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Case Report

Fulminant presentation of a SMARCB1-deficient, anterior cranial fossa tumor in adult

Idan Levitan[1], Suzana Fichman[2], Yosef Laviv[1]

Departments of [1]Neurosurgery, [2]Pathology, Rabin Medical Center, Petha Tiqva, Israel.

E-mail: Idan Levitan - idanlevitan@gmail.com; Suzana Fichman - suzanaf@clalit.org.il; *Yosef Laviv - yossilaviv@gmail.com

***Corresponding author:**
Yosef Laviv,
Department of Neurosurgery,
Rabin Medical Center, Petah Tiqva, Israel.

yossilaviv@gmail.com

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ABSTRACT

Background: Malignant atypical teratoid rhabdoid tumor (ATRT) usually develops in children. ATRTs are rare in adults, with only one case in the literature describing involvement of the anterior skull base. These primary intracranial tumors are characterized molecularly as SMARCB1 (INI1) deficient. Different types of such SMARCB1-deficient tumors exist in adulthood, usually in the form of extracranial tumors. Very few cases of such a new entity, named SMARCB1-deficient sinonasal carcinoma have been described with intracranial penetration and involvement of the anterior cranial fossa.

Case Description: A 36-year-old male presented with acute cognitive deterioration. Over few hours, he developed a fulminant herniation syndrome. Imaging showed a tumor in the anterior cranial fossa surrounded by massive brain edema. The tumor has destroyed the frontal bone with involvement of the nasal cavities and paranasal sinuses. The patient underwent emergent decompressive craniectomy and tumor debulking but could not be saved. Pathological analysis revealed a highly cellular tumor without rhabdoid cells but with areas of necrosis. Further immunohistochemical stains revealed that neoplastic cells were diffusely and strongly positive for epithelial membrane antigen and P63 and negative for SMARCB1 (i.e., loss of expression), confirming the diagnosis of sinonasal carcinoma.

Conclusion: To the best of our knowledge, this is the first report of a fulminant presentation of a SMARCB1-deficient tumor in young adult, involving the anterior cranial fossa and the paranasal sinuses. The main differential diagnosis of aggressive, primary, intracranial SMARCB1-deficient tumors in adults includes ATRT, SMARCB1-deficient sinonasal carcinoma, rhabdoid meningioma, and rhabdoid glioblastoma. Atypical tumors involving the anterior skull base without a clear histopathological pattern should therefore be checked for SMARCB1 expression.

Keywords: Anterior cranial fossa, INI1, Rhabdoid, Sinonasal carcinoma, SMARCB1

BACKGROUND AND SIGNIFICANCE

A number of tumor suppressor genes are important in regulating transcription in eukaryotic neoplastic tissue. SMARCB1 is such a gene which has been named by acronyms for its function: switch/sucrose nonfermentable (SWI/SNF) related, matrix-associated, actin-dependent regulator of chromatin, subfamily B, member 1. It is a tumor suppressor gene that encodes a core subunit of the SWI/SNF chromatin-remodeling complex positively regulating transcription of a particular set of genes involved in differentiation, tumorigenesis, invasion, and apoptosis.[22,33] The gene is located at chromosome 22q11.23 and is also known as HIV integrase interactor1 or INI 1.

SMARCB1 is ubiquitously expressed in the nuclei of all nonneoplastic cells and can be readily identified using immunohistochemistry.[15] Biallelic inactivation of SMARCB1 was originally described in atypical teratoid rhabdoid tumors (ATRTs).[29] However, this gene is not limited to ATRT and can be found in a variety of other tumors, including extracranial soft-tissue tumors (e.g., extrarenal malignant rhabdoid tumor, epithelioid sarcoma, some extraskeletal myxoid chondrosarcomas, some epithelioid malignant peripheral intracranial nerve sheath tumors, and some myoepithelial tumors) as well as few cranial tumors.[15]

The spectrum of non-ATRT, cranial SMARCB1-deficient tumors mostly includes other pediatric tumors, such as the benign cribriform neuroepithelial tumor and rare poorly differentiated chordomas.[9] In adults, they also include the recently described SMARCB1-deficient sinonasal carcinoma[3] and other meningeal SWI/SNF-related, SMARCB1-deficient tumors.[11] The rarity of these pathologies in adults poses significant diagnostic difficulties, which require meticulous clinical, radiological, and above all, pathological evaluation to arrive at the correct diagnosis.

