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Imaging Features of Primary Intracranial Sarcoma with *DICER1* Mutation: A Multicenter Case Series

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ABSTRACT

SUMMARY: Primary intracranial sarcoma, *DICER1*-mutant, is a rare, recently described entity in the fifth edition of the WHO Classification of CNS Tumors. Given the entity's rarity and recent description, imaging data on primary intracranial sarcoma, *DICER1*-mutant, remains scarce. In this multicenter case series, we present detailed multimodality imaging features of primary intracranial sarcoma, *DICER1*-mutant, with emphasis on the appearance of the entity on MR imaging. In total, 8 patients were included. In all 8 patients, the lesion demonstrated blood products on T1WI. In 7 patients, susceptibility-weighted imagining was obtained and demonstrated blood products. Primary intracranial sarcoma, *DICER1*-mutant, is a CNS neoplasm that primarily affects pediatric and young adult patients. In the present case series, we explore potential imaging findings that are helpful in suggesting this diagnosis. In younger patients, the presence of a cortical lesion with intralesional blood products on SWI and T1-weighted MR imaging, with or without extra-axial blood products, should prompt the inclusion of this entity in the differential diagnosis.

ABBREVIATIONS: ASL = arterial spin-labeling; DCE = dynamic contrast enhancement; SDH = subdural hematoma

The WHO Classification of CNS Tumors continues to evolve with the ever-expanding discovery of tumor molecular markers that have a direct impact on our understanding of tumor biology, precise classification, treatment options, prognosis, and outcome. The fifth edition of the WHO Classification of CNS Tumors, published in 2021, reflects new advances in the molecular diagnosis of CNS neoplasms and further builds on the updates of the revised fourth edition, published in 2016, and the recommendations of the Consortium to Inform Molecular and Practical Approaches to CNS Tumor Taxonomy (cIMPACT-NOW).[1-9]

The fifth edition of the WHO Classification introduces 22 new tumor types.[1] One of these entities, under the umbrella of mesenchymal nonmeningothelial tumors, is primary intracranial sarcoma, *DICER1*-mutant,[1] which is a rare highly malignant entity that can be associated with familial *DICER1* syndrome and neurofibromatosis type 1.[10-12] Given the entity's rarity and recent description, imaging data on primary intracranial sarcoma, *DICER1*-mutant, remain scarce. The few published case reports and series describing the entity have alluded to its imaging features; however, detailed imaging descriptions that include larger patient series remain lacking, and the emphasis of the literature has been on molecular diagnosis and treatment options.[12-16]

In this multicenter case series, we aim to present detailed multimodality imaging features, including both anatomic and advanced imaging characteristics of primary intracranial sarcoma, *DICER1*-mutant, with emphasis on the appearance of the entity on MR imaging with an attempt to elucidate imaging characteristics that can help in suggesting the diagnosis.

CASE SERIES

Methods

This retrospective study was approved by the Institutional Review Board at The University of Texas MD Anderson Cancer Center as a collaborative, multi-institutional study.

Patient Selection and Clinical Data. From January 2015 to October 2023, a total of 8 consecutive patients with a diagnosis of primary intracranial sarcoma with *DICER1* mutation were identified (Table 1), as follows: 3 patients diagnosed at the MD Anderson Cancer Center, 3 patients diagnosed at the Texas Children's Hospital, and 2 patients diagnosed at the Washington University School of Medicine in St Louis. Assessments of the pathology, clinical, and imaging data were then performed to assess

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Table 1: Demographic characteristics of patients with primary intracranial sarcoma, *DICER1*-mutant, included in this case series

Demographic Characteristic (Number Assessed)	Finding
Age (8)	Mean: 16 years Standard deviation: 11.57 years Range: 4–33
Presenting symptoms (7)	Headaches: 5 Extremity weakness: 2 Nausea: 2 Numbness: 2 Language deficit: 1 Blurry vision: 1 Unsteady gait: 1
Duration of symptoms (5)	Average: 7 days Range: 1–14 days
DICER1 mutation (8)	Yes
Additional mutations	*ATRX* 5/5 patients *TP53* 7/7 patients
Other *DICER1*-related tumors (7)	None

the following inclusion criteria: all patients have tissue-based diagnosis confirmation at the above study institutions per described guidelines, including DNA methylation profiling and/or sequencing; all patients have clinical and demographic data available for extraction via each institution's electronic medical record system; and all patients have, at a minimum, multisequence pretreatment MR imaging available for assessment.

