

## Desmoplastic Infantile Ganglioglioma: A Case Report of a Rare Supratentorial Tumor

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### Abstract

Desmoplastic infantile gangliogliomas are a rare benign tumor almost always found in the pediatric population with a good prognosis following surgical resection. It usually manifests as a very large supratentorial tumor which can be misinterpreted as malignant. It has a good prognosis as complete surgical resection is curative. Other than the age difference, histopathological and radiological findings of desmoplastic infantile ganglioglioma and desmoplastic non-infantile ganglioglioma are similar and are considered under the same WHO classification. In this case report, we have highlighted a case of a 4-year-old female who presented to the outpatient department with episodes of seizures, and loss of consciousness followed by vomiting. The patient was initially diagnosed with an astrocytoma on MRI, underwent surgery and was histopathologically confirmed as desmoplastic ganglioglioma with an excellent postoperative course.

### Keywords

Desmoplastic infantile ganglioglioma, Desmoplastic non infantile ganglioglioma, MRI, Histology, Supratentorial, Primitive neuroectodermal tumor, Ependymoma

### Introduction

In 1987, Vandenberg introduced the term desmoplastic infantile ganglioglioma, which was previously described as composite cerebral neuroblastoma and astrocytoma [1]. It is a benign tumor with cure following a complete surgical resection and is considered as a WHO grade I tumor [2]. Most commonly, it occurs in children below 2 years of age with a male predilection [2, 3]. Here, we report a case of a 4-year-old female who presented with episodes of seizures and loss of consciousness with vomiting over the past 3 years. Due to the suspicion for some intracranial pathology, MRI was performed and showed a large cystic mass with peripheral solid components in the frontoparietal lobe. Neuroimaging and pathological analysis was done. The MRI study was done at 1.5 Tesla with the following sequences: T1W, T2W, FLAIR, and DWI. Post-gadolinium T1W sequences were acquired in the axial, coronal and sagittal planes.

### Case Report

A 4-year-old female with a GCS of 15/15 presented to the outpatient department with a history of episodes of seizures, loss of consciousness, and vomiting. She was the first child in the family and had no significant family history. Birth history included a home birth by normal vaginal delivery. There was

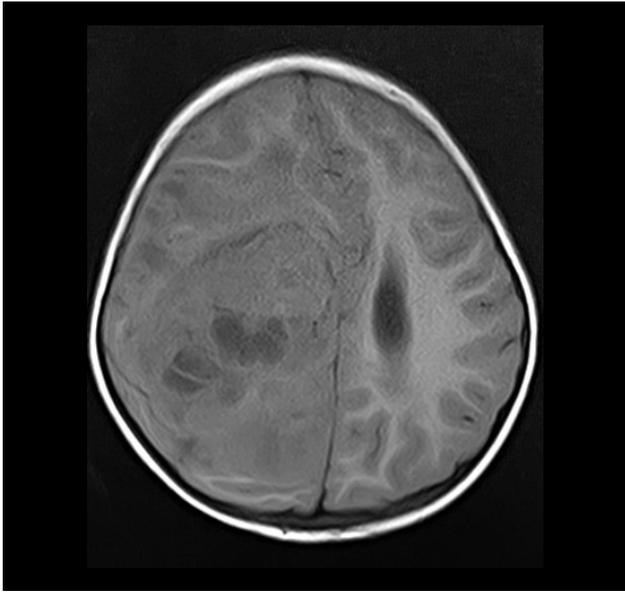


Figure 1: T<sub>1</sub>W axial image showing hypointense to isointense signal in the right.

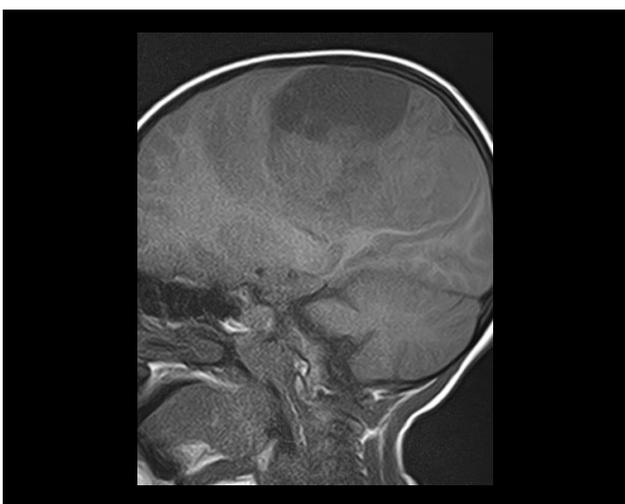


Figure 2: T<sub>1</sub>W sagittal image showing the extent of the tumor.



Figure 3: T<sub>2</sub>W image showing areas of cystic hyperintensity with hypointense solid areas.

