



**Supplementary Figure S1.** Circular genetic map of the result of next-generation sequencing. Circular genetic map revealed a Somatic MGA:c.6565 deletion, a Germline BUB1B:c.948-966 deletion and a Fusion EWSR1(E7): ZNF444(E5).

**Supplementary Table S1.** Review of cases of primary myoepithelioma in mediastinum

Case	Age/ sex	Site	Origin	IHC profile	Genetic alteration	Behavior	Invasive growth	Treatment/ outcome
Case 1 <sup>[1]</sup>	24/M	Left mediastinum	Undefined (Stem cells)	CKAE1/AE3(+), CK7(+), Vimentin(+), MIC2(CD99+), S100(+), α-SMA(+), p63(+), GFAP(+), CD117 (F+), WT1 (cytoplasmic)(F+)	NA	Benign	No	Resection/NED, 6 mo
Case 2 <sup>[2]</sup>	51/F	Right posterior mediastinum	Undefined	EMA(+), SMA(+), S-100(+), CD99(+), calponin(F+), p63(F+), GFAP (F+)	EWSR1 rearrangement	Benign	No	Resection/NED, 24 mo
Case 3 <sup>[3]</sup>	51/F	Inferior top mediastinum	Atrial epicardium or myocardium	CKAE1/AE3(+), CK7(+), Vimentin(+), S-100(+), Calretinin(+), CEA(+)	NA	Benign	Yes	Resection/NED, 7 mo
Case 4 <sup>[4]</sup>	62/F	Left posterior mediastinum	NA	S100 (+), vimentin(+), EMA(F+), CK8/18(F+)	EWSR1 rearrangement	Benign	No	Resection/NA
Case 5 [The present]	9/M	Right posterior mediastinum	Intervertebral foramina (Stem cells)	S-100(+), Desmin(+), CK(+), CK19(+), INI1(+), SMA(F+), CD99 (F+)	EWSR1 rearrangement	Benign	Yes	Resection/REC, 6 mo

**Note.** Abbreviations: M, male; F, female; NA, not available; IHC, immunohistochemistry; CK, cytokeratin; GFAP, glial fibrillary acid protein; F+, focally positive; SMA, alpha-smooth muscle actin; EMA, epithelial membrane antigen; WT, wilm tumor; NED, no evidence of disease; REC, recurrence; EWSR1, Ewing sarcoma breakpoint region1; CEA, carcinoembryonic antigen; mo, month.

**Supplementary Table S2.** Gene alteration in myoepithelioma of different sites

Tumor site	Gene rearrangement						Gene deficiency SMARCB1/ INI1
	EWSR1	FUS	OGT	SRF	PLAG1	RREB1	
Soft tissue	Yes <sup>[5]</sup>	Yes <sup>[5]</sup>	Yes <sup>[4, 6]</sup>	Yes <sup>[7]</sup>	Yes <sup>[8]</sup>	Yes <sup>[9]</sup>	Yes <sup>[10, 11]</sup>
Bone	Yes <sup>[5]</sup>	Yes <sup>[12]</sup>	NR	NR	NR	NR	NR
Salivary gland	No <sup>[5]</sup>	NR	NR	NR	Yes <sup>[13]</sup>	NR	NR
Visceral locations (lung)	Yes <sup>[5]</sup>	Yes <sup>[5, 14]</sup>	NR	NR	NR	NR	NR
Skin	Yes <sup>[15]</sup>	NR	NR	NR	Yes <sup>[16]</sup>	NR	NR
MELTVR	No <sup>[17]</sup>	No <sup>[17]</sup>	NR	NR	NR	NR	Yes <sup>[17]</sup>

**Note.** Abbreviations: NR, not reported; MELTVR, myoepitheliom like tumors of the vulvar region; EWSR1, Ewing sarcoma breakpoint region1; FUS, fused in sarcoma gene; PLAG1, pleiomorphic adenoma gene 1; OGT, O-linked N-acetylglucosamine; SRF, serum response factor; SMARCB, SWI/SNF-related matrix-associated actin-dependent regulator of chromatin subfamily B member 1.