Clinical and Pathologic Data Review. The following clinical data were collected from each patient's electronic medical record: age, sex, presenting symptoms, duration of symptoms, and, when available, the presence of any hereditary cancer syndrome associated with additional extracranial malignancies, such as neurofibromatosis or *DICER1* syndrome.

The following data were extracted from each patient's Pathology Report: confirmation of *DICER1* mutation and the presence of any other clinically relevant molecular alterations, such as a *TP53* mutation.

Imaging Data Review. All imaging characteristics were collected as a consensus, and scans were assessed by either a neuroradiology fellowship-trained radiologist (R.W.E., with 5 years of neuroradiology experience, reviewed the MD Anderson Cancer Center cases; M.S.P., with 17 years of neuroradiology experience, reviewed the Washington University School of Medicine in St Louis cases) or a pediatric neuroradiology fellowship-trained radiologist (T.A.G.M.H., with 32 years of neuroradiology experience, reviewed the Texas Children's Hospital cases).

Patient imaging was reviewed on the participating institutions' PACS. If a CT was available, the following features were assessed: presence of blood products within the tumor and presence of extra-axial blood products, including subdural hematoma (SDH) and SAH. The patients' MR imaging was assessed for the following features: size of tumor in 3 dimensions; location of the tumor, including side and distribution as well as deep versus superficial; appearance on blood-sensitive sequences (T2* or SWI); appearance on DWI; presence of tumoral blood products on T1WI; and presence of SDH or SAH. In addition, the tumor was characterized based on its macroscopic imaging morphology as either solid/mostly solid (if more than 90% solid), cystic (if more than 90% cystic), or mixed (if in between a solid/cystic mixture). Tumor enhancement was recorded as present or absent and then further assessed, if present, for pattern type (solid homogeneous, peripheral, patchy/ill-defined, or mixed) and extent (>75%, <25%, or in between). If advanced imaging was performed, an analysis of PWI and ^{1}H-MR spectroscopy was conducted, and the results were recorded. The type of perfusion technique was noted as DSC, dynamic contrast enhancement (DCE), and/or arterial spin-labeling (ASL), and perfusion maps were generated, thereby allowing for the determination of whether perfusion was altered relative to a normal-appearing brain (eg, elevated). If ^{1}H-MR spectroscopy was performed, a note was made of the type (single versus multi voxel tissue sampling), and a peak analysis was performed, including an assessment of the absolute peak values and ratios. Finally, a general pattern of imaging was extracted and summarized for an assessment of potentially helpful imaging markers in suggesting the diagnosis.

Results

Patient Characteristics. Table 1 summarizes the demographics and clinical characteristics. The average age at diagnosis was 16 years (range, 4–33 years). Five patients were 10 years or younger at diagnosis. Seven patients had presenting symptoms available for assessment, with headaches being the most common presenting symptom. In 5 patients, the duration of symptoms was known. Two patients had a subacute duration of symptoms for 2 weeks. Three patients presented acutely with durations of symptoms ranging from 1 day (2 patients) to 5 days (1 patient).

Pathology Characteristics. All 8 patients had a surgical resection or biopsy with the sequencing of the sampled tissue demonstrating a *DICER1* mutation. Additional commonly identified mutated genes included *ATRX* and *TP53* (Table 1).

Imaging Characteristics. Table 2 summarizes the imaging characteristics.

