REVIEW ARTICLE

Pathognomonic Landmark of Mature Cystic Ovarian Teratomas in HDlive Ultrasound

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ABSTRACT

Mature cystic ovarian teratomas are the most common benign ovarian tumors. For their diagnosis, the following ultrasound (US) images can be considered as pathognomonic:
- A cystic lesion (unilocular, very frequent, or multiple, with a densely echogenic tubercle (Rokitansky nodule).
- A diffuse or partially echogenic mass with the echogenic area usually demonstrating sound attenuation.
- The 'iceberg' sign, very similar to the 'sound attenuation' sign.
- Fluid-fluid/fat-fluid level. Fluid-fluid levels, or fat-fluid interface, result from sebum floating above aqueous, which appears more echogenic than the sebum layer.
- Dermoid mesh with hyperechoic calcifications indicating the presence of bone, teeth, or other ectodermal derivate.
- Multiple mobile spherical structures (fat balls).

Keywords: Ovarian cystic teratomas, Two-dimensional, Three-dimensional, HDlive.

INTRODUCTION

Ovarian teratomas are a group of tumors that contain mature and/or immature tissues, which are origined by the uncontrolled growth of pluripotential germ cells after the first meiotic division.[1]

Ovarian teratomas include different types of histological tumors:

a. Mature cystic teratoma, also called 'Dermoid cyst', which is the most common one. It contains always tissues that arise from, at least, two of the three embryologic layers, as shown:
- Ectodermal: Skin, brain, neural tissue (always present).
- Mesodermal: Muscle, fat, cartilage, bone (present in over 90% of cases). Teeth are seen in 31% of cases.
- Endodermal: Gastrointestinal, bronchial mucinous ciliated epithelium, Thyroid tissue (seen in the majority of cases). Adipose tissue is present in 67 to 75% of cases.
b. Immature teratomas: Very infrequent (<1% of all teratomas). They have a malignant behavior.
c. Monodermal teratomas: In these ones, one type of tissue predominates (e.g. thyroid tissue in struma ovarii, neuroectodermal tissue in carcinoid and neural tumors).

Most of the mature cystic teratomas can be diagnosed by using ultrasonography (US) but they have very different ways of appearance characterized by echogenic sebaceous material, bones, teeth and hair.

About its Pathogenesis

The pathogenesis of balls formation remains unknown and several theories have been postulated:

1. Predominance of large secretory and absorptive layers rather than exfoliated ones would favor the absorption of most of the cysts contents into the general circulation, leaving the remaining material in a solid status and mold into balls.[14]
2. It is speculated that each globule is formed by the aggregation of sebaceous material around a tiny focus of debris, squames, or fine hair shaft while moving around the cystic cavity.
3. The spherules appear to have been modeled into discrete masses rather than remaining as an amorphous mass because of the difference in physical and thermal properties of the material being deposited around each nest. Floating balls require space to be remodeled.
4. This type of growth could be related to an unusual pattern of estrogen and progesterone receptor expression in the cystic teratoma,[27] although consensus has not been reached.[18,21,24]

With magnetic resonance imaging (MRI), the sebaceous component is specifically identified with fat-saturation techniques. With computer tomography (CT), fat attenuation and a calcification inside a cyst are diagnostic.

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Different features are shared by the mature cystic teratomas:

1. Cystic tumors are generally unilocular (88%), filled with sebaceous material, liquid at body temperature and semisolid outside. Cystic dominant lesions may be seen as complex masses. Intracystic fat tissue is visualized as an hyperechogenic image, when solid epithelial rests and hair form conglomerates.[2,3]
2. The wall of the cyst, a smooth-surfaced lesion, is lined by squamous epithelium.
3. Sebaceous material, hair follicles, bones, calcifications, skin glands, muscle, thyroid tissue (exceptionally) or bronchial mucous membranes can be seen inside this wall.

SONOGRAPHIC APPEARANCE OF TERATOMAS

The US appearance of immature teratomas is no specific. These tumors are typically heterogeneous and their sonographic appearance depend on their major content.[3] They can be solid or with a prominent solid component with cystic elements, usually filled with serous, mucinous or fatty sebaceous material. Scattered calcifications and small fat foci within the solid component are usually recognized:

Monodermal teratomas are usually formed by only one type of tissue. Struma ovarii ones show a non-specific heterogeneous, predominantly solid US feature. Carcinoid tumors are solid and indistinguishable from other solid malignancies.

Benign cystic teratomas can produce a wide spectrum of sonographic appearances which depends of their major content.[3]

The US imaging of mature teratomas is predominantly cystic. However, sometimes they appear as a complex mass, or even as a solid tumor if the intracystic fat tissue, epithelial debris and hairs are conglomerated filling the entire cyst.

They can also be seen as an hyperechogenic mass with posterior acoustic shadow, including diffuse or local shiny echoes or a fluid-fluid/fat-fluid level.[2-5]

The three pathognomonic landmark images are:

