Thieme

Subependymomas – Characteristics of a "Leave me Alone" Lesion

Case Series and Literature Overview

Subependymome – Charakteristika einer „leave me alone"-Läsion

Case series und Literaturübersicht

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ABSTRACT

Purpose Intracranial subependymomas are rare, mostly asymptomatic tumours, which are often found incidentally and therefore did not receive much attention in previous literature. By being classified as benign grade I in the WHO classification of tumours of the central nervous system, they are given a special status compared to the other ependymal tumours. Tumor recurrences are a rarity, spinal "drop metastases" do not occur. While etiological, pathological and therapeutic characteristics have been subject of several publications over the last few decades and have meanwhile been well studied, the imaging characteristics are much less well received.

Material and method Retrospective analysis of our relatively large group of 33 patients with subependymoma, including 4 patients with a mixture of subependymomas with ependymal cell fractions in terms of imaging and clinical aspects and with reference to a current literature review.

Results Subependymomas have typical image morphologic characteristics that differentiate them from tumors of other entities, however, the rare subgroup of histopathological mixtures of subependymomas with ependymal cell fractions has no distinctly different imaging properties.

Conclusions Knowing the imaging characteristics of subpendymoma and their differential diagnoses is of particular importance in order to be able to decide between the necessity of follow-up controls, an early invasive diagnosis or, depending on the entity, tumor resection.

Key Points:

- Subependymomas have typical imaging characteristics that are clearly distinguishable from other entities.
- Increased incidence in middle/ older aged men, most frequent localization: 4th ventricle.
- Symptomatic subependymomas, often located in lateral ventricles, are usually characterized by hydrocephalus.
- Radiological identification of mixed subependymoma with ependymal cell fractions is not possible.
- Image based differentiation from other entities is important for the procedure.

Citation Format

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ZUSAMMENFASSUNG

Ziel Intrakranielle Subependymome sind seltene, meist asymptomatische Tumore, die oft als Zufallsbefund auffallen und in der Literatur wahrscheinlich aus diesem Grund wenig

Beachtung finden. In der WHO-Klassifikation der Tumoren des zentralen Nervensystems mit Grad I als benigne klassifiziert, nehmen sie gegenüber den übrigen ependymalen Tumoren eine Sonderstellung ein. Tumorrezidive sind eine Rarität; spinale Abtropfmetastasen kommen nicht vor. Während ätiologische, pathologische und therapeutische Charakteristika seit Jahrzehnten Gegenstand einiger Publikationen und mittlerweile gut untersucht sind, findet der bildmorphologische Aspekt deutlich weniger Beachtung.

Material und Methode Retrospektive Analyse unseres relativ großen institutseigenen Kollektivs von 33 Patienten mit Subependymom einschließlich 4 Patienten mit einer Mischform aus Subependymomen mit ependymalen Zellanteilen hinsichtlich bildgebender und klinischer Aspekte und Bezug auf eine aktuelle Literaturübersicht.

Ergebnisse Subependymome weisen typische bildmorphologische Charakteristika auf, die sie von Tumoren anderer Entität, nicht aber der seltenen Sonderform eines histopathologischen Mischbildes aus Subependymom- und Ependymomanteilen, unterscheiden.

Schlussfolgerung Bildmorphologische Eigenschaften der Subependymome und Differentialdiagnosen zu kennen ist von besonderer Bedeutung, um je nach Entität zwischen Verlaufskontrollen, einer zeitnahen invasiven Abklärung und gegebenenfalls Resektion entscheiden zu können.

Background

The authors are unaware of a large case series on image-based morphological characteristics of subependymomas. Although etiological, pathological and therapeutic characteristics have been the subject of some publications for decades and have now been well studied, the image-based morphological aspect has received much less attention [1 – 3].

Introduction

Subependymomas are a rare, benign, noninvasive entity of ependymal origin representing approximately 0.2 – 0.7 % of all intracranial tumors worldwide [1, 3, 4]. The true incidence remains unclear, since subependymomas are generally asymptomatic and usually appear as incidental findings in autopsies or imaging. Since Scheinker's initial description in 1945 [5], there have been few case series with generally small cohorts [2, 6 – 9]. In addition to the more frequent occurrence of subependymomas of the fourth ventricle (approx. 56 – 60 %), origin of tumor growth can be observed on the lateral ventricle in about 30 – 40 % of cases [2], and more rarely on the spine [4]. The incidence is much more common among middle-aged and older men. Clinical symptoms due to cerebrospinal fluid accumulation correspond to tumor localization; clinical manifestation of tumors in the fourth ventricle is less common, compared with tumors in the lateral ventricles, since CSF accumulation or seizures can occur here more frequently [2, 10]. Tumor recurrence is quite rare [11]; spinal drop metastases have not been described [11].

As initially described by Scheitauer in 1978, there is a seldom-observed special form of a mixed tumor with cell clusters of a subependymoma as well as an ependymoma; that is, morphologically "typical" subependymomas with atypical growth tendency [1].