We present an extremely rare case in which a SMARCB1-deficient tumor, with both extracranial and intracranial components, presented with a fulminant clinical course of cognitive deterioration and subsequent loss of consciousness, resulting in death in a young adult. The differential diagnosis of primary, cranial, and SMARCB1-deficient tumors in adults is discussed.

CASE REPORT

A 36-year-old otherwise healthy male presented to an outside hospital with blurred vision for few days and acute onset of aggressive behavior and agitation. While in the emergency department, he deteriorated rapidly to a Glasgow Coma Scale of 8 requiring intubation. A head computed tomography (CT) revealed a large, bifrontal extra-axial mass of the anterior skull base, measuring 52 mm × 37 mm. The lesion also involved the ethmoid cells and frontal sinuses as well as the right orbit. It was infiltrating the dura. Extended peritumoral brain edema with significant mass effect was also noted [Figure 1]. The patient was emergently transferred to our hospital. On arrival, he had fixed mid-dilated pupils, with minimal flexion movement to pain and intact corneal and gag reflexes. Laboratory tests were within normal limits. He was treated with intravenous push of high-dose dexamethasone (20 mg) and mannitol (70 g) and was immediately transferred to the operating room for a bifrontal decompressive craniectomy and tumor debulking.

At surgery, the tumor was found to be very vascular. On opening the dura, the brain was swollen and of firm consistency with no pulsations. A debulking operation was

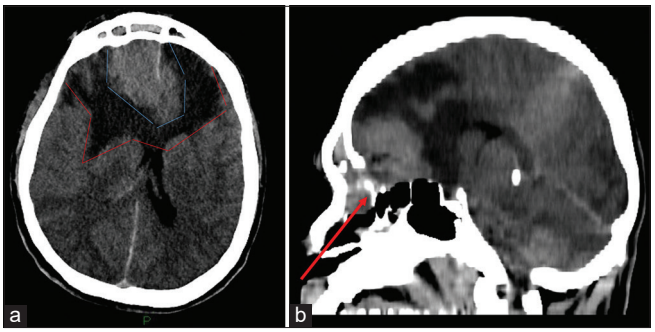

Figure 1: (a) Noncontrast head CT (axial cut) at time of presentation, showing large bifrontal mass (light blue lines) surrounded by brain edema (red lines), causing significant mass effect. (b) Sagittal cut. The tumor has destroyed the frontal bone, extending into the paranasal sinuses (arrow) and intracranially, involving the anterior skull base.

performed along with bifrontal decompressive craniectomy. An intracranial pressure (ICP) monitor was inserted to the parenchyma. The ICP levels were <25 mmHg on average during the postoperative course, the patient was in Glasgow Come Scale 3, with no brain stem reflexes. Postoperative head CT demonstrated persistent, bilateral, massive edema, the patient's condition remained critical and he passed away 1 week later.

Histological examination for hematoxylin and eosin stain showed a tumor comprised small to medium size cells, some of them with clear cytoplasm. Rhabdoid cells were not noted. The tumor shows areas of necrosis and high levels of mitotic and apoptotic activity [Figure 2a and b]. The stain for INI1 was negative (loss of expression) [Figure 2c]. Immunohistochemical stains were diffusely and strong positive for epithelial membrane antigen (EMA) [Figure 2d] and P63, partially positive for smooth muscle antigen (SMA) [Figure 2e], glial fibrillary acidic protein (GFAP) [Figure 2f], B-cell lymphoma 2, and vimentin (VIM), focally positive for pan-KER and KER 8, and negative for OLIG2, S100, desmin, synaptophysin, NEU-N, and chromogranin. The proliferative index Ki-67 was 25–30%. The initial diagnosis was ATRT. Later, the possibility of SMARCB1-deficient sinonasal carcinoma was added as a differential diagnosis.

