Case Report

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

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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. 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. The gene is located at chromosome 22q11.23 and is also known as HIV integrase interactor 1 or INI 1.
SMARCB1 is ubiquitously expressed in the nuclei of all nonneoplastic cells and can be readily identified using immunohistochemistry.\cite{15} Biallelic inactivation of SMARCB1 was originally described in atypical teratoid rhabdoid tumors (ATRTs).\cite{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.\cite{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.\cite{9} In adults, they also include the recently described SMARCB1-deficient sinonasal carcinoma\cite{9} and other meningeal SWI/SNF-related, SMARCB1-deficient tumors.\cite{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 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 Coma 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\cite{26} and are now classified as embryonal tumors Grade IV by the World Health Organization.\cite{26} 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.\cite{12} ATRT in adults has a predilection for midline structures,
particularly the pineal and pituitary glands. Still, they most frequently occur in the cerebral hemisphere and typically appear involving continuity the lateral ventricles.

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. 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. 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. On the other hand, homozygous deletions occur in 20–25% of pediatric cases, but were only reported in 1/17 (6%) adult cases.

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. In addition to vimentin, the rhabdoid cells usually express EMA. The neuroepithelial component is primitive, consisting of sheets of small, poorly differentiated cells. Dardis et al. have summarized the immunostaining patterns of all published adult ATRT cases (n = 35). 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. In addition, in some sellar ATRT, only scattered rhabdoid cells were found, making the diagnosis even more complicated. 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 and although an extracranial metastasis to the scalp and lungs was reported, 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. Distinction from ATRT is based on histopathological and immunohistochemical features. In addition, it retains INI1 expression or shows only focal loss of INI1, limited to the rhabdoid component.

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. 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, only 1 case of skull involvement was found: parietal calvaria (24-year-old male). Two more cases involved the jugular foramen.
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          |
|----------------------|---------|---------------------------------------|-------------------------------------------------|------------------------------------------|-----------------------------|-----------|----------------|--------------------|
| Barresi et al., 2016 | 16/F    | Epistaxis, HA                         | Anterior skull base, sinus, 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  | 38/F    | NA                                    | Sinuses, frontal skull base                      | Plasmacytoid/rhabdoid cells             | NA NA NA NA NA ++ NA ++ + NA | NA        | SMARCB1-def SN | Alive, 4 mo | bone Mets |
| Bell et al., 2015    | 33/F    | Epistaxis, HA                         | Anterior skull base eroding through the cribriform plate | Plasmacytoid/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 | 56/F   | Anosmia, epistaxis, diplopia, facial pain | 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 - | No mutation was detected | 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: Headache, INI1: HIV-integrase interactor1, KER: Keratin, LOC: Loss of consciousness, M: Male, 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 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.[2] Interestingly, this report was later considered by another group as SMARCB1-deficient sinonasal carcinomas.[1]

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.[1] and Bishop et al.[14] 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.[1] Most SMARCB1-deficient sinonasal carcinomas are staged as T4 at the time of diagnosis.[12] 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).[3] 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.[12] 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 cribiform 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]

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**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.\textsuperscript{14}

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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Conflicts of interest

There are no conflicts of interest.

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