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REVIEW

Malignant intraventricular meningioma: literature review and case report

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Abstract

Malignant intraventricular meningiomas (IVMs) are very rare with only a few reported cases. A midline search up to December 2020 selected 40 articles for a total of 65 patients. The inclusion criteria were series and case reports in English language, as well as papers written in other languages, but with abstracts written in English. Malignant IVMs at the first diagnosis (group A, 50 patients) and those with anaplastic transformation from previous WHO grades I and II tumors (group B, 15 patients) were separately analyzed. The unique personal case among 1285 meningiomas (0.078%) is also added. Malignant IVMs mainly occur in women (61%) with a median age of 45 years and are mainly located in the lateral ventricle (93%) and trigonal region (74%), with no cases in the fourth ventricle. Irregular borders (80%), heterogeneous enhancement (83%), and perilesional edema (76%) are the most frequent radiological findings. The histology was mainly pure anaplastic (85%), whereas papillary (7%), rhabdoid (5%), and mixed forms (3%) are very rare. The CSF spread was found in 60% of the cases. The prognosis is very dismal, with an overall median survival of 17.5 months after surgery for the anaplastic forms. Malignant IVMs at initial diagnosis (group A) show better overall survival (25 months) than those occurring from anaplastic transformation of lower grade tumors (group B) (10.1 months).

Keywords Intraventricular malignant meningioma · Lateral ventricle · Third ventricle · Papillary meningioma · Rhabdoid meningioma

Introduction

Intraventricular meningiomas (IVMs) are unusual tumors, which are believed to arise from the stroma of the choroid plexus or tela choroidea [1]. They account for 0.5 to 5% of all intracranial meningiomas [1] and are most commonly located in the lateral ventricle (77.8%), mainly in the trigonal area [2, 3]. The distribution of the intraventricular meningiomas in the different grades according to the 2016 WHO classification [4] is similar to that of the overall meningiomas. In a recent literature review of 608 intraventricular meningiomas with histologic description, Pereira et al. [5] found 89.9% of grade I tumors, 7.4% of grade II, and 2.8% of grade III (17 cases). In another review [6], only 20 reported cases were identified.

The estimated annual, age adjusted, incidence rate for malignant meningiomas is 0.12 per 100,000 individuals in the general population [7]. Thus, the intraventricular location is found with an incidence of 0.0005–0.003 per 100,000 individuals (about one case every year).

This paper reports a case of malignant intraventricular meningioma at recurrence after surgery for an atypical tumor and analyzed 65 cases from a large literature review. Cases of malignant meningiomas at initial diagnosis and those with malignant transformation at regrowth or recurrence after resection of benign or atypical meningiomas are separately analyzed.

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Methods

A midline search from January 1980 to December 2020 was made by using the key phrases "intraventricular malignant meningioma," "intraventricular anaplastic meningioma," "intraventricular papillary meningioma," "intraventricular rhabdoid meningioma," "lateral ventricle meningioma," "third ventricle meningioma," and "fourth ventricle meningioma." The inclusion criteria were case reports, surgical series, and other reviews including cases of intraventricular tumors whose histological diagnosis was malignant meningioma, and articles published in English language, as well as papers written in other languages, but with the abstract in English. Studies with no abstract in English and those including previous already reported cases were excluded.

The analyzed factors are patient age and sex, signs and symptoms at presentation, meningioma location, tumor borders and enhancement, perilesional edema, histologic type, CSF spread and metastases, number of surgeries, adjuvant therapies, and survival. On postcontrast magnetic resonance imaging, the tumor borders were defined as regular or irregular, and the tumor enhancement as homogeneous or inhomogeneous. The perilesional edema was defined as hyperintense signal of the periventricular brain tissue on T2-weighted images. The histology was graded according to the classification reported in the included studies.

The patients were divided in two groups: group A with malignant WHO grade III tumor at initial diagnosis and group B with malignant meningioma at progression or recurrence after resection of a grade I or grade II tumor.

A retrospective review of 1285 patients with meningiomas operated on in our neurosurgical unit from January 1980 through November 2020 disclosed 10 cases of benign WHO I not recurred meningiomas (0.8%) and only one case of malignant WHO III intraventricular meningioma at recurrence (0.078%). The clinical history, magnetic resonance (MR) imaging, surgical description, and histological specimens of the patient were reviewed. The immunohistochemical study included the proliferation index Ki67-MIB1 and the progesterone receptor (PR) expression. The biomolecular analysis was made including the mutation analysis and the methylome analysis. The mutation analysis of the promoter of the gene C228T (TERT) was conducted by amplicon sequencing [8]. The methylome analysis was conducted by EPICarry Illumina to provide information on the methylation state at 850.000 CpG sites, gene copy number variation, and loss and gain of chromosome genetic material. The samples of methylome profiles were compared to those present in Tumor Cancer Genome Atlas database [9].

Results

Case description

In 2004, a 57-year-old woman underwent complete surgical resection of a 2-cm left trigonal intraventricular tumor through left parietal-occipital transcortical approach.

The histopathological and immunohistochemical studies diagnosed an atypical WHO grade II meningioma with Ki67 Li 15% and progesterone receptor (PR) expression 50%.

The patient was again observed in June 2009 because of the appearance of right lateral homonymous hemianopsia, ataxia, short-term memory deficit, and confusion. MRI (Fig. 1a) showed recurrence of the left trigonal meningioma, with heterogeneous enhancement and peritumoral edema.

At reoperation, through the same surgical approach, a soft hemorrhagic tumor was removed. It was attached to the choroid plexus with focal invasion of the ventricular wall.