Seven patients had cortical/subcortical lesions, and 1 patient had a hypothalamic mass. Four patients had a right hemisphere cortical lesion. The parietal lobe was the most involved lobe, with 4 patients. One patient had multifocal disease isolated to the right frontal lobe. The average anteroposterior dimension of the lesions was 4.25 cm (SD, 2.76). The average transverse dimension was 3.79 cm (SD, 2.08). The average craniocaudal dimension was 4.37 cm (SD, 2.51). Five lesions were solid or mostly solid (>90% solid), and 3 lesions were mixed solid and cystic.

NCCT Imaging Findings. Three patients presented acutely and had NCCT that demonstrated hyperattenuated blood products within the lesion, which is suggestive of an acute/recent bleeding episode (Fig 1). One patient had an associated hyperattenuated subdural hematoma, and another patient had a hyperattenuated subarachnoid hemorrhage. The third patient had subarachnoid blood products diagnosed on MR imaging.

Table 2: MR imaging characteristics of patients with primary intracranial sarcoma, *DICER1*-mutant

MR Imaging Characteristic	Finding
Size (cm)	Anteroposterior dimension, 4.25 cm (SD, 2.76) Transverse dimension, 3.79 cm (SD, 2.08) Craniocaudal dimension, 4.37 cm (SD, 2.5)
Location	
Side	4 right, 3 left
Cortical/deep	7 cortical/subcortical, 1 hypothalamic
Cortical lobe	4 parietal, 2 frontal, 1 temporal/insula
Hemorrhage	
Tumor	7/7 hemorrhage on SWI 8/8 hemorrhage of T1WI
Extra-axial	2 SDH 2 SAH
Diffusion restriction	3 patients
Enhancement	
Presence	8/8 present
Pattern	5 solid homogeneous, 2 heterogeneous, 1 peripheral
Extent	5 > 75% enhancement; 2, 25%–75%; 1 < 25%

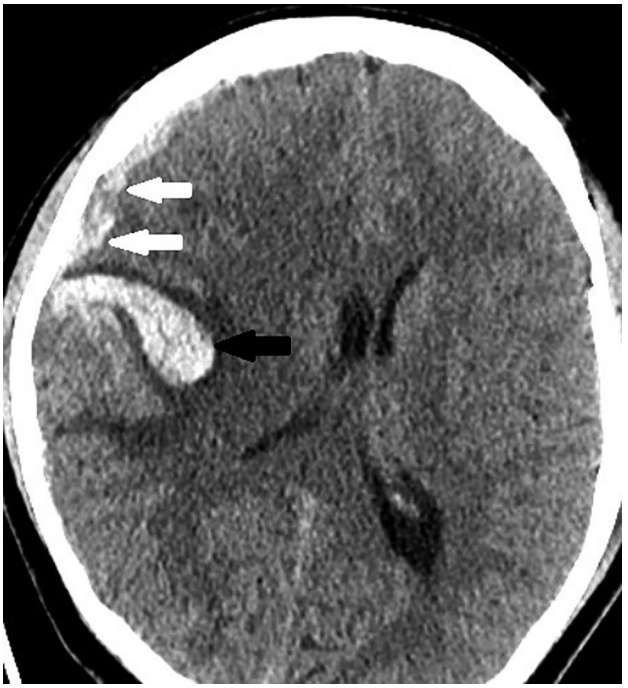

FIG 1. A 28-year-old patient presenting acutely with headaches over a 1-day period. Axial NCCT demonstrates acute hemorrhage within the right frontal lobe lesion (*black arrow*) with overlying right frontal convexity subdural hematoma (*white arrows*). CT also shows secondary leftward midline shift and regional mass effect.

MR Imaging Findings. Seven patients had a SWI sequence available for assessment. In all 7 patients, the lesion demonstrated susceptibility-related signal loss secondary to blood products. All 8 patients had T1WI available for assessment, and in all 8 patients, the lesion demonstrated blood products on T1WI. Three patients had an ADC value of less than 1×10^{-3} mm^2/s. Diffusion restriction was in part explained by the blood products but was also noted in the solid enhancing component and the solid nonenhancing component (Figs 2 and 5). The remaining patients did not demonstrate diffusion restriction. The solid component of the lesions demonstrated enhancement in all 8 patients. In 5 patients, the solid component demonstrated homogeneous enhancement; in 2 patients, the solid component enhancement was heterogeneous; and in 1 patient there was peripheral enhancement and central hemorrhage. In 5 patients, more than 75% of the solid component enhanced; in 2 patients, 25%–75% of the solid component enhanced; and only 1 patient had less than 25% enhancement of the solid component.

