

First author	Year	Age	Primary site	EWS-WT1 fusion	Metastasis at diagnosis	Treatment	Status of disease
Prat <sup>28</sup>	1992	22	Paratesticular	NR	Supraclavicular, retroperitoneal and inguinal LN, lung	Orchiectomy + Chemotherapy (NR)	DOD 17 months after diagnosis
Ordoñez <sup>29</sup>	1993	28	Paratesticular	NR	NR	Orchiectomy + CDDP, VP	AWD (para-aortic LN 12 months after diagnosis)
Farhat <sup>30</sup>	1996	NR	Paratesticular	NR	-	Orchiectomy + CPM, VP, EPI, CDDP + HDC (Carbo, VP, IFO) + ASCR	Mediastinal and retroperitoneal LN, lung, bone metastasis 6 months after orchiectomy. NED 32 months after diagnosis
Bertoldin <sup>25</sup>	1996	28	Paratesticular	NR	-	Orchiectomy (complete resection) + Chemotherapy (NR)	Abdominal and retroperitoneal relapse; DOD 13 months after diagnosis
Cummings <sup>31</sup>	1997	17	Paratesticular	NR	-	Orchiectomy	Lost to follow-up
		28	Paratesticular	NR	-	Orchiectomy + CDDP, Doxo, CPM (Unclear if chemotherapy was given after surgery or after relapse)	Relapse in cervical LN 7 months after diagnosis; DOD 16 months after diagnosis
		32	Paratesticular	NR	Retroperitoneal LN	Orchiectomy	Lost to follow-up
		26	Paratesticular	NR	Multiple LN, lung	Orchiectomy + Chemotherapy (NR) + ASCR	NED 2.5 years after diagnosis
		28	Paratesticular	NR	-	Orchiectomy + Doxo, CPM, VCR (Unclear if chemotherapy was given after surgery or after relapse)	Relapse in lung 2 years after orchiectomy; Alive; NED 3 years after diagnosis
		37	Paratesticular	NR	-	Orchiectomy	Relapse in retroperitoneal LN 3y after orchiectomy; Lost to follow-up
Furman <sup>32</sup>	1997	21	Paratesticular	NR	-	Resection + IFO, Carbo, VP, VCR, CPM, Doxo	NED 33 months after diagnosis
Roganovitch <sup>26</sup> , Bisogno <sup>27</sup>	1999	17	Paratesticular	NR	-	Orchiectomy (complete resection) + IFO, VCR, Actino	NED 63 months after diagnosis
Kawano <sup>33</sup>	1999	43	Paratesticular	Positive (RT-PCR)	NA	NA	NA
García-González <sup>34</sup>	2005	23	Paratesticular	NR	-	Resection + MTX, DAC, CPM, Actino, VCR, Doxo + Radiation therapy (not reported dose and site).	NED 72 months after diagnosis
Thuret <sup>35</sup>	2005	24	Paratesticular	Positive (RT-PCR)	Retroperitoneal LN, lung	Orchiectomy + Chemotherapy (NR)	NR
Saab <sup>3</sup>	2007	19	Paratesticular	NR	-	Orchiectomy (complete resection) + Carbo, VCR, Doxo, CPM, Irino, VP	NED 120 months after diagnosis
Sha <sup>36</sup>	2007	27	Paratesticular	NA	NA	Orchiectomy + CDDP, VP, IFO, EPI	NED 36 months after diagnosis
Rais <sup>37</sup>	2010	27	Paratesticular	Positive (RT-PCR)	Retroperitoneal (para-aortic) LN	Orchiectomy + CDDP, VP	NED 6 months after diagnosis
Cliteur <sup>14</sup>	2012	32	Paratesticular	Positive (COBRA-FISH); UF	Abdominal	Orchiectomy + VCR, IFO, Doxo, VP + Incomplete abdominal debulking after three first cycles of chemotherapy	DOD 24 months after diagnosis
He <sup>10</sup>	2013	27	Testicular <sup>A</sup>	NR	-	Orchiectomy + VCR, Doxo, IFO	DOD 26 months after diagnosis
Yue <sup>38</sup>	2014	25 and 35	Paratesticular	Positive (FISH) in one case	Abdominal	Orchiectomy in one case 1 <sup>st</sup> patient: Hepatic arterial embolization + CPM, Pira, Vind, DAC; 2 <sup>nd</sup> patient: CPM, Vinpo, EPI	Unclear; Probably DOD
Zhang <sup>11</sup>	2014	42	Testicular <sup>B</sup>	Positive (FISH)	Abdominal-pelvic dissemination	Orchiectomy + VCR, Doxo, CPM	AWD (partial response) 9 months after surgery
Manjula <sup>39</sup>	2015	17	Testicular	NR	Left supraclavicular and para-aortic LN	Orchiectomy + CPM, Doxo, VCR, IFO, VP	Progression disease (multiple metastases); DOD.
Herman <sup>12</sup>	2016	65	Testicular <sup>C</sup>	Positive (NGS); UF	-	Orchiectomy (complete resection) + VCR, Doxo, CPM	NED 6 months after diagnosis
Sedig <sup>4</sup>	2017	14	Paratesticular	Positive (WES and RNAseq)	-	Orchiectomy (complete resection with microscopic positivity at tumor margins) + VCR, Topo, CPM, Doxo, VP, IFO + Radiation therapy (50.4 Gy in tumor bed)	NED 24 months after diagnosis
Al-Ibraheemi <sup>18</sup>	2019	31	Paratesticular	Positive (RT-PCR)	NR	Resection	NR
Romagnol	2020	13	Testicular	Positive (NGS)	-	Orchiectomy (complete resection) + Irino, VCR	NED 96 months after diagnosis

**Table 1 - Paratesticular and testicular Desmoplastic Small Round Cell Tumor reported in the literature**

**Legend:** Actino: Actinomycin-D; ASCR: Autologous Stem-Cell Rescue; Carbo: Carboplatine; CDDP: Cisplatin; COBRA-FISH: Combined Binary Ratio labelling Fluorescence In-Situ Hybridization; CPM: Cyclophosphamide; DAC: Dacarbazine; Doxo: Doxorubicin; EPI: Epirubicin; FISH: Fluorescence In-Situ Hybridization; HDC: High Dose Chemotherapy; IFO: Ifosfamide; Irino: Irinotecan; LN: Lymph nodes; MTX: Methotrexate; NGS: Next-Generation sequencing; NA: Not available (only abstracts); NR: Not reported; Pira: Pirarubicin; RNAseq: RNA sequencing; RT-PCR: Reverse Transcription Polymerase Chain Reaction; Topo: Topotecan; UF: Unusual fusion; VCR: Vincristine; Vind: Vindesine; Vinpo: Vinpocetine; VP: Etoposide; WES: Whole Exome Sequencing; AWD: Alive with disease; DOD: Died of disease; NED: No evidence of disease.

<sup>A</sup> With extension to the epididymis

<sup>C</sup> Invasion to rete testis and lymphovascular spaces

<sup>B</sup> With epididymis and spermatic cord involvement