1. A cystic lesion, usually unilocular (Figs 1 to 3). It can be as well multilocular sometimes (Fig. 4) with a dense echogenic tubercle (Rokitansky nodule), with sound attenuation because of sebaceous material and hair within the cystic cavity (Figs 1 and 2). This nodule is also known as 'dermoid plug', which represents a protuberance that arises from the tumor wall and contains hair follicles
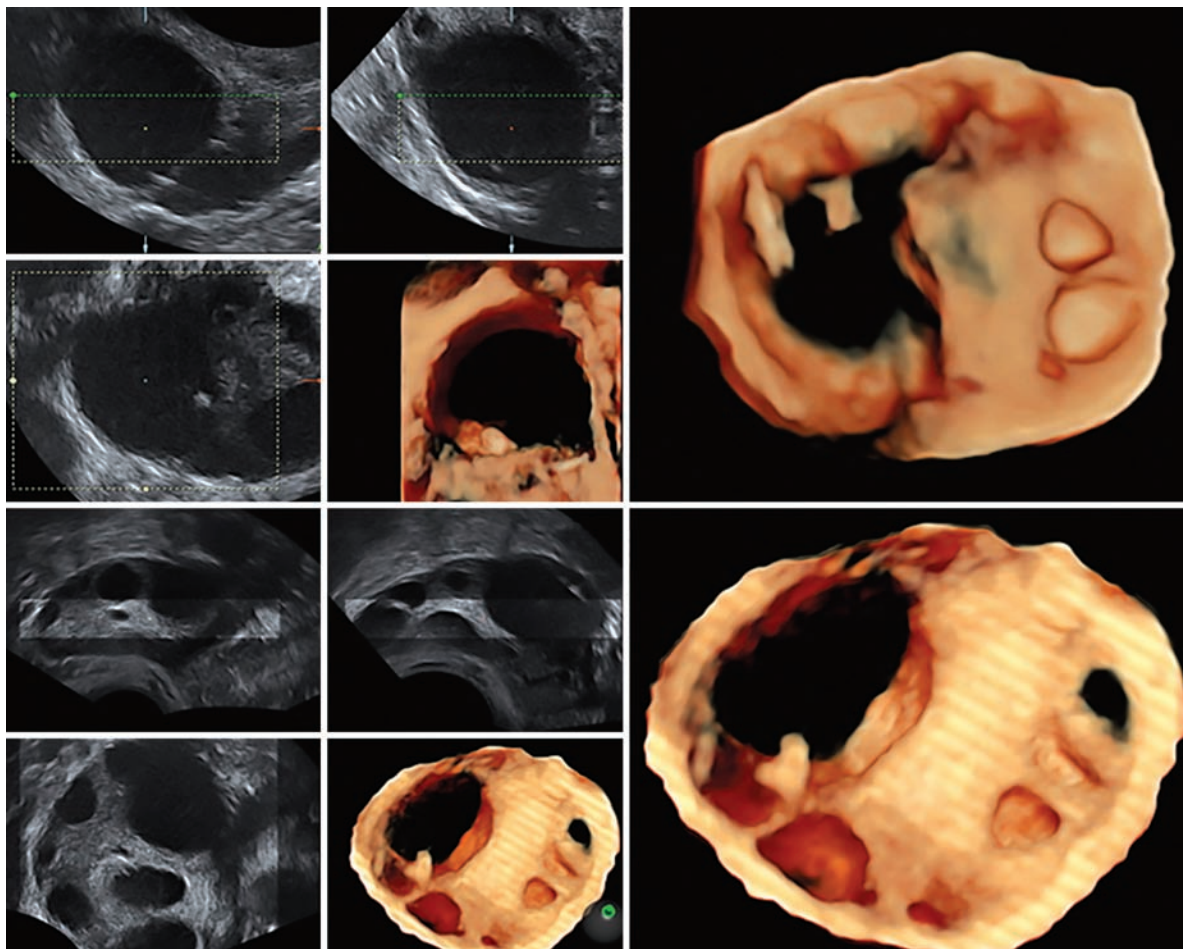
Fig. 1: Pure cystic teratoma with Rokitansky nodule
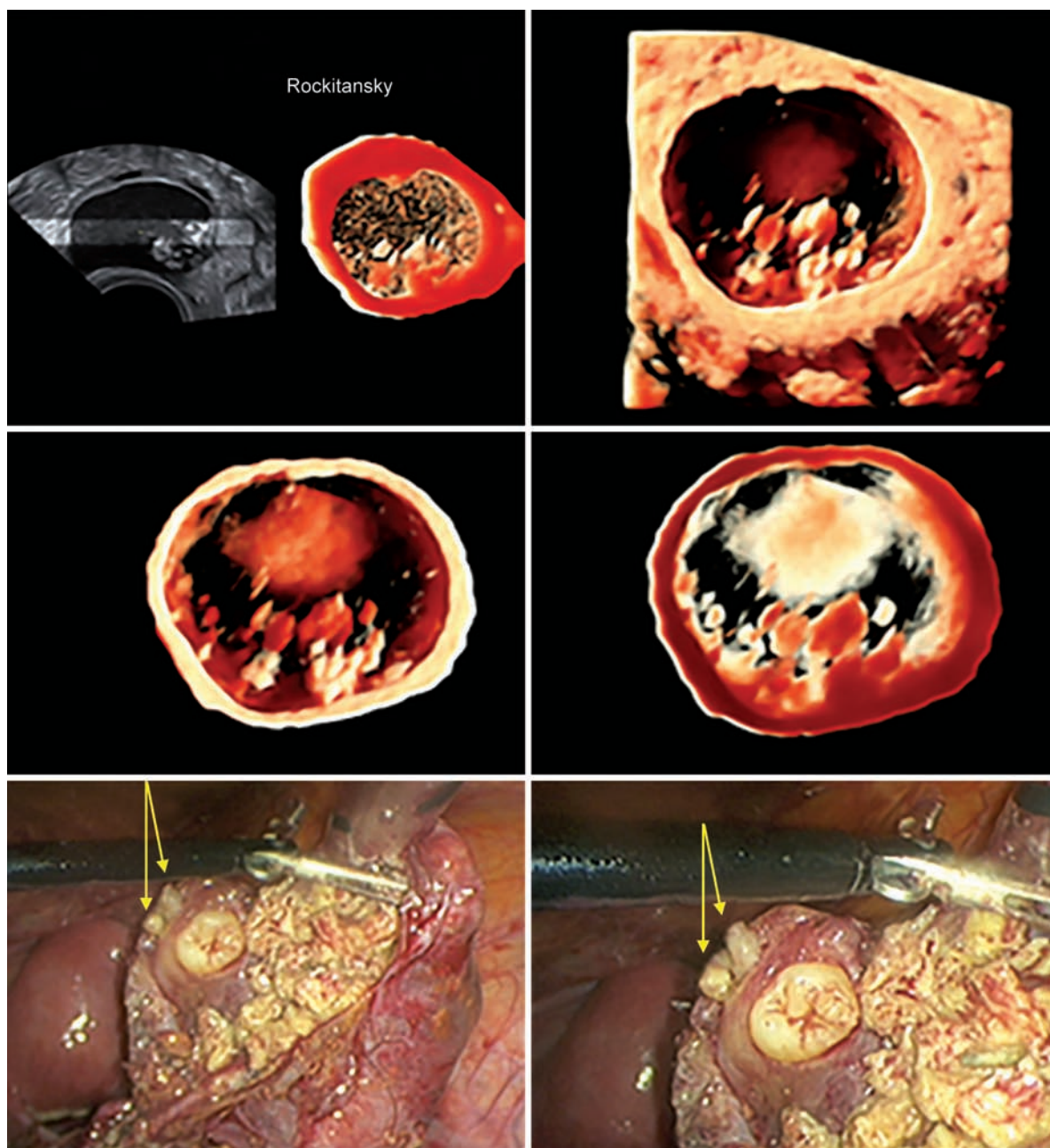
Fig. 2: 2D orthogonal planes, 3D and HDlive of a cystic lesion with a dense echogenic tubercle (Rokitansky nodule) located in the inferior part of the cyst. 3D of the Rokitansky nodule shows bones and teeth. The photo below shows a macroscopic view of the nodule with two teeth (arrows)