Materials and Methods

Patients and techniques

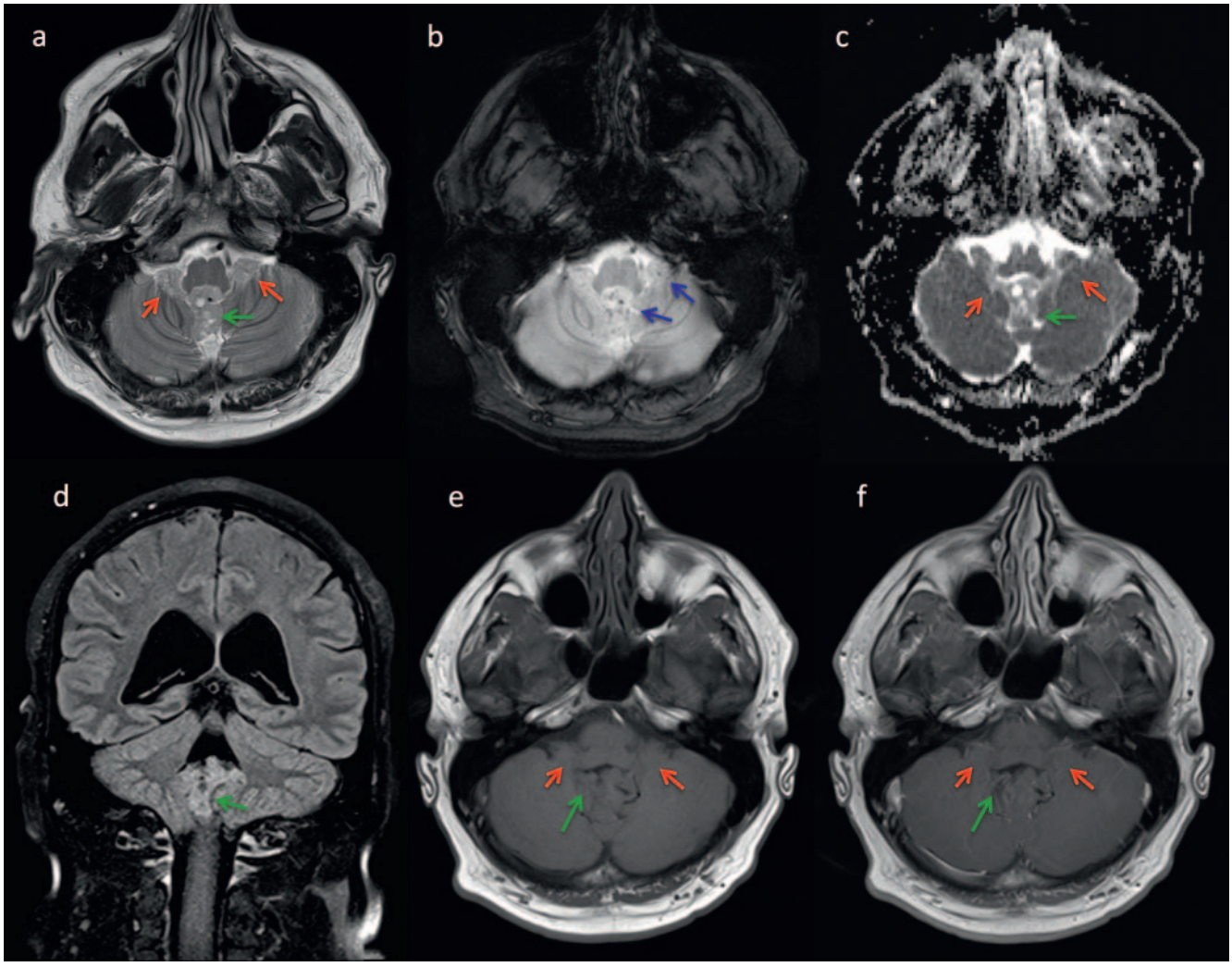
The retrospective study was approved by the local ethics committee. After in-house database research under the keyword "subependymoma" all relevant MRI and CT examinations between January 2009 and January 2017 were included in which subependymomas were morphologically suspected. This corresponded to 33 patients, of whom 22 were male and 11 female. The mean age was 58.6 years (29 – 86 years). Based on clinical indication, and after providing written consent, all patients were examined using 3.0 T magnetic resonance imaging (MRI) equipment (Magnetom Verio, Siemens Healthcare, Erlangen, Germany) or a 1.5 T MRI device (Intera Achieva, Phillips Medical Systems, Eindhoven, Netherlands). Examinations followed a clinically-established protocol including at least axial T2-weighted (w), FLAIR, T1w, T1w after intravenous contrast administration, T2*w, DWI/ADC and coronary T1w after intravenous contrast administration. The contrast medium was administered intravenously in a weight-adapted standard dose (0.1 ml/kg body weight of a 0.1 molar gadolinium-based contrast agent) (Gadovist, Bayer Schering). Morphology, signal and contrast behavior at initial diagnosis as well as the presence of possible growth of the tumors in follow-up studies was consensually described by two experienced neuroradiologists.

Results

Tumor entities

The etiology of 10 tumors was confirmed histologically, of which 6 were subependymomas and 4 intermediate forms between ependymoma and subependymoma.

