ESGO

Pocket Guidelines

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Uterine Sarcomas

THE EUROPEAN VOICE OF GYNAECOLOGICAL ONCOLOGY

ESGO
European Society of Gynaecological Oncology

POCKET GUIDELINES

Based on

ESGO-EURACAN-GCIG guidelines for the management of patients with uterine sarcomas

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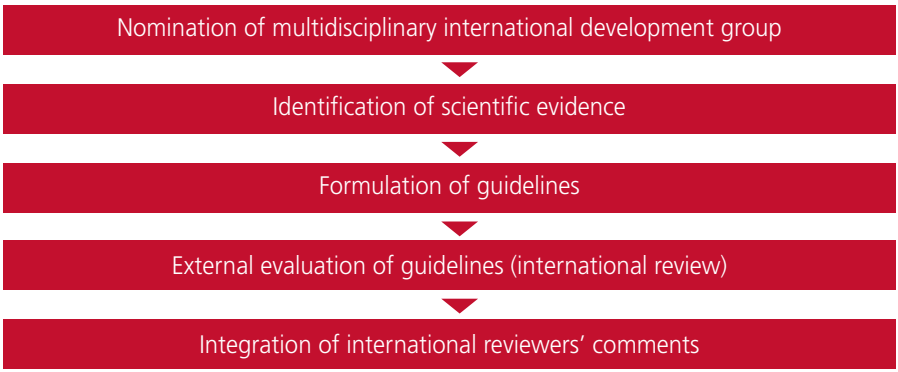
In 2014, the Gynecologic Cancer InterGroup (GCIG) published consensus reviews and recommendations for the management of a number of these rare uterine sarcomas. Given the advances and new evidence that have emerged over the last 10 years which have impacted the management of patients with uterine sarcomas, a collaboration was established between the European Society of Gynaecological Oncology (ESGO), the European Reference Network on Rare Adult Solid Cancers (EURACAN), and GCIG with the specific objective of developing clinically relevant and evidence-based contemporary guidelines to guide the multidisciplinary approach for management of patients with uterine sarcomas from initial diagnosis to relapse.

Attention was given to imaging, pathology, and molecular analyses in addition to clinical management. Guidelines for surgery including specific recommendations at first diagnosis and at relapse for all histological subtypes of uterine sarcomas were developed. Indications for radiation and systemic therapies including chemotherapy options, endocrine therapies, and targeted therapies for the following histological subgroups were developed for: uterine leiomyosarcoma (uLMS), high-grade endometrial stromal sarcoma (HG-ESS), undifferentiated uterine sarcomas (UUS), low-grade endometrial stromal sarcoma (LG-ESS), adenosarcoma, and selected very rare entities. Recommendations for follow-up after treatment are also provided.

The guidelines were developed using a five-step process as defined by the ESGO Guideline Committee:



The objectives of these ESGO/EURACAN/GCIG guidelines are to improve the quality of care for patients with uterine sarcomas across Europe and worldwide. They are intended for use by all health professionals involved in the management of patients with uterine sarcomas across all allied disciplines.

These guidelines do not include any economic analysis of the strategies. Any clinician seeking to apply or consult these guidelines is expected to use independent medical judgment in the context of individual clinical circumstances to determine any patient's care or treatment.

To ensure that the statements were evidence based, the current literature was reviewed and critically appraised. A systematic, unbiased literature review of relevant studies published between April 2013 and April 2023 was carried out.

