

Table S1. Detailed data of the patients treated with chemotherapy in the first-line treatment of MNT1.

PATIENTS TREATED WITH ADJUVANT CHT

patient's no.	author, year of study	patient's age at diagnosis [months], sex	primary tumor site and size [cm]	metastases at diagnosis	initial management (before adjuvant CHT)	adjuvant CHT (cytostatic drugs, duration)	response to CHT	further course of treatment	follow-up from the beginning of treatment	outcome
1.	Cohen 1988 [1]	4, ND	CNS, ND	ND	R2 resection	vincristine, lomustine, methylprednisolone, procarbazine, hydroxyurea, cisplatin, cytosine arabinoside, cyclophosphamideⁱ 12 months	stable disease	progression → RTX	26 months	alive with disease
2.	De Chiara 1992 [2]	17, M	epididymis 2.3x0.9	yes, LN	complete resection → LN resection	cyclophosphamide, doxorubicin, cisplatin, etoposideⁱⁱ 18 months	no recurrence	-	28 months	NED
3.	Hoshino 1994 [3]	5, M	skull 4x4	no	R2 resection → R2 resection	vincristine, lomustine, methylprednisolone, procarbazine, hydroxyurea, cisplatin, cytosine arabinoside, cyclophosphamideⁱⁱⁱ 3 months	complete response	-	≈ 10 months	NED
4.	Patankar 1998 [4]	9, ND	skull ND	ND	biopsy → R2 resection	cyclophosphamide, doxorubicin^{iv} 4 months	complete response	-	≈ 23 months	NED
5.	Kumari 2005 [5]	4, M	skull 9.2x7.8	ND	R2 resection	vincristine, cyclophosphamide^v 6 months	partial response	R0 resection	16 months	NED
6.	Yoo 2014 [6]	28, F	CNS, ND	yes, spinal canal, inguinal sac peritoneum, cardiophrenic region (disseminated through VP shunt)	R2 resection of the inguinal mass	ifosfamide, doxorubicin, vincristine^{vi} 1,5 months	partial response	progression	5 months	DOD
7.	Higashi 2016 [7]	3, F	maxilla, ND	ND	biopsy -> R1 resection	vincristine, cyclophosphamide, pirarubicin, cyclophosphamide, carboplatin^{vii} ≈ 5 months	no recurrence	-	≈ 3 years	NED
8.	Murphy 2016 [8]	3, M	maxilla 3.5x2x1	no	biopsy → R1 resection → R1 resection	vincristine, cyclophosphamide^{viii} 2,5 months	no recurrence	-	3 years	NED
9.	Azari 2016 [9]	6, M	mandible 9.4x6.7x6	yes, cervical LN	biopsy → R0 resection	ifosfamide, vincristine, doxorubicin/dactinomycin^{ix} 7 months	no recurrence	-	4,5 years	NED
10.	current study, patient 3*	3, F	suboccipital area, spinal canal 4x4x3.1	no	biopsy → 2 courses of neoadjuvant CHT → R2 resection	cisplatin, etoposide, vindesine, vincristine, dacarbazine, ifosfamide^x 2 months	stable disease	progression after 3 years	4,5 years	alive with disease