and/or other solid elements. This nodule can be seen as an echogenic mass with distal acoustic shadow.

2. A diffuse echogenic mass with an echogenic area that usually demonstrates sound attenuation, due to sebaceous material and hair within the cyst cavity (Figs 5 and 6).

3. Multiple thin echogenic bands caused by hair in the cyst cavity[6] (Figs 4 and 7).

We also consider the following ultrasonographic 'signs' as specific and patognomonic to establish the diagnosis:

• A pure cystic lesion with bones, teeth or cartilaginous nodules (generally called 'bones', Rokitansky or dermoid plugs): the Rokitansky nodule or papillae, is a raised protuberance with acoustic shadow, projecting into the cyst cavity. Most of the hair, teeth and bones typically arise from this protuberance (Figs 1 to 4 and 8).[10]

• The 'iceberg' sign:[7] This sign is very similar to the 'sound attenuation' one previously described. The author describes it as 'a mass with the amorphous, poorly defined echogenic focus in the near field that causes posterior shadow and thus obscures the posterior portion of the lesion and any structures behind it'. Because these masses are frequently large, the tip of the iceberg sign is identified on both transabdominal and transvaginal US scans.
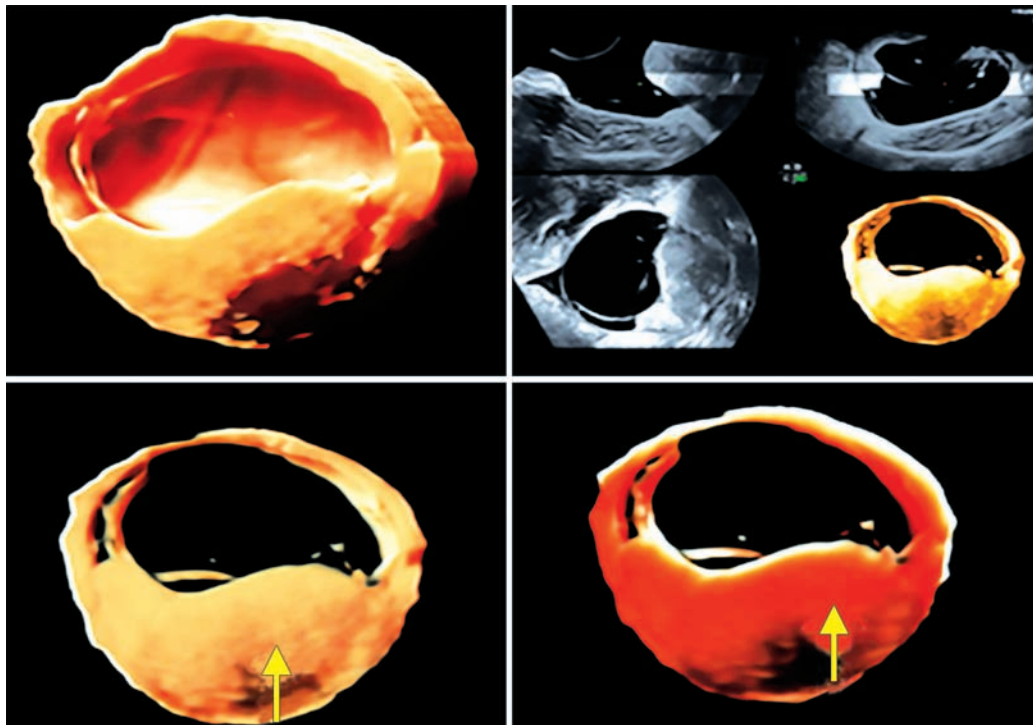
Fig. 3: Pure cystic teratoma with sebaceous tissue in its lower part (arrow)
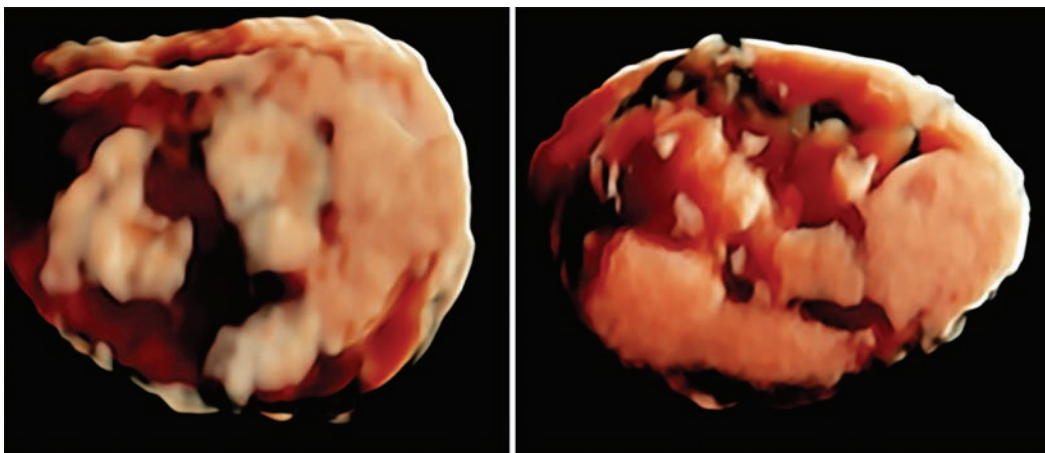

Fig. 4: Multicystic dermoid lesion visualized with maximal luminescence

The echogenic focus appears using US as a solid mass, but it is actually a cyst that contains a mixture of fatty liquid (i.e. sebum) matted hair and cellular debris. The multiple tissue interfaces are responsible of the characteristic acoustic shadow.

