Histological tumour type (Core)

Our knowledge of the different types of mesenchymal tumours that can occur in the uterus has expanded in the past decade, as underlying molecular abnormalities have helped define distinctive clinicopathologic entities. Proper classification of malignant and potentially malignant tumours is crucial as there are important differences in clinical management and outcome. In many instances, additional tumour sampling may be more useful than ancillary techniques; in particular, sampling of the border of tumours can be useful.

All mesenchymal tumours of the uterus should be typed according to the most recent edition of the World Health Organization (WHO) Classification of Tumours of Female Genital Tumours, 5th edition, 2020 (Table 1).[1] The International Collaboration on Cancer Reporting dataset includes 5th edition Corrigenda, June 2021.[2] The most commonly encountered sarcomas - leiomyosarcoma, endometrial stromal sarcoma and Müllerian adenosarcoma - will be discussed first.

Table 1: World Health Organization classification of mesenchymal tumours of the uterine corpus.[1]

Descriptor	**ICD-O codes**[a]
Mesenchymal tumours specific to the uterus	
Smooth muscle tumour of uncertain malignant potential (STUMP)	8897/1
Leiomyosarcoma	8890/3
Endometrial stromal sarcoma, low grade	8931/3
Endometrial stromal sarcoma, high grade	8930/3
Undifferentiated uterine sarcoma	8805/3
Uterine tumour resembling ovarian sex cord tumour (UTROSCT)	8590/1
Perivascular epithelioid cell tumour (PEComa)	8714/3
Inflammatory myofibroblastic tumour	8825/1
Mixed epithelial and mesenchymal tumours	
Adenosarcoma	8933/3

[a] These morphology codes are from the International Classification of Diseases for Oncology, third edition, second revision (ICD-O-3.2).[3] Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade III intraepithelial neoplasia; /3 for malignant tumours, primary site; and /6 for malignant tumours, metastatic site. Behaviour code /6 is not generally used by cancer registries. Incorporates all relevant changes from the 5th Edition Corrigenda June 2021.

Smooth muscle tumours

Classification of smooth muscle tumours primarily relies on histologic assessment of several parameters. This assessment, however, can be challenging as benign, malignant and tumours classified as of uncertain malignant potential can share overlapping morphologies. For example, a high degree of cellularity may be seen in both benign (cellular leiomyoma) and malignant (leiomyosarcoma) tumours. Nevertheless, using the most recent edition of the WHO Classification, most uterine smooth muscle neoplasms are readily diagnosed either as benign or malignant.[4] The type of leiomyosarcoma (spindle, epithelioid, myxoid) should be included in the report.

Leiomyosarcoma with spindle cell differentiation is diagnosed when there are at least two of the following three histological parameters: diffuse, moderate to severe nuclear atypia, mitotic count ≥10 per 2 mm^2 (≥10 mitoses per 10 high power fields (HPF) if field diameter is 0.55 mm) and tumour cell necrosis.[24] The criteria for diagnosis of malignancy in epithelioid and myxoid smooth muscle tumours are stricter. Epithelioid leiomyosarcoma usually contains ≥4 mitoses per 2 mm^2 (≥4 mitoses per 10 HPFs if field diameter is 0.55 mm) moderate to severe nuclear atypia and/or tumour cell necrosis.[5-7] Myxoid leiomyosarcomas usually have an infiltrative border, and either moderate to severe nuclear atypia, tumour cell necrosis, or >1 mitosis per 2 mm^2 (>1 mitoses per 10 HPFs if field diameter is 0.55 mm).[8]

Tumours which show morphological features that exceed the criteria for leiomyoma but fall below the threshold for leiomyosarcoma may be diagnosed as smooth muscle tumour of uncertain malignant potential (STUMP).[9,10] The category of STUMP should be used sparingly and before making a diagnosis of STUMP, every effort should be made to establish a diagnosis of either a leiomyoma subtype, leiomyosarcoma, or one of the recently described mesenchymal tumours with deceptively bland cytology as included in this dataset (i.e., perivascular epithelioid cell tumour, inflammatory myofibroblastic tumour (IMT), and neurotrophic tyrosine receptor kinase (NTRK)-rearranged spindle cell sarcoma). Besides epithelioid and myxoid neoplasms, the most common histologic subtypes of leiomyoma which may give rise to diagnostic difficulties are leiomyoma with bizarre nuclei and cellular leiomyoma. In order to make a distinction from leiomyosarcoma, accurate assessment of the number of mitoses in leiomyoma with bizarre nuclei is important but this is not straightforward because karyorrhectic nuclei may mimic mitoses.[11,12] Fumarate hydratase (FH)-deficient morphology can be seen in leiomyoma with bizarre nuclei as well as conventional and cellular leiomyomata. In FH-deficient leiomyomata, the nuclei are often arranged in chains, have eosinophilic cytoplasmic inclusions, prominent eosinophilic nuclei, and perinucleolar haloes. The presence of these features, often accompanied by staghorn blood vessels and alveolar-pattern edema, particularly in a smooth muscle tumour occurring in a young woman, should prompt consideration of association with fumarate hydratase deficiency. Loss of FH staining by immunohistochemistry supports the diagnosis. Of note, a subset of these tumours are characterised by intact expression of FH (corresponding to the presence of FH protein that is non-functional). With either loss or a non-functional FH protein, accumulation of 2-SC and a positive 2-SC stain confirms the diagnosis. Although the majority of these cases seems to be sporadic, hereditary leiomyoma and renal cell carcinoma syndrome needs to be ruled out in the appropriate clinical setting.[13-16] As high cellularity may be observed in benign and malignant smooth muscle tumours, as well as endometrial stromal neoplasms, tumours considered cellular leiomyoma should not contain microscopic features which exceed the WHO criteria for leiomyoma.[4]

Smooth muscle tumour of uncertain malignant potential (STUMP) is an applicable diagnosis if a spindle cell smooth muscle tumour has focal/multifocal or diffuse nuclear atypia, 5-9 mitoses per 2 mm^2 (5-9 mitoses per 10 HPFs if field diameter is 0.55 mm) and lacks tumour cell necrosis. Approximately 12-17% of such tumours have recurred. The STUMP diagnosis is also applicable to any bland smooth muscle tumour with tumour cell necrosis or necrosis of an uncertain type. Approximately 28% of such tumours have recurred. Tumours lacking cytological atypia and tumour cell necrosis, but with ≥15 mitoses per 2 mm^2 (≥15 mitoses per 10 HPFs if field diameter is 0.55 mm) are also considered STUMPs. Although none of such cases has recurred, the experience with these tumours is limited.[10,17] In addition to the Stanford criteria,[17] other helpful parameters that may be included in the assessment of recurrent potential in smooth muscle neoplasms are atypical mitoses, vascular involvement, and infiltrative/irregular margins.[18] Epithelioid and myxoid STUMPs are rare, and it is important to exclude their respective benign and malignant variants by integrating gross, microscopic and molecular findings.