The guidelines were adopted if they were supported by sufficient high level of scientific evidence and/or when a large consensus among experts was obtained. An adapted version of the "Infectious Diseases Society of America-United States Public Health Service Grading System" was used to define the level of evidence and grade of recommendation for each of the recommendations:

LEVELS OF EVIDENCE

I Evidence from at least one large randomised, controlled trial of good methodological quality (low potential for bias) or meta-analyses of well conducted, randomised trials without heterogeneity

II Small randomised trials or large randomised trials with a suspicion of bias (lower methodological quality) or meta-analyses of such trials or of trials with demonstrated heterogeneity

III Prospective cohort studies

IV Retrospective cohort studies or case-control studies

V Studies without control group, case reports, expert opinions

GRADES OF RECOMMENDATIONS

A Strong evidence for efficacy with a substantial clinical benefit, strongly recommended

B Strong or moderate evidence for efficacy but with a limited clinical benefit, generally recommended

C Insufficient evidence for efficacy or benefit does not outweigh the risk or the disadvantages (adverse events, costs…), optional

D Moderate evidence against efficacy or for adverse outcome, generally not recommended

E Strong evidence against efficacy or for adverse outcome, never recommended

ESGO would like to thank the members of the international development group for their constant availability, work, and for making possible the development of these guidelines for the management of patients with uterine sarcomas (see below). ESGO is also very grateful to the 104 international external reviewers for their participation (list available on the ESGO website).

NAME	SPECIALTY	AFFILIATION
Isabelle Ray-Coquard	Medical oncologist (chair)	Centre Léon Bérard, Lyon (France)
Paolo Giovanni Casali	Medical oncologist (chair)	Fondazione IRCCS Istituto Nazionale Tumori, Milan (Italy) University of Milan, Milan (Italy)
Michael Friedlander	Medical oncologist (chair)	School of clinical medicine, Faculty of medicine and health, Sydney (Australia), Prince of Wales and Royal Hospital for Women, Randwick (Australia)
François Planchamp	Methodologist	Institut Bergonié, Bordeaux (France)
Sabrina Croce	Pathologist	Institut Bergonié, Bordeaux (France)
Fiona M Fennessy	Radiologist	Brigham and Women's Hospital, Harvard Medical School, Boston (United States of America)
Daniela Fischerova	Gynecologic oncologist	First Faculty of Medicine, Charles University and General University Hospital in Prague, Prague (Czech Republic)
Robin L Jones	Medical oncologist	Royal Marsden Hospital and Institute of Cancer Research, London (United Kingdom)
Roberta Sanfilippo	Medical oncologist	Fondazione IRCCS Istituto Nazionale dei Tumori, Milan (Italy)
Ignacio Zapardiel	Gynecologic oncologist	La Paz University Hospital, Madrid (Spain)
Frédéric Amant	Gynecologic oncologist	KU Leuven, Leuven (Belgium), Antoni van Leeuwenhoek Nederlands Kanker Instituut, Amsterdam (The Netherlands)
Jean-Yves Blay	Medical oncologist	Centre Léon Bérard, Lyon (France)
Javier Martin-Broto	Medical oncologist	Fundación Jimenez Diaz University Hospital, Madrid (Spain) University Hospital General de Villalba, Madrid (Spain), Instituto de investigación Sanitaria Fundación Jimenez Diaz, Madrid (Spain)
Antonio Casado	Medical oncologist	University Hospital San Carlos, Madrid (Spain)
Sarah Chiang	Pathologist	Memorial Sloan Kettering Cancer Center, New York (United States of America)
Angelo Paolo Dei Tos	Pathologist	Treviso General Hospital Treviso, Padova (Italy) University of Padua School of Medicine, Padua (Italy)
Rick Haas	Radiation oncologist	The Netherlands Cancer Institute, Amsterdam (the Netherlands) Leiden University Medical center, Leiden (the Netherlands)
Martee L Hensley	Medical oncologist	Memorial Sloan Kettering Cancer Center, New York (United States of America), Weill Cornell Medical College, New York (United States of America)
Peter Hohenberger	Gynecologic oncologist	Mannheim University Medical Center, University of Heidelberg, Mannheim (Germany)
Jae-Weon Kim	Obstetrician & gynecologist	Seoul National University College of Medicine, Seoul (Republic of Korea)
Se Ik Kim	Obstetrician & gynecologist	Seoul National University College of Medicine, Seoul (Republic of Korea)
Mehmet Mutlu Meydanli	Gynecologic oncologist	Medical Point Hospital, Gaziantep (Turkey)
Patricia Pautier	Medical oncologist	Institut Gustave Roussy, Villejuif (France)
Albiruni R Abdul Razak	Medical oncologist	Princess Margaret Cancer Centre, Toronto (Canada) University of Toronto, Toronto (Canada)
Jalid Sehouli	Gynecologic oncologist	Charité-Universitätsmedizin, Berlin (Germany)
Winan van Houdt	Gynecologic oncologist	The Netherlands Cancer Institute, Amsterdam (the Netherlands)

