Case Report

Desmoplastic infantile ganglioglioma: Report of an unusual case with a cranial defect

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ABSTRACT

Desmoplastic infantile ganglioglioma (DIG) is a rare tumor that typically occurs in infants under the age of 24 months. These tumors commonly have a good prognosis after surgical resection despite their aggressive radiological appearances. Clinical signs are due to the large size of the tumor and include increased head circumference, bulging fontanel, sunset sign and seizures. We report an unusual DIG case who presented with parietal bulging associated with a bony defect. The patient was thought to have a leptomeningeal cystic formation, but on his cranial magnetic resonance imaging (MRI), we observed a centrally and homogeneously gadolinium-enhanced lesion fixed to the dura by its solid component. A surgical gross total resection was performed, and no residual tumor was observed on follow-up.

Key words: Bony defect, desmoplastic, ganglioglioma, pediatric, skull deformation, supratentorial tumor

Introduction

Desmoplastic infantile ganglioglioma (DIG) was first described by Vandenberg *et al.* in 1987.[1] DIG is a rare supratentorial neuroepithelial tumor with dense desmoplastic tissue and divergent astrocytic and ganglionic differentiation.[1,2] It primarily occurs in infancy, with a male:female ratio of 1.7:1.0.[2,3] It is classified as a benign tumor and coded as WHO grade 1.[2,4]

The supratentorial region is preferentially involved, especially the frontal and parietal lobes, followed by the temporal lobe.[1,4-6] Most of these tumors have a favorable prognosis after gross total resection,[1,2,5,6] despite their infiltrating pattern and an aggressive radiological appearance. Most DIGs do not require adjuvant therapy, even after incomplete resection.[4,6]

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When they reach a large size, patients with these tumors present with increased head circumference with tense bulging fontanel, headache, seizures and motor delay.[2,6,7]

We report an unusual case with clinical signs of a parietal bony defect and bulging.

Case Report

History and examination

A 9-month-old boy with right parietal bulging presented to the emergency department. Before neurosurgery consultation, the emergency physician suggested a leptomeningeal cyst due to head trauma. However, the boy had no previous definitive head trauma history but had an accompanying increased head circumference (47 cm, 90th percentile), motor delay within 1.5 months and rapid onset of epileptic seizures 2 weeks previously. Cranial computerized tomography (CT) scans showed a cranial bony defect in his right parietal bone [Figure 1]. Magnetic resonance imaging (MRI) showed that the tumor was characterized by a cystic mass with a central homogeneous solid component enhancing with gadolinium attached to the dura, leading to a large effect on the ventricles in the temporoparietal region [Figure 2].

Antiepileptic treatment was commenced for seizure prevention.

Treatment

A frontotemporal craniotomy was performed. A mass was observed to be attached to the dura. The solid component of the tumor was solid, vascularized and barely distinguished from normal glial tissue. The cystic part of the tumor was aspirated. The postoperative period was uneventful. Total removal of the tumor was documented on a control MRI. Because there was subdural hygroma formation squeezing the brain tissue on CT scan on the third day of the postoperative period, a subdural-peritoneal shunt was inserted. His 6-month follow-up revealed no identifiable pathology.

Histological examination

The histological examination revealed a mixed glial-neuronal tumor presenting with a nodular pattern involving the cerebral cortex and subcortex. Astrocyte-like cells had large, round and vesicular nuclei with surrounding large amphiphilic cytoplasm. Neuronal cells had vesicular chromatin and polygonal nuclei with large perikarya. A few multinucleate cells were observed. The cells were surrounded by diffuse reticulin and collagen fibers. There was only atypia, with no mitotic figures, necrosis or vesicular euchromatic nuclei (VEP). Vasogenic edema was present around the neural tissue, with non-gemistocytic gliosis. The conventional histochemical stains Massontrichome (MTC) and Gomori'sreticulin stain were used to stain interstitial collagen fibers and reticulin fibers, respectively [Figure 3]. Tumor cells were immunohistochemically stained for glial fibrillary acidic protein (GFAP) [Figure 4], synaptophysin [Figures 5 and 6], chromogranin, S-100 and epithelial membrane antigen (EMA), and they were negative for CD34, NFP and progesterone. Nearly 2% of tumor cells were labeled with Ki-67/MIB-1[Figure 7], and approximately 1/3-2/3 of cells showed moderate nuclear staining for p53. The diagnosis was desmoplastic infantile ganglioglioma grade I (WHO, 2007).

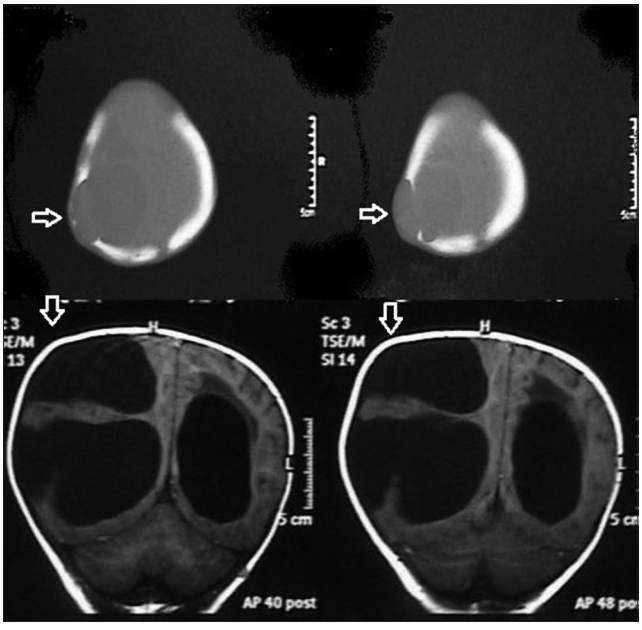

Figure 1: Bone window axial CT images and coronal section of MRI show a bone defect located in the right posterior parietal region with skin bulging