Endometrial stromal sarcoma

The classification of endometrial stromal sarcoma has evolved over time due to a better understanding of its morphologic spectrum and underlying recurrent molecular abnormalities. While it was historically separated into low and high grade categories based on mitotic count, the category of high grade endometrial stromal sarcoma was removed from the 2003 WHO Classification as there was a lack of clinical relevance in separating tumours that morphologically resembled proliferative phase endometrial stroma into low and high grade categories based on mitotic count alone.[19] It is worth noting that the category of high grade endometrial stromal sarcoma at that time represented a heterogeneous group of tumours including those that resembled endometrial stroma and those with more nuclear pleomorphism. Currently, two categories of endometrial stromal sarcoma are recognised by the most recent WHO Classification.[1] While they maintain the same lexicon used in the past - low grade and high grade endometrial stromal sarcoma - they represent two distinct clinicopathologic entities with differing morphology, biologic behaviour and molecular findings.[19-25]

Low grade endometrial stromal sarcoma is composed of cells which morphologically resemble proliferative-phase endometrial stroma, i.e., cells have uniform round to ovoid nuclei and scant cytoplasm and they are associated with a delicate spiral arteriole-like network. This tumour has a characteristic growth pattern as it typically permeates the myometrium in a 'finger-like' or 'tongue-like' fashion; lymphovascular invasion (LVI) is frequent and sometimes prominent. Smooth muscle, sex cord-like, fibrous and myxoid variant morphology is not uncommon. Most, but not all tumours, harbour gene fusions most commonly *JAZF1-SUZ12.* Tumours with sex cord-like differentiation often harbour fusions involving *PHF1*. Patients with low grade endometrial stromal sarcoma typically have an indolent and protracted course.

Some low grade endometrial stromal tumours are classified as having 'limited' infiltration. These represent tumours that lack overt myometrial permeation but have more margin irregularity than allowed for designation as an endometrial stromal nodule.[26,27] Although most behave in benign fashion, a subset of tumours classified as such have metastasized. Thus, these tumours should be regarded as potentially malignant and be classified as low grade endometrial stromal sarcomas with limited infiltration.[28]

High grade endometrial stromal sarcoma encompasses tumours that have distinctive clinical, histologic and molecular findings that differs from low grade endometrial stromal sarcoma.[23,25,29-31] This tumour type occurs over a wide age range and shows a variable morphology but typically contains at least a focal characteristic round cell component (if associated with *YWHAE*-rearrangement), or myxoid spindle cell component (if associated with *BCOR*-rearrangement, most commonly *ZC3H7B-BCOR*, or internal tandem duplications). Patients with high grade endometrial stromal sarcoma more commonly present at higher stage in comparison to patients with low grade endometrial stromal sarcoma. Histologically, they can show expansile, permeative, or more commonly destructive infiltration of the myometrium; LVI can also be prominent. Tumours associated with *YWHAE*-rearrangement often, but not always, have a morphologically low grade component often akin to the fibromyxoid variant of low grade endometrial sarcoma. *BCOR*-associated tumours can closely mimic the appearance of myxoid leiomyosarcoma as tumour cells are often spindled with mild to moderate nuclear atypia and set in a prominent myxoid stroma. Limited clinical data suggest that high grade endometrial stromal sarcomas, regardless of the underlying genetic abnormality, are more likely to pursue an aggressive clinical course with earlier recurrences and metastasis, in comparison to low grade endometrial stromal sarcoma.[23,25,29,31]

Although rare, a scenario worth mentioning is the potential for low grade endometrial stromal sarcoma to 'transform' to a high grade tumour. In this scenario, the tumour may have the appearance of a high grade endometrial stromal sarcoma or undifferentiated uterine sarcoma but harbour translocations characteristic of conventional low grade endometrial stromal sarcoma.[32]

Müllerian adenosarcoma

Müllerian adenosarcoma is a biphasic neoplasm composed of a benign, non-neoplastic Müllerian epithelial component and a malignant sarcomatous component which is usually, but not always, morphologically low grade. These tumours are uncommon, representing less than 1% of all uterine malignancies and approximately 10% of uterine sarcomas. They present over a wide age range, most commonly in postmenopausal women, but a significant subset occurs in younger adults. Patients typically present with abnormal uterine bleeding. Other findings may include an enlarged uterus, pelvic mass, or polyp (either endocervical or endometrial in origin). Gross examination may show multiple large, soft polypoid masses filling the uterine cavity; tumours may invade the myometrium or cervical stroma, a finding more commonly associated with sarcomatous overgrowth. Characteristic histologic findings include a leaf-like growth pattern (also often described as phyllodes-like as the appearance is akin to a phyllodes tumour of the breast), with intraglandular stromal polypoid projections, and cuffing of the glands by hypercellular stroma. However, not all adenosarcomas show phylloidiform growth with some being composed of variably sized rounded glands surrounded by hypercellular stroma ('rigid cysts'); a combination of these appearances is not uncommon. The stromal cells may show variable amounts of nuclear atypia in the form of nuclear enlargement with irregular nuclear contour or nuclear hyperchromasia. Mitoses are typically identified (usually >1 per 2 mm^2 (>1 mitosis per 10 HPFs if field diameter is 0.55 mm)) but may be sparse or, in rare cases, absent. The stromal component is most commonly homologous, i.e., it has the appearance of endometrial or cervical stroma, but may also show heterologous differentiation, most commonly rhabdomyosarcoma. Sex cord-like differentiation may also occur. Sarcomatous overgrowth is defined as the presence of greater than 25% of the tumour composed solely of a neoplastic stromal component without epithelium; sex-cord-like differentiation is not considered in the assessment of stromal overgrowth.[33,34] Sarcomatous overgrowth often shows aberrant p53 immunoreactivity and loss of hormone receptor positivity.[35,36] It is important to note that the non-neoplastic epithelial component typically has a banal appearance, sometimes with various types of epithelia (tubal, endometrioid, mucinous, squamous); occasionally the epithelial component may show some cytologic atypia in the form of nuclear enlargement and hyperchromasia. In this latter scenario, additional sampling may be prudent to exclude carcinosarcoma. Features associated with an unfavourable outcome include sarcomatous overgrowth, deep myometrial invasion, and extrauterine extension; morphologically high grade nuclear atypia (marked nuclear enlargement and hyperchromasia) that shows mutation-type staining pattern for p53 may also be an adverse prognostic feature.[36]

Undifferentiated uterine sarcomas (Unclassifiable sarcomas)