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General recommendations

A	Centralisation of care in specialised centres and referral network is encouraged.
A	Treatment planning should be multidisciplinary (within a tumour board, composed according to local guidelines) and supported by all available evidence including an understanding and appreciation of prognostic and predictive factors, potential adverse effects of treatments, and quality of life.
A	Patients should be carefully counseled on the recommended management plan and potential alternatives, including risks and benefits of all options taking into full consideration their perspectives and wishes.
A	Treatment should be undertaken by an experienced team in the diagnosis and management of uterine sarcomas.
A	Enrollment of patients with uterine sarcomas in clinical trials should be considered if available.
B	International collaboration and prospective registries for this rare group of disease are encouraged.

Diagnosis - Pathology

General diagnosis

A	Pathological diagnosis should be confirmed by a pathologist with expertise and experience in the diagnosis of gynecologic mesenchymal tumours, preferably at a sarcoma reference centre.
C	Molecular testing may be required to confirm pathological diagnosis and to identify therapeutic targets.
B	According to the International Collaboration on Cancer Reporting (ICCR) data set, there is not a minimum recommended number of blocks.
B	Sampling should be based on a careful macroscopic examination and follow ICCR guidelines.
B	For challenging diagnoses additional/extensive sampling is necessary (e.g. differential diagnosis of sarcoma from carcinosarcoma, LG-ESS from HG-ESS, smooth muscle tumour of uncertain malignant potential (STUMP) from LMS).
B	For low-grade tumours, biopsy can be used for diagnosis only in advanced stage disease. For stage I, biopsy is challenging to establish diagnosis (e.g. differential diagnosis between endometrial stromal nodule and LG-ESS).

Immunohistochemistry

B

Immunohistochemistry is recommended to refine the diagnosis and inform therapeutic decisions, especially when molecular diagnostics are not available. However, many markers may not be specific or diagnostic alone and should be interpreted in conjunction with morphology.

B

A non-exhaustive list of diagnostic immunohistochemical markers includes:

- STUMP: desmin, h-caldesmon, smooth muscle actin
- Fumarate hydratase (FH) deficient smooth muscle tumours: FH and 2SC
- LMS: *ATRX, RB, PTEN, p53, DAXX, MTAP, MDM2*
- Endometrial stromal tumours: CD10, IFITM1, Cyclin D1, BCOR
- Perivascular epithelioid cell tumours (PEComa): HMB45, Melan A, TFE3
- neurotrophic tropomyosin-receptor kinase *(NTRK)*-rearranged sarcomas: Pan-TRK, CD34, S100
- p53 useful in distinguishing between complex genomic sarcomas (aberrant expression in LMS, UUS) and simple genomic sarcoma (wild-type expression fusion driven sarcomas such as LG-ESS, HG-ESS, NTRK-rearranged sarcoma)
- ER, PR, ALK, ROS-1, Pan-TRK

Ancillary techniques

B

Molecular tests are recommended to confirm fusion and/or mutation status for diagnosis and/or therapeutic decision making. Suspected therapeutic targets (e.g., NTRK, ALK, ROS fusions) require molecular confirmation. Assays may include FISH, RNA and/or DNA sequencing for fusion confirmation and DNA sequencing for mutation confirmation.

uLMS

B

The diagnosis is based on combination of morphology (2020 World Health organization and ICCR criteria) and immunohistochemistry (for smooth muscle differentiation).

