

# Alveolar Soft Part Sarcoma in the Female Genital Tract: Case Series with Literature Review and SEER Database Analysis

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**Introduction:** Alveolar soft part sarcoma (ASPS) is a rare and distinct subtype of soft tissue sarcoma. This study aims to describe the unique presentation of ASPS in the female genital tract.

**Methods:** Prognostic factors for cancer-specific overall survival (CSS) were evaluated using multivariate analyses.

**Results:** In our case series, we identified a novel TFE3-PRCC gene fusion in a 24-year-old unmarried patient with cervical ASPS who underwent fertility-sparing surgery and remained recurrence-free for 41 months. The other two patients underwent radical hysterectomy and bilateral salpingo-oophorectomy. At the time of writing, the two patients had been disease-free for 49 and 71 months, fluorescence in situ hybridization showed break-apart signals for the ASPL-TFE3 gene. Among the 55 cases with available information from the PubMed/Medline database, most presented with localized disease, and at the last follow-up, all patients were alive and 45 patients showed no evidence of disease. The 5-year CSS rate in the female genital tract cohort from SEER database was 86.2%. Multivariate analysis revealed that older age was associated with a 1.042-fold increased risk of cancer-specific mortality (HR=1.042, 95% CI 1.022–1.063,  $P < 0.001$ ), involvement of soft tissue including the heart was associated with a 4.786-fold higher risk (HR=4.7868, 95% CI 1.681–13.623,  $P = 0.003$ ), and regional infiltration and distant metastasis were associated with approximately 8.6-fold and 18-fold higher risk of cancer-specific mortality compared to local disease, respectively (HR=8.652, 95% CI 2.529–29.63,  $P = 0.001$ ; HR=18.366, 95% CI 6.153–54.817,  $P < 0.001$ ). Patients who underwent radical excision did not show reduced cancer-specific mortality compared to those who underwent local excision (HR=0.492, 95% CI 0.224–1.081,  $P = 0.078$ ).

**Discussion:** Previously unrecognized genetic diversity exists in ASPS. Patients with ASPS in the female genital tract have the lowest likelihood of presenting with a distant disease and are associated with a more favorable survival outcome.

**Keywords:** ASPSCR1, TFE3, alveolar soft part sarcoma, fusion

## Introduction

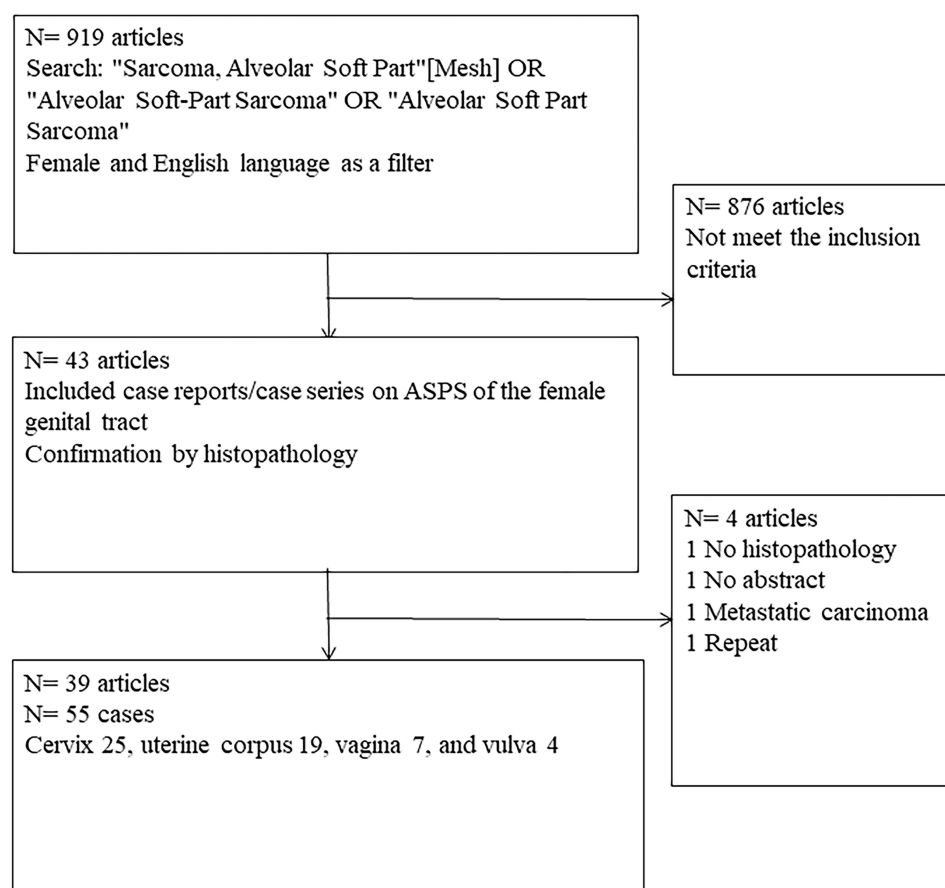
Alveolar soft-part sarcoma (ASPS) is a rare and distinct subtype of soft tissue sarcoma (STS) that constitutes less than 1% of all STS cases.<sup>1</sup> It was first identified in 1952 as a painless mass occurring in the extremities, head, and neck.<sup>2</sup> ASPS exhibits a strong female predominance. Primary ASPS in the female genital tract is a rare histological subtype of STS, with only a few reported cases. This subtype is classified separately by the World Health Organization (WHO) in the 5th edition of the classification of female genital tumors and presents significant challenges in terms of diagnosis and treatment.<sup>3</sup> Despite this, there is limited knowledge about the patient characteristics and prognosis associated with this condition. ASPS shares clinicopathological and histomorphological features with other Xp11 translocation-related tumors.<sup>4</sup> While ASPS patients are prone to distant metastasis, the site of the primary tumor influences the rate of metastasis and prognosis.<sup>5</sup> Prognosis for ASPS in the female genital tract has been reported to be better compared to other locations in the body, but the follow-up duration for survival assessment is short.<sup>6</sup> ASPS predominantly affects young women, and it remains uncertain whether fertility-sparing local excision is a recommended treatment approach. In

order to gain a better understanding of ASPS in the female genital tract, the present study examines the histopathological and molecular pathology profile of ASPS, along with survival outcomes, through the analysis of case series, literature review, and the SEER database. The aim is to provide assistance for clinical treatment decisions.

