teratoma

Mature teratoma is a benign tumor; however, the immature type, which is also benign, has a more aggressive course and a tendency to recur. Immature teratoma consists of three germ cell tissues, which is similar to the composition of mature teratoma. It is difficult to differentiate between mature and immature teratoma, but immature teratoma typically affects younger women (usually during the first 2 decades of life) and it has a worse prognosis [3]. A marked propensity for rapid growth and metastasis is observed even in early-stage lesions [4]. Removal of the uterus, including the fallopian tubes and ovaries, and the omentum is recommended if there is local extension or contralateral involvement. Radiotherapy offers no benefit.

Immature teratoma is extremely rare in adults and should not be confused with mature teratoma in which one tissue element has undergone malignant transformation. The latter is focal, and found incidentally in elderly patients with long-standing tumor (usually squamous cell carcinoma), and is associated with more favorable prognosis [4].

Immature teratoma tends to be larger (mean diameter: 12–25 cm) than mature teratoma (mean diameter: approximately 7 cm) at the time of diagnosis [5]. Similar to mature cystic teratoma, the cystic and fatty

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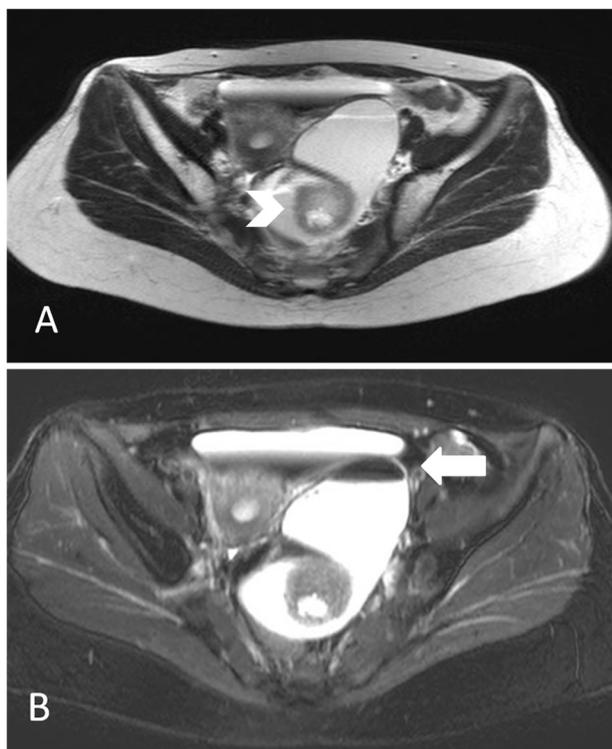


Fig. 1. A 30-year-old female with mature cystic teratoma. (A) Axial T2-weighted image shows Rokitansky nodule at posterior part of left ovarian cystic lesion (arrowhead). (B) Axial T2-weighted image with fat saturation shows low signal intensity of the supernatant fatty layer creating an interface with a high signal intensity aqueous layer (arrow).

component can be identified. However, the prominently enhanced solid component and large size are features that are helpful for differentiating immature from mature cystic teratoma. Tumor margins tend to be regular and smooth in mature teratoma, and irregular and poorly defined in immature teratoma. In immature teratoma, perforating tumor capsule is frequently demonstrated; however, it is not always well-defined [6]. Moreover, fatty areas may be small and difficult to observe as compared to mature teratoma. On CT, intratumoral calcification is another important feature since it is more focally distributed in the Rokitansky nodule or dermoid plug, and it is more curvilinear in

mature teratoma when compared to the random or dispersed pattern found in immature teratoma (Fig. 2).

Mature teratoma is usually treated with surgery, whereas immature teratoma is treated with combination surgery and chemotherapy. A phenomenon that is referred to as ‘retroconversion’ can be observed in immature teratoma treated with chemotherapy. Retroconversion is defined as the process of tissue maturation from immature to mature cystic teratoma. Retroconverted immature teratoma can remain stable for years [7]. Immature teratoma is associated with the presence of mature teratoma; however, it can be sometimes difficult to make preoperative diagnosis on imaging. Ipsilateral typical mature cystic teratoma is found in 26% of immature teratoma cases, and an immature teratoma will be observed in the contralateral ovary in 10% of cases [8] (Fig. 3).

