Review

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Review

Uterine Tumor Resembling Ovarian Sex Cord Tumor (UTROSCT): A Rare Polyphenotypic Neoplasm

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Abstract: Uterine Tumor Resembling Ovarian Sex Cord Stromal Tumor (UTROSCT) is a rare uterine mesenchymal neoplasm, which resembles ovarian sex cord tumors of the ovary. Although, in general, UTROSCT exhibits benign behavior with a favorable prognosis, this neoplasm is nonetheless classified as being of an uncertain malignant potential, given its low rate of recurrence and the fact that it rarely produces metastases (lymph nodes, epiploic appendix, omentum, small bowel, subcutaneous, lungs). Its histogenesis too is uncertain. Typically, UTROSCT occurs in peri-menopausal or menopausal women but can sometimes be observed in young women. Usually, this neoplasm can be found in the uterine corpus as a nodular intramural lesion, less frequently submucosal, subserosal or polypoid/intracavitary. UTROSCT can cause abnormal bleeding, pelvic pain, enlarged uterus, mass sensation, but sometimes it is found purely by chance. This neoplasm can be considered polyphenotypic on morphological, immunohistochemical, and genetic analyses. Generally, on microscopic examination, UTROSCT shows a predominant pattern of the cords, nests, and trabeculae typical of sex cord tumors of the ovary while, immunohistochemically, it is characterized by a co-expression of epithelial, smooth muscle, and sex cord markers. The aim of this review is to report clinical and pathological data and genetic alterations to establish its impact on the prognosis and management of patients affected by this rare entity.

Keywords: uterine tumor resembling ovarian sex cord stromal tumor; uterine mesenchymal neoplasm; polyphenotypic neoplasm; prognosis

1. Introduction

Uterine Tumor Resembling Ovarian Sex Cord Tumor (UTROSCT) is a rare mesenchymal neoplasm of the uterus, accounting for less than 0.5% of all uterine malignancies and 10 - 15% of mesenchymal uterine malignancies [1]. Morphologically, this neoplasm resembles ovarian sex cord tumors, without a component recognizable as an endometrial stroma. In 1945, Morehead and Bowman first described a case of UTROSCT as a uterine neoplasm resembling a granulosa cell tumor of the ovary [2]. Later, in 1976, Clement and Scully described this entity as a uterine neoplasm characterized by the presence of a component with sex cord differentiation and subdivided it into two groups based on morphological and prognostic features [3]. Group I corresponded to an endometrial stromal tumor with foci of sex cord differentiation (ETSCLE) < 50%, associated with recurrences and metastases. On the contrary, Group II was composed predominantly or exclusively by sex cord-like elements and was named Uterine Tumor Resembling Ovarian Sex Cord Tumor (UTROSCT). According to the current World Health Organization (WHO) classification, UTROSCT is included under "Miscellaneous mesenchymal tumors", and is still considered a uterine neoplasm with a component that resembles those seen in ovarian sex cord tumors, but without a component recognizable as endometrial stroma [4].

The histogenesis of these rare neoplasms is still unknown. However, many theories about the histogenesis of UTROSCT have been suggested. According to some authors, the neoplasm could arise from ovarian sex cord cells which have been displaced during embryogenesis. Instead, others think that UTROSCTs could arise from an uncommitted mesenchymal stem cell, from overgrowth of sex

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cord elements within an endometrial stromal neoplasm, adenosarcoma, or within foci of adenomyosis and endometriosis [3,5–10]. However, molecular studies have demonstrated that UTROSCTs have no molecular alterations found in endometrial stromal tumors, such as JAZF1-JJAZ1 or PHF1 fusion [7]. In addition, although UTROSCTs have a similar morphology to ovarian adult granulosa cell tumors and sex cord-stromal tumors, they do not present the same mutations, such as DICER1 and FOXL2 mutations, which can be observed in these neoplasms [11,12]. The association with the use of tamoxifen for the treatment of breast carcinoma and UTROSCTs in some cases reported in the literature could suggest a correlation with this drug [13–18]. Because of its rarity and peculiar morphological findings, this neoplasm can pose many problems for pathological diagnosis. The aim of this review is to report clinical and pathological data and genetic alterations to establish its impact on the prognosis and management of patients affected by this rare entity.

2. Clinical Data

As regards age, this neoplasm often occurs in perimenopausal or menopausal women [19]. However, in a more recent and large series reported by Boyraz et al, the patients' age ranged from 21-84 (mean: 52.4; median: 53) years [20], moreover in the literature many cases have been reported in which the patients can be <40 years [16,21–32] or are very young [33,34].

The most common symptom found is abnormal vaginal bleeding (67.1%) [35]. Other symptoms observed are pelvic pain (5.1%), palpable pelvic masses (2.5%) [35]. In rare instances extra-uterine symptoms have been observed, due to hormonal alterations. Thus, in a case reported by Dimitriadis, the presence of UTROSCT was associated with hyperprolactinemia and galactorrhea, which resolved after tumor resection [36]. Moreover, in the case described by Suzuki et al a patient with UTROSCT had elevated serum calcium and parathormone (PTH-rP) levels and in this case the tumor production of PTH-rP was demonstrated by normalization of serum PTH-rP after a tumorectomy and by the presence of immunoreactivity for PTH-rP in the tumor cells. [37].

In other cases, UTROSCT can be asymptomatic, and can be found by chance [15,20,35].

Most tumors exhibit benign behavior. In fact, we have found small case series in which the patients were free of recurrences and metastases. [15,20,32,38]. As a result, there are many examples of UTROSCT being treated with conservative surgery [16,22–32,39–41] and in only three of these cases [30,31,41] there was a recurrence observed after conservative surgery, which could well be due to previous incomplete surgery. Thus, as suggested by Watrowski et al and Carbone et al, in young women, fertility-sparing surgery should be offered to patients who wish to preserve their fertility [28,32]. That said, it is important to advise such patients that the neoplasm may recur [25].

In addition, close follow-up should be made after conservative surgery while radical surgery should be considered after a pregnancy [28,32]. In fact, although, UTROSCT usually exhibits benign behavior with a favorable prognosis, this neoplasm is classified as being of uncertain malignant potential, given its low rate of recurrence and the fact that it can sometimes cause metastases. In table 1 we have summarized the cases of UTROSCT which we found in literature from 1986 to 2023, that recurred or caused metastases and occasionally also death [19,20,30,31,36,40–61] **(Table 1).**

Table 1. CASES OF UTROSC WITH RECURRENCES AND /OR METASTASES.