Sarcomas which cannot be classified are considered undifferentiated uterine sarcoma. This is a diagnosis of exclusion after other malignancies, such as undifferentiated carcinoma, carcinosarcoma, leiomyosarcoma, and high grade endometrial stromal sarcoma have been excluded. As many of the tumours in the differential diagnosis may have areas of morphologic overlap, the diagnosis of undifferentiated uterine sarcoma is best rendered on a complete excision specimen as a more limited specimen may lack the diagnostic features of other uterine malignancies. It is worth noting that undifferentiated uterine sarcoma can be separated into two different types based on their morphologic appearance: uniform and pleomorphic.[37] With advanced molecular techniques and newly reported molecular abnormalities, the former are increasingly categorised as high grade endometrial stromal sarcomas whereas the pleomorphic type likely mostly represent sarcomas that are so poorly differentiated that cannot be classified.[38] Some of these tumours may represent

carcinosarcomas in which only the sarcomatous component is seen, and thus before making a diagnosis of an undifferentiated uterine sarcoma, additional sampling should be considered which may reveal diagnostic areas. Prior to rendering the diagnosis of undifferentiated uterine sarcoma in an excision specimen, extensive tumour sampling, immunohistochemical staining, and if possible, molecular testing may be needed to exclude other neoplasms.

Potentially malignant mesenchymal tumours of the uterus are those in which prognostication requires assessment of various clinical and pathologic parameters to determine biologic potential. Tumours within this category include uterine tumour resembling ovarian sex cord tumour, perivascular epithelioid cell tumour, and IMT.

Uterine tumour resembling ovarian sex cord tumour (UTROSCT)

Uterine tumour resembling ovarian sex cord tumour (UTROSCT) is an uncommon uterine tumour whose histologic features recapitulates the appearance of an ovarian sex cord tumour. Historically, the term UTROSCT included tumours entirely composed of sex cord elements as well as endometrial stromal tumours with extensive sex cord differentiation; the latter are no longer considered in this category based on morphologic as well as molecular differences. UTROSCT exhibit a wide range of morphologic appearances with diffuse, corded, trabecular, tubular, retiform and/or nested growth. Tumour cells have variable amounts of cytoplasm ranging from inconspicuous to abundant, which may be pale, foamy or eosinophilic; rhabdoid morphology may be seen and can be extensive. Nuclei are usually uniform with minimal cytologic atypia and the mitotic count is low. Nuclear atypia in the form of nuclear enlargement and hyperchromasia, as well as brisk mitotic activity, may be seen. UTROSCT is characterised by recurrent gene fusions involving *NCOA1-3*, *GREB1* and *ESR1*.[39-42] These tumours are considered to be of uncertain malignant potential. Although data is limited, features that may be associated with aggressive behaviour include a mitotic count >2 per 2 mm^2 (>2 mitoses per 10 HPFs if field diameter is 0.55 mm), necrosis, extensive (>50%) rhabdoid morphology and potentially tumours with *GREB1* rearrangement.[40-43]

Perivascular epithelioid cell tumour (PEComa)

Perivascular epithelioid cell tumours (PEComas) are unusual mesenchymal neoplasms that are composed of a distinctive population of cells, termed perivascular epithelioid cells, which co-express smooth muscle and melanocytic markers. These tumours have wide anatomic distribution and the uterus is the most common site when they occur in the female genital tract.[44-48] Most tumours occur sporadically with only a small subset being associated with tuberous sclerosis. Histologically, tumours most commonly are composed of epithelioid and spindle cells but can sometimes be solely or predominantly epithelioid or spindled. Tumours often, but not always, show a characteristic perivascular pattern of growth in which the tumour cells are radially arranged around the vasculature. In some tumours, the neoplastic cells can be seen within the muscular wall of the vessel. Another distinctive aspect of these tumours is their cytologic appearance; the cells are remarkable for abundant granular eosinophilic or clear cytoplasm although predominantly spindled tumours may show less abundant cytoplasm. When epithelioid, the tumour cells grow in sheets, nests and/or trabeculae that are surrounded by a delicate capillary vasculature. Spindled tumours often exhibit fascicular growth and can mimic smooth muscle neoplasia, the distinctive morphologic difference being the granular appearance of the cytoplasm. Of note, *TFE3*-associated PEComas often are composed of epithelioid cells that have predominantly clear cytoplasm; extensive melanin deposition may also occur. Assessment of potential malignant behaviour for PEComa of the female genital tract is based on the following parameters: tumour size ≥50 mm, high nuclear grade, mitotic count of >1 mitosis per 12 mm^2, presence of necrosis and presence of vascular invasion. If a tumour has three or more of these features, it is best classified as malignant. Only tumours that lack all features could potentially be considered benign. Any tumour with one or two features should be considered of uncertain malignant potential.

Inflammatory myofibroblastic tumour (IMT)

Inflammatory myofibroblastic tumour (IMT) is a myofibroblastic/fibroblastic neoplasm characterised by a variably myxoid stroma with an accompanying variably intense inflammatory infiltrate, primarily composed of lymphocytes and plasma cells. This tumour shows a wide anatomic distribution with the uterine corpus being the most common location in the female genital tract; less commonly it involves the cervix.[49-52] Occasionally, IMT may be identified at the time of delivery and in some cases may be adherent to the maternal surface of the placenta or be associated with the placental membranes.[53,54] Microscopically, the tumour borders can be well demarcated or irregular, either showing permeative (stromal sarcoma-like) or infiltrative margins. A number of different morphologies may be seen and are often intermixed: myxoid, leiomyoma-like, or hyalinised. The myxoid pattern is characteristically hypocellular with individual cells dispersed in an abundant myxoid matrix, a feature that imparts a fasciitis-like appearance on low power magnification. The leiomyoma-like areas are composed of spindled cells in intersecting fascicles or showing storiform growth; the former closely mimics smooth muscle neoplasia. The hyalinised pattern is remarkable for an abundant hyalinised and collagenous stroma containing scattered spindled cells. The tumour cells in the fasciitis-like areas are spindled with eosinophilic to amphophilic cytoplasmic processes and ovoid to tapered nuclei with open dispersed chromatin, features that closely resemble the appearance of the spindled cells of nodular fasciitis. The spindled cells in the leiomyoma-like areas have features indistinguishable from smooth muscle neoplasia with eosinophilic cytoplasm and more oblong nuclei with blunt ends. The epithelioid variant of IMT, which typically occurs in the abdominal cavity and is characterised by a predominant component of epithelioid cells with eosinophilic cytoplasm and vesicular nuclei, has yet to be described in the uterus, but has been reported in the ovary.[55,56] An inflammatory infiltrate, often present at the periphery of the tumour but also dispersed throughout, is typically composed of lymphocytes and plasma cells although other inflammatory cells can be seen; the amount and distribution of inflammatory cells vary but inflammation is typically a reproducible finding and characteristic of IMT. Assessment of potential malignant behaviour for IMT of the female genital tract is not well established. Some tumours present at high stage and should be considered malignant. Pathologic features which have been associated with aggressive behaviour include large tumour size (>150 mm), marked nuclear atypia, LVI and tumours with high mitotic counts (>10 per 2 mm^2 which corresponds to >10 mitoses per 10 HPFs if field diameter is 0.55 mm). These features are not invariably associated with adverse outcome and conversely tumours as small as 62 mm or with mitotic counts of only 1 per 2 mm^2 (1 mitosis per 10 HPFs if field diameter is 0.55 mm) have recurred. Recurrence of IMT at all anatomic sites is estimated at 25% and is related to resectability.[57] As complete resection is typically achieved with hysterectomy, this may partially explain the overall good outcome for most patients with uterine neoplasms.