Due to the incidental findings, wait-and-see behavior with imaging controls was selected for 23 tumors. These remained stable over a period of a few months to 10 years, so that the diagnosis of a subependymoma was made based on imaging.



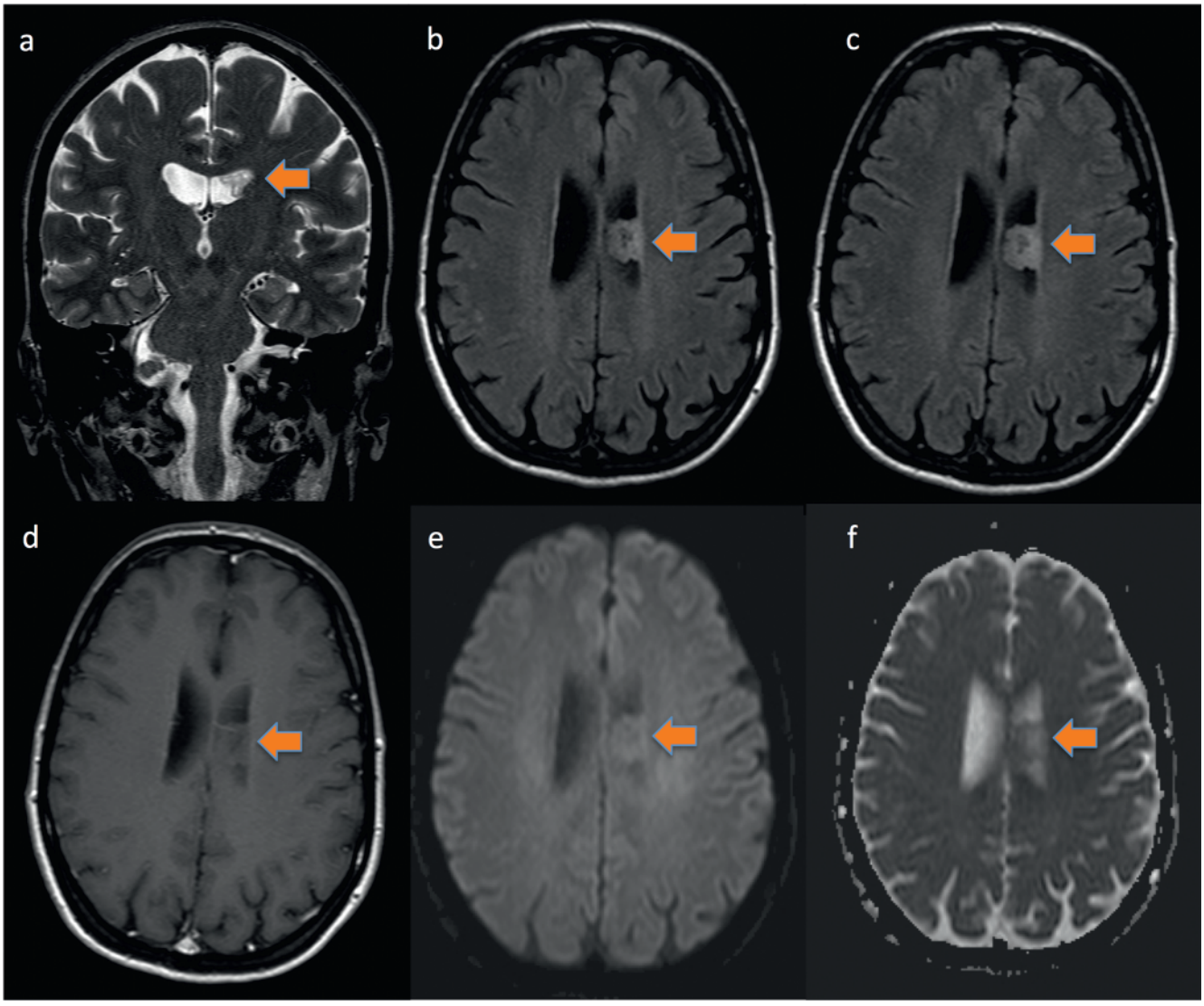
▶ **Fig. 1** Infratentorial Subependymom **a-f**: infratentorial subependymoma in the 4th ventricle with tumor growth over the foramina Lushkae on both sides (red arrows) and the foramen Magendie (green arrow). Susceptibility effects in the T2* sequence indicating small calcifications or bleeding (blue arrows). (**a** T2 TSE, b. T2*, **c** ADC map, **d** FLAIR, **e** T1 before KM and **f** T1 after i. v. gadolinium administration) The images available to us with an interval of 2 years show constant size and shape.

