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Uterine Sarcoma in the World

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Uterine Sarcoma (US) is a rare cancer with an estimated worldwide incidence rate of 1,55–1,95 per 100 000 and comprises less than 5% of all the uterine corpus malignancies. US is a sophisticated topic to study on. It should be always born in mind that the final diagnosis of a uterine myoma patients could be sarcoma.
Key words: *Uterine Sarcoma, incidence rate, final diagnosis.*

Introduction

Uterine Sarcoma (US) is a malignant tumor that arises from the smooth muscle or connective tissue of the uterus. It is a rare cancer with an estimated worldwide incidence rate of 1,55–1,95 per 100 000 [1] and comprises less than 5% of all the uterine corpus malignancies [2]. According to SEER data 7% of all soft tissue sarcomas arise from the uterus [3].

Classification

Although US originates from mesenchymal tissue it has been classified by the World Health Organisation (WHO) in two main groups: pure mesenchymal tumors and mixed epithelial and mesenchymal tumors in 2003 [4]. The pure mesenchymal tumor group includes LMS (63%), ESS (21%) and undifferentiated uterine sarcoma (UUS, 5%) since the mixed group consists of carcinosarcomas and adenosarcomas which are composed of a mixture of epithelial and mesenchymal elements.

The classification is than revised by the International Federation of Gynecology and Obstetrics (FIGO) in 2009 (table 1). According to the FIGO staging system, carcinosarcomas or mixed malignant Mьllerian tumors (MMMT) however, today are regarded as a subset of endometrial carcinoma [5]. After excluding carcinosarcomas (MMMT) LMS has become the most common type of the US.

Also the rabdomyosarcoma which is typically arises in children and adolescents should not be evaluated in context of USs.

Diagnosis

The hystopathologic diagnosis of the US is made classically upon the most recent WHO classification [4] such as:

1. Cytological atypia or pleiomorphysm.
2. More than 5 mitotic figures per 10 high magnification field.
3. Necrosis.

Hypercellularity, infiltrating border, large size (>10 cm), atypical mitotic figures, extrauterine extension and peri- or postmenopausal age are the other supportive clinicopathologic features.

Epidemiology

Sarcomas comprise less than 5% of all cancers [6]. US is a small group of the sarcomas and makes up less than 1% of all gynecological malignancies [3], however it is responsible for more than 15% of deaths from uterine malignancies [7]. This rarity is the main handicap about having data about US. It has been suggested that all US cases and suspects must be referred to a tertiary clinic that has experience of it. Only by this way it would be possible to provide correct information and to make further treatment plans [1].

Although the incidence of uterine body malignancies has increased, USs and carcinosarcomas had a stable incidence within the last decades [8–10].

When it comes to analyse the epidemiological data of the US there are plenty of obstacles. Although its rarity causes difficulty about providing data about, US is a compelling diagnosis to make. First of all US comprises a huge heterogeneous group of pathologic cells originated from different embryonic tissues, which has completely different biological behavior and morphology. Secondly its preoperative diagnosis is challenging.

It has been reported that 94% of carcinosarcomas, 75% of endometrial stromal sarcomas (ESS) and 65% of leiomyosarcomas (LMS) were preoperatively diagnosed as uterine myoma [11]. Moreover with another study, 356 LMS cases were re-examined and only 259 of them got diagnosed as LMS again [12].

Ethiology

In spite of the fact that some genetic tendencies and specific alterations such as translocations [t(7;17)(p15;q21)] and deletions (del 7p) have been found related to US, the ethiology of US is still unknown [13, 14].

It has been detected that Afro-Americans has a two-fold increased risk for having US compared to caucasian Americans [15]. Also use of Tamoxifen for more than 5 years has found that could be associated with US (17 per 100.000) [16]. Pelvic radiation may increase the risk of US, but its relations appear to be stronger for carcinosarcoma (0.5% of irradiated women develop US) [17].

Some hereditary conditions like Hereditary Leiomyomatosis and Renal Cell Carcinoma (HLRCC) Syndrome (OMIM#605839) [18] are found to be predisposing US. Moreover survivors of childhood retinoblastoma are at a high risk for sarcomas, including US [19].

Other risk factors, such as early menarche, parity, late menopause, obesity, diabetes and hypertension, which are responsible for endometrial cancer progress, are inconclusive for US [20].

Survival

Although cytopathologic criteria, such as necrosis and mitotic figure density are crucial. The correctly performed surgery is the most important prognostic factor for the US patients' survival [2, 21]. On the other hand, neither the performance nor the extensity of lymphadenectomy, seem likely to be having an effect on US survival [22]. Additionally overall survival rate is higly affected by tumor grade and patients age [23].

If the 5-years survival rate is less than 30%, it is considered to be low. If this level is more than 65%, it is considered to be good, regarding the survival rates. US is considered as intermediate, with a 5-years overall survival rate of 43,5%, according to the epidemiological data [24]. ESS tends to be the less aggressive US subtype with a favorable outcome [25]. The survival rates of different subtypes of US are summarized at the table below (table 2) [8, 26-28].