Rhabdomyosarcoma

Less common uterine sarcomas include rhabdomyosarcoma and alveolar soft part sarcoma. Different types of rhabdomyosarcoma have been described in the female genital tract; in the uterus, embryonal rhabdomyosarcoma (ERMS) and pleomorphic rhabdomyosarcoma are the most likely to be encountered.[58-61] Histologically, ERMS characteristically shows alternating cellularity with hypocellular myxoid zones and hypercellular foci of spindled rhabdomyoblasts, often condensing underneath the overlying epithelium (cambium layer). Heterologous cartilaginous differentiation is commonly seen. Pleomorphic rhabdomyosarcoma is composed of sheets of highly atypical spindled, polygonal or rhabdoid cells with large irregular, frequently multinucleated cells and eosinophilic cytoplasm. Mitoses are frequent and often atypical. Both subtypes of rhabdomyosarcoma are positive for desmin, myoD1 and myogenin. ERMS is associated with high frequency of *DICER1* mutation which may be somatic or germline. Adult patients with ERMS have a less favourable prognosis than children.[58] Patients with pleomorphic rhabdomyosarcoma have a poor prognosis.[61] Some of these tumours may represent rhabdomyosarcomatous overgrowth in an adenosarcoma or

carcinosarcoma; thus, extensive sampling should be undertaken to exclude an epithelial component before diagnosing a pleomorphic rhabdomyosarcoma.

Alveolar soft part sarcoma (ASPS)

Alveolar soft part sarcoma (ASPS) can occur at any location in the female genital tract; uterine tumours can occur in the corpus or cervix.[62-65] Histologically, they are composed of nests of large, polygonal epithelioid cells with abundant eosinophilic granular cytoplasm containing an eccentric or centrally located nucleus with vesicular nuclei and prominent nucleolus. Characteristically, the nests are enveloped by a delicate sinusoidal vascular network and the cells often show dyscohesion resulting in an alveolar-like appearance. Cytoplasmic clearing and rhabdoid cells may be seen. Intracytoplasmic granules and/or rhomboid crystals may be seen, which can be highlighted by periodic acid-Schiff stain and are diastase resistant. Mitoses are typically sparse. ASPS is characterised by *TFE3* rearrangements as a result of chromosomal translocation t(x;17)(p11;q25). As a consequence, tumour cells typically show strong and diffuse nuclear staining for TFE3. The prognosis of ASPS at all anatomic sites appears to be related to resectability, which may explain the relative better prognosis for ASPS of the gynaecologic tract in comparison to those that arise elsewhere.

It is worth noting that many different types of sarcoma more commonly encountered in other anatomic locations can also rarely arise in the uterus, such as liposarcoma[66] and angiosarcoma.[67-69] Prior to making the diagnosis of a pure unusual type of sarcoma of the uterus, additional sampling of the lesion should be performed to exclude the possibility that it represents the component of a more commonly encountered uterine neoplasm, such as sarcomatous overgrowth of an adenosarcoma or the mesenchymal component of a carcinosarcoma.

Emerging entities

Emerging uterine mesenchymal entities include *NTRK*-rearranged sarcoma, *PDGFR*-rearranged sarcoma and *SMARCA4*-deficient uterine sarcoma. *NTRK*-rearranged sarcoma has been recently described to occur in the uterine cervix and lower uterine segment.[70-73] Histologically, *NTRK*-rearranged sarcomas typically have an infiltrative border and are composed of a proliferation of spindled cells exhibiting either a patternless architecture or showing (often haphazard) fascicular or herringbone growth. Entrapped endocervical glands may be encircled by the neoplastic cells, sometimes with polypoid projections simulating adenosarcoma; however, there is typically no periglandular stromal condensation. The spindle cells have eosinophilic cytoplasm and generally show mild to moderate nuclear enlargement with nuclei that are ovoid with dispersed chromatin and small nucleoli; epithelioid change and foci of marked atypia may be seen. The vascular component can be composed of delicate capillaries or vessels with variably thickened walls often with prominent hyalinisation. The mitotic count is variable ranging from 0 to 50 per 2 mm^2 (0-50 mitoses per 10 HPFs if field diameter is 0.55 mm) atypical mitotic figures and necrosis may be seen. Other findings that may be encountered include focal myxoid matrix, focal hemangiopericytoma-like vasculature and a prominent lymphocytic infiltrate. These tumours show positivity for pan-TRK, but this marker is not specific for the gene fusion. Patients with *NTRK*-rearranged sarcoma typically present with Stage I disease; however, approximately one third have developed recurrence or metastatic disease.[70-73] Targeted therapy against tropomyosine kinase receptors has shown clinical benefit in patients with *NTRK*-associated sarcomas.[74]

RET fusion positive neoplasms may also exhibit fibroblastic or neural-like differentiation and have phenotypic overlap with *NTRK*-related neoplasms.[75] Recently a cervical sarcoma with a novel *RET-SPECC1L* fusion has been described.[76] Rare spindle cell sarcomas with recurrent *MEIS1-NCOA2* fusions have also been recently described.[77]

COL1A1-PDGFB rearranged uterine sarcomas are rare and data is limited.[72,78] These tumours are composed of a cellular proliferation of spindle cells that typically exhibit a storiform or herringbone growth pattern although one tumour has been described as showing a fascicular 'leiomyoma-like' growth pattern. Overall, it is interesting to speculate that these tumours could be the uterine counterpart of dermatofibrosarcoma protuberans, as they show morphological overlap (including fibrosarcomatous areas), a similar immunophenotype (focal loss of CD34 staining in 'fibrosarcomatous areas'), as well as sharing the same gene fusion.