2.2. Monodermal teratoma

A teratoma that consists exclusively of endodermal or ectodermal tissue is referred to as a monodermal teratoma. The types of monodermal teratoma include struma ovarii and carcinoid tumor.

2.2.1. Struma ovarii

Struma ovarii is the most common type of monodermal teratoma. It consists of mature thyroid tissue, and may be associated with symptoms of thyrotoxicosis in 5% of cases (helpful feature) [9]. Pathologic struma ovarii specimens differ from pathologic MCT specimens because struma ovarii consists of thyroid tissue, necrosis, hemorrhage, and fibrosis. Malignant struma ovarii is uncommon, and the clinical behavior is benign. Since 95% of struma ovarii are benign and they usually occur in premenopausal women, preoperative diagnosis is very important to avoid unnecessary surgery, such as hysterectomy and dissection of pelvic lymph nodes [10].

No fatty component is identified in this tumor. Although the imaging features are nonspecific, CT shows multiple cystic and solid areas that represent struma (Fig. 4). Cystic lesion with high CT attenuation (range: 58–98 Hounsfield units) that is caused by thyroid hormone and thyroglobulin in the ovarian tissue that attenuates the X-ray beam can be identified [11].

MRI findings of struma ovarii have been described in only a few cases [12]. These cases demonstrated multilocular cystic tumor with both high and low signal intensity on T1- and T2-weighted images that reflects gelatinous or colloid material. A markedly enhanced solid component was also observed that corresponds with thyroid tissue and

Table 1
Unusual imaging features of ovarian teratoma

Teratoma	Key features	Diagnostic pitfalls
Immature teratoma	<ul style="list-style-type: none"> - Younger age - Larger than mature teratoma - Prominence enhancing solid nodule - Perforation of tumor capsule - Peritoneal implants and lymph node metastases 	<ul style="list-style-type: none"> - Retroconversion - Fat may become invisible in larger mass
Monodermal teratoma – struma ovarii	<ul style="list-style-type: none"> - Benign clinical course despite pathologically confirmed malignancy - Solid part: strongly enhanced solid part, which is indicative of thyroid and vascular tissue - Cystic part: high signal intensity on T1W and T2W representing colloid material 	<ul style="list-style-type: none"> - No fat component - Only 5% associated with thyrotoxicosis
Monodermal teratoma – carcinoid tumor	<ul style="list-style-type: none"> - Postmenopausal woman - Primary ovarian carcinoid is unilateral - Metastatic ovarian carcinoid is bilateral - Hypervascular ovarian mass 	<ul style="list-style-type: none"> - No specific finding - No fat component unless associated with mature cystic teratoma - Carcinoid syndrome is rare
Mature cystic teratoma with minimal or no fat Collision tumor	<ul style="list-style-type: none"> - Identification of minimal fat in the cyst wall or in the Rokitansky nodule, especially when using chemical shift imaging - Most common is mixture of mature cystic teratoma and MCN - Typical finding is two adjacent cystic masses with septations, one of which contains intratumoral fat 	<ul style="list-style-type: none"> - Minimal or no fat component - Differentiation from other epithelial ovarian tumors is difficult - Overlooked nonteratoma or malignant tumor at imaging - Clinical context and serial imaging may help to define the occurrence order of the two components

Abbreviation: MCN, mucinous cystic neoplasm.

