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An High-Grade Stromal Sarcoma Localized in the Left Fallopian Tube: A Diagnostically Challenging Case and a Literature Review

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SUMMARY

High-grade stromal sarcoma (HGSS) confined to the fallopian tube has not been clearly documented and may be mis-classified among other rare tubal sarcomas or malignant mixed Müllerian tumours (MMMT). A systematic search of PubMed, Scopus and Google Scholar (inception to 31 May 2025) using the terms "high-grade stromal sarcoma", "endometrial stromal sarcoma", "fallopian tube", "adnexal sarcoma", "carcinosarcoma", "malignant mixed Müllerian tumour" was performed. Reference lists of relevant articles were screened manually. Eligibility criteria included English-language case reports, series or reviews that described a sarcomatous component arising in the fallopian tube.Thirty-nine primary tubal sarcomas were identified, most commonly leiomyosarcoma (n = 22) and undifferentiated pleomorphic sarcoma (n = 5). No publication described a pure HGSS arising de-novo in the tube. Ninety-four cases of tubal MMMT/carcinosarcoma were retrieved; in four reports the sarcomatous element demonstrated high-grade stromal morphology but was biphasic with carcinoma. One recent case illustrated epithelial–mesenchymal transition of a high-grade serous carcinoma into carcinosarcoma at metastatic sites. Molecular data from extra-uterine high-grade endometrial stromal sarcoma (HG-ESS) highlight recurrent YWHAE-NUTM2 and BCOR alterations, but such profiles have not been demonstrated in tubal lesions. To date, a primary high-grade stromal sarcoma of the fallopian tube has not been confirmed. Sarcomatous differentiation in the tube most often represents MMMT or leiomyosarcoma. Accurate histo-molecular work-up is essential to avoid mis-classification and to guide management.

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Introduction

Sarcomas account for <1 % of gynaecological malignancies, and those of tubal origin are exceedingly rare [1]. High-grade stromal sarcoma (HGSS) is recognised primarily in the uterus (high-grade endometrial stromal sarcoma, HG-ESS) and exceptional extra-uterine sites [2]. A pure HGSS of the fallopian tube has not yet been described; however, tubal sarcomas such as leiomyosarcoma and malignant mixed Müllerian tumours (MMMT, also termed carcinosarcoma) have been reported sporadically [3-6]. Furthermore the symptoms are usually nonspecific and can include pelvic pain, abnormal vaginal bleeding, abdominal bloating, or a mass effect. Nevertheless, these symptoms often resemble those of other more common gynecological conditions, such as ovarian cysts or endometriosis, making it challenging to diagnose early. In addition, due to its rarity, the definitive diagnosis is typically made through biopsy and histopathological examination. In some cases, the diagnosis may not be made until after surgery.

In this article, we performed a review of literature and presented the first case of an incidental diagnosis of primary high grade stromal sarcoma of the fallopian tube. This review consolidates the available evidence on high-grade sarcomatous tumours of the fallopian tube, with emphasis on stromal morphology and molecular features.

Case Presentation

A 68-years-old postmenopausal woman (Gravida 2, Para 2) experienced an abnormal vaginal bleeding episode. Transvaginal ultrasound was performed detecting, in the mid-pelvic region, a voluminous fluid collection of 11 cm cranially continuous with the vaginal vault; this find was consistent with a large hydrometra, likely due to cervical canal stenosis. At gynecological examination, the uterine body was enlarged with reduced consistency, and adnexal areas were free. Speculum examination revealed scar stenosis of the cervical canal as a sequela of prior conization [7].

The patient subsequently underwent second-level investigations. At abdominal MRI the uterus was fibromatous, enlarged, (80x75x120mm) and globular due to homogeneous fluid overdistension of the cavity. The left fallopian tube showed

fluid distension, with a maximum diameter of about 14 cm in the ampullary region.