## Materials and Methods

### Data Source and Patients

Clinicopathological characteristics and relevant data from January 2010 to December 2022 were obtained from the electronic medical record system at our institution. These three patients have signed consent forms for information disclosure and a statement confirming consent to publish has been obtained. All undergone tissue pathology testing and fusion gene testing. Case 3 underwent whole exon sequencing. A search was conducted in the PubMed/Medline databases for case reports and series on ASPS of the female genital tract from January 1975 to May 2022. Two independent reviewers screened the identified studies based on titles and abstracts, with any discrepancies resolved through consensus with a third investigator. The search criteria included “Sarcoma, Alveolar Soft Part” [Mesh] OR “Alveolar Soft-Part Sarcoma” OR “Alveolar Soft Part Sarcoma” Alveolar Soft Part Sarcoma, with filters for females and English language. Patients with histopathologically confirmed ASPS in the female genital tract were included, while those with genital tract metastases were excluded. Among the 43 authors considered for inclusion, 4 were excluded (one without histopathology, one without an abstract, one with vaginal metastasis, and one repeat case), resulting in 39 studies meeting the final inclusion criteria. A total of 55 cases were utilized for analysis (Figure 1).<sup>7–45</sup> To identify eligible female patients with histologically confirmed primary ASPS (ICD-0 coding 9581/3) between 1975 and 2019, the SEER stat 8.4.1 software with Plus database was used. A multivariate analysis was performed to identify potential risk variables



**Figure 1** A flowchart outlining the criteria for patient eligibility.

in alveolar soft part sarcoma, male sex as negative prognostic factors for overall survival, so the SEER database only focuses on the female.<sup>45</sup> Patients with ASPS that was not the first malignant primary indicator were excluded. Cancer-specific survival (CSS) was defined as the time interval between diagnosis and ASPS-related death. The permission to use information from PubMed/Medline and SEER stat 8.4.1 software with Plus database are not required, as it is publicly available and unrestricted re-use is permitted via an open licence. This study received approval from the ethics committee of Chongqing cancer hospital (CZLS2023318-A).

## Histologic and Immunohistochemical Stains

Tumor samples were fixed in 4% formalin, conventionally dehydrated, embedded in paraffin, and sliced to a thickness of 3  $\mu$ m. These procedures were carried out using an automated immunohistochemical stainer. A positive reaction was observed under a microscope, and appropriate positive and negative controls were utilized. The following antibodies were available: TFE3 (RMA-0663, 1:300; Maixin Biotech. Co., Ltd, China), HMB45 (MAB-0098, 1:500; Maixin Biotech. Co., Ltd, China), cathepsin K (3F9, 1:300; Abcam, Cambridge, MA, USA), PAX8 (4H7B3, 1:100; Proteintech Group, Rosemont, IL, USA), Melan-A (MAB-0275, 1:100; Maixin Biotech. Co., Ltd, China), CD10 (MAB-0668, 1:100; Maixin Biotech. Co., Ltd, China), desmin (MAB-0766, 1:100; Maixin Biotech. Co., Ltd, China), SMA (MAB-0890, 1:100; Maixin Biotech. Co., Ltd, China), S100 (Kit-0007, 1:2000; Maixin Biotech. Co., Ltd, China). Paraffin-embedded slices with a thickness of 3  $\mu$ m were stained using the periodic acid-Schiff method.

## Fluorescence in situ Hybridization (FISH)

Fluorescence in situ hybridization (FISH) was conducted using the TFE3 fracture-separation probe, where the fluorescent mark of the TFE3 probe and the nucleic acid of the sample were hybridized in situ. The fluorescent signals were distinguished and counted under a microscope following the manufacturer's instructions. The TFE3 (Xp11.2) gene break probe (F.01106-01, anbiping Co., Ltd, China) was used. Result interpretation: Normal cells displayed two yellow signals (red and green fusion signals). A positive result was determined when the separation distance between the red and green signal widths matched that of two large fused signals. Each sample contained 200 cells, and a sample was considered positive if more than 10% of tumor cells exhibited separation signals.

## RNA Sequencing

RNA sequencing was conducted by gene plus Inc (Beijing, China). The Nebnext rRNA Removal Kit (NEB # z1955e) was used to remove targeted ribosomal RNA. All RNA fragments with a percentage  $> 200$  nucleotides (DV200)  $\leq 50\%$  underwent fragmentation and library preparation. After rRNA depletion and fragmentation, cDNA synthesis and NGS library preparation were performed using the NEBNext<sup>®</sup> Ultra<sup>™</sup> II Directional RNA Library Prep Kit (NEB, E7760L). The library was quantified using Qubit3.0 (Life Invitrogen, USA), and quality assessment was conducted with LabChip GX Touch (PerkinElmer, USA). Terminal adaptor sequences and low-quality data were removed using fastp (version: 0.19.5), and rRNA reads were eliminated by aligning clean reads to the rRNA database (downloaded from NCBI) using bowtie2 (version: 2.2.8). Clean reads without known rRNA were aligned to the reference human genome (hg19) using STAR (version 020201). Fusions were detected using a customized version of Arriba 1.1.0. and annotated using the in-house software annoFilterArriba (version: 1.0.0) with the NCBI release 104 database. All final candidate fusions were manually verified using an integrative genomics viewer browser. RNA-SeQC assessment was utilized to compute a series of quality control metrics. A threshold of  $\geq 80$  million mapped reads and  $\geq 10$  million junction reads per sample was established.

