

Diagnostic steps in adenomyosis – an updated review

Corina Grigoriu^{1,2}, Roxana Elena Bohiltea^{1,3}, Tiberiu-Augustin Georgescu^{4,5}, Lucica Elena Eddan-Visan², Gina Calinescu², Nicolae Bacalbasa¹, Irina Balescu⁶, Maria Irina Vladareanu⁷, Andra Magdalena Balan²

¹Department of Gynecology and Obstetrics, “Carol Davila” University of Medicine and Pharmacy, Bucharest, Romania

²Department of Gynecology and Obstetrics, Emergency University Hospital, Bucharest, Romania

³Department of Gynecology and Obstetrics, Filantropia Clinical Hospital, Bucharest, Romania

⁴Department of Pathology, “Carol Davila” University of Medicine and Pharmacy, Bucharest, Romania

⁵“Alessandrescu-Rusescu” National Institute for Mother and Child Health, Bucharest, Romania

⁶Department of Visceral Surgery, Ponderas Academic Hospital, Bucharest, Romania

⁷Faculty of Medicine, “Carol Davila” University of Medicine and Pharmacy, Bucharest, Romania

ABSTRACT

Adenomyosis is defined by the presence of ectopic endometrial glands and stroma inside the myometrium. Definite diagnosis is made only on hysterectomy specimens, but all the progress in modern imaging techniques has increased the paraclinical diagnostic chances. The diagnostic algorithm is based on patient’s history (main complaints, menstrual, obstetrical and gynecological history), clinical examination, imaging studies – ultrasound and MRI. Definitive diagnosis is certainly histopathological. We present an updated review of diagnostic steps in adenomyosis, based on the existing consensus opinions of experts in the field. The Morphological Uterus Sonographic Assessment (MUSA) group published a consensus statement on terms, definitions and measurements that helps clinicians to characterize uterine pathology, accordingly to the modern classification system for abnormal uterine bleeding implemented by the International Federation of Gynecology and Obstetrics (FIGO). The direct signs of adenomyosis are: the myometrial cysts, the non-homogeneous myometrium (linear hyperchogenic striae inside the myometrium (the venetian blinds sign), the fan shaped shadowing, hypoechogenic subendometrial zones – lines and buds, hyperechogenic islands, hypoechogenic ill-defined pseudonodularities, ill-defined endometrial-myometrial delimitation as irregular or thick junctional zone. The indirect signs of adenomyosis are: globular, enlarged uterus; asymmetrical myometrial walls; linear, translesional vascularization; a retroverted uterus, with a characteristic appearance of the endometrium (the question mark sign). Adenomyosis has nowadays good criteria of non-invasive diagnosis and the appropriate diagnosis algorithm should be applied in all women with complaints of bleeding. Ultrasound imaging requires no special preparation, has no contraindication, is available and cost-effective. Ultrasound accuracy is very good in diagnosing adenomyosis, leading to prompt treatment in younger patients, who are especially interested in conserving their reproductive function.

Keywords: adenomyosis, diagnostic criteria, ultrasound, MRI

INTRODUCTION

Adenomyosis (from Greek: adenos = glands, myos = muscle) assume the presence of ectopic endometrial glands and stroma inside the endometrium. It is a common condition, but although it was first described in 1860 by Professor Rokitansky in Vienna (as „cystosarcoma adenoidis”), it is still poorly understood. Final positive diagnosis is made only on hysterectomy specimens, but all the pro-

gress in modern imaging techniques has increased the paraclinical diagnostic chances. Etiology and pathophysiology are still not entirely understood, but nowadays management overpasses the past dogma of hysterectomy as the unique treatment modality (1).