HG-ESS

C

The diagnosis of HG-ESS is based on morphology (atypia, mitotic count and necrosis, frequent lymphovascular invasion) and immunohistochemistry (frequent Cyclin D1 and/or BCOR overexpression and/or altered ER/PR expression). This can be associated with a low-grade component.

B

Molecular analysis is indicated and encouraged to detect BCOR, YWHAE, or BCORL1 alterations.

UUS

B

Especially in UUS with uniform histology, immunohistochemistry and molecular studies (RNA sequencing) are indicated to exclude other tumour types.

LG-ESS

C

An endometrial stromal tumour with permeative (tongue-like) infiltration of the myometrium (>3 finger-like projections measuring >3 mm from tumour periphery) and/or lympho-vascular invasion is diagnostic of LG-ESS. Detection of LG-ESS associated fusions may be helpful.

Adenosarcoma

B

The diagnosis is based on morphology.

B

The following aggressive prognostic factors should be taken into account:

- Sarcomatous overgrowth
- High-grade component
- Lympho-vascular invasion
- Myometrial infiltration

PEComa

B

The diagnosis is made based on a combination of morphology (perivascular epithelioid cells) and melanocytic markers (HMB45, Melan A).

NTRK

B

Confirmation of *NTRK* fusion status by molecular methods is essential given pan-Trk expression in other uterine sarcoma subtypes (HG-ESS and uLMS).

Principles of pre-/post-operative imaging

General recommendations

B — In cases with atypical uterine fibroid findings on baseline pelvic ultrasound scans, the patient should be referred to magnetic resonance imaging (MRI) or specialised ultrasound.

A — The interpretation should be performed by subspecialised radiologists evaluating specific MRI features.

A — Specialised ultrasound examination should preferably be performed in a specialised cancer centre by an experienced sonographer fully dedicated to the imaging of gynecological cancers.

Tailoring surgery in patients with symptoms, high-risk factors, fertility needs, suspicion on ultrasound or pelvic MRI

B — Pre-operative pelvic MRI (preferably), or transvaginal/transabdominal ultrasound performed only by an expert sonographer at a highly specialised site to assess the mass and to determine if features associated with higher likelihood of sarcoma are present, is recommended.

B — In patients with suspicious imaging features in whom hysterectomy is not immediately feasible, pre-operative imaging-guided biopsy is an option in a specialised cancer centre by an experienced sonographer or interventional radiologist, while also making the patient aware that false negatives may exist with this pathway.

B — Pre-operative ultrasound guided trans-uterine cavity (in-organ) core-needle biopsy (≥14-16G) of the most suspicious lesion should be performed with expert pathologic review using microscopic and molecular analysis as needed.

C — Percutaneous biopsy using coaxial needle (for less accessible masses) may be an option but should be used with caution in a specialised centre.

Patients incompletely resected with malignant uterine smooth muscle tumours and patients with incidental findings of malignant uterine smooth muscle tumours after hysterectomy

B — Chest/abdomen/pelvis contrast-enhanced computed tomography scan or abdominal/pelvic MRI plus chest computed tomography scan are recommended to evaluate for locoregional tumour extension, and for distant metastases.

B — Positron emission tomography with 2-deoxy-2-[fluorine-18] fluoro-D-glucose integrated with computed tomography (18F-FDG PET/CT) in specific case of indeterminate lesions can be considered.

Principles of surgery

General recommendations

B	An open approach should be the preferred route of surgery in most cases.
C	Only in cases when integrity of the uterus can be assured, minimally invasive techniques may be considered.
A	Morcellation should always be avoided in cases with preoperative suspicion of uterine sarcoma (e.g., rapid growing mass or suspicious appearances on MRI). Patients undergoing morcellation for an apparently benign condition must be counseled on the low risk of unsuspected sarcoma even in apparently benign lesion.

Early stage (FIGO I & II)

A	Complete removal of the intact uterus is the gold standard of surgical management.
A	Bilateral salpingo-oophorectomy (BSO) is the standard of care in postmenopausal women.
C	In premenopausal women with stage I disease, ovarian preservation with bilateral salpingectomy could be considered in selected cases regardless the histological subtype to avoid the need for postmenopausal endocrine therapy.
D	Routine systematic lymphadenectomy should not be performed.
B	Suspicious nodes or peritoneal lesions should be removed as well.