The fat mixed with the hair strands is echogenic and often attenuates the ultrasound beam. In addition, *in vitro* scans of surgical specimens have proved that the reflective echo pattern is caused by the multiple tissue interfaces of the hair and sebum within the cyst mass. Acoustic shadow may totally obscure the back of a large, clinically palpable, mass; hence, the term used for describing this image 'tip of the iceberg sign.'

• *Fluid-Fluid/Fat-fluid level*: This type of levels within a cystic ovarian tumor may strongly suggest cystic teratoma and it has been considered pathognomonic.[8] Pure sebum inside the cyst may be hypoechoic or anechoic. Fluid-Fluid levels, or fat-fluid interface, result from sebum floating above aqueous, which appears more echogenic than the sebum layer.

There is a constant horizontal fluid level inside the mass that changes with the patient's position. Associated with the fluid level is usually a dense echogenic structure with faint acoustic shadow behind it (Fig. 8).

• *Dermoid mesh*[9]: Dermoid mesh, with hyperechoic calcifications, indicates the presence of bone, teeth,
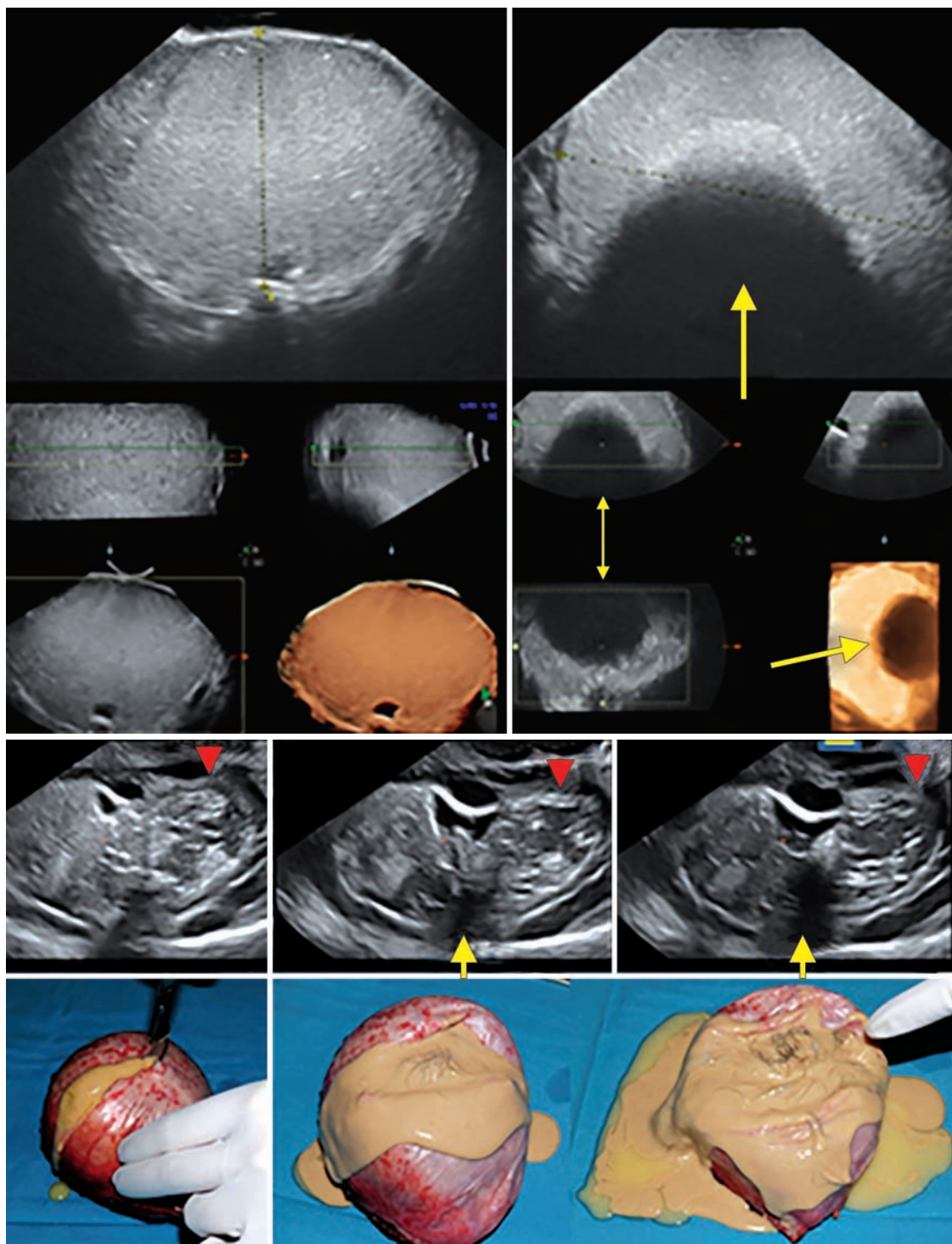
Fig. 5: Tomographic ultrasound image (TUI) of a diffuse echogenic gray mass with the echogenic area showing attenuation due to sebaceous material and hair within the cyst cavity (yellow arrows). At the right (images showed in the second and third TUI lines), sebaceous and hair material within the cystic cavity produce multiple thin echogenic bands (red arrows). Below, macroscopic view with sebum and hairs

or other ectodermal structures. They usually are in a predominantly cystic medium. Hyperechoic solid mural components and hair-fluid levels represent multiple echogenic linear interfaces floating inside a cyst; all these interfaces represent hair fibers (Figs 9 and 10).