Authors and year (malignant cases/t	Age (ys)	Surgery	Gross appearance	Tumor size (cm)	Microscopic appearance (architecture / rhabdoid features)	Nuclear atypia	Mitotic rate	Tumor margins	LVI	Necrosis	Stage	Molecular findings	Site and time of recurrence/ metastasis

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otal of cases)												
Kanteli p et al., 1985 (1/1 case) **ref (42)**	86	Total hysterect omy and bilateral salpingo-oophorect omy	Multiple intamyo metral cysts filled with hemorrag ic and necrotic fluid, surround ed by a fibrotic capsule	Ran ging from 2 to 10	Cords, trabecula e, nests, tubules	Not signifi cant	<1/1 0 HPF	Well circums cribed	Abs ent	Pres ent	NR	NR	One nodule in the left ovary and two epiploic nodules present at the time of diagnosi s
Bierma nn et al., 2007 (1/1 case) **ref (43)**	68	*NR*	Graysh-yellow intramyo metral nodule	4.5	Tubules, pseudoro settes, sheets, cords	Not signifi cant	Ki-67 < 5%	Focally infiltrati ve	Abs ent	NR	NR	NR	Mesenter y and small bowel (48 mo)
O'Mear a et al., 2009 (1/1 case) **ref (44)**	35	Hysterect omy	Soft and partly cystic yellow mass	9.9	Sertolifor m	NR	Ki-67 = 5%	Expansi ve /serosal infiltrati on	NR	NR	NR	NR	Bladder, abdomin al and intestinal wall, peritone um, ovaries, lymph nodes (36 mo)
Umeda et al., 2014 (2/2 cases) **ref (45)**	38	Transvagi nal myomect omy followed by total hysterect omy and bilateral salpingo-oophorect omy + LND	Yellowish -white intramyo metral mass	4.5	Solid, cords, tubules	Not signifi cant	1/10 HPF	Infiltrati ve	Pres ent	Abse nt	NR	NR	Left internal iliac lymph node methasta sis present at the time of diagnosi s
	57	Total hysterect omy and bilateral	Polypoid submucos al mass	6.4	Cords, tubules	Not signifi cant	0/10 HPF	Infiltrati ve	Pres ent	Abse nt	NR	NR	Nodule in the epiploic appendi

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		salpingo-oophorectomy										x at the time of diagnosis
Mačák et al., 2014 (1/1 case) **ref (46)**	53	Polyectomy followed by hysterescectomy and bilateral salpingo-oophorectomy	Polypoid mass and intramyometral mass	1.5 and 1.5	Solid nests, trabeculae, ribbons	Not significant	<1/10 HPF	NR	Absent	Absent	NR	NR	Pelvic limph node methastatis at the time of diagnosis
Liu et al., 2015 (1/6 cases) **ref (40)**	50	Trans-vaginal submucosal myomectomy	Isthmus mass protruding through cervical os	4.5	NR	NR	NR	Well circumscribed	Absent	NR	NR	NR	Local recurrence (10 mo)
Jeong et al., 2015 (1/1 case) **ref (47)**	32	Submucosal resection	Submucosal protruding mass	3.6	Cords, tubules and nests	NR	NR	Infiltrative	NR	NR	NR	NR	Local recurrence (17 mo)
Gomes et al., 2015 (1/1 case) **ref (48)**	53	Supracervical hysterectomy followed by bilateral salpingo-oophorectomy, omentectomy, parametrectomy, uterine cervical resection + LND	Myometrial mass	12	Cords, trabeculae	NR	NR	Infiltrative	Present	Present	NR	NR	Parametrial and right ovarian hilum involvement present at the time of diagnosis
Endo et al., 2016 (1/1 case) **ref (49)**	62	Hysterectomy	NR	NR	Trabeculae, nests, cords	Not significant	Ki-67 < 5%	Myometral infiltration	NR	NR	NR	NR	Pelvic limph-node (23 yr)

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Viau et al., 2017 (1/2 cases) **ref (50)**	43	Total hysterectomy and bilateral salpingo-oophorectomy + debulking	Peduncolated solid mass attached to the uterine fundus, with ruptured surface and whitish-yellow colour and solid subserosal mass in the anterior myometrium	13 and 5.5	Solid, trabeculae	NR	NR	NR	NR	NR	IIB	NR	Pelvis (40 mo). Sigmoid serosa and peritoneal implants present at the time of diagnosis.
	24	Submucosal resection	Submucosal nodule	NR	NR	NR	NR	NR	NR	NR	NR	NR	Local recurrence (9 mo)
Schraag et al., 2017 (2/3 cases) **ref (30)**	28	Partial resction (due to intraoperative complication) followed by fertility sparing surgery. Hysterectomy + partial bilateral salpingectomy after delivery	Cystic-solid tumor in the anterior uterine wall	10	NR	NR	NR	NR	NR	NR	NR	NR	Initial persistence of disease (3 mo). Pelvis, peritoneum, right fallopian tube, ovaries, vagina (39 mo)
Moore and McCluggage, 2017 (8/34 cases)	44	Hysterectomy	NR	12.5	NR	Not significant	≥2/10 HPF	NR	Absent	Absent	NR	NR	Para-aortic limph nodes, peritoneum (11 mo)

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ref (51)												Para-aortic limph nodes, retroperitoneal and sacral metastatis present at the time of diagnosis
	75	NP	NR	NA	NR	Not significant	≤1/10 HPF	NR	Absent	Absent	NR	NR
	62	Hysterectomy	NR	7	NR	Not significant	≤1/10 HPF	NR	Absent	Present	NR	NR	Peritoneum, lung (33 mo)
	43	Hysterectomy	NR	1	NR	Not significant	≥2/10 HPF	NR	Absent	Absent	NR	NR	Pelvis, peritoneum (25 mo)
	47	Hysterectomy	NR	6	NR	Not significant	≤1/10 HPF	NR	Absent	Present	NR	NR	Vertebra, ovary (78 mo)
	68	Hysterectomy	NR	8	NR	Significant	≥2/10 HPF	NR	Absent	Absent	NR	NR	Death (12 mo) Peritoneum, liver (11 mo)
	61	Hysterectomy	NR	12.5	NR	Not significant	≥2/10 HPF	NR	Present	Present	NR	NR	Death (23 mo) Vertebra, clavicle (12 mo)
	72	Hysterectomy	NR	7	NR	Not significant	≥2/10 HPF	NR	Absent	Absent	NR	NR	Death (23 mo) Liver (23 mo)
Kuznicki et al., 2017 (1/1 case) **ref (52)**	49	Cytoreductive surgery post NACT	Intramyometrial mass	6	Trabeculae, rosette-like structures, nests, tubules	NR	High	Myometral invasion / tumor present at 1mm from	Present	NR	NR	NR	Death (15 mo) Liver, peritoneum and pelvis (4 mo). Bilateral ovarian and omental metastasis at the