DISCUSSION

ATRTs are primary rhabdoid tumors of the central nervous system (CNS). They are rare malignant brain tumors usually diagnosed in children younger than 3 years old[26] and are now classified as embryonal tumors Grade IV by the World Health Organization.[20] The presence of ATRT in adults is exceedingly rare. While ATRT is the most common malignant CNS tumor in children aged <1, the lifetime risk in adults (i.e., age >18) is estimated as <1/1,000,000.[12] ATRT in adults has a predilection for midline structures,

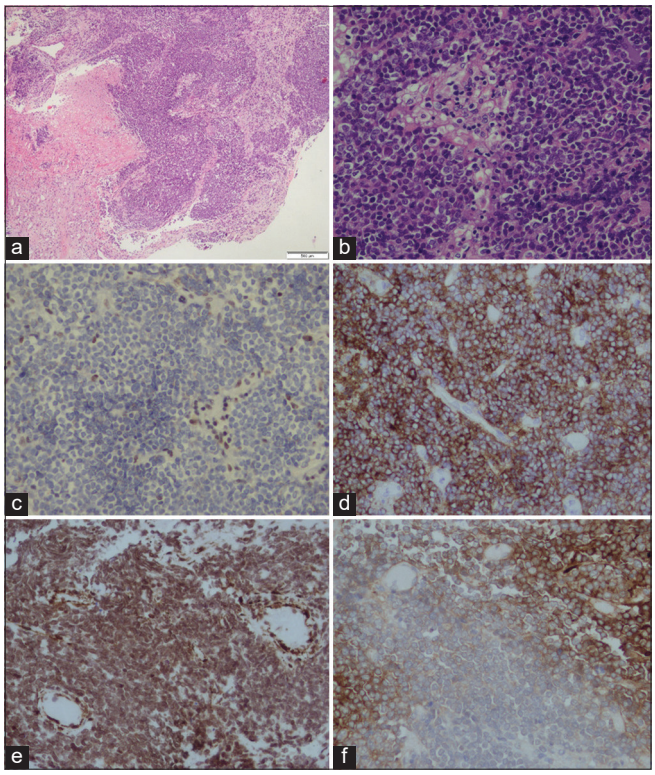

Figure 2: (a) Histological examination for hematoxylin and eosin (H&E) stain showing a tumor comprised small to medium size cells, some of them with clear cytoplasm. Rhabdoid cells were not noted. The tumor shows areas of necrosis. (b) H&E, ×20. (c) The stain for INI1 was negative (loss of expression). Immunohistochemical stains were diffusely and strong positive for EMA (d) and partially positive for SMA (e) and GFAP (f).

particularly the pineal and pituitary glands.[24] Still, they most frequently occur in the cerebral hemisphere and typically appear involving continuity the lateral ventricles.[12]

Immunohistochemical study using antibody against the INI1 gene product or fluorescence *in situ* hybridization to identify loss of the INI1 locus is the current routine workup for diagnostic confirmation of ATRT.[27] In 2016, ATRTs have been molecularly defined by the inactivation of either the INI1/SMARCB1 or BRG1/SMARCA4 genes; however, most cases harbor the former alterations.[17] Genetically, a different mutation in each allele (a compound heterozygous mutation) is extremely rare in children (<1%) but very common in sellar ATRTs in adults.[23] On the other hand, homozygous deletions occur in 20–25% of pediatric cases, but were only reported in 1/17 (6%) adult cases.[16]

ATRTs can exhibit epithelial, primitive neuroepithelial, and mesenchymal differentiation. Histologically, the mesenchymal component of ATRTs is characterized by cells with discrete borders and a rhabdoid morphology, that is, abundant cytoplasm with eosinophilic paranuclear inclusions of intermediate filaments. These filaments are identified as vimentin by immunohistochemistry.[21] In addition to vimentin, the rhabdoid cells usually express EMA. The neuroepithelial component is primitive, consisting of sheets of small, poorly differentiated cells.[25] Dardis *et al*. have summarized the immunostaining patterns of all published adult ATRT cases (n = 35).[12] Of the "classical" features (vimentin, EMA, and SMA), only vimentin was universally positive (33/33). EMA and SMA were positive in 83% and 56% of cases, respectively. Neuronal markers were positive in 33–67% of cases, depending on the marker (neurofilament protein, NFP, the most common). GFAP and synaptophysin were positive in 40% and 27% of cases, respectively. Keratins were variably expressed (40% of cases), with keratin 8 being the most common (75%). Desmin immunopositivity was not observed.