- *Dermoid plugs*: A vast hyperechogenic area with multiple bright linear echoes and spots.[11]

- *Multiple mobile spherical structures* (fat balls) of slightly increased echogenicity floating free in a large cystic mass is one of the rarest patterns (Figs 11 and 12). In all cases, some degree of mobility was noticed when pressure was applied via abdominal. In some cases, as an exception, there was only one big ball between 4 and 7 cm.[2,13,14]
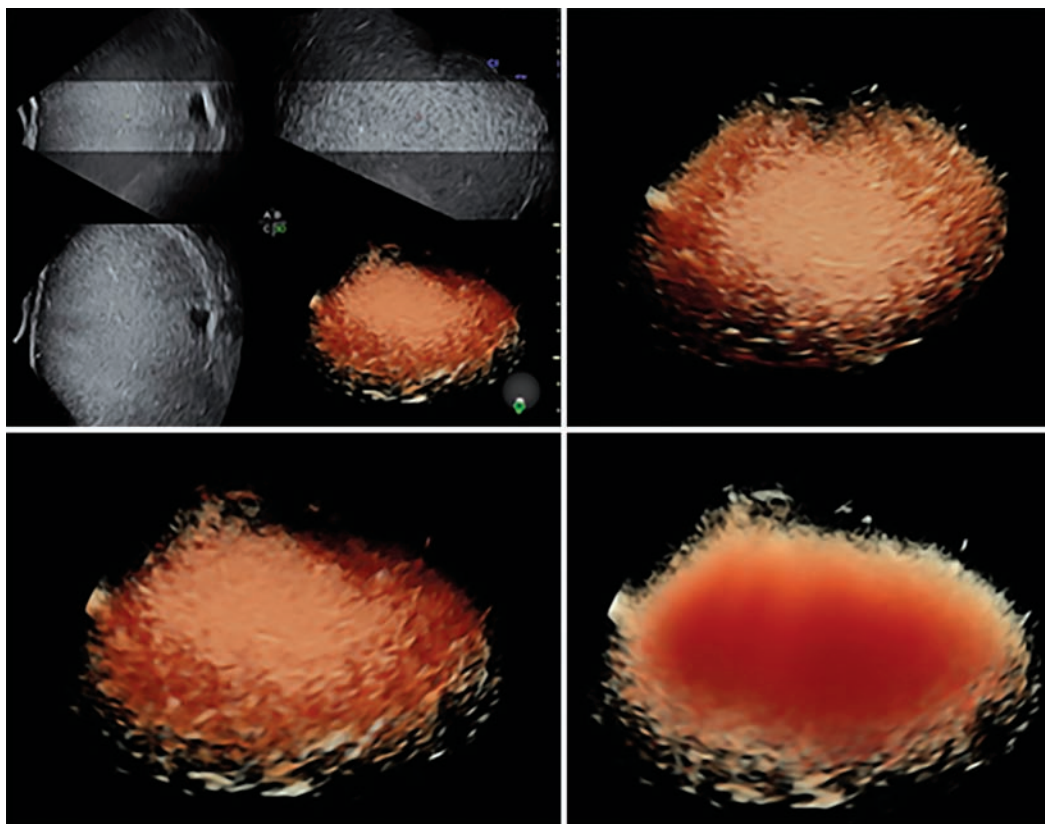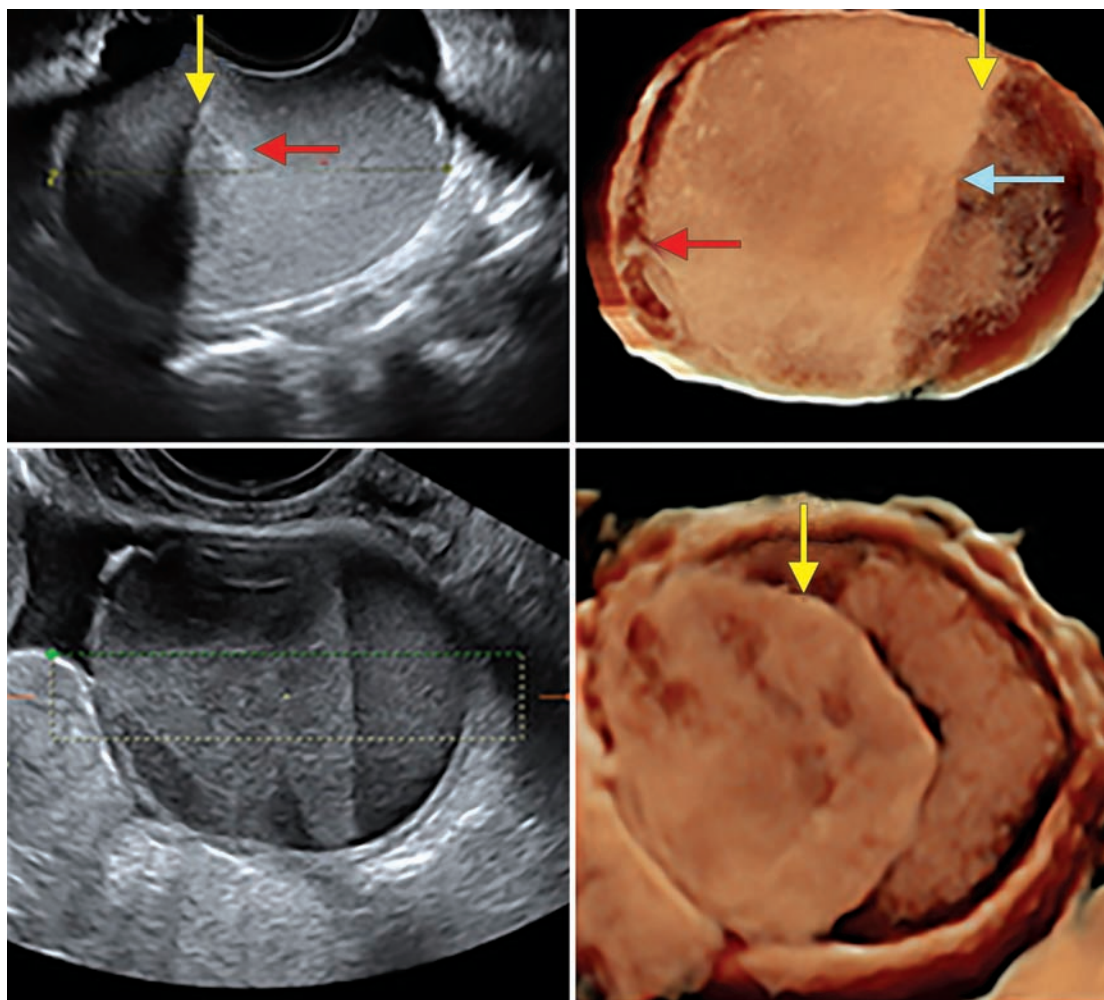
Fig. 6: 2D orthogonal planes a 3D HDlive of a diffuse echogenic gray mass. The echogenic area shows a small cyst and attenuation due to sebaceous material. The teratoma has been delimited using the 'cut off system'
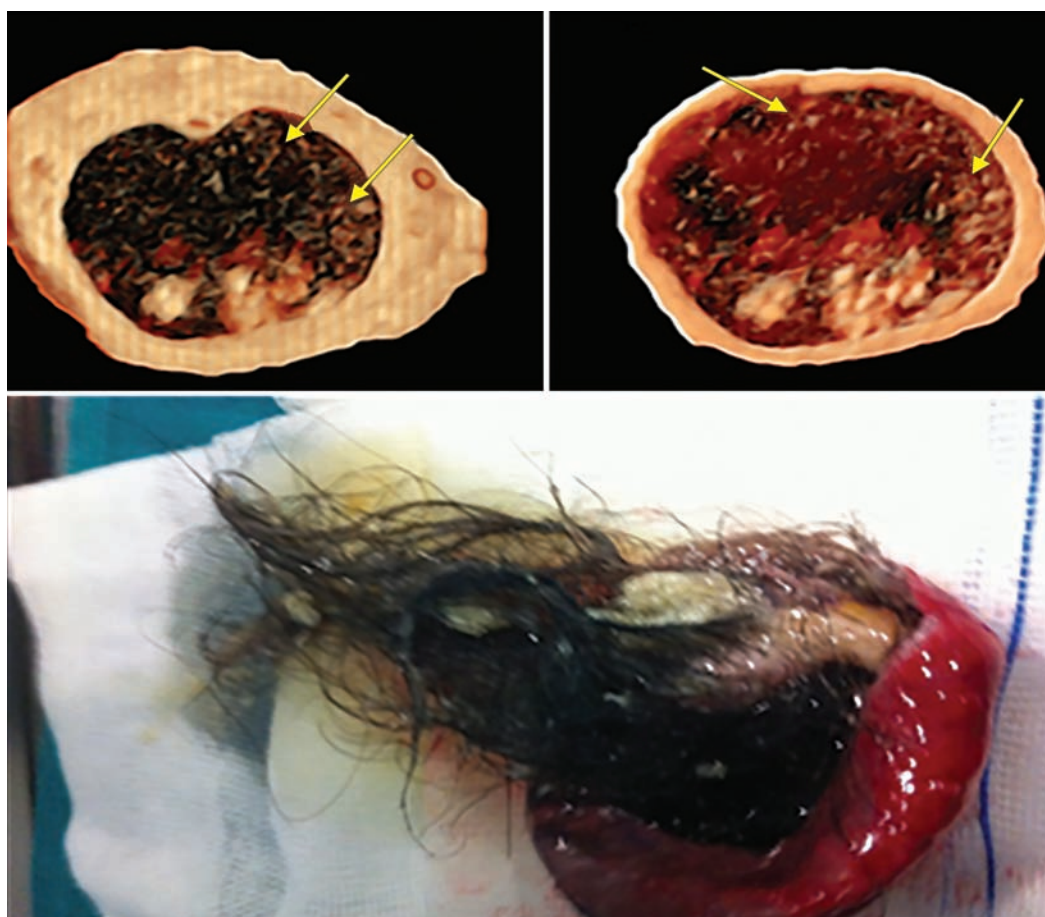
Fig. 7: The use of 3D HDlive allows us to visualize multiple thin echogenic dots and bands caused by hair in the cyst cavity (arrows)