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												time of diagnosis.
Kondo et al., 2018 (1/1 case) **ref (53)**	69	Hysterectomy	NR	NR	NR	NR	NR	NR	NR	NR	NR	Lung (36 mo)
Cömert et al., 2018 (1/1 case) **ref (35)**	61	Total hysterectomy and bilateral salpingo-oophorectomy	Endometrial located tumor, suspect for superficial myometrial invasion	7	Cords, trabeculae, nests, tubules	NR	2/10 HPF	Infiltrative	Absent	Absent	NR	NR	Pelvis, omentum and splenic hilum (60 mo) Pelvis, anterior surface of the abdominal wall (75 mo)
Marrucci et al., 2019 (1/1 case) **ref (54)**	54	Total hysterectomy and bilateral salpingo-oophorectomy	Grayish-yellow intramyometral mass with poorly delineated margins	9	Trabeculae, alveolar-like structures, tubules	NR	3/10 HPF	NR	NR	NR	NR	NR	Vaginal vault (59 mo)
Croce et al., 2019 (1/1 case) **ref (55)**	70	Total hysterectomy and bilateral salpingo-oophorectomy	Yellow, well circumscribed myometrial mass	10	Diffuse, tubules, nests, trabeculae / focal rhadoid cells	Not significant	1/10 HPF	Well circumscribed, with serosal involvement	NR	NR	NR	GREB1-CTNNB1 fusion	Pelvis (17 mo) Lung and peritoneum (29 mo)
Goebel et al., 2020 (1/26 cases) **ref (56)**	74	Hysterectomy	NA	13	Cords, trabeculae, sertoliform, retiform	NR	1/10 HPF	Infiltrative	Absent	NR	NR	GREB1-NCOA2 fusion	Pelvis (66 mo)
Kaur et al., 2020 (1/6 cases) **ref (57)**	47	Total hysterectomy and bilateral salpingo-oophorectomy	Mass in the uterine body	9.3	Cords, nests, trabeculae, sheets	Not significant	1-3/10 HPF	Infiltrative	NR	Absent	NR	NR	Pelvis, retroperitoneal lymph nodes, lungs (7 mo)

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Chang et al., 2020 (1/1 case) **ref (58)**	57	Total hysterectomy and bilateral salpingo-oophorectomy	Intramyometrial, well circumscribed, soft, yellow mass	10	Diffuse sheets, nests, cords, trabeculae, glands	Not significant	3/10 HPF	Infiltrative	NR	Absent	IB	GREB1-NCOA2 fusion	Pelvis (30 mo)
	37	Hysterectomy	Yellow white myometrial mass	NA	Sheets, cords, trabeculae, occasional pseudopapillary apperean ce / extensive Rhabdoid component	NR	0/10 HPF	Infiltrative	Suspect	Absent	I	ESR1-NCOA2 fusion	Pelvis (7 yr)
Bennett et al., 2020 (3/3 cases) **ref (19)**	54	Supracervical hysterectomy followed by complection trachelectomy and staging	Multiple red brown myometrial nodules	Ranging from 1.5 to 6.5	Sheets, cords, trabeculae, occasional pseudopapillary apperean ce /extensive rhabdoid component	NR	4/10 HPF	Infiltrative	Absent	Absent	II	ESR1-NCOA2 fusion	Pelvis (9 yr). Cervical involvement and paratubal soft tissue localization at the time of dagnosis
	30	Hysterectomy	NR	NR	NR	NR	NR	NR	NR	NR	NR	ESR1-NCOA2 fusion	Omentum (32 yr and 34 yr) Rectosigmoid nodule (38 yr)
Dimitriadis et al., 2020 (1/1 case)	46	Hysterectomy followed by bilateral salpingo-	Well circumscribed uterine mass	11	Sheets, cords, nests, trabeculae	NR	6/10 HPF	NR	NR	NR	NR	NR	Intra-abdominal receurrence (24 mo)

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ref (36)	oophorectomy											
Devereaux et al., 2021 (1/1 case) **ref (59)**	42	Myomectomy	Myometrial based lesion	8.8	Clusters, cords	NR	2/10 HPF	NA	NR	Present	NR	GFT2A1-NCOA2 fusion (in recurrent tumor)	Uterus, bilateral ovarian surfaces, peritoneum, large bowel serosa, anterior abdominal wall (6 mo)
Dondi et al., 2021 (1/1 case) **ref (31)**	24	Myomectomy	Submucosal mass	3	NR	NR	1/10 HPF	NR	NR	Absent	NR	NR	Local recurrence (20 mo)
Boyraz et al., 2023 (5/75 cases) **ref (20)**	32	Hysterectomy or excision with negative margins	NA	11	Diffuse, cords	Moderate to severe	6/10 HPF	Infiltrative, with serosal involvement	Absent	Absent	NR	NR	Lung metastasis present at the time of diagnosis
	47		NA	13	Diffuse, cords / extensive rhabdoid component.	Moderate	7/10 HPF	Infiltrative, with serosal involvement	Absent	Present	NR	NR	Peritoneum (60 mo)
	58		NA	7	Diffuse, cords	Moderate	4/10 HPF	Infiltrative	Absent	Absent	NR	NR	Peritoneum (144 mo)
	68		NA	13	Diffuse, cords	Moderate	7/10 HPF	Infiltrative, with serosal involvement	Absent	Absent	NR	NR	Death (96 mo) Peritoneum (60 mo)
	73		NA	3	Diffuse, cords	Moderate to severe	9/10 HPF	Infiltrative	Absent	Present	NR	NR	Death (50 mo) Brain (30 mo) Femour (48 mo)

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Bini et al., 2023 (4/4 cases) ***ref (60)***	NR	NR	NR	NR	NR	NR	4/50 HPF	NR	NR	NR	NR	ESR1-NCOA2 fusion	All patients had metastatic disease and recived several sistemic treatments. After a median of 13.5 years of follow up (6 to 34 years), 3 patients died of disease.
	NR	NR	NR	NR	NR	NR	8/50 HPF	NR	NR	NR	NR	ESR1-NCOA3 fusion	
	NR	NR	NR	NR	NR	NR	5/50 HPF	NR	NR	NR	NR	ESR1-NCOA2 fusion	
	NR	NR	NR	NR	NR	NR	1/50 HPF	NR	NR	NR	NR	NA	
Bi et al., 2023 (7/22 cases, 1 already reported by *Chen et al., 2021*) **ref (40)**	33	Total hysterectomy and bilateral salpingo-oophorectomy + LND	Endometrial thickening, intramyometral mass	2	Retiform, papillae, nests, diffuse, whorls, sex cords	Not significat	<1/10 HPF	Infiltrative	Absent	Absent	III C	GREB1-NCOA2 fusion	Death (177 mo) Retroperitoneal recurrence with abdominal aorta involvement (167 mo) Pelvic lymph nodes involvement presents at the time of diagnosis
	48	Total hysterectomy and bilateral salpingo-oophorectomy	Intramyometral solid mass, partially cystic	13	Nests, sex cords, sertoliform trabeculae	Not significat	1/10 HPF	Well-circumscribed	Absent	Absent	IB	GREB1-NCOA2 fusion	Pelvis and omentum (45 mo)