## Statistical Analysis

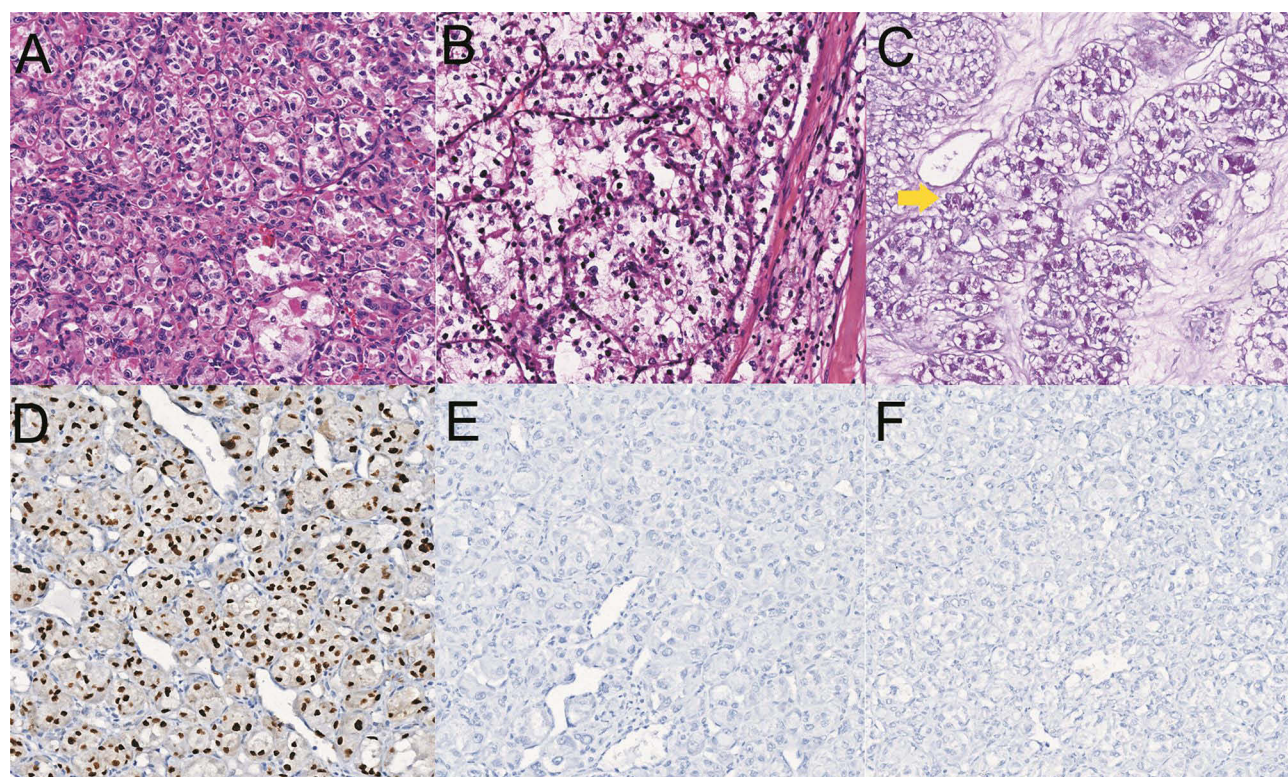
Statistical analysis was performed using SPSS 25 and GraphPad Prism 6.01 software. Categorical variables were compared using the  $\chi^2$  or Fisher's exact test. Measurement data were expressed as means and standard deviation ( $M \pm SD$ ). Skewed distribution data were expressed as median and interquartile range ( $M \pm QR$ ). Univariate and multivariate Cox regression analyses were conducted to identify independent predictors associated with survival, reporting hazard ratios (HR) and 95% CI. Variables included in the multivariate model were those with  $P \leq 0.1$  in the univariate analysis. A two-tailed  $P$  value of  $< 0.05$  was considered statistically significant.