**Supplementary Table S3.** Review of soft tissue myoepithelioma in children below 18 years old<sup>a</sup>

Authors	No. of cases	Age/sex	Site	Gene rearrangement	Behavior	Invasive growth	Recurrence	Metastasis	Treatment/outcome
Kilpatrick et al, 1997 <sup>[18]</sup>	1	9/M	axilla	NA	Malignant*	Yes	Yes	No	Resection/REC, 30 mo
Waldrop et al, 2001 <sup>[19]</sup>	1	11/M	thigh	NA	Malignant	Yes	No	Orbir, 10 mo	Resection/died 4 mo after metastasis amputation <sup>b</sup>
Van den Berg et al, 2004 <sup>[20]</sup>	1	17/M	left lower leg	NA	Malignant	NA	-	LN, L, bone, 2 mo	died 7 mo after amputation Resection/
Harada et al, 2005 <sup>[21]</sup>	1	17/M	forearm	NA	Malignant	Yes	No	L, 6 mo	died 18 mo after metastasis Resection/
Hallor et al, 2008 <sup>[22]</sup>	1	11/F	back	NA	Benign	Yes	Yes, 6 y	L, 7 y	Resection/AwD, 9 y
Herlihy et al, 2009 <sup>[23]</sup>	1	3mo/M	orbit	NA	Benign	No	No	No	Resection/NED, 36 mo
Guedes et al, 2009 <sup>[24]</sup>	1	6/F	forearm	NA	Malignant	No	No	L, 1 mo	Resection+chemo/, NA
Antonescu et al, 2010 <sup>[5]</sup>	7	9/M	arm	EWSR1	Benign	NA	NA	NA	NA
		7/F	hip	EWSR1	Malignant	NA	NA	NA	NA
		11/F	forarm	EWSR1	Malignant	NA	NA	NA	NA
		13/F	peri-ocular	EWSR1	Malignant	NA	NA	NA	NA
		7/F	peri-orbital	EWSR1	Malignant	NA	NA	NA	NA
		1/M	head&neck	EWSR1	Malignant	NA	NA	NA	NA
		2/F	mediastinum	EWSR1	Malignant	NA	NA	NA	NA
Rekhi et al, 2012 <sup>[25]</sup>	1	18/M	inguinal	No	Benign	Yes	NA	NA	NA
Antonescu et al, 2013 <sup>[17]</sup>	3	3mo/M	knee	No	Malignant	NA	NA	NA	NA
		3/M	mediastinum	No	Malignant	NA	NA	NA	NA
		8/F	thigh	No	Malignant	NA	NA	NA	NA
Park et al, 2013 <sup>[26]</sup>	1	18/M	forearm	NA	Benign	No	No	No	Resection/NED, 4 y
Gupta et al, 2016 <sup>[27]</sup>	1	2/F	CNS	NA	Malignant	Yes	Yes	Cervical spine	chemo+Resection+XRT/AwD, 30 mo
Baldovini et al, 2018 <sup>[28]</sup>	1	newborn	neck	NA	Malignant	No	No	No	Resection+chemo+XRT/NE
Bhanvadia et al, 2017 <sup>[29]</sup>	1	14/M	gluteal	NA	Benign	No	No	No	D, 12 mo
De Cates et al, 2020 <sup>[30]</sup>	1	15/M	skull base	No	Benign	No	No	No	Resection/NED, 24 mo
Segawa et al, 2020 <sup>[8]</sup>	2	16/M	groin	EWSR1	Benign	No	No	No	Resection/NED, 24 mo
		5/F	back	EWSR1	Benign	No	No	No	Resection/NED, 3 mo
Trevino et al, 2020 <sup>[10]</sup>	1	12/M	foot plantar	NA	Malignant	No	NA	L, pericardium, RP	Resection/NA
The present case	1	9/M	mediastinum	EWSR1	Benign	Yes	Yes, 6 mo	No	Resection/AwD, 7 mo

**Note.**\* Mitoses numbered eight per ten. <sup>a</sup>Cases with ductal differentiation/structures were excluded. <sup>b</sup>TNF-α, melphalan, perfusion followed by amputation. Abbreviations: chemo, chemotherapy; NA, not available; NED, no evidence of disease; XRT, radiation therapy; AwD, alive with disease; LN, lymph node; L, lung; RP, retroperitoneum; REC, recurrence; CNS, central nervous system; mo, month; y, year.

## References for Supplementary Tables

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