We present an updated review of diagnostic steps in adenomyosis, based on the existing consensus opinions of experts in the field. The Morpholog-

Corresponding author:

Roxana Elena Bohiltea

E-mail: dr.bohiltea@gmail.com

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ical Uterus Sonographic Assessment (MUSA) group published in 2015 (2) a consensus statement on terms, definitions and measurements that helps clinicians in the best possible way to characterize uterine pathology, accordingly to the modern classification system for abnormal uterine bleeding implemented by the International Federation of Gynecology and Obstetrics (FIGO). This system, now well known and constantly used, PALM-COEIN, classifies the causes of abnormal uterine bleeding as follows: polyp, adenomyosis, leiomyoma, malignancy and hyperplasia, coagulopathy, ovulatory dysfunction, endometrial causes, iatrogenic and not yet classified causes (3). In 2019, the MUSA group published another paper, proposing an uniform reporting system for ultrasound examination of adenomyosis.

DIAGNOSTIC ALGORITHM

The diagnostic algorithm is based on patient's history (main complaints, menstrual, obstetrical and gynecological history), clinical examination, imaging studies – ultrasound and MRI. Definitive diagnosis is certainly histopathological (4).

History

Patients with adenomyosis present with a major complaint, which is exaggerate vaginal bleeding – menorrhagia, metrorrhagia or menometrorrhagia. They also may present with dysmenorrhea, profound dyspareunia or chronic pelvic pain. Often the patients describe primary dysmenorrhea, combined with a progressive increase of blood loss during the menstrual period (1). Younger patients (around 30 years of age) may present with a history of infertility or unsuccessful attempts of assisted reproductive treatments. Older patients (older than 40 years) have a history of multiple abortions or birth and uterine surgeries. Associations of other uterine pathology - endometrial polyps, endometrial hyperplasia or previous diagnosis of uterine fibroma are common (5).

Clinical examination

A thorough clinical examination will frequently observe a globular, sometimes enlarged and tender uterus. Some patients may have associated endometriosis, with more or less infiltrating lesions and consequent symptoms and signs.

Imaging

Progresses in imaging techniques, especially in magnetic resonance imaging (MRI), permitted several years ago a better characterization and understanding of adenomyosis in regard of the junctional zone (JZ). Ultrasonography also developed tremen-

dously, is handier and more cost-effective and so we have now valuable tools in the presumptive diagnosis of adenomyosis (6,7).

We examine the uterus transvaginally, but also transabdominally (if the structures are beyond the small pelvis). Imaging in grey-scale provides most information, but color Doppler and three-dimensional ultrasound (adding coronal sections) may also be successful used. The ultrasound image is a reflection of the pathological alterations in myometrial structure (5). Ultrasound exam should be done in an orderly, rigorous manner, following to describe important parameters.

Uterine measurement

Uterine measurement is describing the shape and external contour. It measures the uterine corpus in a sagittal plane. The uterine corpus length is calculated as the sum of the fundal length and the endometrial cavity length. The fundal length is measured from the fundal surface to the fundal tip of the endometrium. The endometrial cavity length is measured from the fundal tip of the endometrium to the cervical internal os. The serosae contour may be described as regular or lobulated (8). The uterine anterior and posterior uterine walls should be measured also in a sagittal plane, from the serosa to the external endometrial outline. The measurement should be perpendicular on the endometrium. It should include the junctional zone, but not the endometrium. The asymmetry of the myometrial anterior and posterior walls has to be noted. The overall echogenicity has to be evaluated: homogeneous or heterogeneous. The transverse diameter of the uterus and myometrium should be also measured (9).

Description and reporting the junctional zone JZ

The JZ is a functional layer situated between the endometrium and the myometrium, less than 5 mm thick. It has a role in the uterine peristalsis and is hormone-dependent. The junctional zone is best evaluated with 3D ultrasound, because we can assess a complete examination in sagittal, transverse and coronal plane. Two-dimensional evaluation is also possible (10). The JZ is described as follows: regular or irregular, interrupted, not visible or not assessable (Figure 1). The maximum and minimum thickness of the JS has to be noted, and also the location (fundus, anterior, posterior, lateral right or lateral left, or global). The irregularities in the junctional zone are pathognomic for adenomyosis; they are considered as the sign of endometrial invasion in the myometrium (Figures 2,3). The irregularities in the JZ are: cystic areas; hyperechogenic dots; hyperechogenic buds and lines. The extend of the irregularities of the JZ may be subjectively reported, meaning more or less then 50% of the entire JZ (2).