In adult patients, it is very difficult to render a diagnosis of ATRT for CNS malignant tumors, even when a predominant rhabdoid cell component is present, because there are more common malignant tumors (primary and metastatic) that show rhabdoid features, such as rhabdoid glioblastoma, rhabdoid meningioma, metastatic melanoma, and metastatic carcinomas with rhabdoid features, all occurring in this age group.[28] In addition, in some sellar ATRT, only scattered rhabdoid cells were found, making the diagnosis even more complicated.[6,23] Of note, our current case had no rhabdoid cells at all.

Tumors resembling ATRT, staining with GFAP, as well as vimentin, SMA, and EMA have been suggested to represent rhabdoid glioblastoma. Rhabdoid glioblastoma (GBM) is an aggressive variant of glioblastoma, which mainly affects young subjects. It can involve the leptomeninges,[10] and although an extracranial metastasis to the scalp and lungs was reported,[4] no bone invasion or sinonasal spreading has been described. This is one of the reasons why this diagnosis was not considered for our case. Rhabdoid GBM shows diffuse staining for EMA and vimentin and focal expression of cytokeratin and GFAP.[10] Distinction from ATRT is based on histopathological and immunohistochemical features. In addition, it retains INI1 expression[10] or shows only focal loss of INI1, limited to the rhabdoid component.[19]

Bone involvement of the skull in ATRT patients is extremely rare, especially in adults. Although hematogenous tumor spread to the skeleton is a rare, it has been a well-known finding in medulloblastomas, though few reports on destruction or invasion of the adjacent skull in medulloblastomas or other CNS primitive neuroectodermal tumors exist.[31] In a series of 91 pediatric ATRT cases, the frequency of skull involvement was 6.6% (2 calvaria, 2 cerebellopontine angle, and 1 clivus). In a review of 54 adult ATRT cases,[34] only 1 case of skull involvement was found: parietal calvaria (24-year-old male).[14] Two more cases involved the jugular foramen[18] and

Table 1: SMARCB1-negative tumors involving the anterior skull base in adults; literature's review.

Ref	Age/ sex	Presentation	Location	H&E	Immunohistochemical staining											K167 (%)	D/T	Prognosis
					EMA	VIM	SMA	GFAP	KER 8	P63	NSE	PAN KER	KER 5/6	DES	INI1			
Barresi *et al.*, 2016[5]	16/F	Epistaxis, HA	Anterior skull base, sinuses, nasal and orbital cavities	Small-medium cells, clear or rhabdoid cytoplasm, eccentric nucleoli	+++	+++	+	-	NA	NA	++	++	-	+	Missense mutation 574G>A in exon 5 of SMARCB1 gene	60	ATRT/ surgery	6 mo; bone and lung Mets
Agaimy *et al.*, 2017[2]	38/F	NA	Sinuses, frontal skull base	Plasmacytoid/rhabdoid cells	NA	NA	NA	NA	NA	++	NA	++	+	NA	Monoallelic deletion	NA	SMARCB1-def SN Ca/ Biopsy + Cx	Alive, 4 mo; bone Mets
	79/M	NA	Nasal cavity, anterior cranial fossa	Plasmacytoid/rhabdoid cells	NA	NA	NA	NA	NA	+	NA	++	NA	NA	-	NA	SMARCB1-def SN/NA	NA
Bell *et al.*, 2015[7]	33/F	Epistaxis, HA	Anterior skull base eroding through the cribriform plate	Microreticulated and basaloid papillary patterns. Some rhabdoid cells.	NA	NA	NA	NA	NA	+	NA	++	NA	NA	No mutation was detected	NA	SMARCB1-def SN/ Surgery + CxRx	Alive, 33 mo
Wasserman *et al.*, 2017[32]	56/F	Anosmia, epistaxis, diplopia, facial paresthesia	Right sinonasal mass with intracranial extension and cavernous sinus invasion	Prominent rhabdoid morphology, empty vacuoles were frequent	NA	NA	NA	NA	NA	-	NA	NA	-	NA	-	NA	SMARCB1-def SN/CxRx	Remission, 12 mo
Current case	36/M	Blurred vision, rapid LOC	Bilateral anterior skull base, frontal and ethmoidal sinuses	Small to medium size cells, some with clear cytoplasm. Basaloid pattern. Rhabdoid cells were not noted. Areas of necrosis and high levels of mitotic and apoptotic activity	+++	++	++	++	+	++	±	+	-	-	-	25–30	SMARCB1-def SN/ emergent DC and tumor debulking	Dead, 1 w