Advanced stage (FIGO III & IV)

A	For stage III, complete surgical removal of disease is the gold standard, including total hysterectomy and BSO and resection of any other suspicious lesions including peritoneal disease and/ or bulky/suspicious nodes.
A	For stage IV, the option for primary resection depends on the number and location(s) of metastases as well as the biology and histological subtype. Surgery should be considered in primary oligometastatic disease together with complete surgical resection of primary tumour if it is deemed feasible with acceptable morbidity.
B D	Lymphadenectomy should only be undertaken if the lymph nodes are grossly enlarged intraoperatively or suspicious for metastases on preoperative diagnostic work-up. There is no indication for routine systematic lymph node dissection.
B	In case of initially unresectable uterine sarcoma, primary systemic treatment is an option followed by re-evaluation for surgery depending on response.

Recurrent disease

A

All cases should be evaluated by a multidisciplinary team to evaluate if surgery is a reasonable and feasible option for all types of uterine sarcomas with the primary goal of complete resection.

B

Selection of the best candidates for surgery should be based on the following criteria:

- Tumour biology and histology
- Localisation of relapse, number of lesions, and tumour burden
- Recurrence-free interval (although there is no validated cut-off)
- Performance status
- Severity of comorbidities
- Patient perspectives
- Potential complications
- Prior treatment

B

In particular, BSO should be considered in premenopausal patient subgroups with low-grade uterine sarcoma (including LG-ESS and adenosarcoma without sarcomatous overgrowth): when the ovaries are *in situ*, to decrease endocrine stimulation.

C

In indolent uterine sarcomas (i.e., LG-ESS, low-grade adenosarcoma without sarcomatous overgrowth, and selected low grade LMS), surgical resection may be a reasonable option in cases with second or third recurrences.

C

Re-laparotomy for recurrence in high-grade uterine sarcoma and adenosarcoma with sarcomatous overgrowth may be a valid option if disseminated disease is not present.

Special situations

B

In case of unexpected sarcoma diagnosis after myomectomy, a hysterectomy should be performed. If extrauterine disease or residual disease is suspected and surgically resectable, complete resection is recommended along with the total hysterectomy.

C

Resection of a uterine sarcoma with preservation of the uterus is a non-standard approach and could only be considered in referral centres for highly-selected cases of LG-ESS and low-grade adenosarcoma without sarcomatous overgrowth with informed consent.

B

Ovarian preservation should be considered in stage I sarcomas where hormonal and gametes preservation is desired.

Uterine leiomyosarcoma

Localised disease (FIGO stage I) - Post-operative systemic therapy

B — Standard approach for uterine confined (non-morcellated) FIGO stage I is surveillance following surgery.

B — Due to conflicting data on the benefit of post-operative chemotherapy and the high risk of relapse, inclusion of patients in randomised controlled trials is recommended.

C — Post-operative chemotherapy is not a standard treatment and may be discussed with a patient within a shared decision-making process due to the uncertainty of benefit.

Localised disease (FIGO stage I) - Role of radiotherapy

D — Post-operative radiotherapy is not the standard of care after hysterectomy.

Locally advanced disease (FIGO stages II-III) - Role of chemotherapy

B — Post-operative chemotherapy can be considered.

C — Available medical regimens include doxorubicin-based regimens or gemcitabine + docetaxel for patients not able to receive doxorubicin.

B — Pre-operative chemotherapy for stage III disease with doxorubicin + trabectedin or doxorubicin + dacarbazine is recommended in an effort to improve surgical resectability.

Locally advanced disease (FIGO stages II-III) - Role of radiotherapy

C — Pelvic radiotherapy may be an alternative option as definitive treatment if surgery is not feasible or after incomplete pelvic surgery (R1/R2).