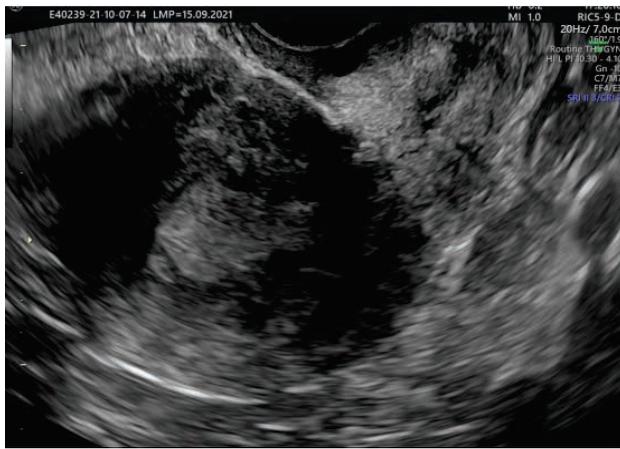


FIGURE 1. Two-dimensional ultrasound image of the longitudinal rotated plane of uterus; the irregular JZ can be noticed as an invasion of the endometrium into myometrium. The both uterine wall presents dispersed hyperechoic buds and lines (personal collection of Roxana Bohîlțea)

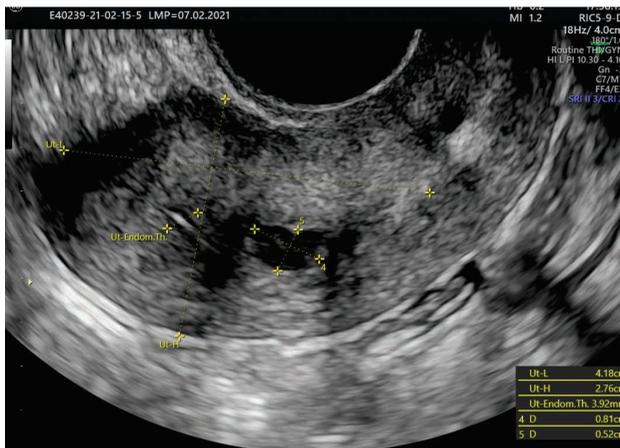


FIGURE 2. Two-dimensional ultrasound image of the uterine longitudinal plane showing a hypoechoic cyst of 8,1/5,2mm that interrupt JZ (personal collection of Roxana Bohîlțea)



FIGURE 3. The subendometrial cyst is better evidenced on 3D static ultrasound image; the interruption of the JZ can be noticed (personal collection of Roxana Bohîlțea)

Description of the myometrium

Myometrial pathology may be localized or diffuse, well- or ill-defined (Figure 4). For each lesion

we should note as follows: location, dimensions, site, echogenicity, shadowing, presence of cysts, hyperechoic islands, subendometrial echogenic lines and buds (11). Location may be anterior, posterior, fundal right or left lateral or global. Dimensions are measured in three perpendicular diameters; volume should be calculated as the product of the three dimensions. Site may be (according to the FIGO classification of myomas) submucosal (with less or more than 50% of myometrium involved), intramural (100%, with or without contact with the endometrium), subserosal (more or less 50% intramural). Outer lesion-free margin (the minimum distance between serosa and the outer border of the lesion) and inner lesion-free margin (the minimum distance between endometrium and inner border of the lesion) should also be measured and reported (5,9). The extension of ill-defined lesions should be appreciated: localized (under 50% of total uterine volume involved) or diffuse (over 50%). Echogenicity of the myometrial cysts should be described as follows: uniform (hypo, iso, hyperechoic) or non-uniform: mixed echogenicity, cystic areas (regular or irregular). The cystic myometrial lesions may be anechoic, of low-level echogenicity, ground glass appearance or of mixed echogenicity (Figures 5-7). The lesion rim may be hypo- or hyperechoic or ill-defined. The lesion shape may be round or oval, lobulated or irregular. Shadowing should be examined: if it is arising from the lesions' edge (present or absent), or if it arises from inside the lesion (internal shadowing). The alternance of hypoechoic and hyperechoic stripes, called "fan shaped shadowing", is an important sign suggesting adenomyosis and should be reported (Figure 4). Degree of shadowing should also be evaluated (slight, moderate or strong). The presence of hyperechoic islands should be reported (with reporting of outline – regular, irregular or ill-defined; size; and number). The presence of subendometrial echogenic lines and buds should be reported. They disrupt the JZ and are perpendicular to the endometrium (12).