ATRT: Atypical teratoid rhabdoid tumor, Ca: Carcinoma, Cx: Chemotherapy, D: Diagnosis, DC: Decompressive craniectomy, DES: Desmin, EMA: Epithelial membrane antigen, F: Female, GFAP: Glial fibrillary acidic protein, H&E: Hematoxylin and eosin, HA: Headaches, INI1: HIV-integrase interactor1, KER: Keratin, LOC: Loss of consciousness, M: Male, Mets: Metastasis, Mo: Months, NA: Nonavailable, NSE: Neuron-specific enolase, Ref: References, Rx: Radiation therapy, SMA: Smooth muscle antigen, SN: Sinonasal; T: Treatment, VIM: Vimentin, W: Week

the internal auditory canal[30] but cannot be regarded as true skull penetration or invasiveness.

In 2016, the first and only report of adult ATRT involving the nasal cavities and anterior skull base was published.[5] Interestingly, this report was later considered by another group as SMARCB1-deficient sinonasal carcinomas.[2]

Sinonasal tract malignancies are uncommon, representing no more than 5% of all head-and-neck cancers.[13] Poorly differentiated sinonasal carcinomas are a heterogeneous group of aggressive neoplasms that encompasses squamous cell carcinoma including basaloid variant, lymphoepithelial carcinoma, sinonasal undifferentiated carcinoma (SNUC), neuroendocrine-type small cell carcinoma, teratocarcinosarcomas, poorly differentiated keratinizing and nonkeratinizing variants of squamous cell carcinoma, and nuclear protein of testis (NUT) midline carcinomas.[1] In 2014, Agaimy *et al*.[3] and Bishop *et al*.[8] independently described a variant of SNUC characterized by loss of nuclear SMARCB1 expression. SMARCB1-deficient sinonasal carcinomas represent 3.3% out of a combined series of 484 sinonasal primary tumors.[7] Most SMARCB1-deficient sinonasal carcinomas are staged as T4 at the time of diagnosis.[32] In 2017, the group of Agaimy *et al*. published the most extensive data on this entity so far, including 39 cases (23 M, 16 F, median age: 52 years old).[2] Histologically, in all 39 cases, mitotic rates were uniformly high and necrosis was common. In many cases, nonspecific, clear, "empty" cytoplasmic vacuoles were seen. Most tumors displayed either a predominantly basaloid ("small, round, blue cells," 61%) or plasmacytoid/rhabdoid morphology ("pink cells," 36%). The plasmacytoid/rhabdoid form consisted of sheets of tumor cells with abundant, eccentrically placed eosinophilic cytoplasm. Despite the aggressive nature of the tumor, it is characterized by minimal pleomorphism.[32] By immunohistochemistry, the tumors were positive for pancytokeratin (97%), CK5 (64%), p63 (55%), and CK7 (48%). Imaging revealed extensive involvement of the paranasal sinuses with or without involvement of the nasal cavity and frequent involvement of the skull base. Of the 39 cases, two were described with extension into the anterior cranial fossa [Table 1] and one with a calvarial extension. All three cases were of the plasmacytoid/rhabdoid type. None of them had a fulminant, aggressive presentation.