Figure MRI

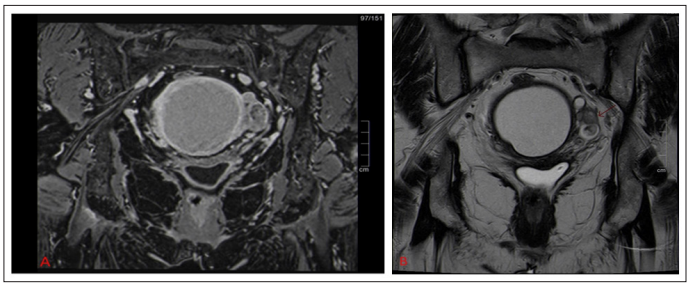

Figure 1: A, B

Protruding lesion within the lumen of the fallopian tube, showing heterogeneous signal on T2-weighted imaging and contrast enhancement. Associated hematometra with distension of the uterine cavity. Size: 28×9 mm.

A minimally invasive approach was attempted, but the hematometra was found to be non-drainable due to stenosis of the vaginal canal.

Total laparohysterectomy with bilateral salpingo-oophorectomy for non drainable hematometra due to cervical scar stenosis was performed without periprocedural complications.

At gross examination the uterus measured 9x6x4.5 cm with adnexa. An intramural myoma measuring 1x1 cm was documented in the anterior wall. The right adnexa consisted of a salpinx measuring 6 cm in length and an ovary measuring 2.2x1.5x0.8 cm. The right adnexa consisted of a salpinx 11 cm in length with a maximum dilation of 2 cm and an ovary measuring 2.5x1.5x1.5 cm.

Haematoxylin and eosin (HE) slides revealed a proliferation of oval/spindle cells with pleomorphic nuclei appearance and atypical mitoses (42/10 HPF). Necrosis was not documented.

The histology indicated no evidence of direct invasion or metastasis into the uterus, left ovary and right adnexa.

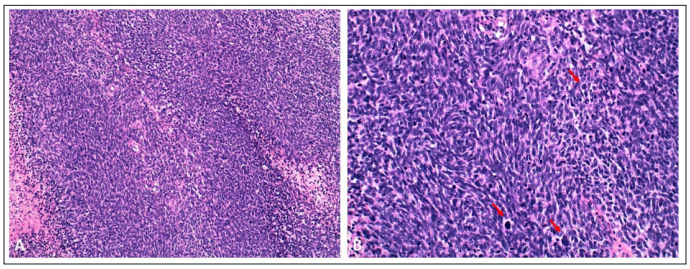

Figure 2: A-B: Proliferation of Uniform Atypical Spindle Cells and Brisk Mitoses (Red Arrows)
A: H&E Magnification 10x
B: H&E Magnification 20x

Immunohistochemistry (IHC) was performed resulting as follows: p53 (+), p16 (+), Cyclin D1 (+), Vimentin (+), CD56 (+), ER (-), PGR (-), Actin1a4 (-), Desmin (-), Caldesmon (-), SOX10 (-), S100 (-), Chromogranin (-), Synaptophysin (-), EMA (-), AE1/AE3 (-), CD99 (-), CAM5.2 (-), and Ki67 proliferation index (95%). IHC staining was performed using a BenchMark XT automated immunostaining system (Ventana Medical System, Tucson, AZ, USA).

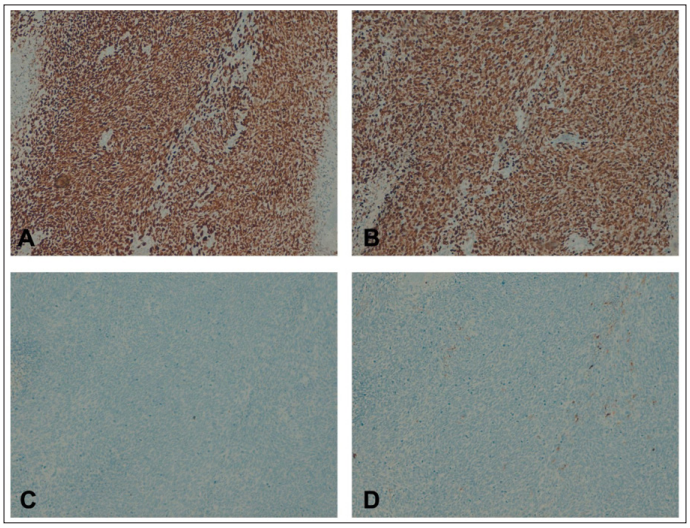

Figure 3: A-D: Immunohistochemistry Performed.
A: p53, **B:** Ki-67, **C:** Caldesmon, **D:** S-100 (A-D: magnification 10x)

A formalin-fixed and paraffin-embedded (FFPE) sample was subjected to DNA extraction. DNA was obtained by manually dissecting 2 × 10 µm sections of FFPE tubal sample. Genomic DNA (gDNA) was extracted using "MagCore genomic DNA FFPE one-step kit" (Diatech-labline pharmacogenetics, Jesi, AN).