Four patients presented with accompanying extra-axial hemorrhage on initial MR imaging. Two patients had a subdural hematoma, and 2 patients had subarachnoid hemorrhage (Figs 2–5).

Advanced imaging was not frequently present on preoperative MR imaging examinations. One patient had ^{1}H-MR at both short TE (35 ms) and intermediate TE (135 ms), with elevated choline, decreased *N*-acetylaspartate, and the presence of a lactate peak on both echo times. One patient had PWI on preoperative MR imaging that demonstrated elevated relative cerebral blood volume.

In summary, imaging demonstrated cortical/subcortical lesions in most patients, with the lesions demonstrating blood products on both SWI and T1WI sequences. Accompanying extra-axial blood products were present in 50% of cases.

DISCUSSION

Intracranial sarcomas are rare, with less than 0.5% of sarcomas occurring in the CNS, making their diagnosis challenging.[16,17] Furthermore, intracranial sarcomas typically occur in older populations and rarely affect children, with primary intracranial sarcoma, *DICER1*-mutant, being an exception.[18,19] In addition, the tissue diagnosis of primary intracranial sarcoma, *DICER1*-mutant, based exclusively on the microscopic evaluation of H&E and immunostained sections, can be challenging, given the morphologic overlap with those of the more frequently occurring high-grade primary CNS tumors, such as gliosarcoma. However, the identification of *DICER1*-mutant CNS sarcoma-associated distinctive features, such as prominent eosinophilic cytoplasmic globules, florid microvascular proliferation in a checkerboard pattern at the tumor/brain interface, focal cartilage nodules, patchy expression of muscle differentiation immunophenotypic markers (desmin, myogenin), loss of the H3 K28 (K27) trimethylation mark (H3 K28me3), and stabilized overexpression of the nuclear p53 protein, greatly increase the index of suspicion.[11,12,20] Definitive diagnosis is provided via next generation sequencing or tumor genetic profiling.[11,13,16] A combination of these factors often results in the misdiagnosis or delayed diagnosis of primary intracranial sarcoma, *DICER1*-mutant. Imaging data are scarce and mostly limited to case reports and small series; thus the role of imaging in the initial diagnosis of the entity at the time of clinical presentation remains challenging, and further investigation is needed.[21]

In this case series, we detail the imaging findings of 8 patients with tissue analysis-confirmed primary intracranial sarcoma, *DICER1*-mutant. First, all tumors presented as supratentorial/cortical lesions. This observation comports with reported data indicating a strong predilection for supratentorial sites, which has been documented in 92% of reported cases.[16,20-22] The cortical/subcortical