In a literature's review of other cases of SMARCB1-deficient sinonasal carcinoma, three more cases involving the anterior skull base with intracranial penetration were reported[5,7,32] [Table 1]. All three cases presented with epistaxis. In the case by Barresi *et al*., a 16-year-old female patient presented with anosmia, epistaxis, and headaches. On imaging, a large extra-axial mass was found, penetrating the anterior skull base and infiltrating the dura mater, ethmoid cribriform plate, as well as nasal and orbital cavities. Light microscopy showed a tumor composed of small/medium sized cells with clear or rhabdoid appearance. At immunohistochemistry, the neoplastic cells were diffusely positive for EMA and vimentin and focally positive for cytokeratin AE1/AE3, SMA, desmin, and NFP. No staining for GFAP was found. Ki-67 was 60%. The authors considered this the third extra-axial case of ATRT overall, and the first one infiltrating the nasal tracts. As mentioned, and despite the absence of cytokeratin expression in the majority of the tumor, the group by Agaimy *et al*. has later considered this case as a SMARCB1-deficient sinonasal carcinoma in their review.[2] According to this group, in the setting of a sinonasal tumor that morphologically resembles SMARCB1-deficient sinonasal carcinoma, SMARCB1 immunohistochemistry should be considered even in the absence of cytokeratin expression. In addition, the mere presence of neuroendocrine differentiation by immunohistochemistry, particularly if the expression is focal, does not exclude the diagnosis of SMARCB1-deficient sinonasal carcinoma.[2]

A subset of SMARCB1-deficient sinonasal carcinomas, particularly the basaloid form, demonstrates diffuse p63 immunoreactivity that may result in a misdiagnosis of nonkeratinizing/basaloid SCC or NUT midline carcinoma.[2] Our case also showed diffuse p63 immunoreactivity. [Table 2]

Table 2: Differential diagnosis of aggressive, intracranial SMARCB1-deficient tumors in adults. Immunohistochemical staining.

HP staining/tumor	ATRT	SMARCB1-def SN Ca	Rhabdoid GBM	Rhabdoid meningioma	Metastatic melanoma
INI1	-	-	+	+	+
GFAP	±	−	+	−	−
SMA	±	−	+ (gliosarcoma)	−	−
EMA	±	±	±	+	−
Pan-KER	±	+	−	±	−
VIM	+	−	+	+	+
P63	−	±	−	±	−
DES	±	−	−	±	−
NSE	±	−	−	−	±

ATRT: Atypical teratoid/rhabdoid tumor, Ca: Carcinoma, DEF: Deficient, DES: Desmin, EMA: Epithelial membrane antigen, GBM: Glioblastoma, GFAP: Glial fibrillary acidic protein, HP: Histopathology, INI1: HIV-integrase interactor1, NSE: Neuron-specific enolase, SMA: Smooth muscle antigen, SN: Sinonasal, VIM: Vimentin

summarizes the relevant histopathological staining according to the discussed differential diagnosis of tumors.

It seems that the diagnosis of SMARCB1-deficient tumors is more frequently made as more and more subtypes are being discovered. In 2017, Dadone *et al*. described two unique cases of meningeal-related tumors.[11]

Based on shared phenotype and genotype features, the authors suggested that these cases are part of an emerging group of primary meningeal SMARCB1-deficient tumors, not described to date. Our case was infiltrating the dura, but showed much more aggressive behavior as compared to the cases described by Dadone *et al*. Unfortunately, due to insufficient material saved for pathology and the absence of fresh frozen samples, we could not perform molecular analysis of the tumor which might have helped reaching more definite diagnosis.

CONCLUSION

Given the radiological, morphological, and pathological data, we believe that the current case represents an extremely aggressive behavior of a SMARCB1-deficient sinonasal carcinoma, possibly of the basaloid subtype. Although it is difficult to explain the positive staining for GFAP, all other data support the diagnosis of sinonasal carcinoma over ATRT. Due to the extreme rarity of reported cases, we cannot suggest that this case represents an aggressive variant of other more recently described primary meningeal SMARCB1-deficient tumors. The fulminant presentation can be explained, perhaps, by infiltration and occlusion of the superior sagittal sinus as well as by mass effect and rapid compression and obstruction of venous outflow, resulting in massive bifrontal brain edema. To the best of our knowledge, this aggressive presentation of a SMARCB1-deficient anterior cranial fossa tumor has never been described before.