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	38	Total hysterectomy and bilateral salpingo-oophorectomy	Intramyometrial solid mass	NA	Nests, diffuse, sex cords / focal rhabdoid cells.	Not significat	NA	Infiltrative	Absent	Absent	NA	GREB1-NCOA2 fusion	Pelvis and omentum (101 mo)
	48	Total hysterectomy and bilateral salpingo-oophorectomy	Protuberant and intramyometrial solid mass	3.5	Diffuse, focal whorls, pseudopapillae, retiform, few cords	Not significat	<1/ HPF	Infiltrative	Absent	Absent	IIB	GREB1-NCOA1 fusion	Pelvis (13 mo). Peritoneal involvement present at the time of diagnosis.
	65	Total hysterectomy and bilateral salpingo-oophorectomy	Intramyometrial solid mass	15	Diffuse, sex cords, whorls, sertoliform trabeculae	Not significat	3/10 HPF	Infiltrative	Absent	Absent	IB	GREB1-NCOA1 fusion	Death (44 mo) Pelvis (35 mo)
	40	Polypectomy	Polypoid mass	4	Sex cords, sertoliform trabeculae / Rhabdoid component	Not significat	<1/10 HPF	Well-circumscribed	Absent	Absent	IA	ESR1-NCOA2 fusion	Local recurrence (21 mo and 64 mo)
	45	Hysterectomy	Intramyometrial solid mass, partially cystic	8	Polypoid mass Diffuse, vague sex cords	Mild to moderate	1/10 HPF	Well-circumscribed	Absent	Absent	IB	ESR1-NCOA3 fusion	Pelvis (56 mo)
Xiong et al., 2023 (6/18 cases) **ref. (61)**	41	Hysterectomy	NR	5.5	Sertoliform, nests, cords	Not significat	3/10 HPF	Infiltrative	Absent	Absent	NR	ESR1-NCOA2 fusion	Peritoneum (144.4 mo)
	46	NA	NR	2.5	Sertoliform, retiform	Not significant	2/10 HPF	Infiltrative	Absent	Absent	NR	Negative for NCOA1-3 fusion and JAZF1/SUZ12/PHF1	Death (26.3 mo) Pelvis, colon (2.5 mo)

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											rearrangement	
19	NA	NR	3	Cords, trabecula e	Not signifi cant	2/10 HPF	Infiltrati ve	Abs ent	Abse nt	NR	NA	Metastas is (site NA) (69.9 mo)
36	Hysterect omy	NR	1.5	Sertolifor m, retiform	Signif icant	10/1 0 HPF	Infiltrati ve	Abs ent	Pres ent	NR	Negative for NCOA1-3 fusion and JAZF1/SUZ 12/PHF1 rearrangem ent	Pelvis, lung (56.5 mo)
55	Hysterect omy	NR	13	Sertolifor m, cords, trabecula e	Signif icant	2/10 HPF	Infiltrati ve	Abs ent	Abse nt	NR	GREB1-NCOA2 fusion	Pelvis, colon (195.3 mo)
48	Hysterect omy	NR	2.2	Sertolifor m, retiform	NA	NA	Infiltrati ve	Abs ent	NA	NR	NA	Lung (21.1 mo)

Table Legend: HPF: High Power Field, LVI: lymphovascular invasion, mo: months, NA: Not Avaiable; NR: Not Reported; NOS: Not Otherwise Specified; NP: Not performed, ys: year.

From our review we observed that many cases with UTROSCT, which recurred or developed metastases, were observed in single case reports [31,35,42–44,46–49,52–55,58,59] or sometimes in very small case series [19,30,40,45,50,57] (Table 1). The most frequent sites of recurrences or metastases in these cases were lymph nodes, peritoneum, omentum, vaginal vault, lung, and liver [44–46,49,52–55]. Interestingly, also in these single case reports and in the small series, there are examples in which the recurrences and the metastases were observed many years later when diagnosing lymph nodes [49] in the pelvis, omentum [19] (Table 1).

Death was not reported in any of these cases, although their metastases were in the liver and lung [49,52] (Table 1).

More recently, from 2017 until now, we have found only five studies with larger series and for these it is possible to establish a recurrence or metastasis rate (Table 1) [20,41,51,56,61]. Moore and McCluggage, in their study, found 8/34 tumors which behaved aggressively as defined by either lymph node metastatic disease at diagnosis (n = 1) or recurrence (n = 7) indicating that this rate had reached 23.5%, with a follow-up ranging from 6 to 135 months (mean 39) [51] (Table 1). Moreover, in this study, three patients (23.5%) developed metastases in the liver, vertebra, and clavicle, respectively after 12 and 23 months and died from their disease [51] (Table 1).

In the series of Goebel et al, out of 26 patients, follow-up information was available for eleven of them (42.3%, 11/26), with a mean follow-up interval of 94.4 (range 1-319) months and in only one case did the neoplasm recur in the pelvis 66 months after the initial diagnosis [56] (Table 1). In a study by Boyraz et al only a minority of the cases showed a malignant outcome (5/58, 8.6%). In this work, five out of 58 patients with a follow-up (22 to 192; mean=73.2 mo) had recurrences/metastases from 30 to 144 months, and 2 died from the disease. Out of three cases with metastases, in one case the metastasis was pulmonary, and this was observed at the time of diagnosis. In the remaining two cases, the metastases developed 60 and 48 months after diagnosis and these involved respectively the peritoneum, brain, and femur. In these cases, death occurred respectively 96 and 50 months after diagnosis [20] (Table 1).

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In a study by Bi et al, the cases with recurrences or metastases were 7/22 (31.81%) and they involved the pelvic lymph nodes, pelvis, and omentum, and in one case the abdominal aorta. Death was observed in two cases 177 and 44 months after diagnosis and in one of these cases the patient already had pelvic lymph node involvement at diagnosis [41] (Table 1).

Xiong et al in their analysis of 19 cases, observed that 6 patients (31.6%, 6/19) had tumor recurrences with a median follow-up of 40.9 months (range, 1.2-195.3 months) [61]. One case was excluded due to molecular translocation suggesting an endometrial stromal neoplasm (JAZF1-SUZ12) [7,61] (Table 1). The sites of these recurrences and metastases were the peritoneum, pelvis, colon, and lung. Only one patient died 23 years after diagnosis. It is extremely interesting to note that the recurrences were observed in two cases many months after diagnosis, respectively 144 and 195 months from diagnosis [61] (Table 1).

3. Pathological Features

For the diagnosis of UTROSCT there are no specific imaging findings, so it is possible to establish that a uterine lesion corresponds to a UTROSCT exclusively on pathological examination along with an accurate morphological and immunohistochemical analysis.

3.1. Macroscopic Findings

On macroscopic examination, UTROSCT is often located in the uterine corpus and can be an intracavitary polypoid lesion mimicking an endometrial polyp which can be removed by hysteroscopy [23,62] or it can appear as a pale-yellow submucous mass, located within the muscle thickness [15,23,30,31,34,45–47,63]. A UTROSCT too can be observed as a yellowish-white intra-myometrial mass, located in the uterine corpus [34,36,41,45,46,48,52,54,55,58,59].

Sometimes the lesion can have hemorrhagic cystic necrotic areas [42] or can be a yellow cystic-solid mass [41,44], and in rare occurrences, can appear as a sub-serosal peduncle solid lesion attached to the uterine fundus [50,64,65] mimicking a sub-serosal leiomyoma. In addition, when the neoplasm shows prominent myxoid features with prominent gelatinous appearance on gross examination, a diagnosis of myxoid leiomyoma or leiomyosarcoma [66] can be suggested. As reported by Liu et al, the lesion can also be located at the isthmus and can protrude through the cervical os [40]. More rarely, a UTROSCT can be observed in the cervix. In fact, in the literature we found only four cases [21,33,37,67] that presented as cervical masses, mimicking a primary cervical carcinoma on instrumental tests, such as Computed Tomography (CT) or pelvic Magnetic Resonance Imaging (MRI), on macroscopic, histologic and cytologic examination. So, as emphasized by Dubruc et al, it is important to keep in mind that UTROSCTs can also be encountered in current cervical screening programs, and in this occurrence, can be responsible for diagnostic pitfalls [67].