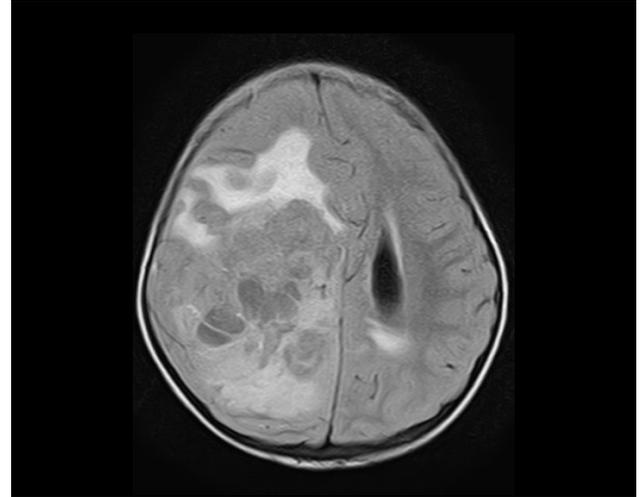


Figure 4: FLAIR sequence showing low signal in the cystic areas with evidence of perilesional edema.

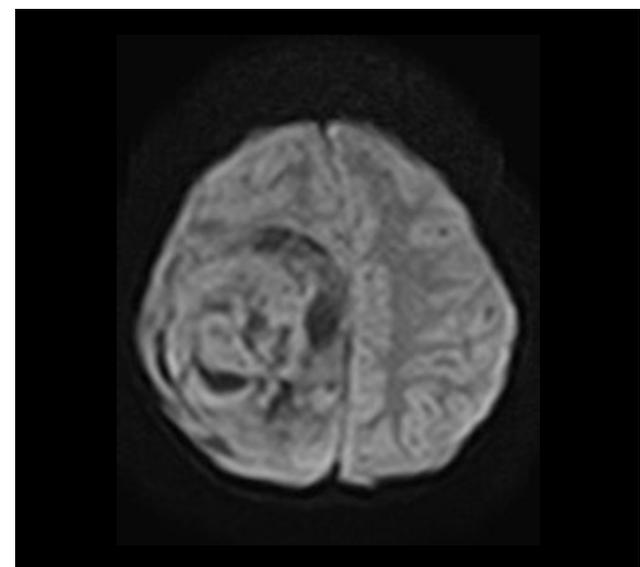


Figure 5: DWI showing no evidence of diffusion restriction.

no history of NICU admission or any perinatal neurological complaints. On ophthalmological examination, the child showed papilledema which aroused suspicion for an intracranial space occupying lesion and thus MRI was advised. MRI was performed and showed a large intra-axial heterogeneous mass in the frontoparietal lobe, which was hypointense to isointense on T<sub>1</sub>, and predominantly hyperintense with solid and cystic components on T<sub>2</sub>.

On FLAIR, the cystic components showed low signal intensity while the solid components were hyperintense. There was no evidence of restriction on DWI. The post-contrast study showed heterogeneous enhancement. The space occupying lesion was surrounded by T<sub>2</sub> and FLAIR hyperintense perilesional edema. The space occupying lesion caused transfalxine herniation with 1.5 cm of midline shift to the left. There was evidence of mass effect on the right lateral ventricle. However, there was no evidence of meningeal enhancement, intralésional calcification, or bone abnormality.

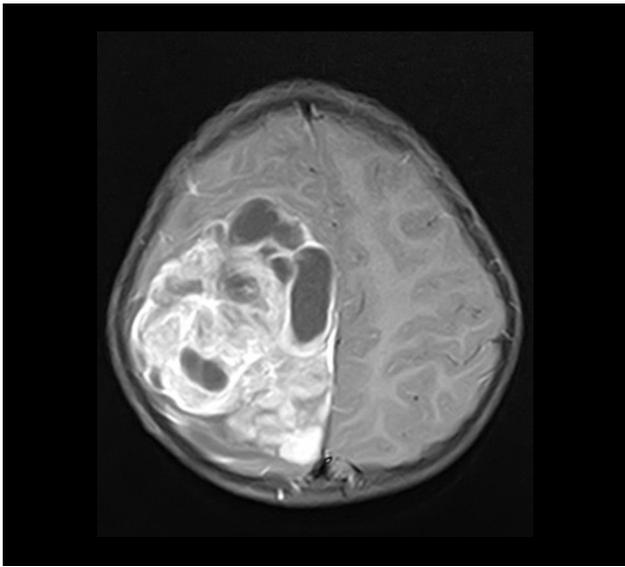


Figure 6: Postcontrast T<sub>1</sub>W sequence showing heterogenous enhancement of the solid component.



Figure 7: Follow up postoperative CT showing craniotomy defect following resection of tumor, with evidence of pneumocephalus.

vasogenic edema in the respective lobes, and midline shift of 1.4 cm to the left with descending tentorial herniation.