The histopathology showed a neoplasm with pleomorphic cells, high mitotic activity, and brain invasion. The diagnosis was atypical meningioma with Ki67 Li 17% and PR expression 30% (Fig. 1b–c). The biomolecular analysis showed mutation of the promoter of the C228T gene (TERT) and changes of the copy number variations of chr4, chr10, and chr22.

After the operation, the lateral homonymous hemianopsia persisted. No residual tumor was observed at 1-month follow-up. Radiotherapy to the surgical field with dose of 54 Gy in 30 fractions was administered.

In November 2011, the patient was again observed. She was unable to walk and presented confusion. Magnetic resonance (Fig. 2a–b) showed a large (39mm) recurrent tumor, with very irregular margins, inhomogeneous enhancement, and diffuse infiltration of the brain parenchyma. The reoperation was decided with the aim to tumor debulking. A soft vascular neoplasm with extensive infiltration of the ventricular wall and brain parenchyma was found. Thus, only a partial resection was possible. The histopathologic studies showed malignant (WHO grade III) meningioma with rhabdoid features, Ki67 Li 22%, and PR expression 5% (Fig. 2c). The biomolecular analysis showed similar results than those of the specimens of the second operation.

Postoperatively, the clinical conditions were substantially stable.

The patient refused chemotherapy. She died for tumor progression in November 2012, 8 years after initial surgery, and 1 year after the diagnosis of malignant transformation, as confirmed through phone contact by her daughter.

Literature review

The literature review has disclosed 40 articles [6, 10–48] for a total of 65 patients. Among the reviewed cases, 50 were malignant WHO III intraventricular meningiomas at initial

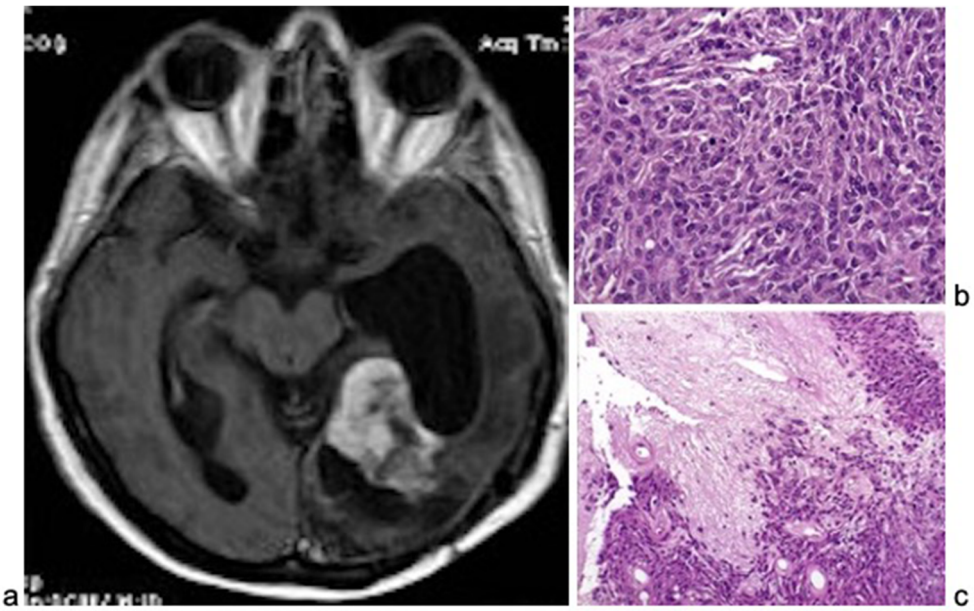

Fig. 1 **a** Cranial magnetic resonance (2009), post-contrast axial T1-weighted sequence: recurrent meningioma of the left trigone with irregular margins and inhomogeneous contrast enhancement; significant enlargement of the left lateral ventricle; **b**–**c** histological specimens, H-E 40× (**b**) and 20× (**c**): neoplasm characterized by hypercellularity and high mitotic activity; the neoplastic cells are pleomorphic with evident nucleolation. Brain invasion is evident (**c**). These features are consistent with an atypical meningioma

diagnosis (group A) [6, 10–13, 15–17, 19–22, 24–26, 29–32, 35–37, 40, 41, 43–48] and 15 were malignant meningiomas at the progression or recurrence after one or more operation for WHO I or II tumors [6, 14, 15, 18, 21, 23, 27, 28, 33, 34, 38, 39, 42].

The data of the 66 cases, including our own, are exposed and summarized in Tables 1, 2, 3, and 4.

Epidemiological and clinical data

The sex was specified in 41 patients; among them, 25 (61%) were females and 16 (39%) were males. The mean age at diagnosis was 45 years (range 3 to 81 years). Only 3 patients were children in the first decade of life. No significant differences of the age and sex were found between group A and B cases (Table 3).

The clinical symptoms and signs were specified in 39 patients. Headache (38%), symptoms of increased intracranial pressure (33%), and focal neurological deficits (33%) were the most frequent clinical manifestations (Table 3). Dizziness and gait ataxia were found in 5 patients (13%). Mild to severe deterioration of the consciousness was present in 7 cases with intracranial hypertension. Epileptic seizures were rare (8%). Only in one patient (3%), the intraventricular meningioma was found as incidental finding.