PATIENTS TREATED WITH NEOADJUVANT CHT

patient's no.	author, year of study	patient's age at diagnosis [mo], sex	primary tumor site and size [cm]	metastases at diagnosis	initial management (before neoadjuvant CHT)	neoadjuvant CHT (cytostatic drugs, duration)	response to CHT	further course of treatment	follow-up from the beginning of treatment	outcome
11.	Pierre-Kahn 1992 [10]	2, F	skull 10x6	ND	biopsy	cyclophosphamide, doxorubicin	stable disease	R0 resection	7 years	NED
12.	Mello 2000 [11]	6, F	maxilla 6	ND	biopsy	vincristine, cyclophosphamide, doxorubicin, vincristine, cisplatin, tenoposide^{xi} 2,5 months	partial response	R0/R1 resection	≈ 22 months	NED
13.	Kumari 2005 [5]	4, F	skull 20.5x18.5	no	biopsy	vincristine, doxorubicin, cyclophosphamide^{xii} 12 months	partial response	R0 resection	20 months	recurrence, lost to follow-up
14.	Hered 2007 [12]	6, M	orbit ND	ND	biopsy	vincristine, etoposide, ifosfamide^{xiii} 5 months	partial response	R2 resection	3,5 years	NED
15.	Choi 2007 [13]	5, M	femur 5x3.8x2,5	no	biopsy	vincristine, doxorubicin, cyclophosphamide^{xiv} 5 months	partial response	resection → CHT	6 months	D (cardiomyopathy)
16.	Rekhi 2011 [14]	12, M	femur 6x4x3	no	biopsy	vincristine, doxorubicin, cyclophosphamide 2 months	stable disease	RTX → R0 resection	ND	ND
17.	Maroun 2016 [15]	4, F	mandible 6.9x4.4x3.3	ND	biopsy	vincristine, dactinomycin, cyclophosphamide^{xv} 2,5 months	partial response	R0 resection	>2 years	NED
18.	Creytens 2017 [16]	3, M	forearm 3	no	biopsy	ifosfamide, vincristine, dactinomycin, doxorubicin^{xvi} 4 months	partial response	R0 resection	28 months	NED
19.	Moreau (a) 2018 [17]	inborn, M	maxilla 4.7x4	no	biopsy	cyclophosphamide, vincristine, etoposide, carboplatin, doxorubicin^{xvii} 3 months	progression → stable disease after last course	R2 resection	>2,5 years	NED
20.	Khemiri 2018 [18]	7, M	mandible 8x4	ND	biopsy	ifosfamide, etoposide^{xviii} ≈ 5 months	partial response	resection	ND	ND
21.	Nicosia 2017 [19]	48, M	skull ND	yes, cervical LN	LN biopsy	topotecan, cyclophosphamide, etoposide, ifosfamide, etoposide^{xix} 5 months	partial response	lymphadenectomy after 3 CHT courses, R2 resection after 5 CHT courses → another R2 resection → CHT + RTX → parents refused treatment → progression	≈ 1 year	DOD
22.	current study, patient 3*	3, F	suboccipital area, spinal canal 4x4x3.1	no	biopsy	vincristine, cyclophosphamide, doxorubicin^{xx} 1 month	stable disease	R2 resection → CHT → progression after 3 years	4,5 years	alive with disease

patient's no.	author, year of study	patient's age at diagnosis [mo], sex	primary tumor site and size [cm]	metastases at diagnosis	initial management	CHT (cytostatic drugs, duration)	response to CHT	further course of treatment	follow-up from the beginning of treatment	outcome
23.	Woessmann 2003 [20]	2, M	maxilla ND	ND	biopsy	vincristine, ifosfamide, dactinomycin → vinblastin, ifosfamide/cyclophosphamide, etoposide, doxorubicin/liposomal daunorubicin, dactinomycin ^{xxi} 10 months	first regimen → progression second regimen → subtotal tumor calcification	-	22 months	alive with stable disease
24.	Sailukar 2007 [21]	3, M	maxilla 8x6	ND	biopsy	vincristine, cyclophosphamide, dactinomycin ^{xxii}	stable disease	parents refused local treatment	ND	ND
25.	Moreau (b) 2018 [22]	4, ND	mandible 5.5	no	biopsy	etoposide, carboplatin	partial response	-	16 months	alive with stable disease
CHT = chemotherapy; CNS = central nervous system; D = died; DOD = died of disease; F = female; HSCT = hematopoietic stem cell transplantation; LN = lymph nodes; M = male; ND = no data; NED = no evidence of disease; R0 = macroscopically complete resection with microscopically negative margins; R1 = macroscopically complete resection with microscopically positive margins; R2 = macroscopically incomplete resection; RTX = radiotherapy * = patients who underwent different types of CHT and have been included in the tables more than once										