Location and size

In most patients (n = 18), the tumor was located in the fourth ventricle, accompanied in a few cases by tumor tissue growth into the foramen Magendie (n = 10) or the foramina Luschka (n = 4). In 11 patients, the tumor was found in the lateral ventricles, 6 of them in the lateral ventricular anterior horn, 4 in the lateral ventricular posterior horn, and in the cella media of one patient. Less common observed localizations included the cisterna magna with a transition into the right foramen Luschka (n = 1) or exclusive tumor location in the foramen Magendie (n = 3). One of the tumors located in the lateral ventricle was classified as a mixed tumor whereas the other 3 tumors originated from mixed tumor portions of a subependymoma and an ependymoma in the 4th ventricle with transition to the foramina Luschka (n = 2) and the fourth ventricle with transition into the foramen Magendie (n = 1). In addition, there was a wide range of the extent of the tumors; 5 masses were smaller than 1 cm (15.15 %); 8 masses exhibited a width between 1 and 2 cm (24.24 %); 10 had a maximum diameter greater than 2 cm (30.3 %). Three of the mixed-form tumors with subependymoma and ependymal cell clusters were assigned to the last group with diameters greater than 2 cm. The fourth tumor with mixed components exhibited a maximum dimension between 1 and 2 cm.

Imaging characteristics

In most cases (n = 21) the tumors showed a sharp boundary with the adjacent brain tissue (▶ **Fig. 1a**); less commonly there was an irregular sharp distinction (n = 11, see ▶ **Fig. 2b–d**), and in one case (n = 1, indicated in ▶ **Fig. 3f**), there was a fuzzy demarcation with respect to the neighboring brain parenchyma. An irregular blurred boundary and in one case a sharp distinction could be observed in the mixed special forms with ependymoma components. An inhomogeneous parenchymal texture was observed in 25 cases, partly T2-hyperintense to cystic internal components, and in the other 8 cases the internal signal was homogeneous. In the majority of cases (n = 28), a T1w cortex isointense signal



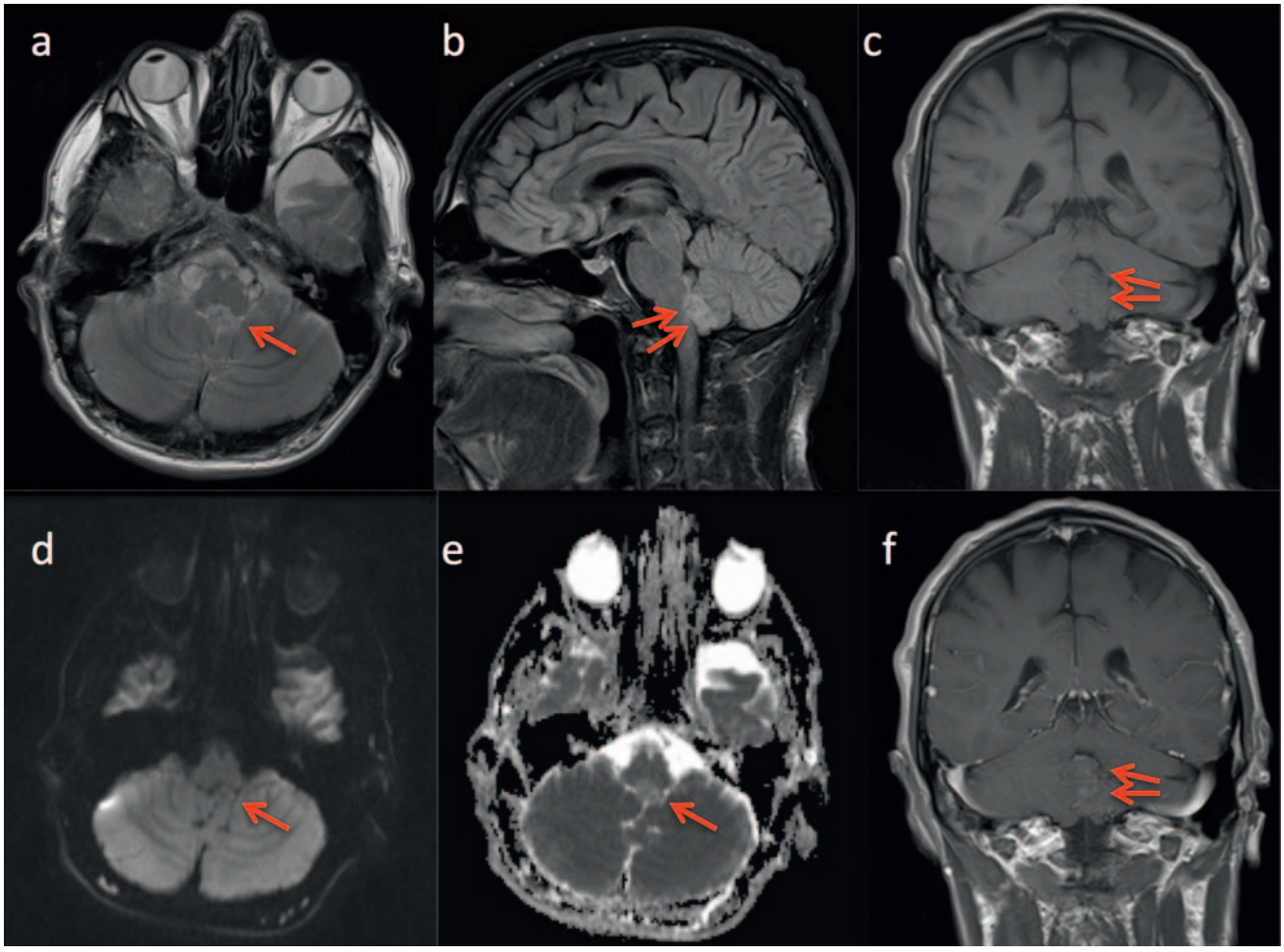
▶ **Fig. 2** **a-f** Subependymoma in the central part of the left lateral ventricle (**a-d** of 2016: **a** T2 TSE, **b** FLAIR, **d** T1 post KM, **e** DWI and **c** of 2007: FLAIR) with a constant size and shape during the 9-year observation period. There is no diffusion restriction (e+f) or contrast enhancement **d**.