After DNA extraction, the sample was sequenced using a 50-gene panel by means of Next Generation Sequencing (Myriapod NGS-L T 50 G Oncopanel Kit, Diatech-Labline Pharmacogenetics, Jesi, Italy) on the Ion S5 System Ion Torrent platform. Analysis of the NGS results and identification of any genetic variants in the target regions were carried out using the "Myriapod NGS Data Analysis Software".

Molecular results and their clinical significance were analysed and interpreted by referencing online public databases (ClinVar and COSMIC). Only referenced pathogenic and disease-related variants were reported.

The analysis identified a pathogenic variant in the TP53 gene: c.713G>Tp.Cys238Phe, with a variant allele frequency of 70.60%.

According to the current surgical staging for tubal cancer, this tumor was staged as FIGO IC. Post surgical assessment with chest and abdominal CT with contrast showed a 3mm micronodule with a faint ground-glass halo in the apicoposterior segment of the left upper lobe, subpleural, indeterminate due to its small size, potentially inflammatory but requiring follow-up. Two additional micronodules along a bronchovascular branch in the same segment, prescisural, likely of similar nature. Given the rarity of the histology and the limited number of cases in the medical literature, a histological review was conducted in a reference center in Padua.

The histological examination confirms the presence of high grade malignant spindle cell mesenchymal neoplasm, consistent with a diagnosis of undifferentiated spindle cell sarcoma G3 sec. FNCLCC. PDQ Adult Treatment Editorial Board. Soft Tissue Sarcoma Treatment (PDQ®): Patient Version. 2023 Jun 2. In: PDQ Cancer [8].

Information Summaries [Internet]. Bethesda (MD): National Cancer Institute (US); 2002–. PMID: 26389216. ref

Immunohistochemistry (IHC) was performed resulting as follows: Cyclin D1 (+), ER (-), BCOR (-) SS18-SSx (-), Actin1a4 (-), Desmin (-), Caldesmon (-), Cytokeratin MNF116 (-) S100 (-).

The case was discussed in a multidisciplinary setting, with a recommendation for an oncology consultation.

The indication, given the limited size of the tumor, did not include any type of complementary treatment. We chose an active surveillance and the patient started her follow up.

The last CT scan assessment was performed in June 2025 and did not show tumor recurrence with stability of the pulmonary micronodes.

Discussion

Primary fallopian tube sarcoma is an exceedingly rare malignancy characterized by high metastatic potential, frequent recurrences and cancer-related deaths [9]. It can occur during the entire life with a median age of incidence of 47 years. Due to its rarity, the prognostic factors are difficult to evaluate. Nevertheless, sarcoma of the fallopian tube is associated with poor prognosis [10]. In 1886 Senger et al. were the first to describe a case of pure fallopian tube sarcoma, and since then, only a small number of cases have been reported in literature.

High-grade stromal morphology observed in tubal tumours is invariably associated with MMMT or represents mis-classified undifferentiated/leiomyosarcoma. Our case represents, to our knowledge, the first documented instance of a molecularly confirmed high-grade endometrial stromal sarcoma arising in the fallopian tube.

We conducted a narrative review of the literature through May 2025, using PubMed, Scopus, and Google Scholar. In our review, 39 cases were identified, most commonly leiomyosarcoma (56%), followed by undifferentiated pleomorphic sarcoma, rhabdomyosarcoma, and fibrosarcoma. None had confirmed molecular features of HG-ESS.

Table 1 summarizes the reported histologic types. Notably, HGSS morphology has been observed only in biphasic tumors like MMMT (carcinosarcomas), not in pure stromal lesions.

Primary Tubal Sarcomas

Thirty-nine eligible primary sarcomas were identified (median age 49 years). Leiomyosarcoma comprised 56% (22/39); other histotypes included rhabdomyosarcoma (n = 3), fibrosarcoma (n = 2) and undifferentiated pleomorphic sarcoma (n = 5). None fulfilled criteria for HGSS or demonstrated YWHAE-NUTM2 fusion testing.