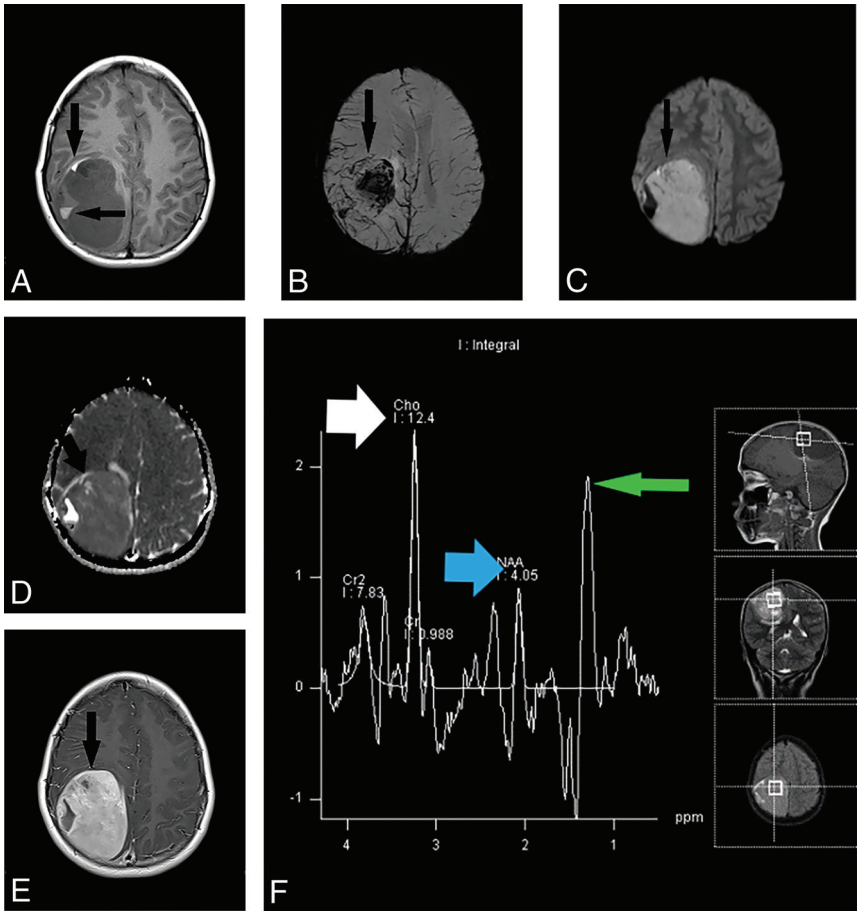

FIG 2. A 10-year-old patient presenting with headaches for 14 days. *A*, Axial precontrast T1 3D MPRAGE demonstrates right superior parietal lobe cortical/subcortical lesion with intrinsic T1 hyperintensity reflective of internal blood products along its anterior and lateral aspects (*arrows*). *B*, SWI sequence demonstrates loss of signal within the lesion, reflective of lesion blood products (*arrow*). *C and D*, DWI and ADC sequences demonstrate diffusion restriction within the lesion that is corresponding to both the solid enhancing component and blood products noted on postcontrast and SWI sequences. *E*, Axial postcontrast T1 MPRAGE demonstrates homogeneous enhancement of the non hemorrhagic component (*arrow*). *F*, Single voxel short TE (35 ms) ^{1}H-MRS demonstrates an elevated choline peak (*white arrow*), decreased N-acetyl aspartate peak (*blue arrow*), and elevated lactate peak (*green arrow*).

location noted in the present case series is consistent with reported data of cortical predilection.[16,21] In 1 patient in the present series, the tumor arose in the hypothalamus, which, to our knowledge, has not been previously reported. There have been reports of cases arising in the pineal and infratentorial regions, which, together with our case, document the rare occurrence of primary intracranial sarcoma, *DICER1*-mutant, in noncortical locations.[16,23]

An interesting observation noted in all patients in our case series was the presence of blood products within the lesion on multiple sequences; in all patients, blood products were noted on both SWI and T1WI. The presence of blood products on T1WI suggested frank hemorrhage within the lesions. This finding serves as a potential imaging phenotypical marker for raising the possibility of primary intracranial sarcoma, *DICER1* mutant, in the imaging differential diagnosis. In addition, 50% of patients presented with associated extra-axial blood products (2 patients with SDH, and 2 with SAH), further suggesting the vascular/hemorrhagic nature of these tumors with their increased propensity to bleed and present urgently with acute neurologic symptoms. This was further supported by the fact that 3 patients presented acutely with NCCT demonstrating acute lesion hemorrhage with/without associated acute extra-axial blood products. This possibly explains the relatively short period of symptoms noted