Figure 4. The asymmetry between the myometrial anterior and posterior walls is evident. The ill-defined diffuse lesion of the anterior wall affects cranially the vacuity line of the endometrium. The linear hyperchogenic striae inside of the myometrium determine the moderate fan shaped shadowing (personal collection of Roxana Bohîlțea)

Description of the myometrial vascularization and of the myometrial lesions vascularization

Uterine and myometrial vascularization are studied using both color and power Doppler. The myometrial vascularization may be uniform or non-uniform. The vascularization of the myometri-



FIGURE 4. The asymmetry between the myometrial anterior and posterior walls is evident. The ill-defined diffuse lesion of the anterior wall affects cranially the vacuity line of the endometrium. The linear hyperchogenic striae inside of the myometrium determine the moderate fan shaped shadowing (personal collection of Roxana Bohîlțea)

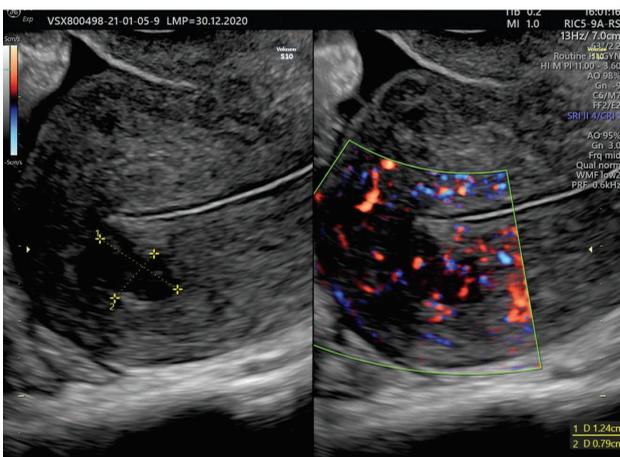


FIGURE 5. The myometrial hypoechoic nodule that has the appearance of a leiomyoma proves to manifest translesional vascularization on color Doppler (personal collection of Roxana Bohîlțea)



al lesions may be circumferential, intralesional or combined. A important sign is the translesional vascularity (vessels perpendicular to the cavity and serosa, crossing the lesion) (Figures 5,6). The amount of color in the lesion should be reported. The vessel morphology should also be reported (large and equal, small and equal, unequal, branching type (regular, irregular or no branching) and direction (perpendicular or not) (13,14).

Adenomyosis may be localized (focal adenomyosis), scattered (diffuse adenomyosis), with cystic transformation (cystic adenomyoma). If the focal adenomyosis is surrounded by hypertrophic myometrium it becomes an adenomyoma (15). The ultrasound appearance depends on the variable proportion between endometrial glands, endometrial stroma and the hypertrophic muscular structures inside the lesions (3).

Ultrasound signs of adenomyosis may be classified in direct and indirect.

The direct signs of adenomyosis are: the myometrial cysts; the non-homogeneous myometrium (linear hyperchogenic striae inside the myometrium (the venetian blinds sign), the fan shaped shadowing, hypoechoic subendometrial zones -lines and buds, hyperechogenic islands, hypoechoic ill-defined pseudonodularities; an ill-defined endometrial-myometrial delimitation, a irregular or thick junctional zone (1,16).