Location and radiological features

Data on tumor location within the ventricular system have been reported in 42 cases; 39 (93%) were in the lateral ventricles and only 3 (7%) in the third ventricle, with no cases in the fourth ventricle. The trigone was the most common location (74% of all cited locations and 31/34 or 91% of all lateral ventricle tumors where the site is specified) (Table 3). The side of the 38 tumors of the lateral ventricles with specified data was more frequently the right (68%). A rather similar

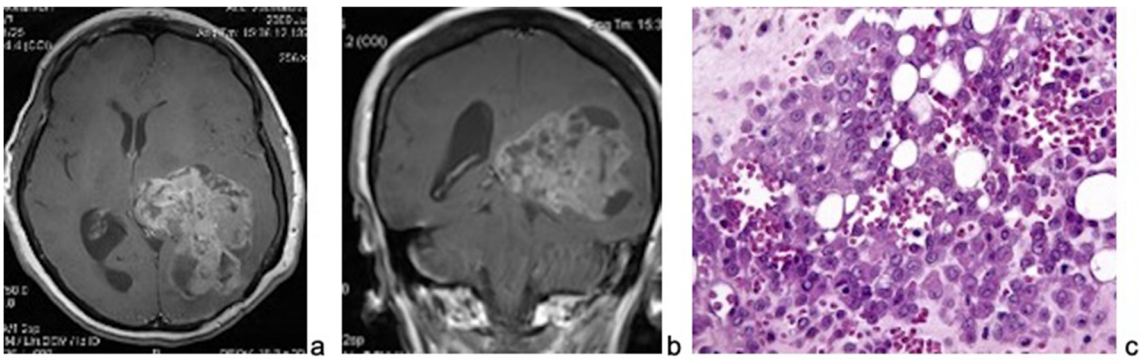

Fig. 2 **a**–**b** Cranial magnetic resonance (2011), post-contrast axial (**a**) and coronal (**b**) T1-weighted sequences: large (39mm) recurrent meningioma with very irregular margins, inhomogeneous contrast enhancement, and diffuse infiltration of the brain parenchyma; **c** histologic specimen: neoplasm characterized by anaplastic cells with high mitotic activity and rhabdoid features (H-E 40×). The diagnosis is in favor of a malignant meningioma

Table 1 Data of 50 reported cases of malignant WHO III intraventricular meningiomas at the first diagnosis

	Authors/year	Sex/age (y)	Symptoms	Location	Attachment	Tumor enhancement	Tumor borders	Perilesional edema	Number of surgeries	Type of WHO III meaning	Adjuvant therapy	Time to progression	Survival	CSF dissemination
1	Kleinschmidt et al.[10] 1985	M, 34	ns	l. trigone	ns	ns	ns	ns	4	A	RT+CHT	7 mo.	24 mo.	ns
2	Criscuolo and Symon[11] 1986	ns	ns	ns	ns	ns	ns	ns	1	A	ns	10 wk	ns	ns
3	Strenger et al.[12] 1987	F, 61	Lethargy, ataxia, urinary incont., l. hemiparesis	3rd ventricle	Choroid plexus	Homogeneous	Regular	No	1	A	None	ns	6 wk	Diffuse
4	Kamiya et al.[13] 1989	M, 67	l. hemiparesis and urinary incont., mydriasis, ataxic respiration, sopor	r. trigone	ns	Heterogeneous	Irregular	Yes	1	A	RT	ns	12 mo.	Spine metastasis (surgery)
5	Greenberg et al.[14] 1993	M, 8	ICH awareness	r. trigone	ns	Heterogeneous	Irregular	Yes	2	A	RT+CHT	2 mo.	6.5 mo.	Multiple brain and spinal
6	Sgouros et al.[16] 1995	M, 6	ICH	3rd ventricle	ns	Homogeneous	Regular	No	1	A	None	Stable residual	12 mo.	None
7	Chen et al.[17] 2003	M, 74	Dizziness, memory loss, gait imbalance, l. hemiparesis	r. frontal horn	None	Heterogeneous	Irregular	Yes	1	A	RT	ns	12 mo.	None
8	Erman et al.[18] 2003	M, 65	Seizures, headache	r. trigone	None	Heterogeneous	ns	ns	1	A	ns	ns	Post-op death	None
9	McDermott et al[19]. 2003	F, ns	ns	ns	ns	ns	ns	ns	ns	A	ns	ns	ns	ns
10	McDermott et al[19]. 2003	F, ns	ns	ns	ns	ns	ns	ns	ns	A	ns	ns	ns	ns
11	Regel et al.[22] 2006	M, 17	ICH, l. leg paresis	r. trigone	ns	Heterogeneous	Irregular	Yes	ns	P	ns	ns	ns	None
12	Li and Zhao[21] 2006	ns	ns	ns	ns	ns	ns	ns	1	A	ns	ns	ns	ns
13	Yang et al.[25] 2007	ns	ns	ns	ns	ns	ns	ns	ns	A	ns	ns	ns	ns
14	Kim et al. [28] 2009	ns	ns	l. trigone	ns	Heterogeneous	Irregular	Yes	ns	A	RT	ns	Alive at 13 mo.	ns
15	Kim et al [28] 2009	ns	ns	ns	ns	ns	Irregular	Yes	7	A	RT	ns		ns

Table 1 (continued)