Table 2
Complications of ovarian teratoma

Teratoma	Key features	Diagnostic pitfalls
Rupture	<ul style="list-style-type: none"> - Discontinuity of the wall - Distorted or flattened shape - Associated with peritonitis - Oily ascites (fat fluid level) 	<ul style="list-style-type: none"> - May mimic peritoneal carcinomatosis or TB peritonitis
Torsion	<ul style="list-style-type: none"> - Identify twisted vascular pedicle - Identify intervening fat in torsed ovary - Thickening of the fallopian tube - Whirlpool sign on color Doppler U/S 	<ul style="list-style-type: none"> - The degree of torsion causes a variety of flow patterns on Doppler U/S, and levels of enhancement on CT
Malignant transformation	<ul style="list-style-type: none"> - Most common is squamous cell carcinoma - Postmenopausal woman - Contrast enhancement and growth of Rokitansky protuberance increases likelihood - Foci of hemorrhage and necrosis is common 	<ul style="list-style-type: none"> - Fat may become invisible - May mimic mixed germ cell tumor
Gliomatosis peritonei	<ul style="list-style-type: none"> - Metastatic implantation of glial tissue on the peritoneal linings - Associated with ovarian mature teratoma or immature teratoma 	<ul style="list-style-type: none"> - May mimic peritoneal carcinomatosis or TB peritonitis

Abbreviations: TB, tuberculosis; U/S, ultrasound; CT, computed tomography.



Fig 2. A 25-year-old female with immature teratoma. (A) Axial non-contrast-enhanced CT shows right ovarian mass with random patterns of intratumoral calcification (arrows) and minimal fat foci (arrowheads). (B) Axial contrast-enhanced CT shows multiple areas of solid enhancement (arrows).

stroma of abundant blood vessels [11,13].

Scintigraphy performed with either iodine-131 or iodine-123 is helpful for diagnosing hyperfunctioning or hyperactive struma ovarii upon higher uptake of the radionuclide by the ovarian mass compared with that of the thyroid gland [14].

The differential diagnosis should include MCT without fatty tissue, cystadenoma or cystadenocarcinoma, and metastasis if it is not



Fig. 3. A 19-year-old female with bilateral ovarian masses. Axial contrast-enhanced CT shows right ovarian mature cystic teratoma (arrows) and left ovarian immature teratoma (arrowheads).

associated with hyperthyroidism.

A case of struma ovarii with ascites and pleural effusion was previously reported [14]. This unusually rare association is referred to as pseudo-Meigs syndrome [14].

