Mature Cystic Teratomas and the most common complications

Poster No.: C-2230  
Congress: ECR 2015  
Type: Educational Exhibit  
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Keywords: Genital / Reproductive system female, Pelvis, Ultrasound, CT, MR, Diagnostic procedure, Neoplasia, Cysts, Acute  
DOI: 10.1594(ecr2015/C-2230

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Learning objectives

• Review epidemiologically relevant facts about ovarian mature cystic teratomas;
• Describe its most common complications;
• Establish the usual differential diagnosis of each complication.
**Background**

Germ cell derived tumors are nearly 20% of all ovarian tumors in adults, and 50% of ovarian tumors in the pediatric population [1].

About 95% of these germ cell tumors are mature cystic teratomas, which are typically benign slow growing lesions that can be bilateral in 10% of the cases. These tumors usually have a prominent cystic constitution and might have fat and calcium components [2].

These lesions usually affect younger women than epithelial ovarian neoplasms, with a mean patient age of 30 years. It is also the most common ovarian mass in children [2].

Although typically incidentally discovered, they are also prone complications, the most common ones being: torsion (3.2-16%), rupture(1.2-3.8%) and malignant transformation(1-2%) - which will be discussed in this poster.

Hence, we review the most common ultrasound, CT and MR features of incidentally discovered lesions and of the above mentioned complications, using data from our institution and from the literature, without forgetting a list of common differential diagnosis.
Findings and procedure details

Ovarian teratomas include a variety of tissues of germ cell (pluripotential) origin. The mature cystic teratoma (MCT - also known as dermoid cyst) is the most common of these lesions, and is composed of well-differentiated tissues, from at least two of the three germ cell layers (ectoderm, mesoderm, and endoderm) [2].

MCT also has a wide spectrum of presentations ranging from the asymptomatic and incidentally detected small mass, to conditions associated with several complications [1]. These complications have unspecific clinical presentations that overlap with many other conditions, and require different therapeutic strategies, which demands an accurate diagnosis to optimize patient care[1].

Some authors advocate nonsurgical management to lesions smaller than 6-cm. MCT requiring removal can also be treated with simple cystectomy [2].

Immature teratomas and monodermal teratomas, although relevant, are beyond the scope of this poster.

**Incidentally discovered lesions:** The constitution of MCT varies, from purely cystic masses to a lesion with a considerable solid component. However, there are some typical findings, like intratumoral fat, that although not constant are highly suggestive of these lesions[1].

The most common US finding of a MCT is an echogenic nodule (Rokitansky nodule or dermoid plug) at its border, projecting into the cystic lumen [1,2]. This nodule may contain hair, teeth, and fat. Other classically described sign occurs when this nodule presents as a highly echogenic mass [1,2] that casts an acoustic shadow, making the so called "tip of the iceberg sign". Heterogenous fluid contents with multiple echogenic bands, caused by hair, and fat-aqueous fluid interface, derived from the non-mixing proprieties of the sebum layer, are also commonly described [1,2]. Mais et al reported an ultrasound sensitivity of 58% and a specificity of 99% in the diagnosis of mature cystic teratoma[2].

Intratumoral fat can be readily detected by CT and MR, which is responsible for the high sensitivity of these methods to the diagnosis of these uncomplicated teratomas[1].
At CT, intratumoral fat (reported in 93% of the cases) has a negative attenuation [1] within a cyst, with or without calcification in the wall (reported in 56% of the cases[2]. A floating debrum of hair might also be identified at the fat-aqueous fluid interface[2].

At MR imaging, although the combination of T1- and T2-weighted images is not specific for fat, the study of a MCT might benefit from fat-saturated T1-weighted sequences and chemical-shift artifact gradient-echo imaging[1,2]. If a fat-aqueous fluid interface is present, the sebaceous layer might present with T1-weighted images signal intensity similar to retroperitoneal fat, but as the T2-weighted images is highly variable this lesions must be distinguished from some hemorrhagic lesions, most promptly endometriomas [2].

Hence, frequency-selective fat saturation will suppress the high signal of intratumoral fat within the teratomas and can help distinguishing them from hemorrhagic lesions[2]. On the other hand, chemical-shift artifact using a gradient-echo sequence, with an echo time in which fat and water are in opposite phase, might be helpful for identifying microscopic fat in tumors that have only a tiny amount of fat[1].

Therefore, it is necessary to be familiar with the imaging features of incidentally discovered mature cystic teratomas and its complications:

**Ovarian torsion:** Adnexal torsion is the fifth most common gynecologic emergency with a reported prevalence of 2.7% [1]. On one hand, 50-80% of these cases have an ovarian mass predisposing to this, and MCT is the most common one. On the other hand, torsion is the most common MCT complication, reaching about 3-16% of the patients with ovarian teratomas[1,3]. MCT affected by torsion have a higher diameter than the average (mean 11 cm vs 6 cm)[2].

Imaging has a key role in the diagnosis, since its clinical symptoms and signs are nonspecific and occur in other acute diseases such as tuboovarian abscess, endometriosis, appendicitis, and ovarian cyst rupture[1].

Ovarian torsion results from blood flow compromise, when the ovary twists around its own vascular pedicle [1].