	Authors/year	Sex/age (y)	Symptoms	Location	Attachment	Tumor enhancement	Tumor borders	Perilesional edema	Number of surgeries	Type of WHO III meaning	Adjuvant therapy	Time to progression	Survival	CSF dissemination
1	Kleinschmidt et al.[10] 1985	M, 34	ns	l. trigone	ns	ns	ns	ns	4	A	RT+CHT	7 mo.	24 mo.	ns
													Alive at 64 mo.	
16	Kim et al.[28] 2009	ns	ns	ns	ns	ns	Irregular	Yes	ns	A	RT	ns	Alive at 13 mo.	ns
17	Kim et al.[28] 2009	ns	ns	ns	ns	ns	Irregular	Yes	ns	A	RT	ns	Alive at 13 mo.	ns
18	Zhi et al.[29] 2009	F, 23	ICH	r. trigone	ns	Heterogeneous/cystic	Irregular	Yes	1	P	ns	ns	ns	Brain, subarachnoid
19	Eom et al.[26] 2009	M, 50	Headache	l. trigone	ns	Heterogeneous	Irregular	Yes	2	P+R	RS	30 mo.	50 mo.	Spinal cord - cauda
20	Rosenberg et al.[30] 2009	ns, 37	ns	r. ventricle	ns	ns	ns	ns	1	A	RT	ns	Alive at 104 mo.	ns
21	Sughrue et al[31]. 2010	ns	ns	Intraventricular	ns	ns	ns	ns	ns	A	RT	ns	ns	ns
22	Sughrue et al.[31] 2010	ns	ns	Intraventricular	ns	ns	ns	ns	ns	A	RT	ns	ns	ns
23	Sughrue et al.[31] 2010	ns	ns	Intraventricular	ns	ns	ns	ns	ns	A	RT	ns	ns	ns
24	Baroncini et al.[32] 2011	ns	ns	ns	ns	ns	ns	ns	ns	A	RT	ns	ns	ns
25	Wu et al.[33] 2011	ns	ns	ns	ns	ns	ns	ns	ns	P+R	ns	ns	ns	ns
26	Tao et al.[36] 2014	F, 51	Headache, visual impairment	r. trigone	ns	heterogeneous	irregular	Yes	3	A	No	12 and 18 mo.	20 mo.	Brain, spine (C2), lung
27	Ma et al.[34] 2014	F, 47	Headache	r. ventricle	ns	ns	ns	Yes	1	A	RT	ns	ns	ns
28	Ma et al.[34] 2014	M, 35	ICH	r. ventricle	ns	ns	ns	Yes	1	A	RT	ns	4 mo.	ns
29		F, 48		r. trigone	ns	Heterogeneous	ns	ns	1	A	ns	ns	24 mo.	ns

Table 1 (continued)

	Authors/year	Sex/age (y)	Symptoms	Location	Attachment	Tumor enhancement	Tumor borders	Perilesional edema	Number of surgeries	Type of WHO III meaning	Adjuvant therapy	Time to progression	Survival	CSF dissemination
1	Kleinschmidt et al.[10] 1985	M, 34	ns	l. trigone	ns	ns	ns	ns	4	A	RT+CHT	7 mo.	24 mo.	ns
	Abedalthagafi et al.[37] 2015		Headache, gait instability											
30	Zhu et al[40]. 2015	ns	ns	ns	ns	ns	ns	ns	ns	A	ns	ns	ns	ns
31	Zhu et al.[40] 2015	ns	ns	ns	ns	ns	ns	ns	ns	A	ns	ns	ns	ns
32	Zhu et al[40] 2015	ns	ns	ns	ns	ns	ns	ns	ns	A	ns	ns	ns	ns
33	Zhu et al.[40] 2015	ns	ns	ns	ns	ns	ns	ns	ns	A	ns	ns	ns	ns
34	Zhu et al.[40] 2015	ns	ns	ns	ns	ns	ns	ns	ns	A	ns	ns	ns	ns
35	Fujimaki et al.[41] 2016	M,81	Visual impairment, r. facial palsy, ataxia, dysphasia	l. trigone	ns	ns	ns	ns	None (autopsy)	A	No	Post-mortem	--	Brainstem, spine
36	Yuce et al.[43] 2016	M,20	headache	l. frontal horn	ns	regular	regular	No	1	R	ns	ns	ns	Subarachnoid
37	Kochanski et al.[42] 2016	F,79	l. homonymous hemianopsia; l. hemiparesis	r. trigone	Choroid plexus	Heterogeneous	Irregular	Yes	1	A	ns	ns	ns	No
38	Hua et al.[45] 2018	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns
39	Hua et al.[45] 2018	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns
40	Hua et al.[45] 2018	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns
41	Hua et al.[45] 2018	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns
42	Hua et al.[45] 2018	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns	ns
43	Li et al.[46] 2018	ns	ns	ns	ns	Heterogeneous	Irregular	ns	1	A	RT	ns	15 mo.	No
44	Chen et al.[6] 2019	F, 61	Incidental	l. trigone	ns	Heterogeneous	Irregular	Yes	1	A	RS	No	No	No
45	Chen et al.[6] 2019	F, 49	Dizziness	r. trigone	ns	Heterogeneous	Irregular	No	1	A	No	No	No	No
46		F, 33	ICH, epilepsy	r. trigone	ns	Heterogeneous	Irregular	Yes	1	A	No	ns	ns	ns

Table 1 (continued)

	Authors/year	Sex/age (y)	Symptoms	Location	Attachment	Tumor enhancement	Tumor borders	Perilesional edema	Number of surgeries	Type of WHO III meaning	Adjuvant therapy	Time to progression	Survival	CSF dissemination
1	Kleinschmidt et al.[10] 1985	M, 34	ns	l. trigone	ns	ns	ns	ns	4	A	RT+CHT	7 mo.	24 mo.	ns
	Chen et al.[6] 2019													
47	Jiang et al.[48] 2020	F, 48	Dizziness	l. trigone	ns	ns	ns	ns	1	A	RT	No (32 mo. follow--up)	Alive at 32 mo.	No
48	Jiang et al.[48] 2020	F, 33	ICH	r. occipital horn	ns	ns	ns	ns	1	A	ns	25 mo.	Alive at 25 mo.	No
49	Jiang et al.[48] 2020	F, 51	Headache	r. trigone	ns	ns	ns	ns	3	A	ns	12 mo.(2nd) 30 mo.(3rd)	31 mo.	Spinal
50	Jiang et al.[48] 2020	M, 42	Numbness	l. trigone	ns	ns	ns	ns	1	A	ns	6 mo.	ns	Multiple brain

l left; *r* right; *WHO* World Health Organization; *ICH* intracranial hypertension; *ns* not specified; *mo.* months; *A* anaplastic; *P* papillary; *R* rabdoid; *RT* radiotherapy; *RS* radiosurgery; *CHT* chemotherapy