C — Post-operative radiotherapy can be considered in high-grade LMS with involvement of cervix/parametria and/or positive margins.

Metastatic disease - Role of chemotherapy

A Systemic therapy is the standard therapy in the metastatic setting.

B Pre- or post-operative chemotherapy can be considered prior to or after local treatment with surgery/radiation dependent on prognostic factors (e.g., extent and number and site of metastases and previous disease-free interval).

For first-line therapy, available regimens include:

B • Doxorubicin-based in combination with
  - o Trabectedin **C**
  - o Dacarbazine **C**

B • Gemcitabine + docetaxel

B • Doxorubicin or gemcitabine or liposomal doxorubicin as single agents (on clinical judgement, e.g. when multi-agent chemotherapy is not feasible, etc.)

For the second-line therapy and more, available regimens include:

B
- The same regimen as above if not used as first-line therapy or if associated with previous response
- Trabectedin
- Gemcitabine ± dacarbazine
- Pazopanib
- Dacarbazine

Low-grade metastatic disease (hormonal receptor positive) - Role of endocrine therapy

B In indolent disease, active surveillance can be offered as initial management.

B In case of progressive disease, endocrine therapy is the first-line recommended treatment.

B Available agents include:

- Postmenopausal patients: aromatase inhibitors or progestins
- Premenopausal patients: luteinizing hormone-releasing hormone (LHRH) agonists ± aromatase inhibitors (providing there is no evidence of transformation to a high-grade LMS)

Relapse/metastatic setting - Role of radiotherapy

C Radiotherapy could be considered as an alternative to surgery or as a pre- or post-operative therapy.

C Radiotherapy can be used for recurrent or metastatic LMS when symptoms of local disease impact quality of life.

High-grade endometrial stromal sarcoma & undifferentiated sarcoma

Early/advanced disease (FIGO stages I-III) - Systemic therapy

D — Adjuvant chemotherapy is not the standard of care for stage I disease.

C — Adjuvant/post-operative chemotherapy could be considered in patients at a high risk of relapse after informed discussion and shared decision-making.

B — In case of morcellation of a HG-ESS or undifferentiated sarcoma, post-operative chemotherapy should be considered due to the high risk of relapse.

Early/advanced disease (FIGO stages I-III) - Radiotherapy

D — Adjuvant radiotherapy is not standard after hysterectomy for localised disease.

C — Post-operative radiotherapy could be considered based on risks of local recurrence.

Relapse/metastatic setting - First-line systemic treatment options

B C — Systemic therapy options in first-line include doxorubicin (if not used in adjuvant setting) combined with ifosfamide or as a single agent; or gemcitabine with docetaxel for patient not able to receive doxorubicin.

B — In case of oligometastatic disease undergoing surgery, pre- or post-operative chemotherapy can be considered in individual patients on the basis of adverse prognostic factors (e.g., number and site of metastases, short previous relapse-free interval).

Relapse/metastatic setting - Systemic options for second-line systemic therapy

B — Systemic therapy options include continuous infusion of high-dose ifosfamide; or gemcitabine and docetaxel; pazopanib or trabectedin as single agent. The choice depends on what was used in first-line setting.

Relapse and palliative setting - Role of radiotherapy

C — Radiotherapy could be considered as an alternative to surgery or as a pre- or post-operative therapy.

C — Radiotherapy can be used for recurrent or metastatic HG-ESS when symptoms of local disease impact quality of life.

Low-grade endometrial stromal sarcoma

Early/advanced disease - Role of adjuvant systemic therapy

D — Adjuvant endocrine therapy is not recommended for stage I uterine LG-ESS.

C — Post-operative endocrine therapy can be considered in patients with stage II, III-IV completely resected ER/PR positive uterine LG-ESS.

C — In the case of morcellation of uterine LG-ESS, consideration could be given to post-operative endocrine therapy due to greater risk of dissemination and recurrence.

C — Endocrine therapy recommended regimens:

- Progestins (megestrol acetate, medroxyprogesterone acetate)
- Aromatase inhibitors (anastrazole, letrozole, exemestane)

D — Tamoxifen is contraindicated.