3.2. Microscopic Findings

Given the rarity of this tumor, the diagnosis of UTROSCT is usually made post-operatively by histopathological and immunohistochemical analysis. Typically, this neoplasm resembles ovarian sex cord-stromal tumors, with sheets, cords, nests, trabeculae (Figure 1A), or tubules (Figure 1B), [51]. Neoplastic cells are epithelioid with scant eosinophilic cytoplasm, their nuclei are bland with minimal atypia (Figure 1A,B).

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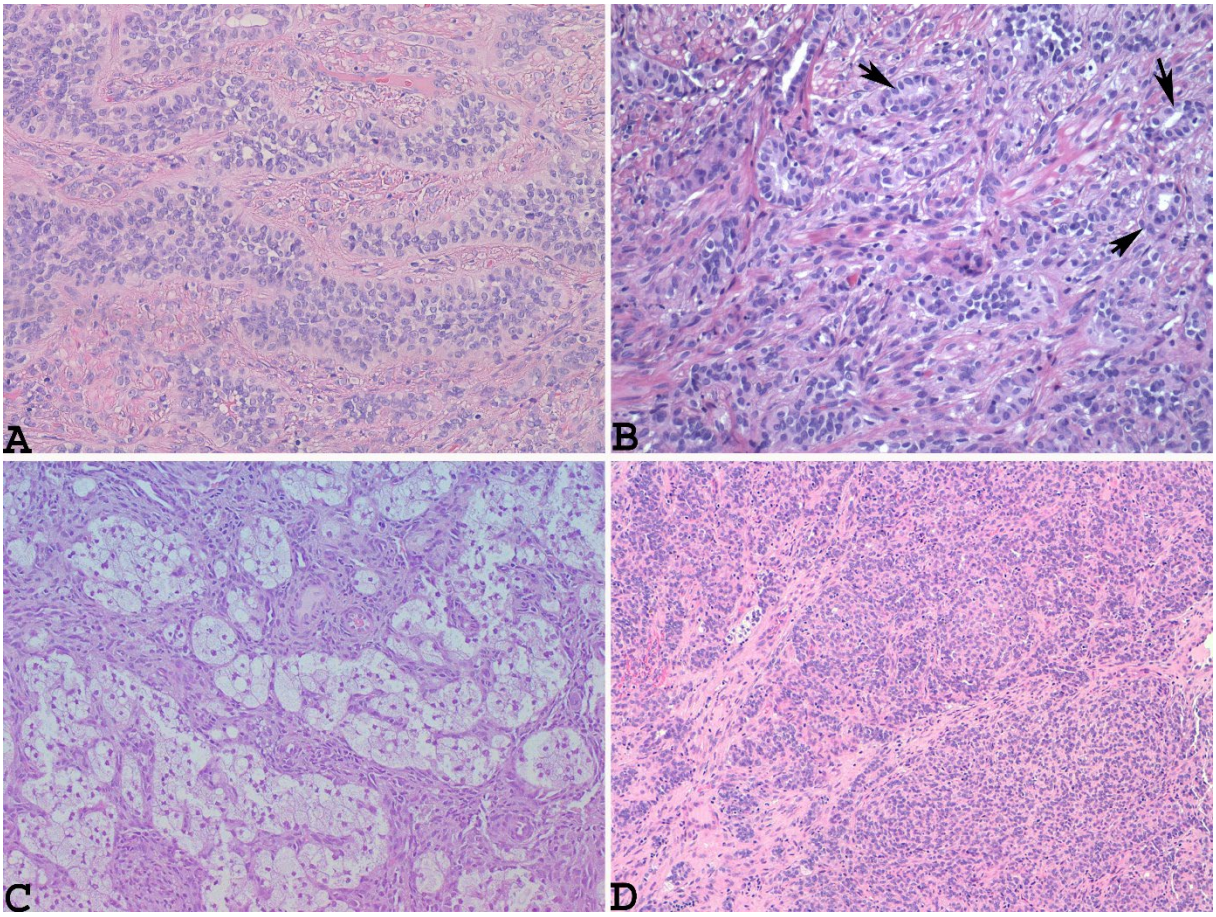

Figure 1. Microscopically, UTROSCT resembles ovarian sex cord-stromal tumors, with sheets, cords, nests, trabeculae, (A: hematoxylin-eosin x 200) or tubules (B: hematoxylin-eosin, Arrows: tubules, x200), scattered foam cells, consisting of single cells and small or larger aggregates of cells with round, central nuclei and abundant clear to foamy cytoplasm resembling foam cells macrophages or Sertoli cells (C: hematoxylin-eosin, x100) and glomeruloid structures (D: hematoxylin-eosin, x100).

Necrosis and mitoses are rare or absent [68]. Sometimes, the neoplasm may show scattered foam cells, consisting of single cells and small or larger aggregates of cells with round, central nuclei and abundant clear to foamy cytoplasm resembling foam cell macrophages or Sertoli cells [34,69,70] (Figure 1C). Glomeruloid structures (Figure 1D), and small nests or micropapillary-like structures [15,16,62,64,70] or micro follicles resembling Call-Exner bodies [20,64] can be observed, too. Another peculiar growth pattern is that of a retiform, with labyrinthine cavities and channels resembling the Rete Ovarii [15,64]. Sometimes this pattern can be prominent, mimicking, on small endometrial biopsy, an adenocarcinoma or myometrial metastasis from a previous breast cancer [15] [Nogales]. Goebel et al also observed microcystic and signet ring cell-like change and retiform patterns [56] [Goebel]. Rhabdoid cells with abundant dense eosinophilic cytoplasm and eccentric nuclei were seen and were diffuse in the example reported by Boyaz et al and Bennett et al [19,20]. Rhabdoid cells with single file growth pattern were found by other authors [55,64].

However, many patterns can be present within the same neoplastic mass causing considerable morphologic heterogeneity [15,56,64,72].

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When the neoplasm shows a predominant tubular pattern or gland-like differentiation with few associated stromal, this can frequently pose diagnostic problems, mimicking Sertoliform endometrial adenocarcinoma [73–75] or extragenital metastatic carcinoma [76]. In addition, when there is both a tubular pattern and lipid-rich cells, UTROSCT can imitate a Sertoli tumor [42].

On immunohistochemical analysis, UTROSCTs characteristically exhibit polyphenotypic immunophenotypes with co-expression of cytokeratin (Figure 2A), and smooth muscle markers, including muscle actin (Figure 2B), desmin (Figure 2 C), and histone deacetylase 8, hormone receptors [64].