SMARCA4-deficient uterine sarcoma (SDUS) is a recently described entity that shares morphologic overlap with undifferentiated endometrial carcinoma but has distinctive clinicopathologic and molecular differences.[79-81] Tumours characteristically show a diffuse growth of large epithelioid cells with round vesicular nuclei and exhibit prominent rhabdoid morphology; other features which may be focally present include phyllodiform architecture, vague cording or nesting associated with stromal hyalinisation, small cell or spindled morphology, and focal myxoid stromal change. Brisk mitoses (usually >20 per 10 HPFs/0.24 mm^2), necrosis and LVI are common. Patients with SDUS have a poor prognosis.[79-81]

References

1 WHO Classification of Tumours Editorial Board (2020). *Female Genital Tumours, WHO Classification of Tumours, 5th Edition, Volume 4*. IARC Press, Lyon.

2 WHO Classification of Tumours Editorial Board (2021). *Female Genital Tumours, WHO Classification of Tumours, 5th Edition, Volume 4 - Corrigenda June 2021*. Available from: https://publications.iarc.fr/Book-And-Report-Series/Who-Classification-Of-Tumours/Female-Genital-Tumours-2020 (Accessed 16th June 2021).

3 Fritz A, Percy C, Jack A, Shanmugaratnam K, Sobin L, Parkin DM and Whelan S (eds) (2020). *International Classification of Diseases for Oncology, Third edition, Second revision ICD-O-3.2*. Available from: http://www.iacr.com.fr/index.php?option=com_content&view=category&layout=blog&id=100&Itemid=577 (Accessed 16th June 2021).

4 Ip PPC et al (eds) (2020). Uterine leiomyoma. In: *Female Genital Tumours, WHO Classification of Tumours, 5th Edition, Volume 4*, WHO Classification of Tumours Editorial Board (ed), IARC Press, Lyon.

5 Prayson RA, Goldblum JR and Hart WR (1997). Epithelioid smooth-muscle tumors of the uterus: a clinicopathologic study of 18 patients. *Am J Surg Pathol* 21(4):383-391.

6 Kurman RJ and Norris HJ (1976). Mesenchymal tumors of the uterus. VI. Epithelioid smooth muscle tumors including leiomyoblastoma and clear-cell leiomyoma: a clinical and pathologic analysis of 26 cases. *Cancer* 37(4):1853-1865.

7 Oliva E (2016). Practical issues in uterine pathology from banal to bewildering: the remarkable spectrum of smooth muscle neoplasia. *Mod Pathol* 29 Suppl 1:S104-120.

8 Parra-Herran C, Schoolmeester JK, Yuan L, Dal Cin P, Fletcher CD, Quade BJ and Nucci MR (2016). Myxoid Leiomyosarcoma of the Uterus: A Clinicopathologic Analysis of 30 Cases and Review of the Literature With Reappraisal of Its Distinction From Other Uterine Myxoid Mesenchymal Neoplasms. *Am J Surg Pathol* 40(3):285-301.

9 Ip PP, Cheung AN and Clement PB (2009). Uterine smooth muscle tumors of uncertain malignant potential (STUMP): a clinicopathologic analysis of 16 cases. *Am J Surg Pathol* 33(7):992-1005.

10 Ip PPC, Croce S and Gupta M (eds) (2020). Smooth muscle tumour of uncertain malignant potential of the uterine corpus. In: *Female Genital Tumours, WHO Classification of Tumours, 5th Edition, Volume 4*, WHO Classification of Tumours Editorial Board (ed), IARC Press, Lyon.

11 Bennett JA, Weigelt B, Chiang S, Selenica P, Chen YB, Bialik A, Bi R, Schultheis AM, Lim RS, Ng CKY, Morales-Oyarvide V, Young RH, Reuter VE, Soslow RA and Oliva E (2017). Leiomyoma with bizarre nuclei: a morphological, immunohistochemical and molecular analysis of 31 cases. *Mod Pathol* 30(10):1476-1488.

12 Chow KL, Tse KY, Cheung CL, Wong KW, Cheung AN, Wong RW, Chan AN, Yuen NW, Ngan HY and Ip PP (2017). The mitosis-specific marker phosphohistone-H3 (PHH3) is an independent prognosticator in uterine smooth muscle tumours: an outcome-based study. *Histopathology* 70(5):746-755.

13 Joseph NM, Solomon DA, Frizzell N, Rabban JT, Zaloudek C and Garg K (2015). Morphology and Immunohistochemistry for 2SC and FH Aid in Detection of Fumarate Hydratase Gene Aberrations in Uterine Leiomyomas From Young Patients. *Am J Surg Pathol* 39(11):1529-1539.

14 Sanz-Ortega J, Vocke C, Stratton P, Linehan WM and Merino MJ (2013). Morphologic and molecular characteristics of uterine leiomyomas in hereditary leiomyomatosis and renal cancer (HLRCC) syndrome. *Am J Surg Pathol* 37(1):74-80.

15 Miettinen M, Felisiak-Golabek A, Wasag B, Chmara M, Wang Z, Butzow R and Lasota J (2016). Fumarase-deficient Uterine Leiomyomas: An Immunohistochemical, Molecular Genetic, and Clinicopathologic Study of 86 Cases. *Am J Surg Pathol* 40(12):1661-1669.

16 Reyes C, Karamurzin Y, Frizzell N, Garg K, Nonaka D, Chen YB and Soslow RA (2014). Uterine smooth muscle tumors with features suggesting fumarate hydratase aberration: detailed morphologic analysis and correlation with S-(2-succino)-cysteine immunohistochemistry. *Mod Pathol* 27(7):1020-1027.

17 Bell SW, Kempson RL and Hendrickson MR (1994). Problematic uterine smooth muscle neoplasms. A clinicopathologic study of 213 cases. *Am J Surg Pathol* 18(6):535-558.

18 Gupta M, Laury AL, Nucci MR and Quade BJ (2018). Predictors of adverse outcome in uterine smooth muscle tumours of uncertain malignant potential (STUMP): a clinicopathological analysis of 22 cases with a proposal for the inclusion of additional histological parameters. *Histopathology* 73(2):284-298.

19 Chang KL, Crabtree GS, Lim-Tan SK, Kempson RL and Hendrickson MR (1990). Primary uterine endometrial stromal neoplasms. A clinicopathologic study of 117 cases. *Am J Surg Pathol* 14(5):415-438.

20 Koontz JI, Soreng AL, Nucci M, Kuo FC, Pauwels P, van Den Berghe H, Dal Cin P, Fletcher JA and Sklar J (2001). Frequent fusion of the JAZF1 and JJAZ1 genes in endometrial stromal tumors. *Proc Natl Acad Sci U S A* 98(11):6348-6353.

21 Nucci MR, Harburger D, Koontz J, Dal Cin P and Sklar J (2007). Molecular analysis of the JAZF1-JJAZ1 gene fusion by RT-PCR and fluorescence in situ hybridization in endometrial stromal neoplasms. *Am J Surg Pathol* 31(1):65-70.

22 Huang HY, Ladanyi M and Soslow RA (2004). Molecular detection of JAZF1-JJAZ1 gene fusion in endometrial stromal neoplasms with classic and variant histology: evidence for genetic heterogeneity. *Am J Surg Pathol* 28(2):224-232.