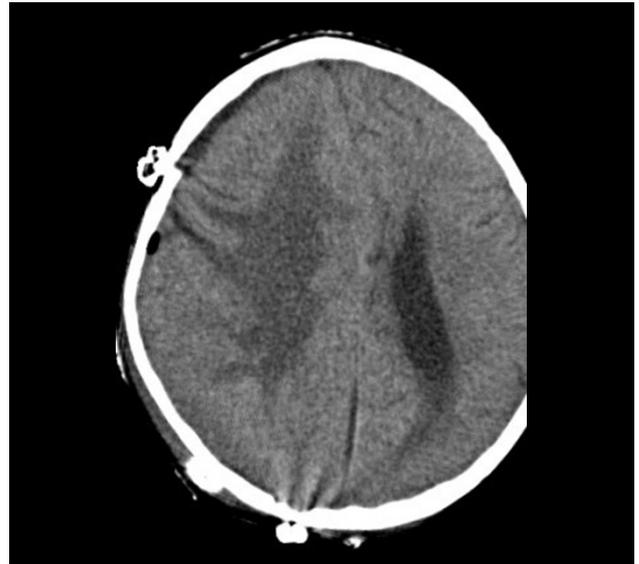


Figure 8: Hypodense area showing vasogenic edema with craniotomy defect and pneumocephalus in right parietal region.

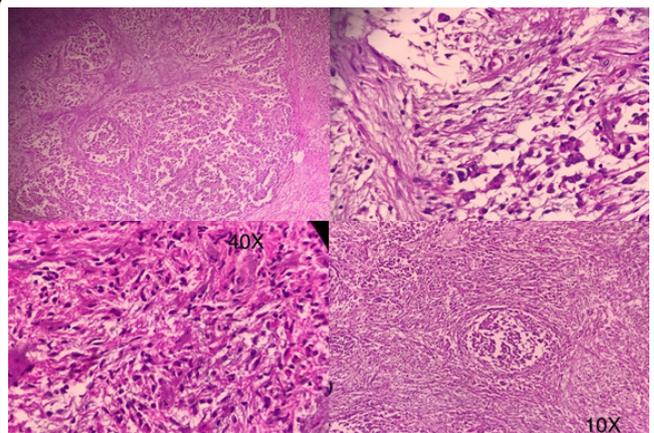


Figure 9: Histology showing neoplastic cells with high pleomorphic vesicular nuclei, and atypical mitotic cells, which are poorly differentiated with stroma demonstrating abundant desmoplasia and focal hyalinization.

The patient was taken for surgical resection and lobectomy was performed. Approximately 90% of the tumor was removed. The specimen was sent for histopathology and the report revealed neoplastic cells arranged in fascicles and a lobular pattern with individual cells demonstrating eosinophilic cytoplasm with high pleomorphic vesicular nuclei containing prominent nucleoli along with atypical mitotic activity. A few poorly differentiated neoplastic cells with small, rounded, deeply basophilic nuclei and stroma were noted. The stroma showed abundant desmoplasia along with focal hyalinization. Masson's trichrome stain showed positive staining of collagen bundles. Postoperative clinical course was unremarkable. The patient had a GCS of 15/15 and no further episodes of seizure. A postoperative CT was performed, which revealed a craniotomy defect in the frontoparietal region with mild

## Discussion

Desmoplastic ganglioglioma is a rare infantile intracranial tumor which arises from glial cells. No known risk factor is linked with the occurrence of this tumor. It however shows a male to female ratio of 2:1 [3]. Usually it is benign and categorized as a WHO Grade I tumor [2]. It usually does not present with devastating symptoms but if symptoms are present, they can include seizures, headache, increased head circumference, bulging fontanelles, and weakness [4]. The tumor has an excellent prognosis and surgical resection is sufficient, however chemotherapy and radiotherapy may be required if complete resection is not achieved [6]. These tumors have no risk of recurrence. Radiological and pathological correlation can lead to diagnosis of this condition. CT and MRI play a crucial role in diagnosis, and usually show a large

mass with cystic and solid components with enhancement of solid components on post-contrast studies. The differential diagnoses include primitive neuroectodermal tumors and ependymomas. However, in these cases histological analysis reveals features of a storiform pattern of spindle cells with evidence of desmoplasia and the presence of ganglion cells. Desmoplasia is GFAP positive, while spindle and ganglion cells are reactive to synaptophysin [7]. The differential diagnosis of histopathology includes pleomorphic xanthoastrocytoma, which is reticulin rich with prominent lipidization and absence of a neural component, as well as gangliofibroma, which also lacks a neural component [5].

## Conclusion

Desmoplastic infantile gangliogliomas are a rare pediatric supratentorial tumor. Despite its large size, due to its less aggressive course and lower grade, it carries an excellent prognosis following resection. Therefore, it must be considered on the differential diagnosis of a large supratentorial mass in the pediatric population.

## Conflict of Interest

Authors declare no conflict of interest.

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