Table 2 Data Of 16 reported cases of malignant WHO III intraventricular meningiomas at progression or recurrence

	Authors/year	Sex/age (y)	Symptoms	Location	Attachment	Tumor enhancement	Tumor borders	Perilesional edema	WHO grade at initial diagnosis	N. of surgeries from WHO grades I-II tumors	PFS from WHO grades I-II to III	N. of surgeries for malignancy	Type of WHO III	Adjuvant therapy	Survival	CSF dissemination and metastasis
1	Peh et al.[15] 1995	F, 34	Headache, r. quadrantopsia	l. trigone	ns	ns	Irregular	No	I	2 (I to II)	60 mo.	2	P	RT	9 mo.	Brain and spine
2	Darwish et al.[20] 2004	F, 53	Dizziness	r. trigone	ns	homogeneous	regular	no	II	1st atypical 2nd atypical	7 mo.	1	A	RT	Alive > 2 mo.	Brain and spine
3	McMaster et al.[23] 2007	F, 60	Headache, tinnitus, l. hemianopsia	l. trigone		ns		ns	I	1st fibroblastic	60 mo.	1	R	RT	6 mo.	None
4	Shintaku et al.[24] 2007	F, 61	Dizziness, ICH	r. trigone	Choroid plexus	Heterogeneous	Irregular	ns	I	1st transitional	52 mo.	1	A	RS	Alive at 4 mo.	Brain (4th ventricle) spine (T11)
5	Eom et al.[26] 2009	F, 42	Headache dysphasia, r. homony-mous hemianopsia	l. trigone	ns	Heterogeneous	Irregular	Yes	II	1st atypical	38 mo.	1	A	RT	21 mo.	Spine
6	Garcia-Conde et al.[27] 2009	M, 4-4	Headache, incontinenc-e, confusion	r. trigone	ns	ns	ns	ns	II	3	ns	1	A	RT	7 mo.	Brain and liver
7	Sun et al.[35] 2014	F, 38	Headache	r. trigone	ns	Heterogeneous	Irregular	Yes	I	1st fibroblastic	33 mo.	1	A	RT	Alive at 12 mo.	ns
8	Sun et al [35] 2014	M, 4-4	Somnolence, fatigue	r. trigone	ns	Heterogeneous	Irregular	Yes	I	1st transitional	43 mo.	1	A	RT	Alive at 3 mo.	ns
9	Karki et al.[38] 2015	M, 4-9	Headache, l. hemiparesis	3rd ventri-cle	Choroid plexus, foramen Monro	Heterogeneous	Regular	Yes	II	1st atypical	20 mo.	2	P	RS	14 mo.	ns
10	Kwee et al.[39] 2015	F, 3	Lethargy, ICH ataxia	r. ventri-cle	ns	Heterogeneous	Regular	No	II	1st atypical	4 mo.	1	A	CHT	ns	1st atypical (C1)
11	Singh et al.[44] 2017	M, 2-5	ICH, seizures	r. trigone	Choroid plexus	Homogeneous	Irregular	ns	I	1st angioma-tous	6 mo.	1	A	RT	ns	Cranial bone
12	Chen et al.[6] 2019	F, 47	ICH, l. homony-mous hemianopsia	r. trigone	ns	ns	ns	ns	II	1st atypical	46 mo.	3	A	RS	8 mo.	Brain
13	Chen et al[6]. 2019	F, 54	headache	l. trigone	ns	Heterogeneous	Irregular	Yes	II	1st atypical	7 mo.	2	A	RS	none	None
14			l. hemianopsia			Heterogeneous	ns	ns	II	WHO II	ns	ns	A	RT	14 mo.	ns

Table 2 (continued)

	Authors/year	Sex/age (y)	Symptoms	Location	Attachment	Tumor enhancement	Tumor borders	Perilesional edema	WHO grade at initial diagnosis	N. of surgeries from WHO grades I-II tumors	PFS from WHO grades I-II to III	N. of surgeries for malignancy	Type of WHO III	Adjuvant therapy	Survival	CSF dissemination and metastasis
1	Peh et al.[15] 1995	F, 34	Headache, r. quadrantopsia	l. trigone	ns	ns	Irregular	No	I	2 (I to II)	60 mo.	2	P	RT	9 mo.	Brain and spine
	Lesmiewsky et al.[47] 2019	M, 5-3		r. ventri-cle												
15	Jiang et al. [48] 2020	F, 54	Headache	l. trigone	ns	ns	ns	ns	II	1st atypical	7 mo.	1	A	RT	21 mo.	Brain multiple
16	Present case	F, 57	ICH, r. lateral hemianopsia	l. trigone	Choroid plexus	Heterogeneous	Irregular	Yes	II	1st atypical 2nd atypical	70 mo.	1	R	RT	12 mo.	None

PFS progression-free survival; *l* left; *r* right; *WHO* World Health Organization; *ICH* intracranial hypertension; *ns* not specified; *A* anaplastic; *P* papillary; *R* rabdoid; *RT* radiotherapy; *RS* radiosurgery; *CHT* chemotherapy

distribution of meningioma location was found in groups A and B. The site of tumor attachment was specified in only 8 cases and was at the choroid plexus in 6, where 2 tumors had not defined attachment at surgery.