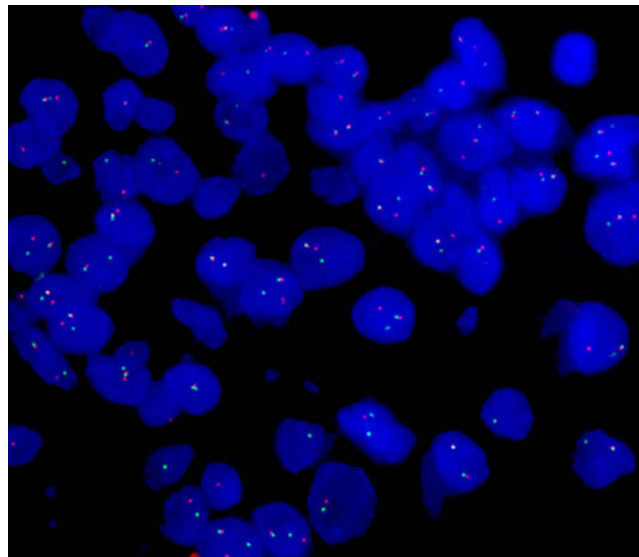
## Results

### ASPS in the Female Genital Tract at Our Institution

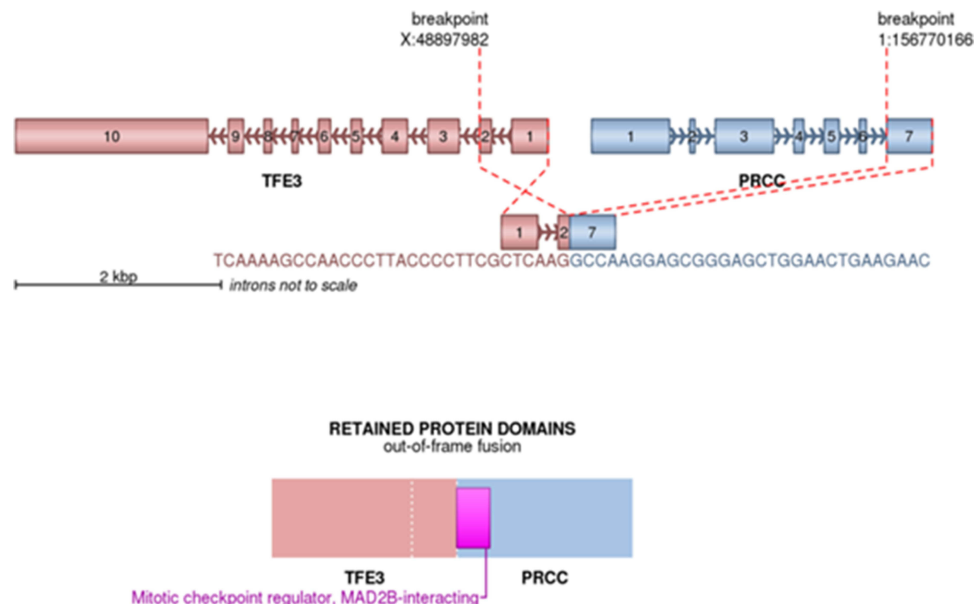
Case 1: A 31-year-old married woman presented with vaginal bleeding. Hysteroscopic examination revealed a 2.5 cm-sized mass protruding from the uterine corpus. The patient underwent total hysterectomy. Fluorescence in situ hybridization demonstrated break-apart signals for the ASPL-TFE3 gene. At the time of writing, the patient had been disease-free for 71 months. Case 2: A 61-year-old woman presented with vaginal bleeding. Gynecologic examination revealed a 4.5 cm-sized mass protruding from the cervix. Fluorescence in situ hybridization showed break-apart signals for the ASPL-TFE3 gene. The patient underwent radical hysterectomy and bilateral salpingo-oophorectomy. At the time of writing, the patient had been disease-free for 49 months. Immunohistochemical staining was positive for TFE3 but negative for HMB45, Melan-A, and Pax-8 in both Case 1 and Case 2. Case 3: A 24-year-old unmarried woman, who had never been pregnant, presented with increased menstrual bleeding for six months. Gynecologic examination revealed a polypoid mass measuring 3×2.5×0.5 cm protruding from the cervical canal. The mass was removed via dilatation and curettage. Gross examination showed a friable, tan-pink surface. Pathological evaluation revealed large, round, or polygonal tumor cells growing in an organoid pattern (Figure 2A) or a nest-like arrangement (Figure 2B), separated by delicate fibrovascular septa. The tumor cells exhibited abundant cytoplasmic and eosinophilic particles. Mitotic activity was infrequent. Periodic acid-Schiff-positive purplish-red crystals were observed in the cytoplasm (Figure 2C). Immunohistochemical staining was positive for TFE3 (Figure 2D) and CD56, but negative for HMB45 (Figure 2E), Melan-A (Figure 2F), Desmin, S-100, Syn, CgA, CD10, and Pax-8. Fluorescence in situ hybridization showed break-apart signals for the TFE3 gene (Figure 3). Whole exon sequencing identified a novel gene fusion between PRCC (NM\_005973.4) exon 7 and TFE3 (NM\_006521.4) exon 2 (Figure 4). Following curettage, magnetic resonance imaging revealed a residual nodule measuring 1.1×0.8 cm on the uterine isthmus. The patient underwent hysteroscopic resection of the residual nodule, which showed the same pathological features as before with negative margins. Positron emission tomography-computed tomography revealed no tumor after hysteroscopic



**Figure 2** Histological findings and the results of special staining and immunostaining in case 3 are as follows: The tumor cells exhibit an organoid pattern (A) or a nest-like arrangement (B), with delicate fibrovascular septa separating them. PAS staining reveals the presence of purplish-red crystals in the cytoplasm (indicated by the yellow arrow) (C). Immunostaining shows diffuse positivity for TFE3 (D), while HMB-45 (E) and Melan-A (F) are negative. The original magnification for all images is 200×.



**Figure 3** Fluorescence in situ hybridization demonstrates a break-apart signal indicating TFE3 rearrangement in case 3.



**Figure 4** A diagrammatic representation is provided to illustrate the two variant gene fusions identified through targeted RNA sequencing analysis. The PRCC-TFE3 transcript consists of PRCC exon 7 fused to exons 2 of TFE3 in case 3.

resection. The patient did not receive additional postoperative treatment. At the time of writing, the patient had been disease-free for 41 months, and the last follow-up date was May 20, 2023. Table 1 summarizes the characteristics of the three patients with ASPS arising in the female genital tract at our institution.

## Systemic Review of Literature for ASPS of the Female Genital Tract

Table 2 presents the basic information and clinical characteristics of the 55 patients. The median age of the patients was 31 years (range: 8–68 years). The most common symptom was abnormal vaginal bleeding (61.5%). Common sites of involvement included the cervix (45.5%), uterine corpus (34.5%), vagina (12.7%), and vulva (7.3%). The median tumor size was 3 cm (range: 0.16–8.7 cm). Exophytic growth was observed in 55.6% of lesions, such as polypoid, submucosal, and pedicle lesions. The majority of histological growth patterns displayed the classic organoid pattern with well-formed

**Table 1** Summary of Clinicopathological Features of ASPS in the Female Genital Tract at Our Institution

Case	Age (Years)	Symptoms	Site	Tumor Size (cm)	TFE3 Gene Translocation	Treatment	Follow Up (months)	Status at Follow Up
1	31	Vaginal bleeding	Uterus corpus	2.5	ASPL-TFE3	Total hysterectomy	71	NED
2	61	Vaginal bleeding	Cervix	4.5	ASPL-TFE3	Radical hysterectomy+bilateral salpingo-oophorectomy	49	NED
3	24	Vaginal bleeding	Cervix	3	TFE3-PRCC	Local excision	41	NED