behavior of the tumor with respect to the brain parenchyma was observed as a special signal characteristic in the individual sequences; more rarely a comparatively hypointense internal signal (n = 4) was seen in T1-weighting. The T2-weighted signal behavior was predominantly hyperintense (n = 32), less frequently combined with iso- to hypointense components in the case of an overall inhomogeneous signal. Diffusion restrictions were not observed. Contrast medium absorption showed large fluctuations from completely absent to partially nodular or occasionally homogeneously flat contrast accumulation of the tumor tissue (▶ **Fig. 1f, 2, 3f**; ▶ **Table 1**). Small calcifications could be observed in all 6 patients who additionally had undergone skull CT (▶ **Fig. 1b**). Of the tumors with mixed histology, there was only one case of computed tomography which, however, also exhibited calcification of the tumor.

On the whole, there were no MR or CT morphological differences between the "pure" subependymomas and the tumors consisting of the mixed form of subependymoma and ependymal cell clusters.

Clinical presentation

In our retrospective study, 4 patients presented with clinical symptoms of headache, nausea and dizziness due to supratentorial cerebrospinal fluid accumulation caused by the tumor. In these cases, the infratentorial tumor was located in the fourth ventricle with transition into the foramen Magendie in only one instance; in 3 cases the mass was located in the lateral ventricle. With regard to the clinical symptoms in the other patients, we have only limited information based on neurosurgeon reports; in 29 cases, the lesion was discovered by chance. Indications for MRI here were a clarification of various, mainly nonspecific, symptoms such as undirected dizziness, occasional headache, paresthesia, hypoaesthesia and occasional visual disturbances (double vision, flicker, blurred vision), and in another patient there was a gait disturbance



▶ **Fig. 3** Mixed form of subependymoma with ependymal cell fractions **f**: preoperative imaging of an infra-temporal tumor in the 4th ventricle, which histopathologically corresponds to a mixed form of subependymoma with ependymal cell fractions. (**a** T2 TSE, **b** FLAIR, **c** T1 SE native, **d** DWI, **e** ADC-map and **f** T1 SE after i. v. gadolinium administration).

without evidence of CSF accumulation, so a connection between symptoms and tumor was not clear on the whole. In the remaining 4 patients, the indication for imaging could not be clearly determined in hindsight.

Discussion

Subependymomas are rare, low-grade (World Health Organization WHO grade 1) glial neoplasia with an ependymal origin. Their rarity is reflected in the limited number of reports in the scientific literature and low number of cases [3, 12].

In this study, we present a relatively large group of 33 patients with typical imaging subependymomas, of which 4 were histopathologically associated with the mixed special form, and in addition we describe their imaging properties.

In the current WHO classification of 2016 [13], there is, in addition to subependymomas and ependymomas, a special form of mixed subependymomas/ependymomas for which the ependymal components are evaluated according to the WHO classification [1, 3]. Prognostic differences between the subependymomas and mixed subependymomas/ependymomas are still unknown and could not be determined in the previously largest cohort of Rushing et al. [3].

Consistent with the available data in the literature, a hypo-isointense signal behavior of the subependymomas was observed in T1 weighting for the brain parenchyma in all cases of our patient data [3, 10, 14]. For example, in 12 cases of MRI examinations, Rushing et al. and Jain et al. described iso- to hypointensity to the cortical brain parenchyma in T1w, hyperintensity on T2w and a predominant gadolinium enrichment in 80 % of the reviewed cases [3, 14]. However, this observation of the high number of contrast-enhancing subependymomas does not correspond to the majority of the morphological properties of subependymomas described in the literature. In this regard, a wide range of variations has been previously described, ranging from slight [9, 10, 15 – 17] to strong and irregular enhancement [18] as well as individually described and controversially discussed incidence of accumulations [19], in some cases also depending on tumor localization [12]. In our cases, partially nodular enhancement was predominantly evident; rarely an inhomogeneous contrast enhancement was observed. Occasionally a small area enrich-

▶ **Table 1** Overview of the patient population with morphological images with suspicion of subependymoma.