Sufficient radiological data were reported in 29 of the 66 cases (Table 3). The contrast enhancement of the tumor was mainly heterogeneous (83%), because of the presence of necrotic and rarely cystic areas [47]; a homogeneous enhancement, typical of benign meningiomas, was found in only 5 cases (17%). The tumor borders were mainly irregular, with lobulated appearance (23 out of 29 cases or 80%), while in 6 (20%), the tumor had round and regular shape. Variable mild to extensive perilesional edema was evidenced in 22 cases (76%). No significant differences of the radiological features between groups A and B meningiomas were evidenced.

Management and pathology

Among the 66 reviewed patients, including ours, 50 (76%) were operated on for malignant WHO III meningiomas (group I), whereas 16 (24%) underwent previous operations for non-malignant tumors (group B) (Table 4). Among 16 patients of group B, 12 (75%) had one previous surgery, 3 (18%) had two previous surgeries, and one had three operations. The histology at initial surgery was WHO grade I in 6 cases (37.5%) (two transitional and fibroblastic, one angiomatous) and WHO grade II or atypical in 10 (62.5%). In this group, the time to progression from the WHO grade I or grade II tumors to the anaplastic form was rather variable but it occurred in all but one cases within 5 years and in more than one-third within the first year (Table 4). In the overall group of 66 patients, the number of surgeries for WHO III malignant meningiomas was specified in 42; most patients (32 or 76%) underwent one operation, 5 (12%) had two operations, and 5 (12%) had three or more surgeries. No significant differences were found between groups A and B (Table 4).

The most frequent type of WHO III meningiomas, specified in 61 patients, was the classic anaplastic form (52 cases or 85%); on the other hand, the papillary (7%), rhabdoid (5%), and mixed papillary-rhabdoid (3%) types account for 15% of the reviewed cases (Table 4).

The adjuvant therapies are specified in 41 among the 66 patients (Table 4), and particularly in all cases of group B and in 25 of group A. Conventional external radiotherapy was the most frequent treatment (28 patients); it was used in 25 cases after the resection of malignant tumors and in 3 cases after resection of an atypical meningioma. Stereotactic radiosurgery for the treatment of residual nodules was employed in 6 patients and chemotherapy in 3.

Cerebrospinal fluid spread and metastases

The finding of cerebrospinal fluid spread and metastases was specified in 33 patients. Secondary localizations due to tumor spread through subarachnoid space and hematogenous route were reported in 20 cases (60%); brain metastases, including single or multiple subarachnoid nodules or enhancement of dura mater, cranial nerve, or cisternal spaces, were found in 11 patients, and spinal localizations at various levels in 13. Two patients also presented extra-neural metastases to the liver and lung, respectively.

Outcome and survival

The patient outcome and survival after the surgery of the WHO III malignant intraventricular meningiomas is specified in 33 out of the 66 reviewed patients (Table 4). One patient died postoperatively. Twenty died during the follow-up, 6 weeks to 50 months after the operation (median 15 months), whereas 12 were still alive after 2 to 104 months (median 22 months). The overall survival was 17.5 months. In the overall series, only 7 out of the 32 patients with specified follow-up were still alive at 2 years and only one at 5 years. A longer survival time was found among patients with malignant tumors at initial diagnosis (group A) (25 versus 10.1 months of group B).

Discussion

Malignant WHO grade III intraventricular meningioma is a very rare pathological entity, with only 65 cases found in our review. For this reason, the natural history, biological progression, the best management, and the outcome of this exceptional meningioma location are not still defined. We have separately analyzed “de novo” anaplastic meningiomas, where the histological diagnosis of WHO grade III was made at initial surgery, and secondary anaplastic meningiomas, where the anaplastic transformation occurred from the progression or recurrence of WHO grades I and/or II meningioma. This stratification, which was also used in some recent series of anaplastic meningiomas of all locations [49–51], seems to have pathological significance.

The mean age at diagnosis in our review is 45 years, rather similar to that of some reviews of all intraventricular meningiomas [1, 5] and significantly lower than that of two series of anaplastic meningiomas (59 and 60 years) [50, 51]. Three out of 41 patients (7%) with malignant intraventricular meningiomas were children, with an incidence slightly higher than that of all meningiomas in childhood (1.5–3.5%) [52]. As for all anaplastic meningiomas [50], there is not age difference between de novo and secondary tumors.

Table 3 Summary of epidemiological, clinical and radiological data of the 66 reported cases of malignant WHO III intraventricular meningiomas

Covariates	Overall series (66 patients)	Group A (50 patients) WHO III IVMs at initial diagnosis	Group B (16 patients) WHO III IVMs at progression or recurrence	Statistical significance group A vs group B
Age	41* 3–81 years (median 44.3 years old)	25* 6–81 years (median 44.5 years old)	16* 3–61 years (median 45 years old)	ns
Sex	41*	25*	16*	ns
F	25 (61%)	14 (56%)	11 (69%)	
M	16 (39%)	11 (44%)	5 (31%)	
Clinical symptoms and signs	39*	23*	16*	ns
Headache	15 (38%)	7 (30%)	8 (50%)	
ICH	13 (33%)	7 (30%)	6 (37%)	
Focal deficits	13 (33%)	5 (22%)	8 (50%)	
Dizziness	5 (13%)	3 (13%)	2 (12.5%)	
Gait ataxia	5 (13%)	3 (13%)	2 (12.5%)	
Seizures	3 (8%)	2 (9%)	1 (6%)	
Incidental	1 (3%)	1 (4%)	---	
Location	42*	26*	16*	ns
Trigone	31 (74%)	18 (70%)	13 (81%)	
Frontal horn	2 (5%)	2 (7.5%)	---	
Occipital horn	1 (2%)	1 (4%)	---	
3^{RD} Ventricle	3 (7%)	2 (7.5%)	1 (6%)	
Lateral ventricle	5 (12%)	3 (11%)	2 (13%)	
Side	38*	23*	15*	ns
Right	24 (68%)	15 (65%)	9 (60%)	
Left	14 (32%)	8 (35%)	6 (40%)	
Site of attachment	8*	4*	4*	ns
Choroid plexus	6 (75%)	2 (50%)	4 (100%)	
None	2 (25%)	2 (50%)	---	
Tumor borders	29*	18*	11*	ns
Regular	6 (20%)	3 (17%)	3 (27%)	
Irregular	23 (80%)	15 (83%)	8 (73%)	
Tumor enhancement	29*	18*	11*	ns
Homogeneous	5 (17%)	3 (17%)	2 (18%)	
Heterogeneous	24 (83%)	15 (83%)	9 (82%)	
Perilesional edema	29*	20*	9*	ns
Present	22 (76%)	16 (80%)	6 (67%)	
Absent	7 (24%)	4 (20%)	3 (33%)	