Declaration of patient consent

Patient's consent not obtained as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Agaimy A. The expanding family of SMARCB1(INI1)-deficient neoplasia: Implications of phenotypic, biological, and molecular heterogeneity. Adv Anat Pathol 2014;21:394-410.
2. Agaimy A, Hartmann A, Antonescu CR, Chiosea SI, El-Mofty SK, Geddert H, *et al*. SMARCB1 (INI-1)-deficient sinonasal carcinoma: A series of 39 cases expanding the morphologic and clinicopathologic spectrum of a recently described entity. Am J Surg Pathol 2017;41:458-71.
3. Agaimy A, Koch M, Lell M, Semrau S, Dudek W, Wachter DL, *et al*. SMARCB1(INI1)-deficient sinonasal basaloid carcinoma: A novel member of the expanding family of SMARCB1-deficient neoplasms. Am J Surg Pathol 2014;38:1274-81.
4. Babu R, Hatef J, McLendon RE, Cummings TJ, Sampson JH, Friedman AH, *et al*. Clinicopathological characteristics and treatment of rhabdoid glioblastoma. J Neurosurg 2013;119:412-9.
5. Barresi V, Branca G, Raso A, Mascelli S, Caffo M, Tuccari G. Atypical teratoid rhabdoid tumor involving the nasal cavities and anterior skull base. Neuropathology 2016;36:283-9.
6. Barresi V, Lionti S, Raso A, Esposito F, Cannavo S, Angileri FF. Pituitary atypical teratoid rhabdoid tumor in a patient with prolactinoma: A unique description. Neuropathology 2017;6:12440.
7. Bell D, Hanna EY, Agaimy A, Weissferdt A. Reappraisal of sinonasal undifferentiated carcinoma: SMARCB1 (INI1)-deficient sinonasal carcinoma: A single-institution experience. Virchows Arch 2015;467:649-56.
8. Bishop JA, Antonescu CR, Westra WH. SMARCB1 (INI-1)-deficient carcinomas of the sinonasal tract. Am J Surg Pathol 2014;38:1282-9.
9. Bodi I, Giamouriadis A, Sibtain N, Laxton R, King A, Vergani F. Primary intracerebral INI1-deficient rhabdoid tumor with CD34 immunopositivity in a young adult. Surg Neurol Int 2018;9:45.
10. Byeon SJ, Cho HJ, Baek HW, Park CK, Choi SH, Kim SH, *et al*. Rhabdoid glioblastoma is distinguishable from classical glioblastoma by cytogenetics and molecular genetics. Hum Pathol 2014;45:611-20.
11. Dadone B, Fontaine D, Mondot L, Cristofari G, Jouvet A, Godfraind C, *et al*. Meningeal SWI/SNF related, matrix-associated, actin-dependent regulator of chromatin, subfamily B member 1 (SMARCB1)-deficient tumours: An emerging group of meningeal tumours. Neuropathol Appl Neurobiol 2017;43:433-49.
12. Dardis C, Yeo J, Milton K, Ashby LS, Smith KA, Mehta S, *et al*. Atypical teratoid rhabdoid tumor: Two case reports and an analysis of adult cases with implications for pathophysiology and treatment. Front Neurol 2017;8:247.
13. Haerle SK, Gullane PJ, Witterick IJ, Zweifel C, Gentili F. Sinonasal carcinomas: Epidemiology, pathology, and management. Neurosurg Clin N Am 2013;24:39-49.
14. Han L, Qiu Y, Xie C, Zhang J, Lv X, Xiong W, *et al*. Atypical teratoid/rhabdoid tumors in adult patients: CT and MR imaging features. AJNR Am J Neuroradiol 2011;32:103-8.
15. Hollmann TJ, Hornick JL. INI1-deficient tumors: Diagnostic features and molecular genetics. Am J Surg Pathol 2011;35:e47-63.
16. Horiguchi H, Nakata S, Nobusawa S, Uyama S, Miyamoto T, Ueta H, *et al*. Adult-onset atypical teratoid/rhabdoid tumor featuring long spindle cells with nuclear palisading and perivascular pseudorosettes. Neuropathology 2017;37:52-7.
17. Judkins AR, Eberhart EC, Wesseling P. Atypical teratoid/rhabdoid tumour. In: Louis DN, Ohgaki OH, Wiestler OD,