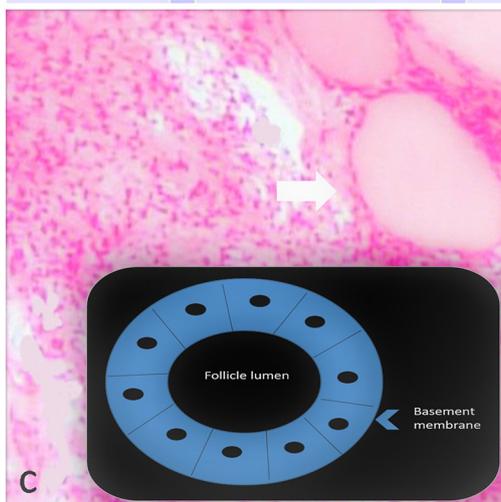
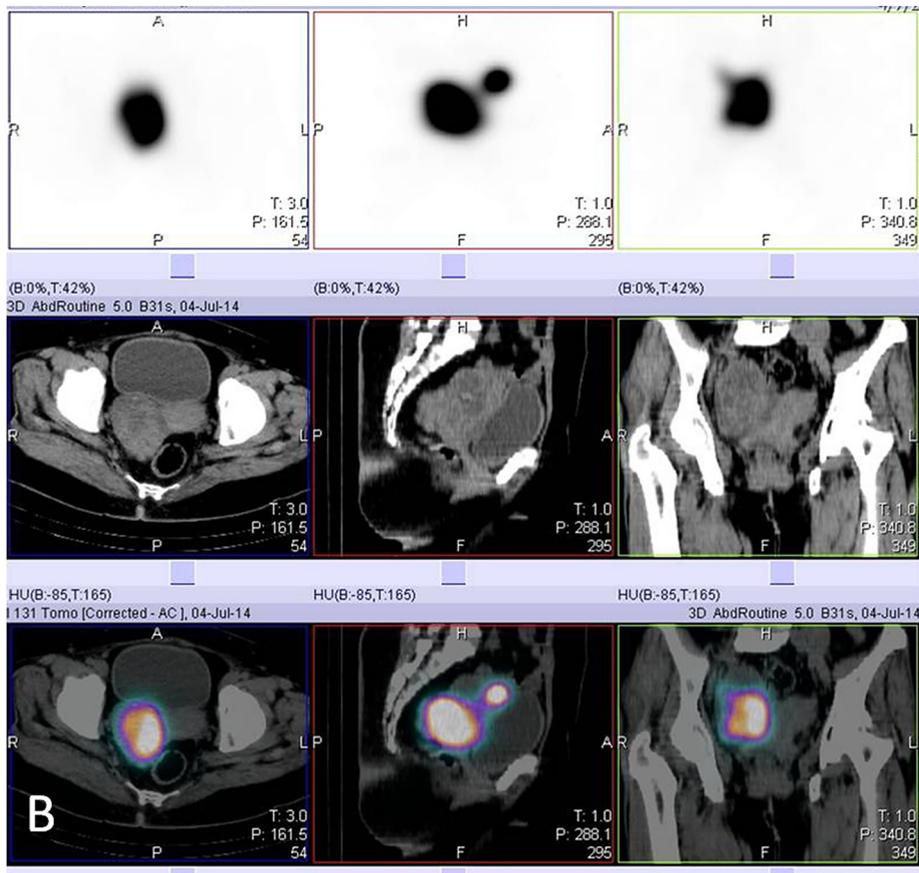
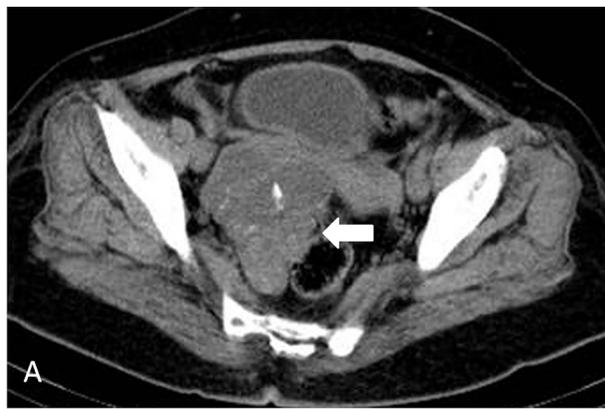
Meigs syndrome is defined by the presence of ascites and pleural effusion in association with a benign ovarian tumor, which is typically an ovarian fibroma. Other tumors associated with this syndrome include thecomas and Brenner tumors [15].

Pseudo-Meigs syndrome refers to a clinical syndrome of ascites and pleural effusion in association with an ovarian tumor that is not a fibroma or a fibroma-like tumor [16].

2.2.2. Carcinoid tumor

Ovarian carcinoid tumors can be categorized as either primary or metastatic, and they usually affect postmenopausal women [17]. Primary ovarian carcinoid is commonly unilateral, it metastasizes only in a minority of cases, and it should be managed as an ovarian tumor with low malignant potential. Differentiating between primary ovarian carcinoid and metastatic carcinoid is a challenge; however, bilaterality, peritoneal spreading, absence of teratomatous component, and lymphovascular invasion are suggestive of metastatic carcinoid [18] (Fig. 5).

The four types of primary ovarian carcinoid are insular, stromal, trabecular, and mixed. Insular type is the most common, followed by stromal type. Carcinoid syndrome occurs only in insular type, and in only about 30% of cases [19]. Carcinoid syndrome normally does not



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Fig. 4. A 45-year-old female with thyrotoxicosis and struma ovarii. (A) Axial non-contrast-enhanced CT shows mixture of cystic and solid areas, and small calcifications at right ovary (arrow). (B) Whole body I-131 imaging shows significantly increased uptake at right ovary, which is consistent with confirmed struma ovarii. (C) Histologic examination revealed a few small areas where microfollicles of thyroid tissue were evident (arrow). Inset image shows a schema of a thyroid follicle.