23 Lee CH, Mariño-Enriquez A, Ou W, Zhu M, Ali RH, Chiang S, Amant F, Gilks CB, van de Rijn M, Oliva E, Debiec-Rychter M, Dal Cin P, Fletcher JA and Nucci MR (2012). The clinicopathologic features of YWHAE-FAM22 endometrial stromal sarcomas: a histologically high-grade and clinically aggressive tumor. *Am J Surg Pathol* 36(5):641-653.

24 Hoang LN, Aneja A, Conlon N, Delair DF, Middha S, Benayed R, Hensley ML, Park KJ, Hollmann TJ, Hameed MR, Antonescu CR, Soslow RA and Chiang S (2017). Novel High-grade Endometrial Stromal Sarcoma: A Morphologic Mimicker of Myxoid Leiomyosarcoma. *Am J Surg Pathol* 41(1):12-24.

25 Lewis N, Soslow RA, Delair DF, Park KJ, Murali R, Hollmann TJ, Davidson B, Micci F, Panagopoulos I, Hoang LN, Arias-Stella JA, 3rd, Oliva E, Young RH, Hensley ML, Leitao MM, Jr., Hameed M, Benayed R, Ladanyi M, Frosina D, Jungbluth AA, Antonescu CR and Chiang S (2018). ZC3H7B-BCOR high-grade endometrial stromal sarcomas: a report of 17 cases of a newly defined entity. *Mod Pathol* 31(4):674-684.

26 Tavassoli FA and Norris HJ (1981). Mesenchymal tumours of the uterus. VII. A clinicopathological study of 60 endometrial stromal nodules. *Histopathology* 5(1):1-10.

27 Dionigi A, Oliva E, Clement PB and Young RH (2002). Endometrial stromal nodules and endometrial stromal tumors with limited infiltration: a clinicopathologic study of 50 cases. *Am J Surg Pathol* 26(5):567-581.

28 Moore M and McCluggage WG (2020). Uterine Endometrial Stromal Tumors With Limited Infiltration: First Report of a Case Series Indicating Potential for Malignant Behavior. *Int J Gynecol Pathol* 39(3):221-226.

29 Mariño-Enriquez A, Lauria A, Przybyl J, Ng TL, Kowalewska M, Debiec-Rychter M, Ganesan R, Sumathi V, George S, McCluggage WG, Nucci MR, Lee CH and Fletcher JA (2018). BCOR Internal Tandem Duplication in High-grade Uterine Sarcomas. *Am J Surg Pathol* 42(3):335-341.

30 Hoang L, Chiang S and Lee CH (2018). Endometrial stromal sarcomas and related neoplasms: new developments and diagnostic considerations. *Pathology* 50(2):162-177.

31 Micci F, Heim S and Panagopoulos I (2021). Molecular pathogenesis and prognostication of "low-grade'' and "high-grade" endometrial stromal sarcoma. *Genes Chromosomes Cancer* 60(3):160-167.

32 Zou Y, Turashvili G, Soslow RA, Park KJ, Croce S, McCluggage WG, Stewart CJR, Oda Y, Oliva E, Young RH, Da Cruz Paula A, Dessources K, Ashley CW, Hensley ML, Yip S, Weigelt B, Benayed R, Antonescu CR, Lee CH and Chiang S (2020). High-grade transformation of low-grade endometrial stromal sarcomas lacking YWHAE and BCOR genetic abnormalities. *Mod Pathol* 33(9):1861-1870.

33 Clement PB and Scully RE (1989). Müllerian adenosarcomas of the uterus with sex cord-like elements. A clinicopathologic analysis of eight cases. *Am J Clin Pathol* 91(6):664-672.

34 Stolnicu S, Molnar C, Barsan I, Boros M, Nogales FF and Soslow RA (2016). The Impact on Survival of an Extensive Sex Cord-like Component in Mullerian Adenosarcomas: A Study Comprising 6 Cases. *Int J Gynecol Pathol* 35(2):147-152.

35 Blom R and Guerrieri C (1999). Adenosarcoma of the uterus: a clinicopathologic, DNA flow cytometric, p53 and mdm-2 analysis of 11 cases. *Int J Gynecol Cancer* 9(1):37-43.

36 Hodgson A, Amemiya Y, Seth A, Djordjevic B and Parra-Herran C (2017). High-grade Müllerian Adenosarcoma: Genomic and Clinicopathologic Characterization of a Distinct Neoplasm With Prevalent TP53 Pathway Alterations and Aggressive Behavior. *Am J Surg Pathol* 41(11):1513-1522.

37 Kurihara S, Oda Y, Ohishi Y, Iwasa A, Takahira T, Kaneki E, Kobayashi H, Wake N and Tsuneyoshi M (2008). Endometrial stromal sarcomas and related high-grade sarcomas: immunohistochemical and molecular genetic study of 31 cases. *Am J Surg Pathol* 32(8):1228-1238.

38 Cotzia P, Benayed R, Mullaney K, Oliva E, Felix A, Ferreira J, Soslow RA, Antonescu CR, Ladanyi M and Chiang S (2019). Undifferentiated Uterine Sarcomas Represent Under-Recognized High-grade Endometrial Stromal Sarcomas. *Am J Surg Pathol* 43(5):662-669.

39 Dickson BC, Childs TJ, Colgan TJ, Sung YS, Swanson D, Zhang L and Antonescu CR (2019). Uterine Tumor Resembling Ovarian Sex Cord Tumor: A Distinct Entity Characterized by Recurrent NCOA2/3 Gene Fusions. *Am J Surg Pathol* 43(2):178-186.

40 Bennett JA, Lastra RR, Barroeta JE, Parilla M, Galbo F, Wanjari P, Young RH, Krausz T and Oliva E (2020). Uterine Tumor Resembling Ovarian Sex Cord Stromal Tumor (UTROSCT): A Series of 3 Cases With Extensive Rhabdoid Differentiation, Malignant Behavior, and ESR1-NCOA2 Fusions. *Am J Surg Pathol* 44(11):1563-1572.

41 Goebel EA, Hernandez Bonilla S, Dong F, Dickson BC, Hoang LN, Hardisson D, Lacambra MD, Lu FI, Fletcher CDM, Crum CP, Antonescu CR, Nucci MR and Kolin DL (2020). Uterine Tumor Resembling Ovarian Sex Cord Tumor (UTROSCT): A Morphologic and Molecular Study of 26 Cases Confirms Recurrent NCOA1-3 Rearrangement. *Am J Surg Pathol* 44(1):30-42.