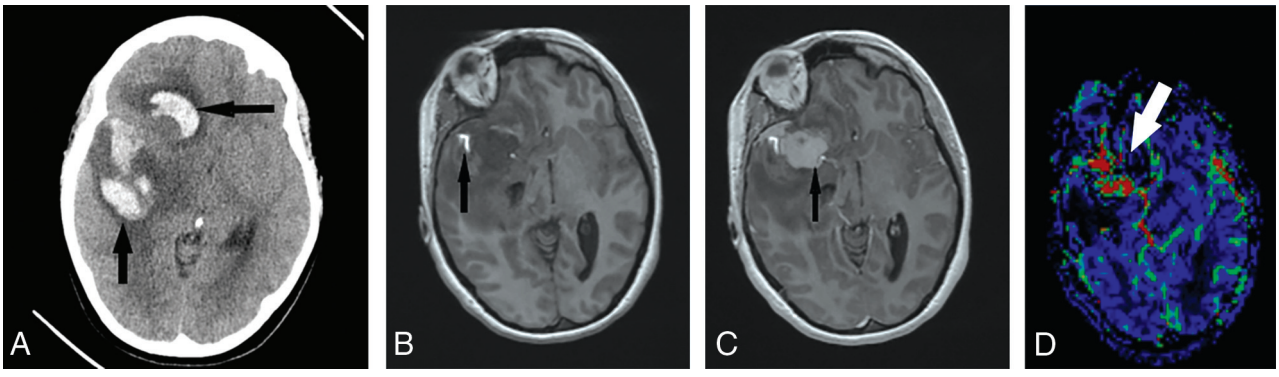

FIG 3. A 33-year-old patient presenting with 5-day history of worsening headaches with a rapid decrease in mentation. *A*, NCCT at the time of presentation demonstrates a large parenchymal hemorrhage involving the right frontal and temporal lobes with associated regional mass effect, leftward midline shift, and right ventricle effacement. *B*, Axial pre contrast T1 MPRAGE demonstrates areas of intrinsic T1 hyperintensity within the right temporal/insular lesion reflective of internal blood products (*arrow*). *C*, Axial postcontrast T1 MPRAGE demonstrates the homogeneous enhancement of the solid component (*arrow*). *D*, Dynamic susceptibility contrast relative cerebral blood volume color map demonstrating elevated relative cerebral blood volume within the enhancing component (*arrow*).

in all 5 patients with a known symptomatic duration of an average of 7 days from onset to presentation. The hemorrhagic nature of the lesion has been described in the pathologic assessment of tumor samples and in a few case reports and case series.[10,12,13,16,21] However, the associated extra-axial hemorrhages have not been described before, and the relative frequency of hemorrhage of the lesion has not been assessed. The combination of these 2 imaging characteristics can serve as a potential clue for suggesting the diagnosis in pediatric patients and young adults.

Finally, the tumors demonstrate a variable degree of solid and cystic components as well as corresponding patterns of enhancement and diffusion restriction. However, in most cases, most of the solid component showed solid homogeneous enhancement.

In summary, primary intracranial sarcoma, *DICER1*-mutant, typically presents as a cortical/subcortical lesion with tumoral blood products and occasionally with extra-axial blood products in younger patients. The presence of these imaging findings should suggest the entity as a possible diagnosis.

Limitations include the retrospective nature of the study as well as the small sample size. However, relative to prior reports, this is the largest dedicated study assessing the imaging features of primary intracranial sarcoma, *DICER1*-mutant. Given the rarity of primary intracranial sarcoma, *DICER1*-mutant, a large data set is difficult to achieve. Last, few patients have had an advanced tumor imaging assessment, with only 1 patient having MR spectroscopy data and 1 patient having perfusion data. However, the findings of this case series serve as a promising tool for larger, multicenter studies in the future that specifically aim to assess the imaging findings of primary intracranial sarcoma, *DICER1*-mutant. Furthermore, these imaging findings can serve as extractable features for machine learning models to aid in the diagnosis and prognosis of this entity.