Fig. 5. A 57-year-old female with metastatic ovarian carcinoid and carcinoid syndrome. (A) Axial non-contrast-enhanced CT shows complex solid cystic mass at both ovaries (arrows). (B) Axial contrast-enhanced CT shows hypervascular solid enhancement at both ovaries (arrows).

occur with intestinal carcinoid unless there is liver metastasis (deficient hepatic detoxification). However, ovarian carcinoid can cause carcinoid syndrome, because venous drainage bypasses the portal venous system into the systemic circulation. Imaging finding shows no specific findings other than a solid mass that is indistinguishable from solid malignant ovarian lesions [19]. Demonstration of prominent hypervascularity of the ovarian carcinoid following contrast administration may be a helpful diagnostic feature. However, hypervascular solid components can be observed in other malignant ovarian lesions along with mixed solid and cystic appearance and areas of necrosis. Serotonin-induced stromal proliferation is generally a pathological feature of ovarian carcinoid [6]. Low signal intensity on T2-weighted images, which is rarely observed in other malignant ovarian tumors, may be a characteristic imaging finding in ovarian carcinoid [19].

2.3. Mature cystic teratoma with minimal or no fat

A small percentage of MCT may present with minimal fat or no fat in the lumen of the cyst, and this may make it difficult to establish a diagnosis of MCT (Fig. 6). However, in some cases, a minimal amount of

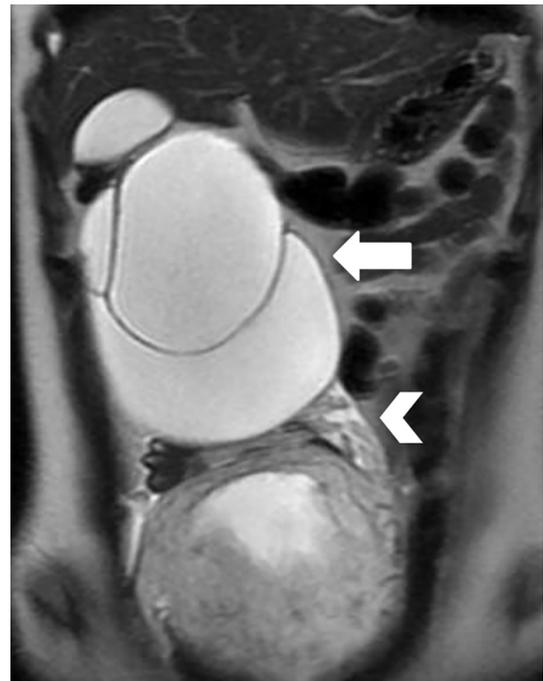


Fig. 6. A 35-year-old pregnant woman with mature cystic teratoma without a fat component. Coronal T2-weighted image shows cystic lesion with homogeneous high signal intensity (arrow) with low signal intensity thin septations. Pathologically confirmed cystic-sebaceous material with less fatty tissue than would normally be observed in a mature cystic teratoma. Torsion is also detected (arrowhead).



Fig. 7. A 36-year-old female with minimal fat ovarian teratoma. Axial contrast-enhanced CT shows cystic lesion at left ovary with minimal streak of fat at the anterior wall (arrow).

fat can be observed in the cyst wall or in the Rokitansky nodule on CT and MRI (Fig. 7). Accordingly, the cystic wall must be thoroughly examined in order to make a correct diagnosis [20].

2.4. Collision tumor

Collision tumor is a term that may be confused with combination tumor or composite tumor [21]. Collision tumor is defined as the