**Abbreviation:** NED, No Evidence of Disease.

necks. Among the cases, 72.7% exhibited positive periodic acid-Schiff (PAS) and/or Periodic Acid Schiff Diastase (PAS-D) staining. TFE3 immunohistochemical staining was positive in 49.1% of cases, HMB45 in 46.1%, Melan-A in 32.7%, and desmin in 34.6%. TFE3 gene fusion was identified in 34.6% of cases. Only 32.7% of cases underwent both supportive immunohistochemistry and confirmatory genetic analysis. At initial diagnosis, 96.4% of patients presented with localized disease and 3.6% presented with regional disease. Radical resection and local lesion resection were performed as initial treatments in 71.1% and 23.1% of patients, respectively. Node dissection was carried out in 29.4% of patients, and two cases exhibited positive lymph node metastases (pelvic lymph node micrometastasis and para-aortic lymph node metastasis, respectively). Postoperative radiotherapy and/or chemotherapy were administered to 11.76% of patients. Preoperative radiotherapy was given in one case. The median follow-up period was 18 months. No cases of distant metastasis were reported. Local recurrence occurred in four patients, and only two patients experienced relapse, which involved local excision with biopsy and cryotherapy. No deaths were observed at the end of the follow-up, and 45 patients were alive without disease.

Analysis of ASPS in SEER Database

We reviewed a total of 174 cases in women and excluded nine cases due to non-first primary tumors, leaving us with 165 patients for analysis. The characteristics of the ASPS are summarized in Table 3. The common sites of occurrence were soft tissue, including the heart (n=134, 81.2%), and the female genital tract (n=17, 10.3%). The median age of the patients was 24±14.054 years (range: 2–78 years). The majority of patients were white (93, 56.4%) and unmarried (101, 61.2%). Regarding disease stage, 83 patients (50.3%) had localized/regional disease. Local excision was performed in 57 patients (34.5%), while radiation and chemotherapy were administered to 76 (46.1%) and 54 (32.7%) patients, respectively. In the female genital tract cohort, the 5-year cancer-specific survival (CSS) rate was 86.2%, whereas in the soft tissue, including the heart, it was 70.5%, and in other parts, it was only 47.1%. Univariate analysis demonstrated significant associations between CSS and age, tumor size, primary site, extent of surgery, stage, radiation, and chemotherapy (*P* < 0.05), as presented in Table 4. Multivariate analysis revealed that older age was associated with a 1.042-fold increased risk of cancer-specific mortality (HR=1.042, 95% CI 1.022–1.063, *P* < 0.001). Soft tissue, including the heart, exhibited a 4.786-fold higher risk of cancer-specific mortality (HR=4.786, 95% CI 1.681–13.623, *P*=0.003). Regional infiltration and distant metastasis carried approximately 8.6-fold and 18-fold higher risks of cancer-specific mortality compared to local disease, respectively (HR=8.652, 95% CI 2.529–29.63, *P* = 0.001; HR=18.366, 95% CI 6.153–54.817, *P* < 0.001). Patients who underwent radical excision did not show a reduced risk of cancer-specific mortality compared to those who underwent local excision (HR=0.492, 95% CI 0.224–1.081, *P* = 0.078), as indicated in Table 4.

Discussion

For the first time, we have identified a novel PRCC-TFE3 gene fusion in ASPS occurring in the female reproductive system. We first report the case of a 24-year-old unmarried patient with cervical ASPS who underwent fertility-sparing surgery without recurrence for 41 months. The median age of the patients was 31 years. The most common symptom was abnormal vaginal bleeding. Common sites of involvement included the cervix and uterine corpus. The median tumor size was 3 cm. Exophytic growth was observed in 55.6% of lesions. ASPS patients with primary involvement of the female genital tract were least likely to present with distant disease and exhibited a favorable survival advantage. The 5-year



**Table 2** Summary of Clinicopathological Features of Previously Published Cases of ASPS in the Female Genital Tract

Case	Authors and Years	Age (Years)	Tumor Location	Tumor Size (cm)	Gross	Histological Growth Pattern	Immunohistochemistry	ASPSCR1–TFE Fusion	PAS/PAS-D	Treatment	Follow-Up (Months)	Status at Follow Up
1	Kasai et al (1980) <sup>7</sup>	34	Vaginal	7.5	Exogenous	Organoid alveolar	NA	NA	+	RH+Node dissection of the right groin	NA	NA
2	Shen et al (1982) <sup>8</sup>	62	Right labium minus	4	NA	Organoid alveolar	NA	NA	NA	Radical vulvectomy+Node dissection of the right groin	24	NED
3	Chapman et al (1984) <sup>9</sup>	26	Vaginal	6	NA	Nested	NA	NA	+	Local excised +Radiotherapy, chemotherapy; Local recurrence, complete excision	25	NED
4	O'Toole et al (1985) <sup>10</sup>	18	Vaginal	6	Ulcerate	Organoid alveolar	NA	NA	+	Radiotherapy, RH	36	NED
5	Flint et al (1985) <sup>11</sup>	37	Cervix	2	Polypoid mass	Nests, Alveolar pattern	NA	NA	+	RH+PLND	NA	NA
6	Gray et al (1986) <sup>12</sup>	27	Cervix	0.7 (after biopsy)	Submucosal nodule	Organoid	NA	NA	+	RH+BSO+PLND+PALND	36	NED
7	Gray et al (1986) <sup>12</sup>	43	Uterus corpus	0.4	Intramymetrial nodule	Organoid	NA	NA	+	TH	9	NED
8	Kopolovic et al (1987) <sup>13</sup>	26	Cervix	4	Multinodular	Alveolar pattern	NA	NA	+	RH+PLND	12	NED
9	Foschini et al (1989) <sup>14</sup>	32	Cervix	1.3	Polyp	Nested	NA	NA	+	Cryotherapy; Recurred four months later, TH	6	NED
10	Abeler and Nelsond (1989) <sup>15</sup>	35	Endocervix	1	NA	Alveolar pattern	NA	NA	+	TH	12	NED
11	Sahin et al (1989) <sup>16</sup>	30	Cervical canal	4	Friable mass	Alveolar pattern	NA	NA	+	Radiation	192	NED
12	Sahin et al (1989) <sup>16</sup>	38	Cervix	2	Submarginal nodule	Alveolar pattern	Desmin(-)	NA	+	TH+BSO	42	NED
13	Nolan and Gaffney (1990) <sup>17</sup>	14	Endometrial	7	Exogenous	Organoid	NA	NA	+	D&C, HT+BSO	78	NED
14	Carinelli et al (1990) <sup>18</sup>	27	Vagina	3	NA	NA	NA	NA	NA	Surgery+radiotherapy	204	NED
15	Guillou et al (1991) <sup>19</sup>	40	Uterus corpus	3	Intramymetrial nodule	Organoid arrangement	HMB45 focal(+);Desmin (+)	NA	+	TH	96	NED
16	Morimitsu et al (1993) <sup>20</sup>	8	Cervix	2	Polypoid mass	Nests, Alveolar pattern	Desmin(+)	NA	+	Polypectomy and postoperative chemotherapy	12	NED
17	Chang et al (1994) <sup>21</sup>	18	Vaginal	7	Exogenous	NA	NA	NA	+	Excision biopsy, recurred, RH+Lymph node sampling of the groin+Radiation	64	NED