In an early-stage or partial torsion, the blockage of the venous flow and lymphatic return occurs, leading to a massive ovarian edema, resulting in the diffuse enlargement of the ovarian parenchyma and distention of peripheral follicles. As this process progresses, persistent edema and increased pressure on the twisted pedicle will cause arterial
ischemia. If diagnosis and reduction of ovarian torsion are delayed, hemorrhagic infarction may occur and can cause a severe peritonitis or even worst[1,3]. A partial torsion may also occur intermittently with spontaneous untwisting, making the symptoms persist, in some cases subsisting for weeks[3].

Early diagnosis might help preventing irreversible damage of the adnexal structures, allowing a conservative, ovary-sparing treatment in young women[1].

Ultrasound may identify some significant adnexal alterations which include cystic, solid, or complex mass appearance, with or without pelvic fluid, and cystic hemorrhage, but still, these gray-scale findings are not specific and for a more accurate diagnosis a Doppler US revealing a twisted vascular pedicle ("whirlpool sign") is necessary and strongly suggestive [1].

We should keep in mind that most ovaries with torsion can demonstrate a wide variety of flow patterns, including normal arterial and venous flow [1].

Common CT and MR imaging features of ovarian torsion include an enlarged ovarian mass with cortical follicles, smooth wall thickening of the twisted MCT, a twisted vascular pedicle and an ipsilateral fallopian tube enlargement - these last two signs are the most specific for the diagnosis according some authors [1]. This process is also associated with peritumoral infiltration seen as ill-defined linear or reticular shadows in the peritumoral spaces, hemorrhage within the mass,[3] ascites, enlarged veins, and uterine deviation to the twisted side[1].

**Rupture:** Rupture occurs in 1%-4% of ovarian teratomas [1,4]. Due to the thick capsule, MCT rarely rupture. When it happens it is more commonly associated with torsion, trauma, infection, or labor. The acute leakage of liquefied sebaceous contents irritates the peritoneum, resulting not only in acute peritonitis, but also in shock and hemorrhage. Chronic and recurrent leakage is more common than an acute episode of leakage and causes chronic granulomatous peritonitis, known as gliomatosis[1,3]. The prognosis of chronic rupture is also more favorable, although this chronic inflammation may cause dense peritoneal adhesions and other related secondary complications such as bowel obstruction [1].

At US, CT, and MR imaging an accurate diagnosis of a ruptured MCT might be accomplished if a discontinuity of the wall is noted [1]. Acute or chronic peritonitis may manifest as ascites, hazy omental and bowel inflammatory infiltration (difuse or focal). Although these findings might mimic peritoneal carcinomatosis and tuberculous peritonitis,
the presence of ascites and a distorted or flattened shape tumor suggests MCT rupture, indicating the need for surgical evaluation[1,3].

**Malignant transformation**: occurs in 1%-2% of ovarian teratomas and represents 1% of all ovarian malignancies[1]. The term *malignant transformation* is reserved for malignancies arising de novo within a preexisting benign MCT[2]. Although MCT are more common in younger women, malignant transformation usually happens in postmenopausal women[4] and occurs more frequently in the 6th or 7th decade of life[2].

Malignant transformation may occur from any of the three germ cell layers, but it is more commonly derived from the squamous lining of the cyst, accounting for 80% of the reported cases, and giving rise to a squamous cell carcinoma[1]. Risk factors for malignant transformation are: patient age greater than 45 years; tumor diameter greater than 9.9 cm; and serum squamous carcinoma antigen level greater than 2 ng/mL according to some authors [1].

The already described soft-tissue protuberance typical of MCT, also known as a Rokitansky nodule or dermoid plug is the most common site of malignant transformation, and although this might be solid and heterogeneous, it should never show transmural growth, which is highly suggestive of malignant transformation. Contrast enhancement of a Rokitansky nodule also raises the possibility of malignant transformation but it does not always necessarily indicate malignancy [1].

Making a differential diagnosis between benign and malignant tumors is crucial, since the surgical procedure performed is quite different. Hence, laparoscopic surgery is often performed for the mature cystic teratoma, but is quite risky when malignant transformation is suspected. [1]

At CT and MR imaging features of malignant transformation includes: size; irregularly marginated soft-tissue components through the tumor, invasive growth towards other structures, the presence of an enhancing soft-tissue component and an obtuse angle between the soft tissue and the cyst inner wall[1,3].
Fig. 1: Axial contrast material-enhanced CT scan- Mature cystic teratoma.

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**Fig. 2:** Axial contrast material-enhanced CT scan - Mature cystic teratoma.

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**Fig. 4:** Axial non-enhanced CT scan after bowel opacification with oral contrast - the cyst cavity demonstrates fat attenuation and a round Rokitansky nodule with calcified elements.

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Fig. 3: Axial non-enhanced CT scan - Mature Cystic Teratoma - fat stranding and liquid outside the formation suggest rupture.

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Fig. 5: Image courtesy of MD Eric K. Outwater. Axial T1-weighted spin-echo MR image shows a heterogeneous mass (black arrow) with high-signal-intensity foci (white arrow). Infarcted mature cystic teratoma resulting from ovarian torsion.

Conclusion

Recognizing the various presentations of mature cystic teratomas and its alarm signs are important to make the correct management of the patient condition.
References