Conclusion

US is a sophisticated topic for further study and the clinician must go over it with a fine tooth. It should be always born in mind that the final diagnosis of uterine myoma could be sarcoma. Because of the rarity and peculiarity of this disease the gynecologist must be prone to refer these patients to a tertiary clinic. Only by this way proper management and recording could be done for US.

Table 1

FIGO staging for uterine sarcomas (2009)

Stage	Definition
	*(1) Leiomyosarcomas and endometrial stromal sarcomas (ESS)**
I	Tumor limited to uterus
IA	Less than or equal to 5 cm
II	Tumor extends to the pelvis
B	More than 5 cm
IIA	Adnexal involvement
IIB	Tumor extends to extrauterine pelvic tissue
III	Tumor invades abdominal tissues (not just protruding into the abdomen).
IIIA	One site
IIIB	More than one site
IIIC	Metastasis to pelvic and/or para-aortic lymph nodes
IV IVA	Tumor invades bladder and/or rectum
IVB	Distant metastasis
	(2) Adenosarcomas
I	Tumor limited to uterus
IA	Tumor limited to endometrium/endocervix with no myometrial invasion
IB	Less than or equal to half myometrial invasion
IC	More than half myometrial invasion
II	Tumor extends to the pelvis
IIA	Adnexal involvement
IIB	Tumor extends to extrauterine pelvic tissue
III	Tumor invades abdominal tissues (not just protruding into the abdomen).
IIIA	One site
IIIB	More than one site
IIIC	Metastasis to pelvic and/or para-aortic lymph nodes
IV IVA	Tumor invades bladder and/or rectum
IVB	Distant metastasis
	(3) Carcinosarcomas
	Carcinosarcomas should be staged as carcinomas of the endometrium.

**Note:* Simultaneous tumors of the uterine corpus and ovary/pelvis in association with ovarian/pelvic endometriosis should be classified as independent primary tumors.

Table 2

Survival averages of US subtypes among their FIGO stages

5 years Overall Survival	LMS & ESS	Adenosarcoma	Carcinosarcoma
Stage I	55%	88%	52%
>Stage I	22%	54%	5–16%

Саркома матки в мире

Мурат Гюльтекин

Саркома матки – редкая онкологическая патология, составляющая менее 5% среди всех новообразований матки, частота встречаемости которой составляет 1,55–1,95 на 100 000 населения. Саркома матки является довольно актуальной темой для исследования. Доктора не должны забывать о том, что окончательный диагноз у пациенток с фибромиомой матки может оказаться саркомой.

Ключевые слова: *саркома матки, частота встречаемости, окончательный диагноз.*

Саркома матки у світі

Мурат Гюльтекін

Саркома матки є редкісною онкологічною патологією, яка складає меньш ніж 5% серед усіх новоутворювань матки, частість зустрічальності яких складає 1,55–1,95 на 100 000 населення. Саркома матки є довольно актуальною темою для дослідження. Лікарі не повинні забувати про те, що заключний діагноз у пацієнток з фіброміомою матки може опинитися саркомою.

Ключові слова: *саркома матки, частість зустрічальності, заключний діагноз.*

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ИДЕЯ ПИТАНИЯ ПО ГРУППЕ КРОВИ – АНТИНАУЧНА

На протяжении более 10 лет реклама предлагает якобы действительно научно обоснованную диету - выбор максимально полезных продуктов зависит от группы крови "худеющего". Исследователи из Канады тщательно изучили принципы этой диеты - и отвергли ее.

Идею питания в зависимости от группы крови придумал в конце XX века американский врач-натуропат Питер Д'Адамо (Peter D'Adamo), который изложил свои идеи в ставшей бестселлером книге "Eat Right 4 Your Type" ("Питайся правильно - в зависимости от твоей группы крови"). Книга была переведена на 52 языка и напечатана общим тиражом более 7 миллионов экземпляров.

Автор утверждал, что появление в процессе эволюции человека 4-х групп крови было непосредственно связано с характером питания обитателей тех или иных регионов планеты - таким образом, если питаться "правильными" продуктами, соответствующими группе крови, то они принесут организму значительно больше пользы.

Однако эта теория сразу же подверглась критике со стороны многих ученых как лженаучная.

Очередное "разрушение мифа" произвели канадские исследователи Challenges New сотрудники университета города Торонто (University of Toronto).

Авторы наблюдали почти 1 500 молодых и здоровых жителей Канады, которые представили ученым подробную информацию о характере их питания. С помощью анализа были определены группы крови всех испытуемых и показатели уровней глюкозы, холестерина и инсулина.

Средние показатели состояния здоровья членов подгруппы, чье питание максимально соответствовало советам Питер Д'Адамо, ничем не отличались от показателей тех участников, которые вели здоровый образ жизни, но питались с точки зрения писателя-натуропата абсолютно неправильно.

Аналогичные результаты получили в начале этого года сотрудники бельгийской некоммерческой медицинской организации Rode Kruis-Vlaanderen, которые провели мета-анализ более 1 400 научных публикаций посвященных связи рациона питания с группой крови.

Ни в одном из исследований не было получено достоверное подтверждение правильности идей Питера Д'Адамо.

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