(Continued)

Table 2 (Continued).

Case	Authors and Years	Age (Years)	Tumor Location	Tumor Size (cm)	Gross	Histological Growth Pattern	Immunohistochemistry	ASPSCR1–TFE Fusion	PAS/PAS-D	Treatment	Follow-Up (Months)	Status at Follow Up
18	Burch et al (1994) <sup>22</sup>	47	Endometrial	3	Polypoid mass	NA	NA	NA	+	TH	48	NED
19	Nielsen et al (1995) <sup>23</sup>	26	Cervix	1.5	Endogenous	NA	NA	NA	+	D&C, HT+BSO+LND	9	NED
20	Nielsen et al (1995) <sup>23</sup>	31	Cervix	4	Endogenous	NA	NA	NA	+	HT	10	NED
21	Nielsen et al (1995) <sup>23</sup>	37	Uterus corpus	1	Intramural nodule	Organoid appearance	NA	NA	+	HT	66	NED
22	Nielsen et al (1995) <sup>23</sup>	30	Uterus corpus	3.5	Submucosal nodule alus	Organoid appearance	NA	NA	+	HT	66	NED
23	Heimann et al (1998) <sup>24</sup>	10	Cervix	3.5	Endogenous	Nested	Desmin(+)	Detected	+	Local excised,48m recurrence, HT+PLND	12	NED
24	Radig et al (1998) <sup>25</sup>	50	Uterus	3	Intramural nodule	Pseudoalveolar and trabecular pattern	Desmin(+)	NA	+	HT+BSO	84	NED
25	Radig et al (1998) <sup>25</sup>	36	Uterus	4.5	Intramural nodule	Pseudoalveolar pattern	Desmin(+)	NA	+	HT	8	NED
26	Roma et al(2005) <sup>26</sup>		Cervix	0.2	Endogenous	Nest-like or organoid pattern	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	NA	+	TH	18	NED
27	Kasahima et al (2007) <sup>27</sup>	50	Endometrium	2	Exophytic nodule	Nested	TFE3(++);HMB45 (-)	NA	+	TH+BSO+PLND	38	NED
28	Gitau et al (2008) <sup>28</sup>	19	Cervix	4	Exogenous	NA	HMB45 (-)	NA	+	RH+L-SO+PLND, Radiotherapy	18	NED
29	PeterssonAnd Michal (2009) <sup>29</sup>	29	Endocervix	0.16	NA	Alveolar pattern	TFE3(+)	Detected	+	Conization	12	NED
30	Guntupalli et al (2009) <sup>30</sup>	38	Endocervix	0.8	Exogenous	NA	TFE3(+)	NA	+	TH+BSO+PLND+PALND	60	NED
31	MU et al (2009) <sup>31</sup>	25	Cervix canal	2.9	Polypoid mass	NA	Des, S-100, HMB-45, MelanA, were positive, EMA, CgA, Syn were negative	NA	NA	RH+BSO+PLND, chemotherapy, Radiotherapy	35	NED
32	Williams et al (2009) <sup>32</sup>	31	Cervix	3	NA	NA	TFE3(++)	Detected	NA	Excision	36	NED
33	Hasegawa et al (2011) <sup>33</sup>	57	Cervix	8	Endogenous	Solid alveolar pattern	TFE3(++);HMB45 focal(+)	Detected	+	TH+ BSO	66	NED
34	Kang et al <sup>34</sup>	52	Uterine cervix	3	NA	NA	TFE3(++)	NA	+	RH+ BSO	NA	NA
35	Zhang et al (2012) <sup>35</sup>	57	Lower uterine segment	2.4	Exophytic tumor	Nest-like or organoid pattern	TFE3(++);HMB45 I(-); Desmin(-)	NA	+	TH+BSO+PLND+PALND	9	NED
36	Feng et al (2014) <sup>36</sup>	21	Cervix, near internal cervical	5	Exogenous, pedicle	Organ-like, alveolar or nest-like	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	Detected	+	TH+BSO+PLND	3	NED