Localised disease - Role of radiotherapy

D — Adjuvant radiotherapy is not recommended.

Relapse/metastatic setting - First-line systemic treatment options

C — Endocrine therapy is recommended for unresectable recurrent tumours.

C — Reassessment after at least 3 months of neoadjuvant endocrine therapy may identify a subset of patients with sufficient tumour response to consider debulking surgery.

C — Endocrine therapy recommended regimens:

- Progestins (megestrol acetate, medroxyprogesterone acetate)
- Aromatase inhibitors (anastrazole, letrozole, exemestane)
- Premenopausal with ovarian function: LHRH agonists ± aromatase inhibitors
- Leuprolide

D — Tamoxifen is contraindicated (due to potential agonistic effect on ER positive ESS).

Relapse/metastatic setting - Systemic treatment options for second--line therapy

B — Second-line endocrine therapy with an aromatase inhibitor/progestin/gonadotrophin-releasing hormone (GnRH) analogues or an ER antagonist such as fulvestrant should be offered to patients with recurrent/metastatic LG-ESS with disease progression after first-line endocrine therapy.

C — Following disease progression and/or high-grade transformation on endocrine therapy (including several lines), chemotherapy regimens as per high--grade tumours could be considered in selected cases.

Relapse/metastatic setting - Role of radiotherapy

B — Radiotherapy can be used for recurrent or metastatic LG-ESS for palliation.

Adenosarcoma & miscellaneous

Early/advanced disease - Systemic therapy - Low-grade adenosarcoma

D — Adjuvant endocrine therapy is not recommended for stage I uterine adenosarcoma.

C — Post-operative endocrine therapy can be considered in patients with stage II, III-IV completely resected ER/PR positive uterine adenosarcoma.

C — In the case of morcellation of uterine adenosarcoma with a low-grade sarcomatous component such as LG-ESS, adjuvant endocrine therapy could be considered due to the high risk of dissemination and recurrence.

Early/advanced disease - Systemic therapy - High-grade adenosarcoma or sarcomatous overgrowth

D — Post-operative chemotherapy is not the standard of care for stage I disease.

C — Adjuvant chemotherapy can be considered as an option in patients with stage II, III-IV completely resected uterine adenosarcoma due to the poor prognosis.

C — In the case of morcellation of a uterine adenosarcoma with a high-grade component/sarcomatous overgrowth, post-operative chemotherapy could be considered.

Localised disease - Radiotherapy

D — Adjuvant radiotherapy is not recommended for stage I uterine adenosarcoma.

C — Post-operative radiotherapy could be considered in stage II-IV uterine adenosarcoma for local control although there is no evidence to support a survival benefit.

Relapse/metastatic setting - First-line systemic treatment options - Low-grade adenosarcoma

C — Endocrine therapy with an aromatase inhibitor, progestogen, or GnRH analogues should be offered to patients with recurrent/metastatic ER/PR positive low-grade uterine adenosarcoma.

E — Tamoxifen should not be administered due to potential agonistic effect on ER positive adenosarcomas.

Relapse/metastatic setting - First-line systemic treatment options - High-grade adenosarcoma with sarcomatous overgrowth

B
C

Systemic therapy options include doxorubicin as a single agent or combined with ifosfamide; or gemcitabine in combination with docetaxel for patients who cannot receive doxorubicin.

Relapse/metastatic setting - Systemic treatment options for second--line therapy - Low-grade adenosarcoma

C

Second-line endocrine therapy with an aromatase inhibitor/progestagen/GnRH analogues/fulvestrant could be considered with the choice depending on what was used in first-line setting. Referral to clinical trials should be considered if available.

Relapse/metastatic setting - Systemic treatment options for second-line therapy - High-grade adenosarcoma with sarcomatous overgrowth

B
B
C

Systemic therapy options include doxorubicin or gemcitabine as a single agent or in combination with docetaxel; high dose ifosfamide (continuous infusion), trabectedin or pazopanib as single agents. The choice depends on what was used in first-line setting.