42 Croce S, Lesluyes T, Delespaul L, Bonhomme B, Pérot G, Velasco V, Mayeur L, Rebier F, Ben Rejeb H, Guyon F, McCluggage WG, Floquet A, Querleu D, Chakiba C, Devouassoux-Shisheboran M, Mery E, Arnould L, Averous G, Soubeyran I, Le Guellec S and Chibon F (2019). GREB1-CTNNB1 fusion transcript detected by RNA-sequencing in a uterine tumor resembling ovarian sex cord tumor (UTROSCT): A novel CTNNB1 rearrangement. *Genes Chromosomes Cancer* 58(3):155-163.

43 Moore M and McCluggage WG (2017). Uterine tumour resembling ovarian sex cord tumour: first report of a large series with follow-up. *Histopathology* 71(5):751-759.

44 Folpe AL, Mentzel T, Lehr HA, Fisher C, Balzer BL and Weiss SW (2005). Perivascular epithelioid cell neoplasms of soft tissue and gynecologic origin: a clinicopathologic study of 26 cases and review of the literature. *Am J Surg Pathol* 29(12):1558-1575.

45 Schoolmeester JK, Howitt BE, Hirsch MS, Dal Cin P, Quade BJ and Nucci MR (2014). Perivascular epithelioid cell neoplasm (PEComa) of the gynecologic tract: clinicopathologic and immunohistochemical characterization of 16 cases. *Am J Surg Pathol* 38(2):176-188.

46 Schoolmeester JK, Dao LN, Sukov WR, Wang L, Park KJ, Murali R, Hameed MR and Soslow RA (2015). TFE3 translocation-associated perivascular epithelioid cell neoplasm (PEComa) of the gynecologic tract: morphology, immunophenotype, differential diagnosis. *Am J Surg Pathol* 39(3):394-404.

47 Bennett JA, Braga AC, Pinto A, Van de Vijver K, Cornejo K, Pesci A, Zhang L, Morales-Oyarvide V, Kiyokawa T, Zannoni GF, Carlson J, Slavik T, Tornos C, Antonescu CR and Oliva E (2018). Uterine PEComas: A Morphologic, Immunohistochemical, and Molecular Analysis of 32 Tumors. *Am J Surg Pathol* 42(10):1370-1383.

48 Vang R and Kempson RL (2002). Perivascular epithelioid cell tumor ('PEComa') of the uterus: a subset of HMB-45-positive epithelioid mesenchymal neoplasms with an uncertain relationship to pure smooth muscle tumors. *Am J Surg Pathol* 26(1):1-13.

49 Haimes JD, Stewart CJR, Kudlow BA, Culver BP, Meng B, Koay E, Whitehouse A, Cope N, Lee JC, Ng T, McCluggage WG and Lee CH (2017). Uterine Inflammatory Myofibroblastic Tumors Frequently Harbor ALK Fusions With IGFBP5 and THBS1. *Am J Surg Pathol* 41(6):773-780.

50 Parra-Herran C, Quick CM, Howitt BE, Dal Cin P, Quade BJ and Nucci MR (2015). Inflammatory myofibroblastic tumor of the uterus: clinical and pathologic review of 10 cases including a subset with aggressive clinical course. *Am J Surg Pathol* 39(2):157-168.

51 Bennett JA, Croce S, Pesci A, Niu N, Van de Vijver K, Burks EJ, Burandt E, Zannoni GF, Rabban JT and Oliva E (2020). Inflammatory Myofibroblastic Tumor of the Uterus: An Immunohistochemical Study of 23 Cases. *Am J Surg Pathol* 44(11):1441-1449.

52 Rabban JT, Zaloudek CJ, Shekitka KM and Tavassoli FA (2005). Inflammatory myofibroblastic tumor of the uterus: a clinicopathologic study of 6 cases emphasizing distinction from aggressive mesenchymal tumors. *Am J Surg Pathol* 29(10):1348-1355.

53 Devereaux KA, Fitzpatrick MB, Hartinger S, Jones C, Kunder CA and Longacre TA (2020). Pregnancy-associated Inflammatory Myofibroblastic Tumors of the Uterus Are Clinically Distinct and Highly Enriched for TIMP3-ALK and THBS1-ALK Fusions. *Am J Surg Pathol* 44(7):970-981.

54 Makhdoum S, Nardi V, Devereaux KA, Kunder CA, Nielsen GP, Oliva E, Young RH and Roberts DJ (2020). Inflammatory myofibroblastic tumors associated with the placenta: a series of 9 cases. *Hum Pathol* 106:62-73.

55 Fang H, Langstraat CL, Visscher DW, Folpe AL and Schoolmeester JK (2018). Epithelioid Inflammatory Myofibroblastic Sarcoma of the Ovary With RANB2-ALK Fusion: Report of a Case. *Int J Gynecol Pathol* 37(5):468-472.

56 Mariño-Enríquez A, Wang WL, Roy A, Lopez-Terrada D, Lazar AJ, Fletcher CD, Coffin CM and Hornick JL (2011). Epithelioid inflammatory myofibroblastic sarcoma: An aggressive intra-abdominal variant of inflammatory myofibroblastic tumor with nuclear membrane or perinuclear ALK. *Am J Surg Pathol* 35(1):135-144.

57 Gleason BC and Hornick JL (2008). Inflammatory myofibroblastic tumours: where are we now? *J Clin Pathol* 61(4):428-437.

58 Ferguson SE, Gerald W, Barakat RR, Chi DS and Soslow RA (2007). Clinicopathologic features of rhabdomyosarcoma of gynecologic origin in adults. *Am J Surg Pathol* 31(3):382-389.

59 McCluggage WG, Lioe TF, McClelland HR and Lamki H (2002). Rhabdomyosarcoma of the uterus: report of two cases, including one of the spindle cell variant. *Int J Gynecol Cancer* 12(1):128-132.

60 Li RF, Gupta M, McCluggage WG and Ronnett BM (2013). Embryonal rhabdomyosarcoma (botryoid type) of the uterine corpus and cervix in adult women: report of a case series and review of the literature. *Am J Surg Pathol* 37(3):344-355.

61 Ordi J, Stamatakos MD and Tavassoli FA (1997). Pure pleomorphic rhabdomyosarcomas of the uterus. *Int J Gynecol Pathol* 16(4):369-377.

62 Schoolmeester JK, Carlson J, Keeney GL, Fritchie KJ, Oliva E, Young RH and Nucci MR (2017). Alveolar Soft Part Sarcoma of the Female Genital Tract: A Morphologic, Immunohistochemical, and Molecular Cytogenetic Study of 10 Cases With Emphasis on its Distinction From Morphologic Mimics. *Am J Surg Pathol* 41(5):622-632.

63 Nielsen GP, Oliva E, Young RH, Rosenberg AE, Dickersin GR and Scully RE (1995). Alveolar soft-part sarcoma of the female genital tract: a report of nine cases and review of the literature. *Int J Gynecol Pathol* 14(4):283-292.