37	Lee et al(2014) <sup>37</sup>	17	Cervix	1.6	Exogenous	Nested	TFE3(++);HMB45 (-)	NA	+	LEEP Conization	24	NED
38	Jabbour et al(2014) <sup>38</sup>	24	Right labium majus	6	NA	Nested	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	Detected	+	Wide radical local excision, radiation therapy	12	NED
39	Giordano et al (2016) <sup>39</sup>	66	Uterine corpus	5	Intramural nodule	Organoid pattern with nests surrounded	TFE3(++);HMB45 (-)	NA	+	TH+BSO	7	NED
40	Zhang et al (2017) <sup>40</sup>	68	Cervix	1	Cervical stroma	Nested	TFE3(++);HMB45 (-);PAX8 (-)	NA	+	RH	NA	NA
41	Schoolmeester et al (2017) <sup>41</sup>	23	Uterine corpus	NA	NA	Nested	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	Detected	NA	Curettage	9	NED
42	Schoolmeester et al (2017) <sup>41</sup>	21	VULVA	4	NA	Nested	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	Detected	NA	Wide local excision	NA	NA
43	Schoolmeester et al (2017) <sup>41</sup>	37	Cervix	2.2	NA	Nested	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	Detected	NA	TH+BSO+PLND	1	NED
44	Schoolmeester et al (2017) <sup>41</sup>	33	Uterine corpus	NA	NA	Solid	TFE3(++);HMB45 (-); Melan-A focal(+);Desmin (-)	Detected	NA	NA	NA	NA
45	Schoolmeester et al (2017) <sup>41</sup>	45	Cervix	0.7	NA	Nested	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	Detected	NA	TH	Recent	NA
46	Schoolmeester et al (2017) <sup>41</sup>	NA	Vaginal	NA	NA	Nested	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	Detected	NA	NA	NA	NA
47	Schoolmeester et al (2017) <sup>41</sup>	15	Vaginal	8.5	NA	Nested	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	Detected	NA	NA	NA	NA
48	Schoolmeester et al (2017) <sup>41</sup>	31	Uterine corpus	NA	NA	Solid	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	Detected	NA	TH	35	NED
49	Schoolmeester et al (2017) <sup>41</sup>	68	Uterine corpus	NA	NA	Nested	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	Detected	NA	TH	15	NED
50	Schoolmeester et al (2017) <sup>41</sup>	32	Uterine corpus	NA	NA	Nested	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	Detected	NA	TH+BSO	8	NED
51	Aiba T (2017) <sup>42</sup>	20	Perineum	8.7	NA	NA	NA	NA	NA	NA	NA	NA
52	Gomez et al (2020) <sup>43</sup>	20	Lower uterine segment	3.5	Polypoid	Nested	TFE3(++);Melan-A(-)	Detected	+	TH	NA	NED
53	Vishwajeet V et al (2021) <sup>44</sup>	24	Myometrial lesion	2.4	Exogenous	Nests and small clusters	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	NA	+	Mass excision	NA	NED
54	Lee Y et al (2022) <sup>45</sup>	27	Lower uterine segment	2.5	Polypoid	Sheets, nests and solid	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-); PAX8(-)	Detected	+	TH	2	NED
55	Lee Y et al (2022) <sup>45</sup>	10	Cervix	2.9	Polypoid mass	Solid	TFE3(++);HMB45 (-); Melan-A(-);Desmin(-)	Detected	+	TH	4	NED

**Abbreviations:** PAS, periodic acid-Schiff; PAS-D, Acid Schiff Diastase; "+", positive; "-", negative; RH, radical hysterectomy; TH, total hysterectomy; HT, hysterectomy; BSO, bilateral salpingo-oophorectomy; PALND, Para-aortic lymphadenectomy; PLND Pelvic lymphadenectomy; LND, lymphadenectomy; D&C, Dilatation curettage; NED, No Evidence of Disease; NA, Not Available.

**Table 3** Summary of Demographic and Treatment Information for ASPS Female Patients in the SEER Database

Variable	n (%)
Medians Age (years)	24±14.054(2–78)
Race	
White	93(56.4)
Black	38(23.0)
Other	34(20.6)
Margin status	
Unmarried	101(61.2)
Married	47(28.5)
Other	17(10.3)
Tumor size (cm)	
< 8.0	83(50.3)
≥ 8.0	82(49.7)
Primary site	
Female genital tract	17(10.3)
Soft tissue including heart	134(81.2)
Other	14(8.5)
Summary stage	
Localized	62(37.6)
Regional	21(12.7)
Distant	67(40.6)
Unstaged/Blank(s)	15(9.1)
Extent of surgery	
Local excision	57(34.5)
Radical excision	69(41.8)
Other	39(23.6)
Radiation	
No	89(53.9)
Yes	76(46.1)
Chemotherapy	
No	111(67.3)
Yes	54(32.7)

**Table 4** Univariate and Multivariate Survival Analysis for ASPS Patients in the SEER Database

Variable	Univariate		Multivariate	
	HR (95% CI)	P	HR (95% CI)	P
Medians Age (years)	1.024(1.012–1.035)	<0.001	1.042(1.022–1.063)	<0.001
Race				
White	1			
Black	0.941 (0.499 –1.775)	0.850		
Other	0.713 (0.354 –1.436)	0.344		
Margin status		0.139		
Unmarried	1			
Married	1.710 (0.999 –2.925)	0.050		
Other	1.075 (0.418 –2.762)	0.881		

(Continued)

Table 4 (Continued).

Variable	Univariate		Multivariate	
	HR (95% CI)	P	HR (95% CI)	P
Tumor size (cm)				
<8	1		1	
≥8	3.334(1.851–6.002)	<0.001	1.062(0.531–2.125)	0.865
Unkown	2.198 (1.093–4.423)	0.027	1.116(0.454–2.744)	0.811
Primary site				0.008
Female genital tract	1		1	
Soft tissue including heart	2.828(0.874–9.155)	0.083	4.786(1.681–13.623)	0.003
Other	4.315(1.138–16.359)	0.032	0.954(0.232–3.929)	0.948
Summary stage				<0.001
Localized	1		1	
Regional	6.552(2.142–20.037)	0.001	8.652(2.529–29.63)	0.001
Distant	18.851(7.319–48.553)	<0.001	18.366(6.153–54.817)	<0.001
Unstaged/Blank(s)	5.190(1.393–19.340)	0.014	2.122(0.425–10.597)	0.359
Extent of surgery				0.036
Local excision	1		1	
Radical excision	0.494 (0.247–0.989)	0.046	0.492(0.224–1.081)	0.078
Other	3.150 (1.756–5.651)	<0.001	1.403(0.712–2.763)	0.327
Radiation				
No	1		1	
Yes	1.722(1.034–2.867)	0.037	1.586 (0.859–2.930)	0.141
Chemotherapy				
No	1		1	
Yes	3.241(1.941–5.411)	<0.001	1.457 (0.795–2.671)	0.223

cancer-specific survival (CSS) rate was 86.2% in the female genital tract cohort. The patients who underwent local resection did not have increased cancer-specific mortality; however, older age, involvement of soft tissue including the heart, and metastasis at presentation were adverse prognostic factors.