*Cases with specified data; *WHO* World Health Organization; *IVMs* intraventricular meningiomas; *ICH* intracranial hypertension; *ns* not significant

Two recent studies [50, 51] have found differences of sex repartition, with overall de novo malignant meningiomas more frequent in women and secondary ones more frequent in men. On the other hand, those located in the ventricular system show female prevalence in both groups (Table 3).

Malignant intraventricular meningiomas occur almost exclusively in the lateral ventricles (93%), with significant predilection for the trigone, and rarely in the third ventricle (7%), with an incidence slightly different from that of all intraventricular meningiomas (80% and 15%, respectively) [5, 32, 53]. This distribution is proportional to the extension of the choroid plexus, the known site of origin of these meningiomas [54]. However, differently from the WHO grade I and II tumors, we did not find cases of malignant meningiomas of the fourth ventricle.

Table 4 Summary of the pathological data, management, and outcome of 66 reviewed patients with malignant WHO III intraventricular meningiomas

Covariates	Overall series (66 patients)	Group A (50 patients) WHO III IVMs at the first diagnosis	Group B (16 patients) WHO III IVMs at progression or recurrence	Statistical significance group A vs group B
WHO grade at first diagnosis	66*	50*	16*	
WHO I	6 (9%)	---	6 (37.5%)	---
WHO II	10 (15%)	---	10 (62.5%)	
WHO III	50 (76%)	50 (100%)	---	
N. of surgeries for WHO grades I-II			16*	
One			12 (75%)	---
Two			3 (18%)	
Three			1 (7%)	
Time to progression from WHO I or II to WHO III			14*	
- < 12 months			5 (35%)	---
- 12–36 months			2 (14%)	
37–60 months			6 (44%)	
> 60 months			1 (7%)	
No. of surgeries for WHO grade III	42*	27*	15*	
One	32 (76%)	21 (78%)	11 (73%)	---
Two	5 (12%)	2 (7%)	3 (20%)	
Three or more	5 (12%)	4 (15%)	1 (7%)	
Type of WHO grade III	61*	45*	16*	
Anaplastic	52 (85%)	40 (89%)	12 (75%)	
Papillary	4 (7%)	2 (4.5%)	2 (12.5%)	ns
Rabdoid	3 (5%)	1 (2%)	2 (12.5%)	
Mixed (papillary-rabdoid)	2 (3%)	2 (4.5%)	---	
CSF spread, metastases	33*	21*	12*	
Yes	20 (60%)	11 (52%)	9 (75%)	
brain	11	8	3	ns
spinal	13	8	5	
extracranial	2	1 (lung)	1 (liver)	
None	13 (40%)	10 (48%)	3 (25%)	
Adjuvant therapies	41*	25*	16*	
Yes	35 (%)	19 (%)	16 (100%)	
RT	28	17	11	---
SRS	6	2	4	
CHT	3	2	1	
None	6 (80%)	6 (%)	---	
Survival	34*	21*	13*	
Dead	20 (med. 15 mo.)	11 (med. 17 mo.)	9 (med. 12.4 mo.)	
Alive	12 (med. 22 mo.)	8 (med. 36 mo.)	4 (med. 5 mo.)	ns
Postoperative death	1	1	---	
Autoptic	1	1	---	
OS (median)	17.5 mo.	25 mo.	10.1 mo.	ns

*Cases with specified data; *WHO* World Health Organization; *IVMs* intraventricular meningiomas; *ns* not significant; *med.* median; *mo.* months; *RT* radiotherapy; *SRS* stereotactic radiosurgery; *CHT* chemotherapy

The attachment of the meningioma to the choroid plexus is specified in only six of the reviewed cases, and particularly in 4 out of 16 secondary meningiomas and only in 2 out of 50 "de novo" ones. We have found in our patient this finding at the operation for the atypical form. It is likely that the adherence and invasion of the ventricular walls in malignant meningiomas do not allow to define the true initial tumor attachment.

The most frequent magnetic resonance features specified in the reviewed cases of malignant intraventricular meningiomas include irregular tumor borders (80%), heterogeneous contrast enhancement with necrotic areas (83%), and variable perilesional edema (76%). All these features are compatible with the diagnosis of malignant meningioma [55], although other neoplasms may be suspected.

Pure intraventricular tumors include meningioma, choroid plexus tumors, and metastases. The choroid plexus carcinoma shows papillary appearance, very irregular margins, large necrotic areas, and frequent brain infiltration [56, 57]; besides, differently from anaplastic meningiomas, they occur almost exclusively in children [56]. Intraventricular metastases account for 0.9 to 4.6% of all cerebral metastases [58], and result from tumor seeding to the choroid plexus. Although their MRI features may be similar to that of a meningioma, the history of a primary neoplasm suggests the correct diagnosis.