Fig. 8: Two dermoid tumors with fluid-fluid Level (yellow and blue arrows).
One Rokitansky nodule or dermoid plugs (red arrows)

CONCLUSION

The following tissues are considered as pathognomonic of ovarian teratomas (they usually indicate a benign behave, but sometimes they have a malignant one):

- Bones, teeth and catilagenous tissue (*see* Figs 1, 2 and 11)
- Sebaceous material with or without attenuation and hair.

DISCUSSION

This type of ovarian tumor was first described one century ago; in fact, many references in German, English and French medical literature describe this entity using curious names, for instance (all mentioned in their original language[14]: Boules de graisse; Butter Kugeln; Caviarlike bodies; Dermoid Kugeln; Epithelial balls; Erbsenartige Köper; Fatty concretions; Fett Kugeln; Floating balls; Inclusions; lipid globules; Pilllike bodies; Rounded balls; Sebum balls; Solid concretions and Spherules.

The last clinical case report was published in 1935,[15] and so, the radiologic and ecographic descriptions start

thereafter. The first case using CT and MR was reported in 1991[16] and several ultrasonographic images have been published thereafter.[4,6,11-14,17-29]

The most common denomination in medical literature is 'intracystic fat balls' even though fat tissue is only a minimal component of the tumor. The microscopic sometimes shows spherules that contain desquamate keratin, fibrin, hemosiderin, and sebaceous debris with squamous skin and fine hair shafts but only a small amount of a fat component.