age	symptoms	location	side	size (mm)	boundary	texture	T1 signal	DWI	T2*	T2 signal	CM absorption	miscellaneous	progression	CSF accum.	CT	therapy	histo available?
77	incidental findings	foramen magendi 4th ventricle		4.6 × 8.7	sharp	homogeneous	iso	iso	hyper	hyper	none		n.a.	no	n.a.		
71	incidental findings	lat. ventricle	left	7.5 × 14.3	indistinct	inhomogeneous with cystic components	iso	iso	hypo	hyper to iso	n.a.		n.a.	no	smallest calcifications peripheral		
SO	incidental findings	lat. ventricle	left	12.2 × 17.1	irregular sharp	inhomogeneous with T2 hyperintense components	iso	iso	n.a.	hyper to iso	none		since 2011 constant	no	n.a.		
40	incidental findings	4th ventricle and foramina luschka	bilateral	20.5 × 48.8	irregular sharp	inhomogeneous with T2 hyperintense components	iso	iso	hyper	hyper to iso	partially nodular		postoperatively constant	no	partially calcified	part. resection	WHO I
78	incidental findings	lat. ventricle	right	11 × 12	irregular sharp	inhomogeneous	iso	iso	iso	hyper to iso	none		since 2011 constant	no	n.a.		
63	incidental findings	4th ventricle		4.5 × 6.3	sharp	homogeneous	iso	iso	hyper	hyper	none		n.a.	no	n.a.		
56	incidental findings	4th ventricle and foramen luschka	left	16 × 29	sharp	inhomogeneous	iso	iso	n.a.	hyper to iso	partially punctate		since 2011 constant	no	n.a.		
83	unknown	4th ventricle and foramen luschka	left	30.5 × 34.7	irregular sharp	inhomogeneous	hypo	n.a.	hypo	hyper	inhomogeneous			no	n.a.	part. resection	WHO I/II
30	double vision, flicker	lat. ventricle	right	20 × 23	sharp	homogeneous	hypo	n.a.	n.a.	hyper	n.a.	strong T2 hyper, FLAIR iso	postoperatively constant	yes	n.a.	full resection	WHO I

▶ **Table 1** (Continuation)

age	symptoms	location	side	size (mm)	boundary	texture	T1 signal	DWI	T2*	T2 signal	CM absorption	miscellaneous	progression	CSF accum.	CT	therapy	histo available?
65	incidental findings	4th ventricle		7.1 × 9.9	sharp	homogeneous	iso	n.a.	n.a.	hyper	partially nodular		since 2011 constant	no	n.a.	full resection	WHO I
72	headaches, nausea	lat. ventricle	right	42 × 43	sharp	inhomogeneous	iso	iso	hypo	hyper to hypo	inhomogeneous	partial hyperperfusison	postoperatively constant	yes	partially calcified	full resection	WHO I/II
82	none	right foramen luschka/cisterna magna	right	16.5 × 17.9	sharp	inhomogeneous	iso	iso	n.a.	hyper	none	centrally punctate T2 hypo, T1 hyper	since 2011 constant	no	central calcifications		
48	none	4th ventricle		8.4 × 13	sharp	inhomogeneous	iso	iso	n.a.	hyper	none		n.a.	no	n.a.		
72	none	lat. ventricle	right	35 × 65	sharp	inhomogeneous	hypo	n.a.	partially hypo	hyper to hypo	inhomogeneous		postoperatively constant	yes	n.a.	full resection	WHO I
61	incidental findings	foramen magendi		5 × 6	irregular sharp	inhomogeneous	iso	iso	hyper	hyper	none		constant	no	n.a.		
53	headache, blurred vision, deafness	foramen magendi		10 × 21	irregular sharp	inhomogeneous	iso	iso	n.a.	hyper to iso	partially punctate		since 2007 constant	no	n.a.		
42	incidental findings	foramen magendi 4th ventricle		4.9 × 6.7	irregular sharp	homogeneous	iso	iso	n.a.	hyper	slightly flat		3 months constant	no	n.a.		
31	incidental findings	foramen magendi 4th ventricle		18.6 × 15.9	sharp	inhomogeneous with cystic components	iso	iso	hyper, singly hypo	hyper to hypo	partially nodular		constant	no	n.a.		
51	incidental findings	Foramen magendi 4th ventricle		15.5 × 17.1	irregular sharp	inhomogeneous	iso	iso	n.a.	hyper	partially nodular		postoperatively constant	no	n.a.	full resection	WHO I/II

▶ **Table 1** (Continuation)