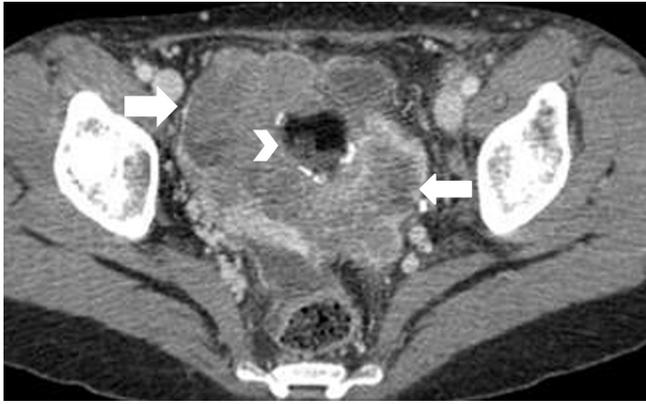


Fig. 8. A 43-year-old female with tumor within tumor pattern of collision tumor. Axial contrast-enhanced CT shows smaller fatty component at the center (arrowhead) and in the larger enhancing solid component (arrows), which was surgically proven to be collision tumor (ovarian teratoma and cystadenocarcinoma).



Fig. 9. A 39-year-old female with kissing tumor pattern of collision tumor. Axial contrast-enhanced CT shows enhancing solid components (arrowheads) abutting a smaller fatty component with calcifications (arrow). The pathologic findings revealed collision tumor (ovarian teratoma and undifferentiated carcinoma).

coexistence of two adjacent, but histologically different tumor components that are separated from each other by stroma with no histological admixture. In contrast, combination tumors are composed of a mixture of tissue fractions that originate from a single type of stem cell, such as mixed germ cell tumor [22]. Composite tumors contain a mixture of tissues with different histology and no clear separable interface. A composite tumor may form from the simultaneous growth of tumors in different tissues of the same organ or from sarcomatous changes in the stroma of the carcinoma (carcinosarcoma).

Ovarian collision tumors are rare, but the most common type is made up of mature cystic teratoma and mucinous ovarian neoplasm (cystadenoma or cystadenocarcinoma) [23,24]. The smaller component is usually situated within the larger component (tumor within tumor) (Fig. 8), or is located at the tumor wall (kissing tumor) [25,26] (Fig. 9). The prognosis or clinical course depends on the characteristics of the tumor. CT and MRI findings include part typical feature of teratoma and part nonteratoma, such as multiloculated cystic lesion, solid-cystic complex mass, or solid content. The possibility of a collision tumor should be considered when an ovarian teratoma has imaging findings that cannot be explained solely via satisfaction of the diagnostic imaging criteria for ovarian teratoma.

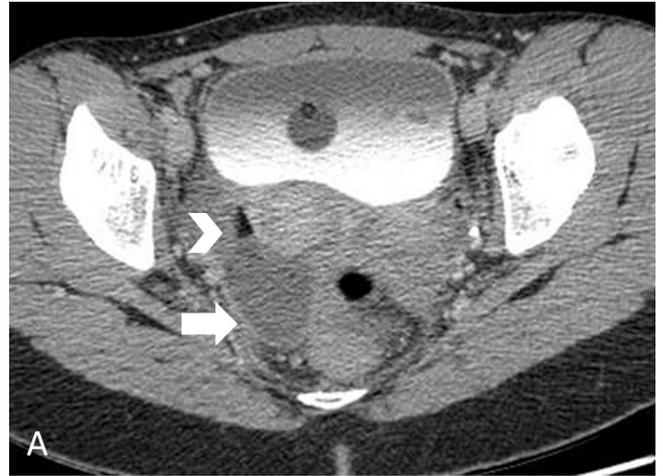


Fig. 10. A 42-year-old female with ruptured ovarian teratoma. (A and B) Coronal contrast-enhanced CT shows fatty mass-like infiltrations at peritoneal linings in perihepatic regions (arrowheads).

3. Complications of ovarian teratoma (Table 2)

3.1. Rupture

Rupture occurs in 1–4% of ovarian teratomas [28]. Chronic rupture of ovarian teratoma is more common than acute rupture. This type of complication causes leakage of sebaceous fluid or particles into the peritoneal cavity, which most often results in peritonitis. Acute peritonitis usually occurs after sudden tumor rupture, whereas chronic peritonitis is more likely to result from chronic tumor leakage.