The indirect signs of adenomyosis are: a globular, enlarged uterus, asymmetrical myometrial walls, the linear, translesional vascularization, a retroverted uterus, with a characteristic appearance of the endometrium (the question mark sign) (2,15).

MRI is still considered to be the best tool in diagnosing adenomyosis, because it differentiates other myometrial lesions (leiomyoma) and gives a very good evaluation of the JZ (17).

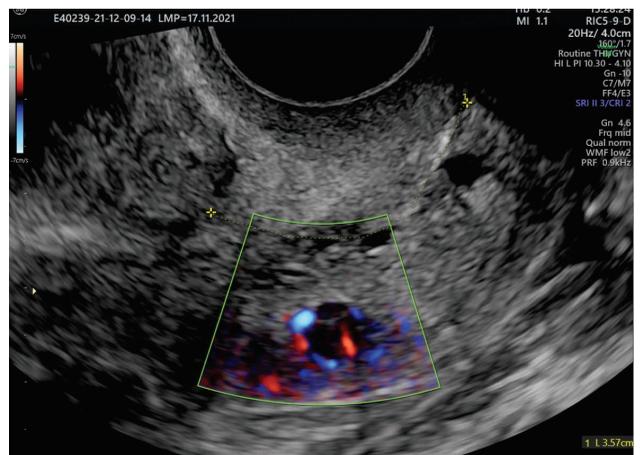


FIGURE 6. The vessels, easily visible on color Doppler, cross the anechoic lesion situated at the isthmus level (personal collection of Roxana Bohîlțea)



FIGURE 7. Myometrial hypoechoic lacunae with irregular contour and largest diameter 9.4 mm (personal collection of Roxana Bohilțea)

The most important MRI finding is thickening of the JS above 12 mm (pathognomonic for adenomyosis). A thickness less than 8 mm excludes adenomyosis. The islands of endometrial glands inside the myometrium are seen as bright foci or as linear striations on T2-weighted images (18). Adenomyoma appears as hypointense masses on T2-weighted images. Margins are ill-defined, with minimal mass effect. The smooth muscle hypertrophy surrounding the adenomyomatous lesions, appears as a low-signal area on T2-weighted images (6,14).

DIAGNOSING REPORT ON ADENOMYOSIS

The specialists' panel from the MUSA group returned 2019 by publishing a paper on more accurate reporting system of adenomyosis, using ultrasound as single diagnostic procedure. They propose seven steps in developing a diagnosing report on adenomyosis:

1. Documentation of the presence of adenomyosis (with the characteristics discussed above)
2. Description of location;
3. Differentiation of diffuse or focal adenomyosis/ adenomyoma or mixed (both diffuse and mixt);
4. Presence of cystic alterations (the presence of a myometrial cyst larger then 2 mm);
5. The involvement of the uterine layers (junctional zone, myometrium and serosa);
6. Assessment of the extent (proportion of uterus affected by adenomyosis), permitting the classification of adenomyosis: mild (under 25% affected), moderate (25-50% affected) and severe (more then 50% affected).
7. Size of lesions (measurement of the largest diameter of lesion).

In a systematic diagnostic approach, using grey scale US, but also 3D US, color and power Doppler, specialists are able to provide an appropriate characterization of the uterus, appreciating disease extent and as such giving a more accurate perspective of therapeutical management possibilities.

CONCLUSIONS

Adenomyosis has nowadays good criteria of non-invasive diagnosis and the appropriate diagnosis algorithm should be applied in all women with complaints of bleeding. Ultrasound imaging requires no special preparation, has no contraindication, is available and cost-effective. Ultrasound accuracy is very good in diagnosing adenomyosis, leading to prompt treatment in younger patients, who are especially interested in conserving their reproductive function.

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