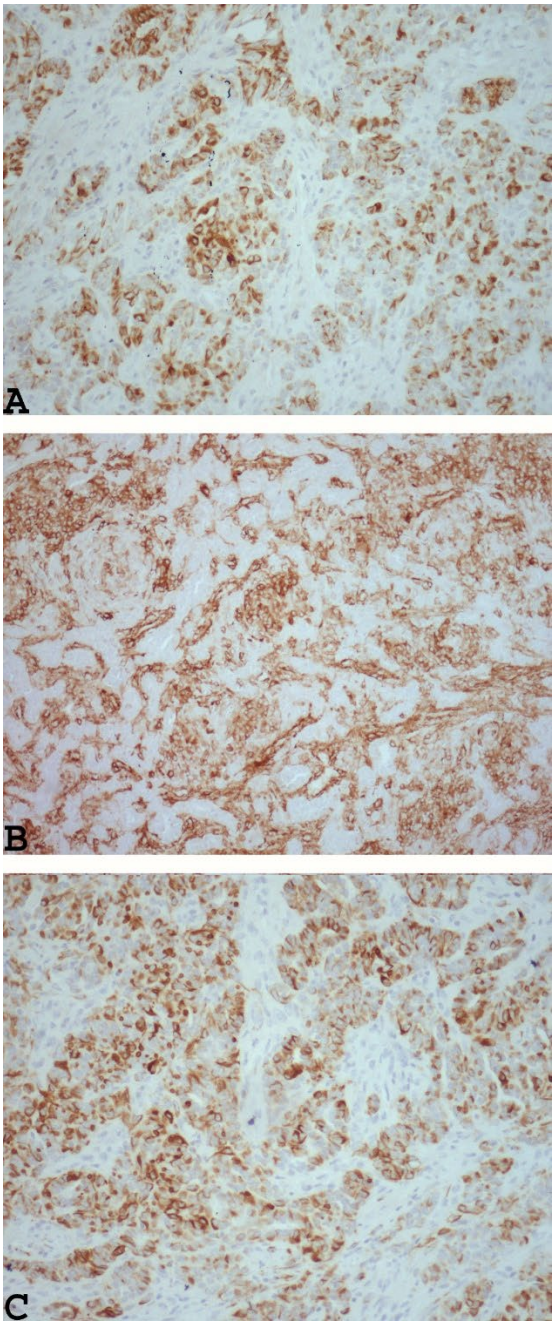

Figure 2. Example of UTROSCT exhibiting polyphenotypic immunoreactivity with co-expression of cytokeratin (A: x 200), smooth muscle markers, including muscle actin (B: x200) and desmin (C: x200).

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In addition this neoplasm show positivity for markers, which are commonly positive in ovarian sex cord-stromal neoplasms, such as inhibin (Figure 3 A**)**, calretinin (Figure 3B), CD99 Wilm's tumor protein 1 (WT1), and MART-1, [16,62,64,68] and others markers such as CD10, S100, and CD117 [63,69]. In addition, as reported by Croce et al, characteristically UTROSCT can show nuclear staining with steroidogenic factor-1 (SF-1) (Figure 3D) [55] similarly to ovarian sex cord tumors (Figure 3D).

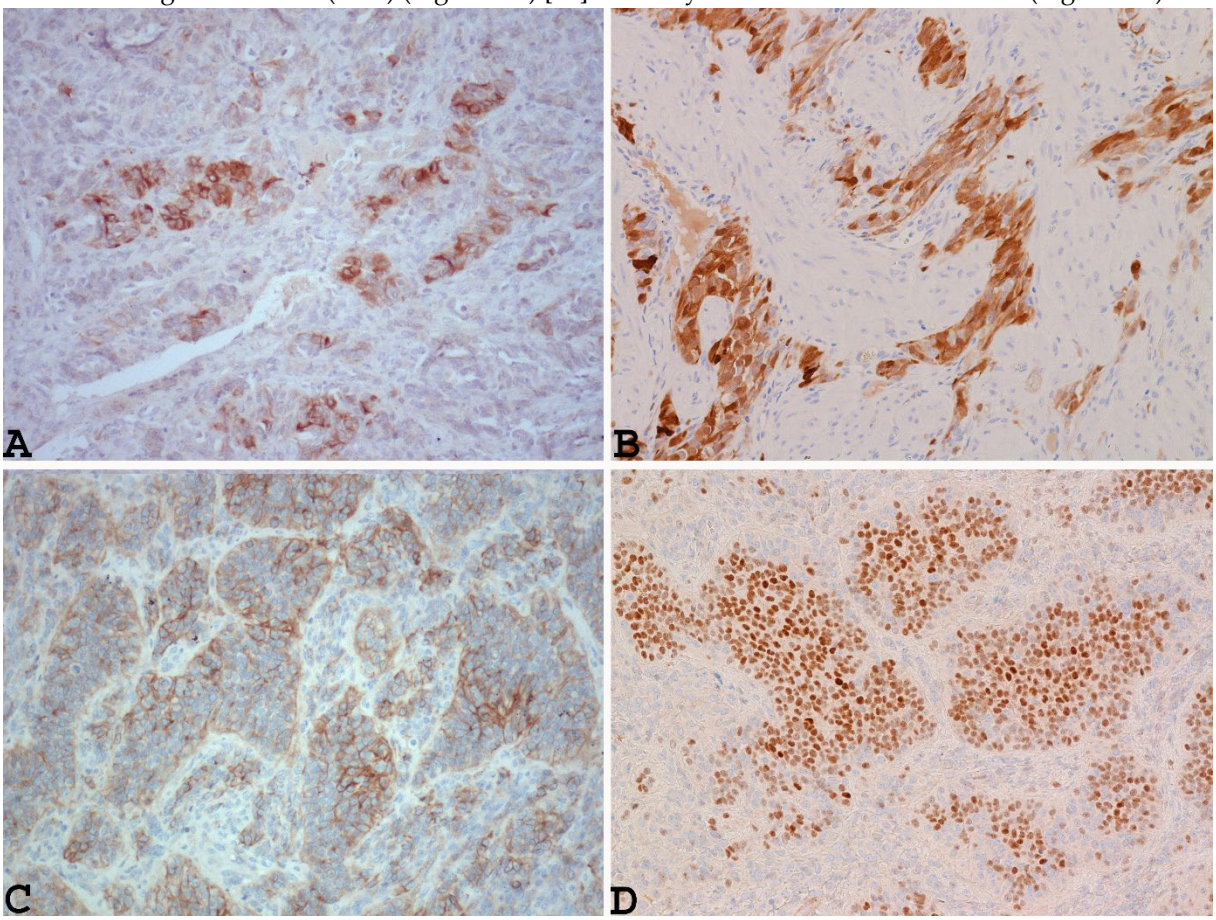

Figure 3. Example of UTROSCT with positivity for markers, which are commonly positive in ovarian sex cord-stromal neoplasms, such as inhibin (A: x 200), calretinin (B: x 200), CD99 (C: x200) and nuclear staining with steroidogenic factor-1 (SF-1) (D: x200).