Wall discontinuity, ascites, and distorted shape are key imaging findings on CT and MRI [27]. Sebaceous content causes irritation of the peritoneum in both acute and chronic peritonitis. The presentation is localized or diffuse peritoneal stranding and mass-like infiltration (Fig. 10). A potential diagnostic pitfall is that these findings mimic both tuberculous peritonitis and peritoneal carcinomatosis [28]. To minimize potential confusion, thoroughly check the integrity of the wall in the setting of ovarian teratoma and observe the fat fluid level of the ascitic fluid (oily ascites) (Fig. 11). Necrotic lymph nodes associated with other granulomatous features, and solid enhancing peritoneal mass with primary site of cancer are the helpful features in tuberculous peritonitis and peritoneal carcinomatosis respectively.

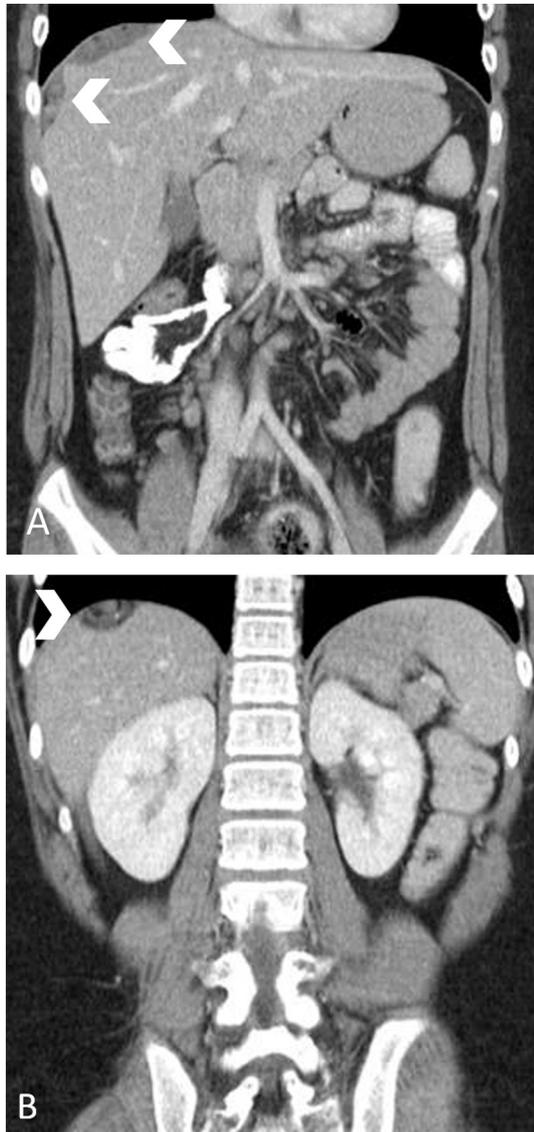


Fig. 11. A 40-year-old female with ruptured ovarian teratoma. (A) Axial contrast-enhanced CT shows right ovarian teratoma with discontinuity of wall enhancement and slight loss of spherical contour (arrowhead). Intact and smooth enhancing wall at posterior aspect is seen (arrow). (B) Axial contrast-enhanced CT (superior to A) shows ascites with fat fluid level (oily ascites) (arrowheads).

3.2. Torsion

Ovarian teratoma is the most common ovarian mass associated with torsion, with a prevalence of torsion of 3–16% [29]. Torsion is defined as partial or complete twist of the ovary on its supportive ligament. If complete torsion occurs, gangrenous or hemorrhagic infarction will result. An ipsilateral twisted pedicle is the most specific imaging finding, and thickened fallopian tube was reported in 84% of cases. U/S is the first-line imaging modality. The aim is to identify the twisted vascular pedicle (whirlpool sign) on color Doppler U/S [30].