64 Kasashima S, Minato H, Kobayashi M, Ueda Y, Oda Y, Hashimoto S and Inoue M (2007). Alveolar soft part sarcoma of the endometrium with expression of CD10 and hormone receptors. *Apmis* 115(7):861-865.

65 Radig K, Buhtz P and Roessner A (1998). Alveolar soft part sarcoma of the uterine corpus. Report of two cases and review of the literature. *Pathol Res Pract* 194(1):59-63.

66 McDonald AG, Dal Cin P, Ganguly A, Campbell S, Imai Y, Rosenberg AE and Oliva E (2011). Liposarcoma arising in uterine lipoleiomyoma: a report of 3 cases and review of the literature. *Am J Surg Pathol* 35(2):221-227.

67 Roma AA, Allende D, Fadare O, Forscher C and Rutgers JK (2017). On Uterine Angiosarcomas: 2 Additional Cases. *Int J Gynecol Pathol* 36(4):369-371.

68 Kruse AJ, Sep S, Slangen BF, Vandevijver NM, Van Gorp T, Kruitwagen RF and Van de Vijver KK (2014). Angiosarcomas of primary gynecologic origin: a clinicopathologic review and quantitative analysis of survival. *Int J Gynecol Cancer* 24(1):4-12.

69 Schammel DP and Tavassoli FA (1998). Uterine angiosarcomas: a morphologic and immunohistochemical study of four cases. *Am J Surg Pathol* 22(2):246-250.

70 Rabban JT, Devine WP, Sangoi AR, Poder L, Alvarez E, Davis JL, Rudzinski E, Garg K and Bean GR (2020). NTRK fusion cervical sarcoma: a report of three cases, emphasising morphological and immunohistochemical distinction from other uterine sarcomas, including adenosarcoma. *Histopathology* 77(1):100-111.

71 Chiang S, Cotzia P, Hyman DM, Drilon A, Tap WD, Zhang L, Hechtman JF, Frosina D, Jungbluth AA, Murali R, Park KJ, Soslow RA, Oliva E, Iafrate AJ, Benayed R, Ladanyi M and Antonescu CR (2018). NTRK Fusions Define a Novel Uterine Sarcoma Subtype With Features of Fibrosarcoma. *Am J Surg Pathol* 42(6):791-798.

72 Croce S, Hostein I, Longacre TA, Mills AM, Pérot G, Devouassoux-Shisheboran M, Velasco V, Floquet A, Guyon F, Chakiba C, Querleu D, Khalifa E, Mayeur L, Rebier F, Leguellec S, Soubeyran I and McCluggage WG (2019). Uterine and vaginal sarcomas resembling fibrosarcoma: a clinicopathological and molecular analysis of 13 cases showing common NTRK-rearrangements and the description of a COL1A1-PDGFB fusion novel to uterine neoplasms. *Mod Pathol* 32(7):1008-1022.

73 Hodgson A, Pun C, Djordjevic B and Turashvili G (2021). NTRK-rearranged Cervical Sarcoma: Expanding the Clinicopathologic Spectrum. *Int J Gynecol Pathol* 40(1):73-77.

74 Demetri GD, Antonescu CR, Bjerkehagen B, Bovée J, Boye K, Chacón M, Dei Tos AP, Desai J, Fletcher JA, Gelderblom H, George S, Gronchi A, Haas RL, Hindi N, Hohenberger P, Joensuu H, Jones RL, Judson I, Kang YK, Kawai A, Lazar AJ, Le Cesne A, Maestro R, Maki RG, Martín J, Patel S, Penault-Llorca F, Premanand Raut C, Rutkowski P, Safwat A, Sbaraglia M, Schaefer IM, Shen L, Serrano C, Schöffski P, Stacchiotti S, Sundby Hall K, Tap WD, Thomas DM, Trent J, Valverde C, van der Graaf WTA, von Mehren M, Wagner A, Wardelmann E, Naito Y, Zalcberg J and Blay JY (2020). Diagnosis and management of tropomyosin receptor kinase (TRK) fusion sarcomas: expert recommendations from the World Sarcoma Network. *Ann Oncol* 31(11):1506-1517.

75 Antonescu CR, Dickson BC, Swanson D, Zhang L, Sung YS, Kao YC, Chang WC, Ran L, Pappo A, Bahrami A, Chi P and Fletcher CD (2019). Spindle Cell Tumors With RET Gene Fusions Exhibit a Morphologic Spectrum Akin to Tumors With NTRK Gene Fusions. *Am J Surg Pathol* 43(10):1384-1391.

76 Weisman PS, Altinok M, Carballo EV, Kushner DM, Kram JJF, Ladanyi M, Chiang S, Buehler D and Dickson Michelson EL (2020). Uterine Cervical Sarcoma With a Novel RET-SPECC1L Fusion in an Adult: A Case Which Expands the Homology Between RET-rearranged and NTRK-rearranged Tumors. *Am J Surg Pathol* 44(4):567-570.

77 Kao YC, Bennett JA, Suurmeijer AJH, Dickson BC, Swanson D, Wanjari P, Zhang L, Lee JC and Antonescu CR (2021). Recurrent MEIS1-NCOA2/1 fusions in a subset of low-grade spindle cell sarcomas frequently involving the genitourinary and gynecologic tracts. *Mod Pathol* 34(6): 1203-1212.

78 Grindstaff SL, DiSilvestro J, Hansen K, DiSilvestro P, Sung CJ and Quddus MR (2020). COL1A1-PDGFB fusion uterine fibrosarcoma: A case report with treatment implication. *Gynecol Oncol Rep* 31:100523.

79 Kolin DL, Quick CM, Dong F, Fletcher CDM, Stewart CJR, Soma A, Hornick JL, Nucci MR and Howitt BE (2020). SMARCA4-deficient Uterine Sarcoma and Undifferentiated Endometrial Carcinoma Are Distinct Clinicopathologic Entities. *Am J Surg Pathol* 44(2):263-270.

80 Kolin DL, Dong F, Baltay M, Lindeman N, MacConaill L, Nucci MR, Crum CP and Howitt BE (2018). SMARCA4-deficient undifferentiated uterine sarcoma (malignant rhabdoid tumor of the uterus): a clinicopathologic entity distinct from undifferentiated carcinoma. *Mod Pathol* 31(9):1442-1456.

81 Lin DI, Allen JM, Hecht JL, Killian JK, Ngo NT, Edgerly C, Severson EA, Ali SM, Erlich RL, Ramkissoon SH, Vergilio JA, Ross JS and Elvin JA (2019). SMARCA4 inactivation defines a subset of undifferentiated uterine sarcomas with rhabdoid and small cell features and germline mutation association. *Mod Pathol* 32(11):1675-1687.