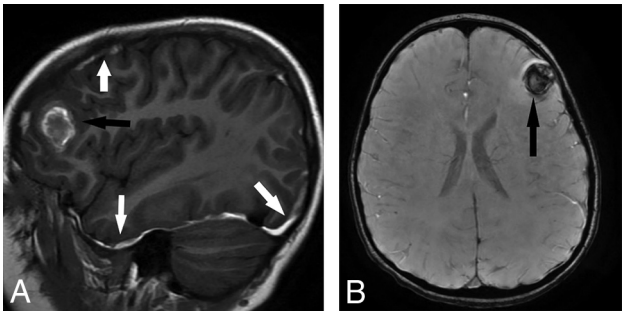

FIG 4. A 5-year-old presenting with headaches for 14 days. *A*, Sagittal precontrast T1 3D MPRAGE demonstrates a left frontal cortical/subcortical T1 intrinsically hyperintense mass that is reflective of internal blood products within the lesion (*black arrow*). Patient also has a left hemispheric thin subdural hematoma (*white arrows*). *B*, SWI sequence demonstrates a loss of signal within the lesion that is reflective of blood products (*black arrow*).

CONCLUSIONS

Primary intracranial sarcoma, *DICER1*-mutant, is a rare, recently described CNS neoplasm that primarily affects pediatric and young adult patients. Detailed, dedicated imaging characterization of the entity is currently lacking in the radiology literature, and the few data that are available are primarily drawn from single case reports. In the present case series, we explore potential imaging findings that are helpful in suggesting this diagnosis. The presence of a cortical lesion with intralesional blood products on SWI and T1-weighted MR imaging, with or without extra-axial blood products, in younger patients should prompt the inclusion of this entity in the differential diagnosis.

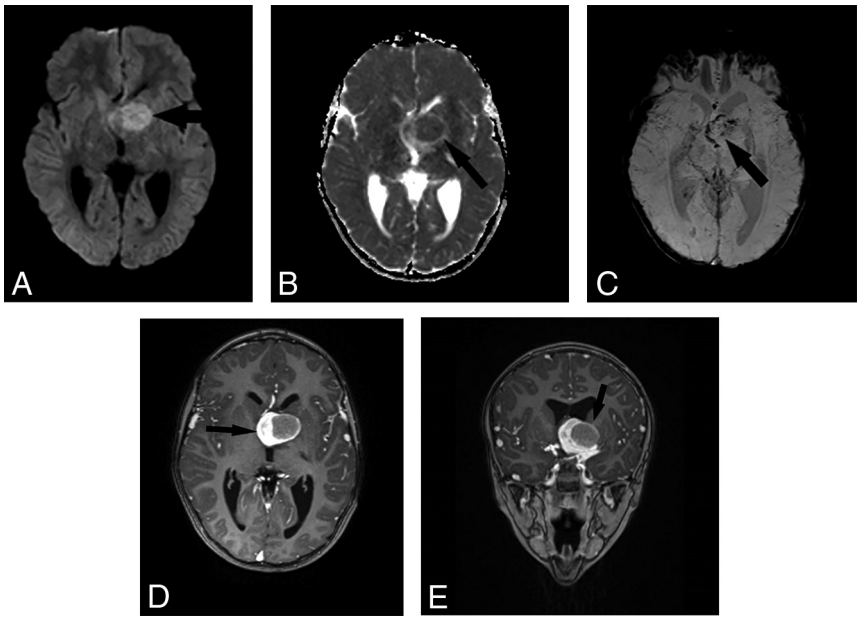

FIG 5. An 8-year-old patient with hypothalamic primary intracranial sarcoma, *DICER1*-mutant. *A and B*, DWI and ADC map demonstrating diffusion restriction of the left hypothalamic lesion nonenhancing component. This is partially explained by the blood products noted on SWI and the cellularity of the nonenhancing component. *C*, SWI sequence demonstrates a loss of signal within the lesion that is suggestive of blood products (*arrow*). *D*, Axial postcontrast T1 MPRAGE demonstrates a mixed enhancing and nonenhancing lesion with homogeneous enhancement of the solid component (*arrow*). *E*, Coronal postcontrast T1 MPRAGE demonstrates a mixed enhancing and nonenhancing lesion with homogeneous enhancement of the solid component (*arrow*).

Disclosure forms provided by the authors are available with the full text and PDF of this article at www.ajnr.org.

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