However, twisted pedicle is found in less than one-third of patients on axial CT or MRI [31] (Fig. 12). The recommendations include: 1) use multiplanar reformation to increase the visualization of the spiral configuration of the vascular pedicle; and, 2) identify ancillary features, such as beak sign, ipsilateral uterine deviation, contralateral or midline ovarian displacement, stranding at periannexal fat, and smooth eccentric wall thickening of the ovarian mass [31]. Importantly, the absence of the aforementioned features does not conclusively exclude a



Fig. 12. A 46-year-old female with torsion of mature cystic teratoma without fat component. (A) Axial T2-weighted image shows left ovarian cystic mass (arrowhead) and twisted vascular pedicle (arrow). (B) Sagittal T2-weighted image shows beak sign (arrow) and twisted vascular pedicle (arrowhead). (C) Axial T2-weighted image shows ipsilateral uterine deviation (arrowhead) with elongation of right round ligament (arrow).

diagnosis of torsion.

The authors have also observed some cases with a target-like lesion located adjacent to the ovarian mass that appears to reflect the presence of torsion, but surgical confirmation revealed neither twisted pedicle nor ovarian torsion. Intervening fat within the torted ligamentous support is the helpful finding for making a more accurate diagnosis.



Fig. 13. A 67-year-old female with malignant transformation of ovarian teratoma. (A and B) Axial contrast-enhanced CT shows large solid enhancing mass arising from left ovary with contiguous sacral spinal and left sacroiliac joint invasion (arrow in A) The mass also extends inferiorly to the perianal region with anal invasion. A minimal amount of fat is detected at the right inferolateral aspect of the mass (arrow in B).

3.3. Malignant transformation

Malignant transformation usually occurs in postmenopausal women and may occur in any of the three germ cell lines (ectoderm, mesoderm, and/or endoderm). Squamous cell carcinoma that arises from the cyst wall that lines the squamous epithelium is the most common type (80%), followed by carcinoid tumor and adenocarcinoma [2,3].

Soft tissue formation in a MCT, namely the Rokitansky nodule or dermoid plug, is commonly observed, but benign teratoma never shows transmural interval growth of the protuberance [32]. Malignant transformation normally originates from the Rokitansky nodule or dermoid plug. Therefore, the appropriate sections must be selected and analyzed during histopathologic investigation [33].

Regarding the analysis of gynecologic oncology, clinical, and laboratory reports, findings associated with malignant transformation include patient age greater than 45 years, tumor diameter greater than 9.9 cm, and serum squamous carcinoma antigen level greater than 2 ng/ml [34–36].

The imaging findings on CT and MRI of malignant transformation include invasive growth of large irregularly marginated soft tissue lesions at the wall of or within the tumor, and adjacent organ invasion in some cases [37] (Fig. 13). The fat component may not be visible or may disappear as the mass grows to a large size.

Importantly, although laparoscopic surgery has gained in popularity

for the management of benign ovarian teratoma, the use of laparoscopic surgery to manage malignant transformation is risky and is not recommended due to the higher risk of tumor spillage of dermoid contents during laparoscopic procedures. Successful laparoscopic removal of adherent tumor without spilling its contents is frequently problematic, so a laparotomy should be performed to prevent inadvertent tumor upstaging by iatrogenic intraperitoneal tumor rupture [38].

3.4. Gliomatosis peritonei

Gliomatosis peritonei is a rare occurrence that is almost exclusively associated with ovarian mature teratoma or immature teratoma [39], with a few cases associated with ventriculoperitoneal (VP) shunting that was performed to manage hydrocephalus. Gliomatosis peritonei is characterized by metastatic implantation of glial tissue on the visceral or peritoneal linings. Glial tissue implants are normally 1–10 mm in size, there is no fatty component, and they are grossly indistinguishable from tuberculous peritonitis and peritoneal carcinomatosis. Omental caking and ascites can also be observed. Therefore, diagnosis of gliomatosis peritonei is made histologically [40]. If the peritoneal implants contain immature glial tissue, one must rule out metastasis of immature teratoma [41]. The treatment decision should be based on the grade of the primary tumor – not on the glial tissue implants.

4. Conclusion

Familiarization with the unusual imaging features of and complications associated with ovarian teratoma can improve the timeliness and accuracy of diagnosis, the management of the disease, and the outcomes of patients with ovarian MCT.

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Declaration of Competing Interest

None.

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