Cavenee WK, editors. WHO Classification of Tumours of the Central Nervous System Revised. 4th ed. Lyon: IARC Press; 2016. p. 209-12.
18. Kanoto M, Toyoguchi Y, Hosoya T, Kuchiki M, Sugai Y. Radiological image features of the atypical teratoid/rhabdoid tumor in adults: A systematic review. Clin Neuroradiol 2015;25:55-60.
19. Kleinschmidt-DeMasters BK, Alassiri AH, Birks DK, Newell KL, Moore W, Lillehei KO. Epithelioid versus rhabdoid glioblastomas are distinguished by monosomy 22 and immunohistochemical expression of INI-1 but not claudin 6. Am J Surg Pathol 2010;34:341-54.
20. Louis DN, Perry A, Reifenberger G, von Deimling A, Figarella-Branger D, Cavenee WK, *et al*. The 2016 World Health Organization classification of tumors of the central nervous system: A summary. Acta Neuropathol 2016;131:803-20.
21. Makuria AT, Rushing EJ, McGrail KM, Hartmann DP, Azumi N, Ozdemirli M. Atypical teratoid rhabdoid tumor (AT/RT) in adults: Review of four cases. J Neurooncol 2008;88:321-30.
22. Medjkane S, Novikov E, Versteege I, Delattre O. The tumor suppressor hSNF5/INI1 modulates cell growth and actin cytoskeleton organization. Cancer Res 2004;64:3406-13.
23. Nakata S, Nobusawa S, Hirose T, Ito S, Inoshita N, Ichi S, *et al*. Sellar atypical teratoid/rhabdoid tumor (AT/RT): A clinicopathologically and genetically distinct variant of AT/RT. Am J Surg Pathol 2017;41:932-40.
24. Nishikawa A, Ogiwara T, Nagm A, Sano K, Okada M, Chiba A, *et al*. Atypical teratoid/rhabdoid tumor of the sellar region in adult women: Is it a sex-related disease? J Clin Neurosci 2018;49:16-21.
25. Raisanen J, Hatanpaa KJ, Mickey BE, White CL 3rd. Atypical teratoid/rhabdoid tumor: Cytology and differential diagnosis in adults. Diagn Cytopathol 2004;31:60-3.
26. Rorke LB, Packer RJ, Biegel JA. Central nervous system atypical teratoid/rhabdoid tumors of infancy and childhood: Definition of an entity. J Neurosurg 1996;85:56-65.
27. Shitara S, Akiyama Y. Atypical teratoid/rhabdoid tumor in sellar turcica in an adult: A case report and review of the literature. Surg Neurol Int 2014;5:2152-7806.
28. Takei H, Adesina AM, Mehta V, Powell SZ, Langford LA. Atypical teratoid/rhabdoid tumor of the pineal region in an adult. J Neurosurg 2010;113:374-9.
29. Versteege I, Sevenet N, Lange J, Rousseau-Merck MF, Ambros P, Handgretinger R, *et al*. Truncating mutations of hSNF5/INI1 in aggressive paediatric cancer. Nature 1998;394:203-6.
30. Wang X, Liu X, Lin Z, Chen Y, Wang P, Zhang S. Atypical teratoid/rhabdoid tumor (AT/RT) arising from the acoustic nerve in a young adult: A case report and a review of literature. Medicine (Baltimore) 2015;94:e439.
31. Warmuth-Metz M, Bison B, Gerber NU, Pietsch T, Hasselblatt M, Fruhwald MC. Bone involvement in atypical teratoid/rhabdoid tumors of the CNS. AJNR Am J Neuroradiol 2013;34:2039-42.
32. Wasserman JK, Dickson BC, Perez-Ordonez B, de Almeida JR, Irish JC, Weinreb I. INI1 (SMARCB1)-deficient sinonasal carcinoma: A clinicopathologic report of 2 cases. Head Neck Pathol 2017;11:256-61.
33. Wilson BG, Roberts CW. SWI/SNF nucleosome remodellers and cancer. Nat Rev Cancer 2011;11:481-92.
34. Wu WW, Bi WL, Kang YJ, Ramkissoon SH, Prasad S, Shih HA, *et al*. Adult atypical teratoid/rhabdoid tumors. World Neurosurg 2016;85:197-204.

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