The vast majority of authors, including our own group, consider ultrasonographic images as pathognomonic of this condition.[6,11,12,14,23,30] Only pelvis affection by equino-coccus—an exceptional condition—may be included in the differential diagnosis. In this situation, the vesicles are seen as hypoechogenic images,[23,25,26,29] opposite to the hyperechogenic pattern seen in teratoma cases.[30]

All the described images are shown as 2D US, but are adaptable to 3D. There is only one 3D US description showing wonderful images[12] and this is the first one using HDlive.
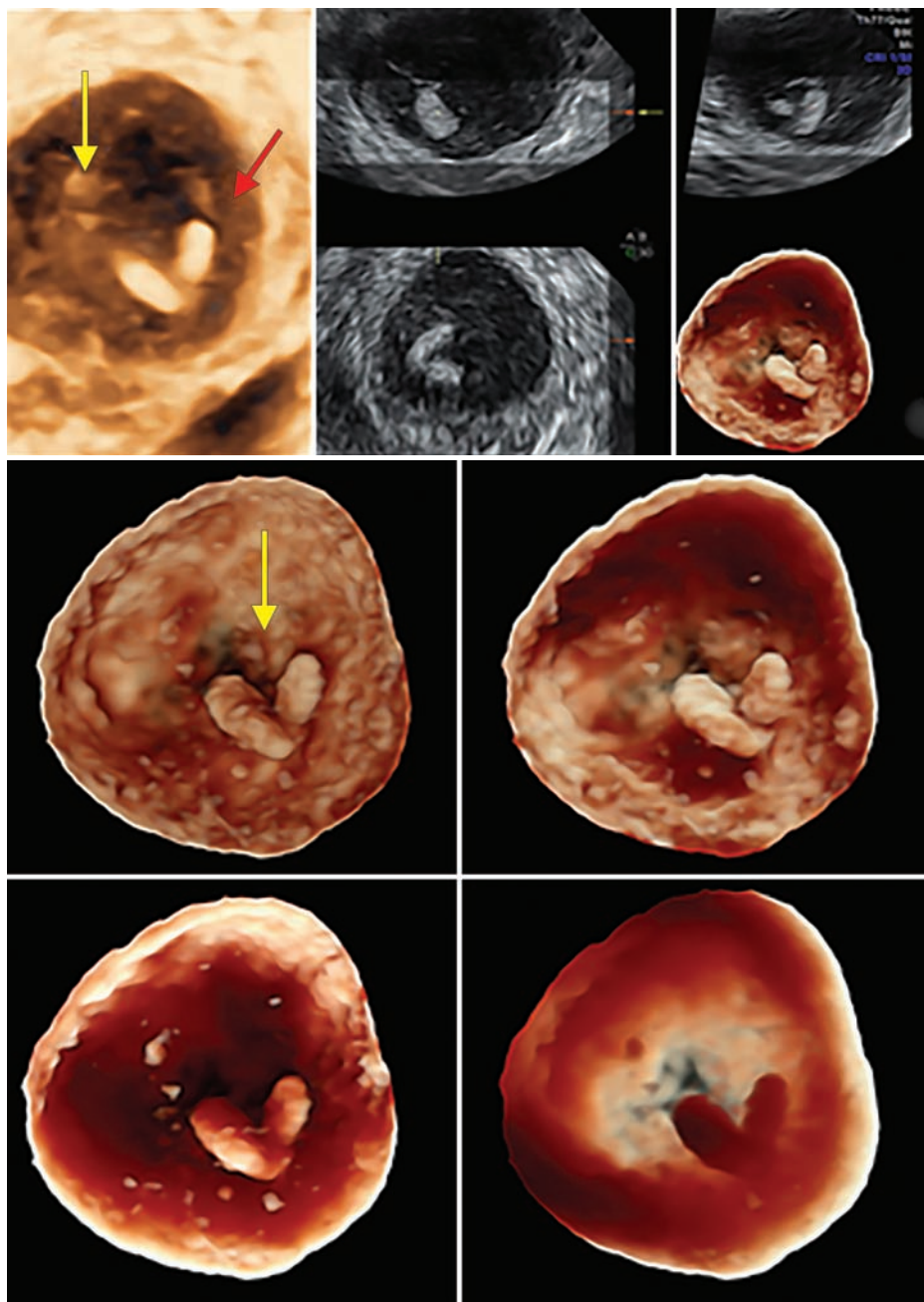