Moreover, other authors [63,78], in their study inadvertently found in all the cases reported, a strong, diffuse BCL-2 positivity that could be related to peculiar genetic alterations which are translocations on chromosomes t(4; 18) (q21.1; q21.3) and t(x; 6) (p22.3; q23.1) [78] and consequently they suggested that also this marker could have a potential value in the diagnosis of UTROSCT. These peculiar immunohistochemical aspects of this neoplasm are of considerable importance in differentiating an UTROSCT from other uterine neoplasms that could present morphological similarities. The most frequent uterine neoplasms that could mimic an UTROSCT and that should be considered, are some histopathological variants of leiomyomas, such as leiomyoma with tubules, epithelioid leiomyoma, or vascular plexiform leiomyoma. Uterine leiomyoma with tubules is a biphasic neoplasm, composed of epithelial and mesenchymal elements, with intersecting fascicles of smooth muscle, tubular and gland-like structures lined by plump cells with indistinct cytoplasm [79]. Thus, histologically, this lesion simulated uterine tumors resembling ovarian sex cord tumors (UTROSCTs), but the immunophenotype was not consistent with true sex-cord differentiation, in its negativity to inhibin, CD99, CD10 and Melan-A [79]. Epithelioid leiomyoma is a subtype of leiomyoma that macroscopically appears as a well-circumscribed, intramural mass with a soft consistency, and yellow to tan cut surfaces. Microscopically, it is characterized by the presence of more than 50% round-polygonal cells and immunohistochemically shows immunoreactivity to

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epithelial and smooth muscle markers but negativity to typical sex-cord markers of UTROSCT [80]. Histologically, vascular plexiform leiomyoma is a well-circumscribed, intramural nodule with anastomosing cords and trabeculae of 2 to 3 cell layers with eosinophilic cytoplasm, indistinct cell borders and plump, slightly hyperchromatic nuclei. In addition, cord lumens contain red blood cells. On immunohistochemical analysis, the neoplastic cells in this subtype of leiomyoma are positive to smooth muscle actin (SMA), caldesmon and CD99, but are negative for inhibin-α [81].

Another uterine neoplasm that could mimic a UTROSCT is a low-grade endometrial stromal sarcoma (LGESS) in a biopsy or curettage specimen. This malignancy represents the second most common uterine sarcoma [82] with a wide range of age, but with a predilection for pre-menopausal and peri-menopausal women [83]. Moreover, LGESS, like UTROSCT, may be related to tamoxifen treatment [84]. Histologically, LGESS is composed of permeative tongue-like islands of tumor cells consisting of monotonous oval to spindle cells with minimal cytologic atypia with a whorl pattern of growth around blood vessels. Immunohistochemical analysis is particularly useful since LGESS shows positivity to CD10 and negativity for sex cord markers [85]. Moreover, these endometrial sarcomas along with ESTSCLE, show genetic alterations, such as JAZF1-JJAZ1 or PHF1 fusion that are absent in UTROSCT [7].

Plexiform tumorlets are rare tumors affecting patients with an average age of 48 to 60 years.

A plexiform tumorlet is a rare lesion usually found in the myometrium and is considered to be a variant of epithelioid leiomyoma. Multiple plexiform tumorlets may have an infiltrative pattern and mimic endometrial stromal sarcoma.

Uterine Sertoli form endometrioid adenocarcinoma, this is a rare subtype of an endometrial carcinoma, which can mimic; on morphological [73,74] and immunohistochemical analysis, an UTROSCT due to the presence of tubules and glandular structures and for its positivity to sex cord markers such as inhibin, CD99, (calretinin, WT-1, and Melan-A [75]. In such an occurrence, for a correct differential diagnosis from UTROSCT and Uterine Sertoli form endometrioid adenocarcinoma, it is important to keep in mind that in this subtype of endometrial adenocarcinoma, the remaining endometrium is affected by atypical complex hyperplasia [75] and within the neoplasms can be observed a typical endometrioid component [73].

3.3. Electron Microscopy Findings

In addition, on ultrastructural analysis, UTROSCT exhibits polyphenotypic features, with both epithelial structures such as desmosome-like junctions, tonofilaments, lumina formation, and microvilli and sex-cord-like features including nuclear indentation, abundant intracellular filaments, sparse to moderate rough endoplasmic reticula and abundant intracytoplasmic lipids [86].

4. Impact of Pathological Features on Recurrences or Metastases in UTROSC

Given that, in the literature there are many single case reports of UTROSCT [31,35,36,42–44,46–49,52–55,58,59] (Table 1) and few small case series [19,20,40,45,50,51,56,57,61], it is extremely difficult to establish which morphological aspects could predict its aggressive behavior, and with a good probability, a poor prognosis.

Moreover, in some cases with recurrences or metastases, morphological findings, which could be signs of malignancy, such as sizes [19,30,45,51,53,58], LVI [19,30,31,36,44,47,50,53–55,57–59], necrosis [19,30,36,40,43–45,47,50,52–56], number of mitoses [19,30,40,41,47,48,50,53,61], nuclear atypia [30,31,35,36,40,44,47,48,50,52–54,56,61], these are not always reported (Table 1). In addition, in the small series reported by some authors, it seems that there are cases which recurred or caused metastases, even though they presented benign morphological features such as well-defined margins [30,31,35,36,40,44,47,48,50,52–54,56,61], absence of necrosis [19,31,35,41,45,46,51,57,58,61], a low or not significant number of mitoses, [19,20,31,35,41–46,49,51,54–59,61], absence of nuclear atypia [31,42,43,45,46,49,51,55,57–59,61] and absence of LVI [19,40,42,43,46,51,56,61,2035].

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Regarding the microscopic appearance, the neoplasms did not show any particular architectural pattern, in fact. the majority of the cases reported in the literature that recurred or developed metastases, revealed the presence of cords, trabeculae, nests or tubules, [20,30,31,36,40,54,56–59,61] except for one case reported by Croce et al which also presented focal rhabdoid cells [55], or a case observed by Bennett which was characterized by the presence of an extensive rhabdoid component [19].

Although there are no clear criteria to establish a firm prognosis and malignancy level for UTROSCT, more recently, Boyraz et al in their large study with 75 cases, relating all morphological features and follow-up available for 58 women, affirmed that it is important to simultaneously evaluate many features for every neoplasm. In fact, these Authors observed that malignant tumors which developed recurrences/metastases, compared with benign neoplasms, showed > 3 of the following 5 features: size > 5 cm, moderate nuclear atypia, ≥ 3mitoses /10 (High Power Field) HPF, infiltrative border and necrosis and probably also, an extensive rhabdoid component [20]. In addition, Boyraz et al emphasized that sometimes it is impossible to define the prognosis of an UTROSCT since occasionally the entire neoplasm is not examined on microscopic examination, but only some fragments from curettage specimens. Thus, it is perfectly possible that some features of malignancy such as infiltration of myometrial tissue can be missed [20].

5. Molecular Alterations of UTROSC and its Impact on Prognosis

Initially, molecular biology studies for this neoplasm were performed to demonstrate that this entity, although it has similar morphological features to ovarian sex cords tumors, due to its peculiar genetic alterations it must be considered another type of tumor [11,12]. In addition, some authors have demonstrated by molecular analysis that UTROSCTs do not have the molecular alterations found in endometrial stromal tumors, such as JAZF1-JJAZ1 or PHF1 fusion [7,45].

In our review, we also evaluated the presence of other molecular alterations in order to establish whether these could have a prognostic significance. We found a few recent studies which reported that using fluorescence in *in situ* hybridization (FISH) and RNA- Sequencing validated by RT-PCR [19,41,55,56,58–61,72,78]. (Table 1).

Wang et al in a single case report, using fluorescence in situ hybridization (FISH) observed, in cultured cells, two balance translocations: t(4;18)(q21.1;q21.3) and t(X;6)(p22.3;q23.1) [78]. The translocations t(4;18)(q21.1;q21.3) are related to gene bcl2 and the development of particular tumors, such as more aggressive squamous cell carcinoma and some forms of acute leukemia or follicular lymphomas. The translocation t(X;6) (p22.3;q23.1) instead involved antigen regulator gene (H-Y R) which is located at p22.3 and is responsible for gonadal organogenesis. However, the molecular results in this study did not provide information on prognostic significance since the patient after a short follow-up of twelve months was well with no signs of disease [78].