ASPS originating from the female genital tract is a rare condition that poses significant diagnostic challenges.<sup>3</sup> ASPS and Xp11 translocation PEComa have very similar microscopic features and immunoprofiles, being characteristically diffusely positive for TFE3 and negative for PAX-8. Clues to the diagnosis of ASPS include negative labeling for melanotic markers (HMB45 or Melan-A) and the fusion gene of ASPSCR1-TFE3.<sup>46</sup> TFE3 translocation RCCs are positive for PAX-8 and CD10 and usually do not express pigment markers.<sup>47</sup> The ASPSCR1-TFE3 fusion gene has long been known as a disease-defining molecular event that underpins the diagnosis of ASPS;<sup>41</sup> however, a novel CSCP-TFE3 gene fusion was identified using whole exon sequencing at our institution. Dickson et al<sup>48</sup> have identified three novel TFE3 fusion partners, namely HNRNP3, DVL2, and PRCC, associated with ASPS in their soft tissue counterparts. These 'MiT family translocation-associated neoplasms' contain diverse TFE3 fusion products. The fusion of DVL2 and PRCC with TFE3 has been discovered in RCC,<sup>49</sup> and a ASPSCR1 fusion partner has been identified in TFE3-rearranged PEComa.<sup>50</sup> The potential under-recognized genetic diversity of ASPS. The next-generation sequencing strategy for fusion gene evaluation in the diagnosis of ASPS is more comprehensive and accurate.

A systematic analysis of the reported 55 cases in the female genital tract revealed that consistent with the report by Schoolmeester,<sup>41</sup> ASPS arising in the female genital tract predominantly affected young individuals (median age 31 years). These tumors showed a strong preference for the uterine cervix, uterine corpus, and vagina, and typically exhibited small sizes (mean 3 cm). In half of the patients, exophytic growth of lesions, such as polypoid, submucosal, and pedicle lesions, was observed. 70.9% of the cases described typical morphology and PAS and/or PAS-D staining, however, Schoolmeester et al have noted these crystals are not always present.<sup>41</sup> Only 32.7% of the cases had either supportive immunohistochemistry and confirmatory genetic analysis. All patients were alive at the last follow-up.

Compared to previous reports,<sup>1</sup> patients with ASPS of the female genital tract presented at earlier stages and had higher rates of resection. Unlike most patients with ASPS in soft tissue sites who remain asymptomatic,<sup>46</sup> it is interesting to note that half of the female genital tract tumors appeared as exogenous masses and were associated with abnormal vaginal bleeding, which can aid in early diagnosis. The standard treatment for uterine sarcoma is total hysterectomy.<sup>51</sup> In our review of the PubMed/MEDLINE databases, we found 12 cases of ASPS of the female genital tract in which lesion removal was performed. Local tumor recurrence data were available for four patients, and only two patients experienced relapse, both of whom underwent local excision with biopsy and cryotherapy. Lieberman et al reported that local recurrence of ASPS was only observed in cases with residual disease left behind.<sup>52</sup> These results suggest that achieving R0 excision is crucial (and perhaps sufficient) for satisfactory local control of ASPS.

The 5-year cancer-specific survival (CSS) rate was 86.2% in the female genital tract cohort, indicating a favorable prognosis consistent with previous literature.<sup>33</sup> Ogura reported one of the largest series based on data from a single center and identified smaller tumor size and absence of metastasis as favorable prognostic factors.<sup>53</sup> Other studies have also suggested differences in prognosis based on primary site, sex, and age.<sup>54</sup> Multivariate analysis using the SEER database confirmed that primary site was an independent risk factor for prognosis. The favorable prognosis of ASPS in the female genital tract may be attributed to early detection facilitated by bleeding and exogenous masses, leading to improved resectability. However, older age was identified as an adverse prognostic factor based on the SEER database. Differences in the biological characteristics between pediatric and adult ASPS may explain this observation.<sup>55</sup> Our analysis also indicated that the prognosis of local resection was not statistically different from radical resection. At our institution, we treated a 24-year-old unmarried patient with ASPS of the cervix who underwent fertility-sparing surgery and remained recurrence-free for 41 months. Fertility-sparing local excision may be considered as a clinical decision for patients with localized disease, although long-term follow-up for prognosis is necessary. Consistent with previous reports, there was no survival benefit from traditional radiotherapy or chemotherapy.<sup>56</sup>

Our study has several limitations. Firstly, some cases may have been misdiagnosed due to the absence of immunohistochemistry and fusion gene detection in the PubMed/MEDLINE databases, which could have affected the accuracy of the differential diagnosis. Secondly, the SEER database lacks information on pediatric patients, limiting our understanding of ASPS in this population. Thirdly, important details such as the surgical margin and information on disease progression or recurrence were not available in the SEER database. Lastly, it is unclear whether patients in the study underwent metastasectomy.

In conclusion, the potential under-recognized genetic diversity of ASPS. The favorable prognosis observed in ASPS of the female genital tract can be attributed to early detection and improved resectability. Furthermore, our findings indicate that patients who underwent local resection did not experience increased cancer-specific mortality.

## Acknowledgments

We thank Ms Lingling Yang, for contributing to the statistics analysis.

## Author Contributions

The contribution of the individual authors: All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

## Funding

The present study was supported by the Beijing Kanghua Foundation for the Development of Traditional Chinese and Western Medicine “Le foundation” (KH-2020-LJJ-044, Xingtao Long).

## Disclosure

The authors have no conflict of interest that could have appeared to influence the work reported in this paper.



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