ⁱ **vincristine** (1,5mg/m2), **lomustine** (15mg/m2), **methylprednisolone** (300mg/m2 x3), **procarbazine** (75mg/m2), **hydroxyurea** (1500mg/m2), **cisplatin** (60mg/m2), **cytosine arabinoside** (300mg/m2), **cyclophosphamide** (300mg/m2) every 4-6 weeks

ⁱⁱ alternating courses

ⁱⁱⁱ 2 courses

^{iv} „alternate courses” every 21 days

^v **vincristine** (0.05 mg/kg), **cyclophosphamide** (750 mg/m2 initially started in dose and then slowly increased), 8 courses every 3 weeks,

^{vi} 2 courses

^{vii} 1 course of **vincristine** (1.5 mg/m2), **cyclophosphamide** (600 mg/m2) and **pirarubicin** (30 mg/m2) followed by 5 courses of **vincristine** (1.5 mg/m2) , **cyclophosphamide** (900 mg/m2) and **carboplatin** (450 mg/m2)

^{viii} weekly **vincristine** until week 6 and then 3 x weekly **vincristine** and 3 x weekly **cyclophosphamide** courses

^{ix} alternating **doxorubicin/dactinomycin**, 9 courses

^x 2 courses of **cisplatin, etoposide, vindesine** and 1 course of **vincristine, dacarbazine, ifosfamide**; details in case description

^{xi} **vincristine** (day 1; 0.7 mg/m2), **cyclophosphamide** (days 1-7; 75 mg/m2), **doxorubicin** (day 8; 18 mg/m2), **vincristine** (day 8; 0.7 mg/m2), **cisplatin** (day 15; 40 mg/m2), **tenoposide** (day 17; 125 mg/m2), 3 courses

^{xii} **vincristine** (0.05 mg/kg), **doxorubicin** (40 mg/m2 initially started in dose and then slowly increased), **cyclophosphamide** (750 mg/m2 initially started in dose and then slowly increased), 1 course every 3 weeks for 12 months

^{xiii} 7 courses every 3 weeks

^{xiv} **vincristine** (0.05 mg/kg), **doxorubicin** (40 mg/m2), **cyclophosphamide** (750 mg/m2), 6 courses every 3 weeks

^{xv} **vincristine** (0.025 mg/kg/dose), **dactinomycin** (0.025 mg/kg/dose), **cyclophosphamide** (36 mg/kg/dose), 3 courses every 3 weeks

^{xvi} RMS 2005 protocol (standard risk group, subgroup D); **doxorubicin** was added after patient reached 6 months of age

^{xvii} 1 course of low-dose **cyclophosphamide** and **vincristine**, 2 courses of **etoposide** and **carboplatin**, 1 course of **cyclophosphamide, doxorubicin** and **vincristine**

^{xviii} 6 courses of **ifosfamide** (100 mg/kg for 3 days) and **etoposide** (5 mg/kg for 3 days).

^{xix} **topotecan** (days 1-5; 0.375 mg/m²) and **cyclophosphamide** (days 1-5; 125 mg/m²), alternating with **cyclophosphamide** (day 1; 4 g/m²) and **etoposide** (days 2-4; 200 mg/m²) or **ifosfamide** (days 1-3; 3 g/m²) and **etoposide** (days 1-3; 150 mg/m²), 5 courses

^{xx} 2 courses, details in case description

^{xxi} **vincristine, ifosfamide, dactinomycin** 2 courses → **vinblastin, ifosfamide/cyclophosphamide, etoposide, doxorubicin/liposomal daunorubicin** and **dactinomycin** 13 courses every 3 weeks

^{xxii} 1 course

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