age	symptoms	location	side	size (mm)	boundary	texture	T1 signal	DWI	T2*	T2 signal	CM absorption	miscellaneous	progression	CSF accum.	CT	therapy	histo available?
52	incidental findings	lat. ventricle	right	4 × 5	sharp	homogeneous	iso	iso	n.a.	hyper	none		n.a.	no	n.a.		
30	incidental findings	lat. ventricle	right	13.1 × 19.8	sharp	homogeneous	hypo	hypo	hypo	hyper	partially nodular	strong T2 hyper, FLAIR iso	n.a.	no	n.a.		
85	dizziness, headaches	foramen magendi 4th ventricle		15.6 × 24.1	sharp	inhomogeneous						no MRI due to HSM	since 2011 constant	yes	partially calcified		
52	incidental findings	lat. ventricle	right	9.9 × 17	sharp	inhomogeneous	iso	iso	hyper	hyper	partially nodular		since 2012 constant	no	no calcifications		
73	incidental findings	lat. ventricle	right	9.7 × 12.7	sharp	inhomogeneous	iso	iso	hyper	hyper to iso	none		since 2011 constant	no	n.a.		
55	incidental findings	foramen magendi 4th ventricle		26 × 34.9	irregular sharp	inhomogeneous	iso	iso	n.a.	hyper to iso	inhomogeneous		postoperatively constant	no	n.a.	part. resection	WHO I/II
55	incidental findings	4th ventricle foramen magendi and foramen luschka	right	10 × 27	irregular sharp	homogeneous	iso	iso	hypo	hyper	none		constant	no	n.a.		
54	incidental findings	4th ventricle	left	15 × 15	sharp	inhomogeneous	iso	iso	centrally hypo	hyper, centrally hypo	none		n.a.	no	n.a.		
84	incidental findings	lat. ventricle	right	7 × 10.3	sharp	inhomogeneous	iso	iso	hyper	hyper to iso	none		constant	no	n.a.		
53	incidental findings	foramen magendi 4th ventricle		10 × 13.8	irregular sharp	inhomogeneous	iso	iso	hypo	hyper	none		postoperatively constant	no	n.a.	full resection	WHO I

▲ **Table 1** (Continuation)

age	symptoms	location	side	size (mm)	boundary	texture	T1 signal	DWI	T2*	T2 signal	CM absorption	miscellaneous	progression	CSF accum.	CT	therapy	histo available?
48	incidental findings	foramen magendi 4th ventricle	bilateral	26.8 × 30.7	sharp	inhomogeneous	iso	iso	n.a.	hyper	partially punctate		postoperatively constant	no	n.a.	full resection	WHO I
63	incidental findings	foramen magendi 4th ventricle		11.3 × 14.2	sharp	inhomogeneous	iso	iso	n.a.	hyper	none		2 months constant	no	n.a.		
87	incidental findings	foramen magendi		12.9 × 14	sharp	inhomogeneous	iso	iso	n.a.	hyper to iso	inhomogeneous	strong T2 hyper, FLAIR iso	since 2007 constant	no	n.a.		
49	incidental findings	4th ventricle		9 × 12	sharp	inhomogeneous	iso	iso	hyper	hyper	none		constant	no	n.a.	–	

ment was seen. No contrast medium absorption occurred in 15 other tumors, and in 3 cases there were no contrast-based sequences. There was no morphological difference between the subependymomas and the histologically proven mixed subependymomas/ependymomas [7, 12], in particular there was no difference in opacification behavior.

In 1997 Maiuri et al. described the slowly progressive growth of subependymomas in 40 % of tumors. For the most part, subependymomas are detected as incidental findings in MRI; this was also true in our cohort. The presence of clinical symptoms is basically dependent on tumor location. If clinical symptoms occur, this is almost always due to cerebrospinal fluid accumulation; focal neurological abnormalities or epileptic seizures due to the subependymoma were not described by Maiuri et al. and were also not evident in our cohort. Localizations that more frequently lead to cerebrospinal fluid involve tumors on the septum pellucidum in the lateral ventricular anterior horn or posterior horn and interventricular foramen, in contrast to tumors in the cella media of the lateral ventricle or in the fourth ventricle, which relatively rarely lead to a corresponding symptomatology [8, 9]. This could be based on the fact that subependymomas are usually small which in fact may result in displacement of the interventricular foramen; in the fourth ventricle, however, the three CSF drainage possibilities rarely lead to decompensation by blocking all three foramina. Furthermore, in the course of our study, in some cases tumor size constancy over a longer period was found in 25 of 33 cases, including 9 completely or partially resected tumors without recurrence or progression of the residual tumor. In 2 cases, observed retroactively since the first available study, size constancy was evident over a period of 10 years; in 7 cases size remained consistent over the entire recorded evaluation period of 8 years. In 16 other cases, constancy was observed over shorter periods of a few years or months. In the remaining 8 cases there was only a single examination; thus progression evaluation was not possible. Likewise in tumors with mixed pathology of a subependymoma with ependymal component, only a partial resection was performed in 2 cases, and no size constancy could be observed in the postoperative course of several months. A macroscopically complete resection is possible in most cases, due to the predominantly sharp delimitation of the subependymomas in relation to the adjacent brain parenchyma and intraventricular localization [9, 14]; in our cases of partial resection, this was restricted due to adhesions on the adjacent brain stem or inaccessible location in the foramina Luschka. With respect to patients with partial resections, we only had short-term follow-up intervals; Rushing et al. however found no significant difference in survival based on the extent of resection in their 83-patient cohort [3].