On the contrary, Croce et al, in a 70-year-old patient affected by UTROSCT with ruptured uterine serosa and a focal rhabdoid component, which recurred with widespread pelvic nodules seventeen months after surgery and then also one year later developed lung metastases, despite a treatment with aromatase inhibitors, demonstrated a novel translocation t(2;3)(p25;p22) involving the GREB1 (intron 8) and CTNNB1 (exon3), using RNA- Sequencing validated by RT-PCR. This peculiar molecular alteration, observed both in primary and recurrent neoplastic tissue, was responsible for nuclear over-expression of hypo-phosphorylated and truncated beta-Catenin, which, thanks to the involvement of GREB1, was due to a response to estrogens and caused the activation of the Wtn/beta-Catenin signal pathway with a major oncogenic effect [55].

In 2019, Dickson et al, using RNA-Sequencing confirmed by (FISH), in four cases of UTROSCT first observed that this entity presents peculiar genetic alterations [72]. These genetic alterations when identified corresponded to *NCOA2/3* gene fusions in 4 cases of UTROSCT, including *ESR1-NCOA3*

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(N=2), *ESR1-NCOA2* (N=1), and *GREB1-NCOA2* (N=1) [72]. Characteristically, these neoplasms did not reveal conspicuous mitotic activity. On the contrary, one tumor was circumscribed, while the remaining three cases showed myometrial infiltration as a low-grade endometrial stromal sarcoma. The Authors, given the genetic alterations found, suggested that these could be used for an accurate diagnosis of UTROSCT [72].

Although the authors suggested that these genetic alterations may be related to malignant mesenchymal neoplasms, such as mesenchymal chondrosarcoma [87] congenital spindle cell rhabdomyosarcoma [88,89], alveolar rhabdomyosarcoma [90], Ewing sarcoma [91] or human leukemia [92], as well as some uterine sarcoma with variable sex cord differentiation [93,94] they gave no information on the follow-up of patients and consequently their prognostic significance [72].

More recently, it has been demonstrated in some studies that UTROSCTs with Growth Regulating Estrogen Receptor Binding-1 (GREB1) rearrangement may have a more aggressive biological behavior with high risks of recurrence or metastases [56,58]. Likewise, it seems that GREB1 rearranged tumors tend to be larger and more mitotically active [58].

As well as neoplasms with Estrogen Receptor-1 (ESR1)-NCOA2 fusions are more likely related to recurrences and the presence of infiltrative margins and sometimes to the presence of an extensive rhabdoid component, as demonstrated by Bennett et al [19]. In addition, tumors with GREB1-NCOA2 fusion could be more frequently related to recurrences than those with other genetic alterations [41,55].

These data suggest that these gene fusions probably cause aberrant activation of estrogen signaling pathways, with a major oncogenic effect due to the increase in proliferation and activation of neoplastic cells.

In fact, gene fusions involving three nuclear receptor coactivators, such as NCOA1, NCOA2 and NCOA3, have been demonstrated in many cases of UTROSCT [56,59]. The NCOA genes belong to a p160 family of steroid receptor coactivators, which interact with ligand-dependent hormone nuclear receptors, including the estrogen receptor-alpha (ERα), to mediate transcriptional programs, can promote a wide range of signaling pathways including cellular proliferation, metabolism, growth, and survival [92].

Considering that Programmed Cell Death Ligand 1 (PD-L1), which is a trans-membrane protein considered to be a co-inhibitory factor of the immune response and plays an important role in various malignancies, attenuating the host immune response to tumor cells, Xiong et al first correlated the expression of this marker with mitotic activity and NCOA-2 gene alteration in a small series of UTROSCTs [61]. Thus, they discovered that UTROSCT with significant mitotic activity, gene alteration of NCOA2 and a high expression of stromal PDL-1, represents the subset of a neoplasm with aggressive behavior and shorter disease-free survival. [61]. Consequently, from this study, Xiong et al suggested that UTROSCTs with aggressive behavior have a peculiar tumor microenvironment and could be treated with immune therapy in line with other neoplasms [61].

6. Conclusions

In conclusion, through our review of the literature, UTROSCT is rare neoplasm which should be considered in the differential diagnosis of other uterine masses. Similarly, to many uterine intracavitary and intramural lesions, the most common symptom of this neoplasm is abnormal vaginal bleeding. To the best of our knowledge, for pre- or perioperative diagnosis in UTROSCT, there are no studies which have evaluated serum inhibin levels, a marker for sex cord differentiation. Moreover, imaging studies such as MRI cannot be useful to identify this neoplasm because they provide findings, such as cystic degeneration, intra-tumoral hemorrhage and necrosis which are common to leiomyoma [95–97] or adenomyosis [32,98].

On the contrary, for a pathological diagnosis many studies over the last few years have contributed to improving pathological diagnosis using immunohistochemical analysis with specific markers, revealing that UTROSCT is a polyphenotypic neoplasm with variable positivity for

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epithelial, smooth muscle, neuroendocrine, sex cord markers and hormone receptors and with molecular heterogeneity [19,41,55,58,60,61].

Although UTROSCT usually exhibits benign behavior with a favorable prognosis, this neoplasm is classified as being of uncertain malignant potential, given that it has a low rate of recurrence and can sometimes cause metastases. However, close follow-up is required for all patients due to the lack of prognostic biomarkers and a long follow up is needed to evaluate the safety of conservative surgery.

A conservative surgical approach can be offered to young women who wish to preserve their fertility. However, it is important to advise the patients that the neoplasm may recur [25]. In addition, a close follow-up should be made after conservative surgery and radical surgery should be considered after a pregnancy given that, in some instances, the recurrences or metastases were observed many months after diagnosis [40,61].

Although in this rare neoplasm it is extremely difficult to establish which morphological aspects could predict its aggressive behavior, recently some authors have suggested evaluating multiple factors [20]. More recently, some molecular biology studies have revealed that UTROSCTs with GREB1::NCOA1-3 fusions [51,58] and PD-L1 molecule expression appear to be predisposed to more aggressive behavior and recurrences, GREB1::NCOA2 being the most common gene fusion in recurrent tumors [61]. As well as, recently Yin et al observed a case of UTROSC with aggressive histologic features harboring a *GREB1-NCOA2* fusion, such as increased mitotic figures (up to 3/10 high power fields), geographic necrosis, and LVI [99].

Thus, it is especially important in the case of this peculiar neoplasm to perform molecular investigations to define the most aggressive forms and to select patients with a higher risk of recurrences or metastases. Additionally, further, more numerous studies should be performed to correlate morphological findings, molecular data and clinical data with long follow up, to identify subtypes with worse prognosis.

Author Contributions: For research articles with several authors, a short paragraph specifying their individual contributions must be provided. The following statements should be used "Conceptualization, GG. and ET; methodology, DG; validation, GG and DG; formal analysis ET and DG; GGwriting—review and editing. All authors have read and agreed to the published version of the manuscript.

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