Fig. 9: Dermoid mesh with two hiperechoic calcifications indicating the presence of bone and teeth (red arrow) and the mesh (yellow arrow), characterized by multiple small echogenic linear interfaces floating inside the cyst (hair fibers)
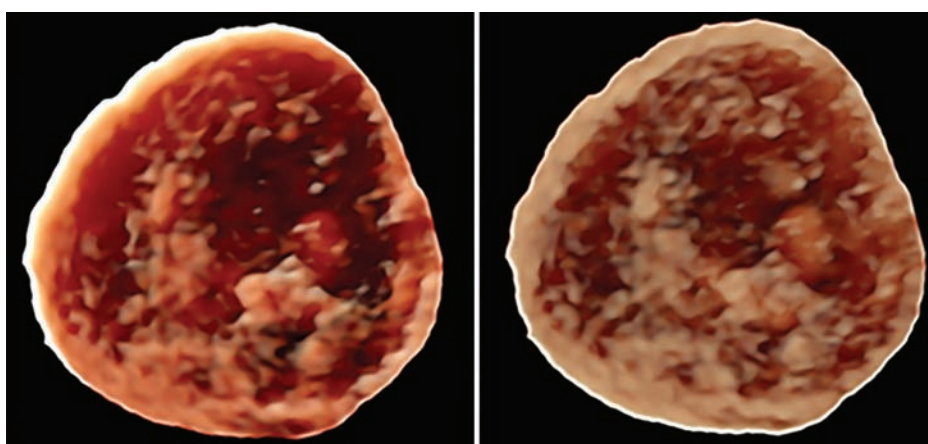
Fig. 10: Multiple small echogenic linear interfaces (mesh) and 'plugs' floating inside the cyst. The small linear dots are hair fibers
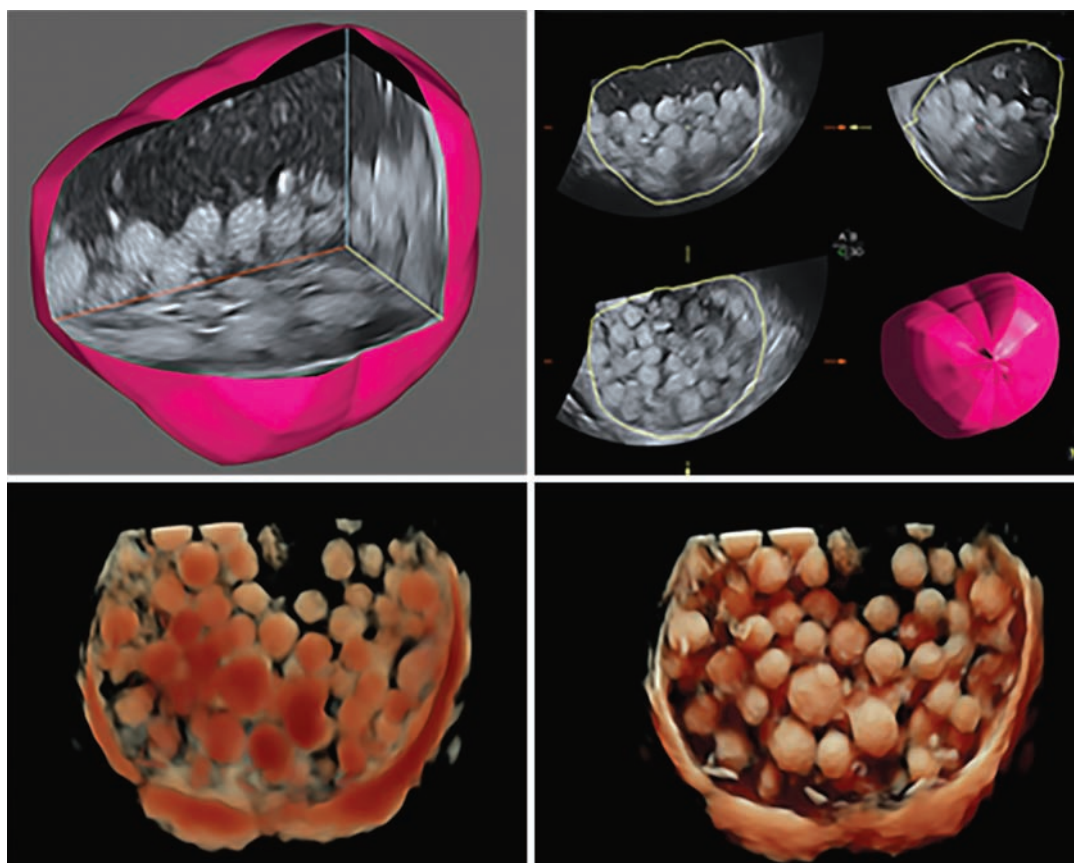
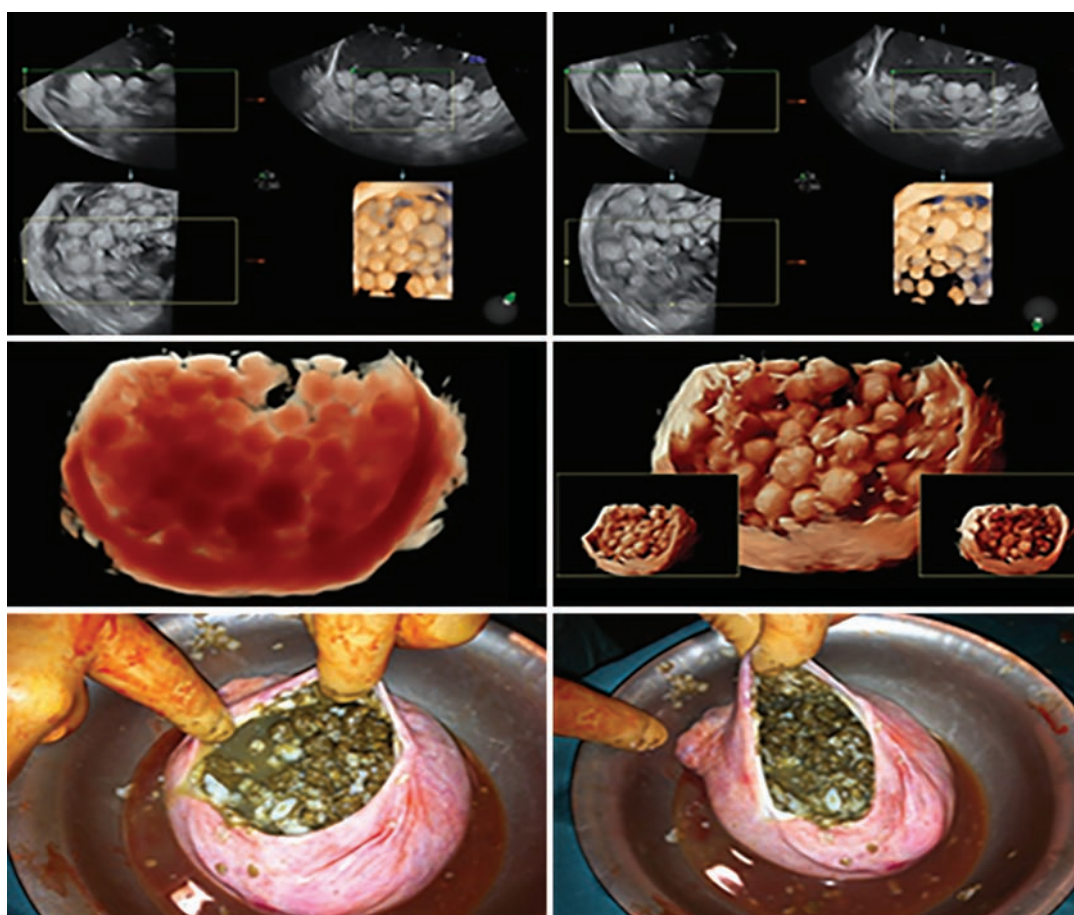**Fig. 11:** Fat balls in 2D shell mode and HDlive with and without maximal luminescence

Fig. 12: 2D, orthogonal planes, shell mode and HDlive with and without maximal luminescence of fat balls in a mature cystic teratoma

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