Malignant Mixed Müllerian Tumours / Carcinosarcomas

Ninety-four MMMT cases were reported between 1986 and 2024 [5, 6, 8].

Sarcomatous components encompassed homologous high-grade spindle cell sarcoma and heterologous elements (cartilage, skeletal muscle). Four reports described a dominant high-grade stromal component, but always within a biphasic tumour [5, 8]. A 2024 case illustrated transformation from high-grade serous carcinoma (HGSC) to carcinosarcoma at metastatic sites via EMT [8].

Carcinosarcomas of the tubes are not associated with a specific clinical presentation. Patients can be either asymptomatic in the early stages of the disease, or the disease may begin with abdominal pain or, less frequently, with abnormal uterine bleeding. Furthermore, it is not associated with the increase of tumor markers such as serum carcinoma antigen125 [11]. High-grade stromal morphology observed in tubal tumours is invariably associated with MMMT or represents mis-classified undifferentiated/leiomyosarcoma. Our case represents, to our knowledge, the first documented instance of a molecularly confirmed high-grade endometrial stromal sarcoma arising in the fallopian tube. In this case the diagnosis was accidental as the tumor was associated with hydrometra,a common gynecological disease, especially in postmenopausal women Inflammation is regarded as the most important cause of hydrometra Jianfa Wu et al investigated the predictive value of hydrometra in gynecological tumors resulting as a risk factor for endometrial cancer and cervical cancer [12, 13]. No mention has been made regarding sarcomas.

Furthermore, in this case we found a somatic mutation of TP53. TP53 mutations are the most frequent alteration, found in 20% of sarcomas. Disease-free survival of localized sarcomas was shorter in TP53 mutated sarcomas [14].

Also in gynecological sarcomas, TP53 mutations, particularly in uterine sarcomas, can have significant implications for diagnosis, prognosis, and treatment [15]. Due to the rarity of this neoplasm, there are no specific guidelines and no validated genomic marker to guide decisions of peri-operative systemic treatments. Management follows uterine sarcoma protocols: radical surgery and adjuvant chemotherapy for high-risk disease, although evidence remains anecdotal [4, 5]. Similar to the treatment for uterine sarcoma, lymphadenectomy is not necessary because hematogenous metastases are the major diffusion pathway.

Sarcoma in general has a low sensitivity to radiation therapy. Randomized trial has failed to show survival benefit for adjuvant RT in stage I or II sarcoma. Various chemotherapeutic regimens used for the treatment of fallopian tube sarcoma include vincristine, actinomycin D, cyclophosphamide, doxorubicin, 5-fluorouracil, dacarbazine, and cisplatin. Furthermore chemotherapy regimens include doxorubicin/cisplatin gemcitabine/docetaxel, pirarubicin/ifosfamide (You D. et al). Collaboration through rare tumour registries will be pivotal to improving knowledge and outcomes.

Table 1: Summary of Reported Primary Sarcomas of the Fallopian Tube (n = 39)

Histologic Type	Number of Cases (n)	Age Range (yrs)	Immunohistochemistry (where available)	Molecular Testing	Comments
Leiomyosarcoma (LMS)	22	35–72	Desmin+, SMA+, h-caldesmon+	Not reported	Most common subtype; variable outcomes
Undifferentiated Pleomorphic Sarcoma (UPS)	5	45–70	CD10+, vimentin+, cytokeratin−	Not performed	High-grade pleomorphic morphology
Rhabdomyosarcoma (RMS)	3	Pediatric–adult	Myogenin+, desmin+, MyoD1+	Not reported	Rare, usually embryonal subtype
Fibrosarcoma	2	Adult	Vimentin+	Not performed	Poorly described in historical reports
Mixed / Not Otherwise Specified (NOS)	7	40–68	Variable	Not available	Often from older literature or incomplete data
TOTAL	39				

Conclusion

Though individually uncommon, rare cancers collectively consist in a significant population: around one in five cancers is rare. Efforts should focus on increasing data collection, multidisciplinary expert assessment and collaborative research. Data collection efforts are essential and should include establishing rare cancer registries.

Scientific evidence is crucial to build clinical practice guidelines. Collecting clinical cases of rare cancers is extremely important for advancing research, improving diagnostics, and developing better treatment strategies for these complex diseases. Vigilant histopathological and molecular assessment is essential to distinguish MMMT, leiomyosarcoma and potential HG-ESS-like tumours. Prospective genomic studies are warranted.