There is no consensus in the literature regarding growth tendency; most sources, particularly older, describe mixed tumors as having a particularly strong growth tendency corresponding to the ependymal cell component [7, 12, 18, 20, 21], whereas a more recent study by Rushing et al. [3] could not establish a correlation between survival rate and mixed tumors, so that in typical image findings (see below) even though the presence of ependymal cell clusters cannot be ruled out, confirmation of such a mixed tumor may not have any further consequence than

▶ **Table 2** Overview of differential diagnoses.

ependymomas	▪ younger patients ▪ heterogeneous, CM enhanced lesions with edema ▪ typically in 4th ventricle with hydrocephalus ▪ supratentorial frequently with parenchymal expansion
medulloblastomas	▪ slight to moderate CM enhancement ▪ areas with cystic and necrotic degeneration (among children strong CM enhancement, no necrotic areas) ▪ generally among patients < 10 years, second peak age, 20 – 40 years
subependymal giant cell astrocytomas	▪ moderately CM enhanced lesion in interventricular foramen ▪ frequently calcification ▪ (patients with tuberous sclerosis, subependymal nodes, cortical glioneural hamartoma and medullary layer lesions)
zentral neurocytomas	▪ young patients ▪ typical cystic appearance ▪ lesion in lateral ventricle with relation to septum pellucidum or interventricular foramen ▪ slight to moderate CM enhancement ▪ frequently calcification
meningiomas	▪ strong homogeneous contrast accumulation
choroid plexus papillomas (CPP)	▪ typically pediatric tumors of the lateral ventricle ▪ frequently in 4th ventricle among adults ▪ CM enhanced papillary lesions ▪ Frequently hydrocephalus
hemangioblastomas	▪ cystic lesions with CM-enhanced mural nodes ▪ typically cerebellar, frequently at the pial surface ▪ rarely intraventricular
metastases	▪ primary tumor known as a rule ▪ frequently numerous lesions in the borderline between the marrow and cortex ▪ frequently intraventricular inclusion of the choroid plexus
cavernous malformations	▪ frequently calcifications and T2 hypointense edge of hemosiderin ▪ variable contrast accumulation ▪ rarely intraventricular

would be the case of a subependymoma. Morphologically, the lesions in our cohort primarily exhibited sharp demarcations (96.9 %); 34.4 % demonstrated an irregularly sharp delineation, similar to what has already been described [7, 10, 18]. Cystic interior elements could be observed in 4 cases of our population, just as described in individual studies [18]. Partial T2* signal reduction as a sign of calcification or hemosiderin deposits were evident in 9 of 33 cases (it should be noted that no T2* sequences of the tumor region were available for 15 of the 33 patients); both calcifications and hemorrhages have already be described for individual cases [10, 12, 16, 18, 22]. In addition, no calcifications could be determined in the 5 available CT examinations. In our cases, we could not observe exclusive occurrence of calcifications in infratentorial tumors as described by Chiechi et al.; however more infratentorial than supratentorial tumors with calcifications were observed. Six of the cases with T2* signal voids concerned infratentorial tumors, and 3 concerned supratentorial masses. There was decreasing incidence of tumor localization in the fourth ventricle (54.5 %), in the lateral ventricles (33.3 %), predominantly in the hind horns, and occasionally in the cisterna magna (3 %) or exclusively the foramen Magendie (9 %) corresponding to the frequency distribution in medical literature [4, 12]; this was analogously similar to the frequency distribution noted by Ernestus et al. with 58 % of tumors in the fourth ventricle, and 38 % in the lateral ventricle [2], or the frequency distribution observed by Smith et al. with tumor localization in the fourth ventricle in 50 – 60 % of cases, and 30 – 40 % in the lateral ventricles [18]. Similar to the more commonly observed infratentorial ependymomas, the mixed tumors in our study with subependymoma and ependymoma components primarily demonstrated an infratentorial location; occurrence was in the lateral ventricle in only a single case.

Corresponding to data in the literature, our cohort reflected a distribution of two-thirds males to one-third females affected by subependymomas [2, 18]. The mean age of onset, 58.6 years of age, affects the middle-aged and elderly population, which likewise corresponds to the distribution frequency among the elderly described in the literature [2, 3, 9, 10, 12]; wide variations were observed in our cohort. We did not observe individual mixed tumors among younger patients as described in the literature [20, 21, 23].

Essential differential diagnoses for intracranial subependymomas should include ependymomas, medulloblastomas, astrocytomas, central neurocytomas and meningiomas. Central neurocytomas and ependymomas should be considered in the area of the interventricular foramen; whereas in the fourth ventricle differential diagnosis should take in to account medulloblastomas and

choroid plexus papillomas (CPP) [15]. Refer to ▶ **Table 2** for the main morphological imaging differentiation options [24, 25].

SUMMARY

- Subependymomas are rare, benign, predominantly asymptomatic, intraventricularly localized glial tumors of ependymal origin with no growth tendency with typical morphological characteristics that distinguish them from tumors of other entities and with different dynamics in the same localization.
- It is of particular importance to know this fact, as the suspicion of the chance finding of a subependymoma justifies a wait-and-see attitude and imaging follow-up at greater intervals, whereas other tumors in the same location may require timely invasive investigation and, if necessary, resection.
- The even rarer special form of a histopathological mixed picture of subependymoma and ependymal components does not differ with regard to the image-based morphological criteria.
- Since WHO II tumor components with a somewhat stronger growth tendency can be present, imaging follow-up monitoring is in every case indispensable.

Conflict of Interest

The authors declare that they have no conflict of interest.

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