Relapse/metastatic setting - Role of radiotherapy

C

Radiotherapy could be considered in stage IV uterine adenosarcoma for local control or palliation.

Special considerations - Extra-uterine adenosarcoma

B

Extrauterine adenosarcomas are very rare and potentially more aggressive and a similar approach to management of patients with uterine adenosarcoma is recommended.

NTRK gynecological sarcomas

B

The primary treatment of NTRK-fusion sarcomas arising in the genital tract is complete surgical resection for localised disease.

D

The efficacy of radiotherapy and chemotherapy is unknown in the neoadjuvant/adjuvant setting. The role of NTRK inhibitors following surgery for localised disease is unclear and investigational and thus they are not recommended.

C

In patients with locally advanced disease, systemic treatment or radiotherapy can be considered to enable a subsequent resection with a curative intent. In this situation, the best response rates reported are with NTRK inhibitors.

B

NTRK inhibitors should be offered in patients with recurrence after primary treatment.

Miscellaneous, PEComas

B — The primary treatment of perivascular epithelioid cell tumour arising in the genital tract is complete surgical resection for localised disease.

C — There are no data to support adjuvant chemotherapy or radiotherapy.

B — For locally advanced disease, mTOR inhibitors can be considered to avoid surgery with the potential for significant morbidity.

B C — For metastatic disease, mTOR inhibitors are recommended as first-line treatment; hormone blockade treatment for selected patients with ER/PR positive tumours could be considered.

Follow-up & survivorship

Follow-up

B — There is a lack of evidence to guide a precise follow-up protocol but due to the possibility of developing asymptomatic/oligometastatic disease, regular follow-up is advised.

B — Patients should be informed about symptoms that could suggest recurrence and the importance of seeking prompt medical attention.

B C B — A reasonable approach for patient for whom we anticipate feasible treatment could be as follows:

- History + physical examination
  - o Every 3 to 4 months for the first 3 years → Every 6 to 12 months thereafter
  - o Or low-grade sarcoma patients are usually followed for local relapse every 4-6 months for the first 3-5 years, then yearly
- 18F-FDG PET/CT is not recommended but may add information in clinically inconclusive situations.
- Imaging surveillance: tailored according risk of systemic metastases with high grade. Systematic imaging surveillance can be discontinued after 10 years.
  - o Computed tomography chest/abdominal/pelvis:
    - High grade: every 3-4 months at least in the first 3 years, then every 4-6 months, and then from the 5th year annually
    - Low grade: every 4-6 months in the first 3 years, then annually
  - o MRI Abdominal/Pelvis + computed tomography chest as an alternative

C — Long-term follow-up is advised as late and distant relapses are not uncommon, particularly for low-grade tumours.

Survivorship

B — Long-term follow-up/survivorship should be tailored to the treatment received.

D
B — New imaging abnormality identified on follow-up imaging should not be automatically assumed to be a recurrence of gynaecological sarcoma. Biopsy should be performed.

Hormone replacement therapy

LG-ESS:

B
- Hormone replacement therapy is contraindicated due to the potential risk of recurrent disease.

C
- Hormone replacement therapy could be considered in selected symptomatic cases in whom menopausal symptoms cannot be controlled and there is a significant impact on quality of life.

B
- Close collaboration with gynaecological/endocrine team is recommended.

HG-ESS + uterine sarcoma:

C
- Hormone replacement therapy could be considered following discussion with individual patients.

LMS:

C
- ER/PR negative expression: hormone replacement therapy could be considered.
- Given heterogeneity of clinical disease for ER/PR positive LMS, hormone replacement therapy could be considered on a case-by-case basis.

Access full ESGO Guidelines: www.esgo.org/explore/guidelines

ESGO
Avenue Emmanuel Mounier 83/11
1200 Woluwe-Saint-Lambert
Belgium
Email: adminoffice@esgo.org
www.esgo.org