Our case is, to date, the only one with both immunohistochemical and molecular confirmation of HG-ESS in the fallopian tube. This expands the spectrum of extra-uterine HG-ESS and underscores the need for genomic profiling in diagnostically ambiguous adnexal tumors.

Declaration of Competing Interests

The authors declare no conflict of interest

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Authorship Contribution Statement

Conceptualization: Roberta Renda, Eleonora Nardi, Maria Cristina Petrella, Francesca Castiglione
Data acquisition: Roberta Renda, Eleonora Nardi
Writing: Roberta Renda, Eleonora Nardi
Review: Roberta Renda, Eleonora Nardi, Marco Vangelisti, Massimiliamo Fambrini, Maria Cristina Petrella, Francesca Castiglione

Ethical Considerations

The present study complied with the Ethical Principles for Medical Research.

Involving Human Subjects according to the World Medical Association Declaration of Helsinki; all samples were anonymized before histology and immunohistochemistry; no further ethical approval was necessary to perform the retrospective study.

References

1. Di You, Qilin Wang, Wei Jiang, Lin Lin, Tianjin Yi, Lingjun Zhao, et al. (2018) Primary leiomyosarcoma of the fallopian tube: a case report and literature review. Medicine (Baltimore) 97: e0536.
2. Kommoss FKF, Mar LM, Howitt BE (2023) High grade endometrial stromal sarcomas with YWHAE-NUTM2 gene fusion exhibit recurrent CDKN2A alterations. Mod Pathol 36: 100044.
3. Di You, Qilin Wang, Wei Jiang, Lin Lin, Tianjin Yi, et al. (2020) Incidental primary leiomyosarcoma of the fallopian tube: Review of recent literature. Int J Gynecol Pathol 39: 378-384.
4. Gupta R, Jenison EL (2011) Carcinosarcoma of the fallopian tube presenting with torsion, rupture and hemoperitoneum. Gynecol Oncol Case Rep 2: 4-5.
5. Cozlea AL, Gheorghe M, Kiss SL (2022) Malignant mixed Mullerian tumour of the fallopian tube: Case report and literature review. Exp Ther Med 23: 177.
6. Jeong YY, Lee SJ, Ryu JM (2024) Next generation sequencing analysis and TP53 mutation in fallopian tube carcinosarcoma: A case report. Eur J Gynaecol Oncol 45: 177-181.
7. Zagouri F, Dimopoulos MA, Thomakos N, Chrysikos D, Papadimitriou CA (2011) Sarcomas of the fallopian tube: Disentangling a rare entity. Onkologie 34: 132-138.
8. Taninaka N, Ishida K, Takada Owada A (2024) High grade serous carcinoma of the fallopian tube transforming into carcinosarcoma at peritoneal dissemination via EMT. Int J Surg Pathol 33: 731-737.
9. Senger H (1886) Primary salpingosarcoma: First description. Zentralbl Gynakol 10: 123-127.
10. Zagouri F, Dimopoulos MA, Thomakos N, Chrysikos D, Papadimitriou CA (2011) Sarcomas of the fallopian tube: Disentangling a rare entity. Onkologie 34: 132-138.
11. Agarwal R, Rajanbabu A, Nair IR (2019) Undifferentiated sarcoma of fallopian tube managed with robotic assisted surgery. J Obstet Gynaecol India 69: 63-66.
12. Wu J, Wang S, Zhang L, Wu S, Liu Z (2023) Epidemiological analysis of hydrometra and its predictive value in gynecological tumors. Front Oncol 12: 1028886.
13. Doorji N, Tshering S, Chhetri N (2024) Presentation of a locally advanced cervical cancer with hydrometrocolpos: A case report. Bhutan Health J 10.
14. Nassif EF, Auclin E, Bahleda R, Honore C, Mir O, et al. (2021) TP53 mutation as a prognostic and predictive marker in sarcoma: Pooled analysis of MOSCATO and ProfiLER precision medicine trials. Cancers (Basel) 13: 3362.
15. Kobel M, Kang EY (2021) The many uses of p53 immunohistochemistry in gynecological pathology. Int J Gynecol Pathol 40: 32-40.