Anaplastic intraventricular meningiomas may occur from the transformation of previously treated WHO grade I or grade II tumors [59, 60]. This review first studies their natural history before the anaplastic transformation. Al-Mefty et al. [59]. found that the time to tumor progression of meningiomas from atypical to anaplastic form is significantly shorter than that required for the transformation from benign to anaplastic form (40.8 months vs 99.7 months in women and 177.5 in men, respectively). In the present review, this average rate of transformation is 42 months for tumors with initial diagnosis of WHO grade I, and 30.5 months for tumors with initial WHO II diagnosis. The shorter time of histological progression of intraventricular meningiomas was ascribed to their often large size [1, 21]. This could favor the occurrence of genetic aberrations. However, it is also possible that at least several de novo anaplastic meningiomas could correspond to the anaplastic transformation of previously misdiagnosed low-grade meningiomas. In this review, two cases with anaplastic component at the initial diagnosis and thus classified as de novo also had tumor component of grades WHO I [17] and WHO I and II [10].

The histological progression from benign through atypical to anaplastic meningioma is regulated by several genetic mutations, mainly by TERT promoter mutation [8–10, 51, 59–62]. TERT gene is mutated in meningiomas with histological progression; besides, the TERT promoter mutation is significantly associated with shorter time to progression in all WHO grades [9] and is a prognostic marker of unfavorable progression-free survival in the secondary anaplastic meningioma subgroup [51]. The biomolecular analysis was reported only in one reviewed case of intraventricular malignant meningioma including all WHO I, II, and III tumor components [10]; the TERT mRNA was found to be low in grade I areas and progressively increased in high-grade areas. We have found in both specimens of the second and third operations of our patient TERT mutation and gene alterations of chr4, chr10, and chr12. Thus, although the histological diagnosis was atypical in the second and malignant in the third specimen, according to the values of Ki67-LI (17% and 22%, respectively), their biomolecular profiles suggest malignant behavior in both.

Surgery was the first line treatment in all reported intraventricular malignant meningiomas. About one-third of the patients underwent two or more operations (up to 4 and even 7). This may be justified by the peculiar tumor location in an enlarged ventricular cavity.

Malignant WHO III intraventricular meningiomas are mainly of the classic anaplastic form. Papillary, rhabdoid, and mixed papillary-rhabdoid forms account for 15%, with no difference between de novo and secondary anaplastic tumors.

Radiation therapy is mandatory after resection of anaplastic meningiomas and for incompletely resected or recurrent atypical WHO II meningiomas [63–65]. In this review, only 3 patients with secondary anaplastic tumor received the radiotherapy after the resection of the initial atypical meningioma; other 24 patients were irradiated after resection of the malignant meningioma. We have irradiated our patient after the WHO grade II recurrence. Others suggest radiotherapy also for patients with completely resected atypical meningiomas to reduce the recurrence risk and improve the survival [66–68]. However, it is not defined whether the irradiation may prevent the secondary anaplastic transformation of benign and atypical meningiomas.

The stereotactic radiosurgery, used in only 6 reviewed cases, should be limited to residual or recurrent atypical and anaplastic nodules [65, 69, 70].

Malignant intraventricular meningiomas are at high risk of cerebrospinal fluid spread, found in 20 cases of this review (60% of those where this finding is specified). This high rate is due to both the tumor location and the repeated operations for atypical and malignant tumors. This also explains the slightly higher rate of CSF spread in secondary anaplastic meningiomas (75% vs 52%), which underwent more operations. However, the spread may also occur with no prior surgery [17].

The outcome and survival of malignant meningiomas is dismal, with an overall survival ranging from 2.5 to 5 years [50, 51]. Several studies on malignant meningiomas of all locations [49–51, 62] have demonstrated that de novo anaplastic meningiomas present with better outcome than secondary tumors occurring from progression of WHO I and II forms. This review confirms this finding also for reported intraventricular meningiomas (median survival 25 versus

10.1 months). It has been postulated that the difference of survival may be explained by the greater difficulty to obtaining gross total resections in secondary anaplastic meningiomas after one or often more previous operations for benign-atypical forms.

Malignant intraventricular meningiomas seem to have poorer prognosis than those of other locations, with an overall survival of 17.5 months. This may be due to the higher incidence of local tumor seeding in the ventricular wall and CSF spread.

Conclusion

Malignant intraventricular meningiomas are very rare and mostly occur as primary anaplastic tumors. The tumor location within the ventricular cavities favors the CSF spread, found in more than half of the reported cases. In spite of the repeated operations and adjuvant treatments, the prognosis is worse than malignant meningiomas of other locations, with a shorter survival. Secondary anaplastic intraventricular meningiomas resulting from malignant transformation of WHO grades I and II tumors show worse prognosis and shorter survival than those "de novo" anaplastic.

Advantages

This review discusses all epidemiological, clinical, and pathological aspects, the management, and the outcome of a very rare pathological entity. It includes a significantly higher number of cases than other even recent reviews on this topic. The difference between de novo and secondary anaplastic meningiomas was not studied in previous reports.

Limitations

This study has several limitations. Although the literature review is wide and accurate, the number of cases is rather small, because of the rarity of this pathological entity. Besides, many reviewed studies do not include complete data on several aspects. Thus, the overall rates and the correlation between de novo and secondary anaplastic meningiomas are available only for a low number of cases.

Availability of data and material Not applicable.
Code availability Not applicable.

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Declarations

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Conflicts of interest The authors declare no competing interests.

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