Discussion

Desmoplastic infantile gangliogliomas are rare cerebral tumors classified as WHO grade I, with an indolent prognosis. They present within the first 18 months of life and have a male predominance.[2] There are a few cases in the literature regarding CSF dissemination and malignant transformation.[2,4,8,9] DIGs are well documented to show the presence of numerous mitoses accompanied by increased MIB-1 indices and the presence of necrotic foci.[2] Thus, CSF examination is recommended in every patient with deeply located and subtotally resected DIGs.

The most common presenting symptoms are enlarged head circumference, seizure, symptoms of increased intracranial pressure, and hemiparesis.[6,7]

Patients can also present to a physician with complaints that appear over time, such as macrocrania and swelling over the fontanel. Some cases of bone abnormalities adjacent to a tumor have been reported in the literature.[4,7] However, thinning of the cranial bones or defects of the bones as a result of elevation of the intracranial pressure is not a common sign. Only one of six cases had a skull deformation in a study by Guillaume.[4] In our case, the patient was referred to a

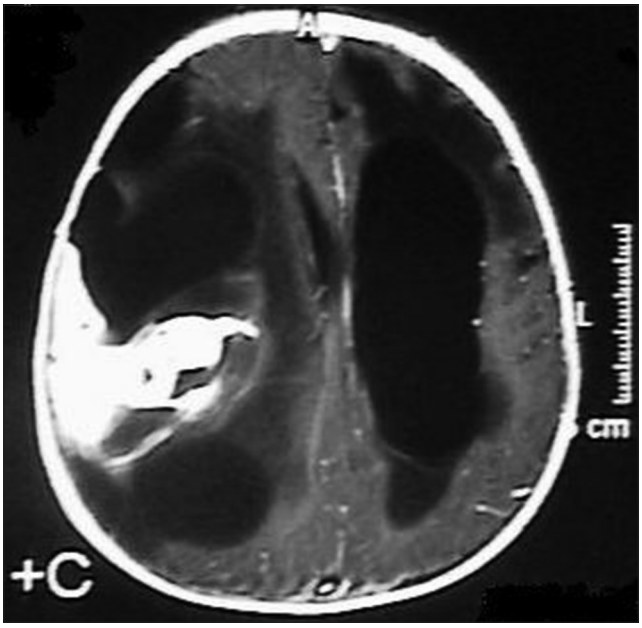

Figure 2: Axial T1-weighted MRI with gadolinium enhancement reveals a mass lesion with irregular borders with heterogeneous contrast enhancement extending posterior and superior towards the temporoparietal structures accompanied by a non-enhancing cystic component

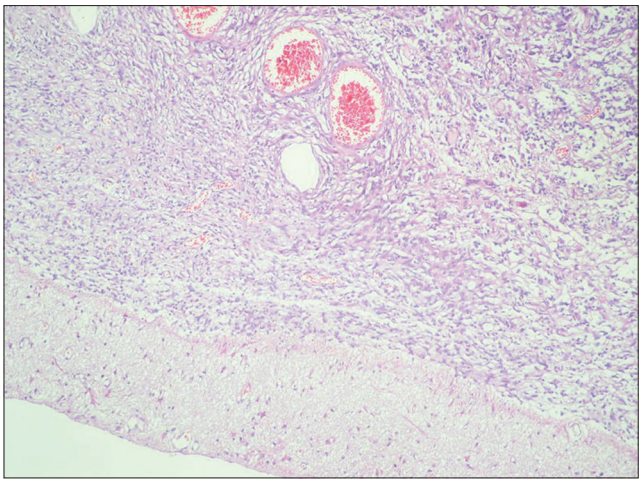

Figure 3: Tumor having a nodular pattern involving cerebral cortex and subcortex (H&E, ×100)

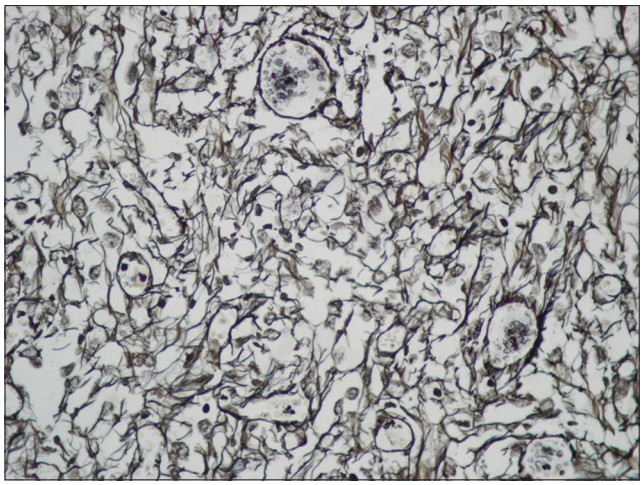

Figure 4: Diffuse interstitial reticulin fibers (Gomori'sreticulin, ×200)

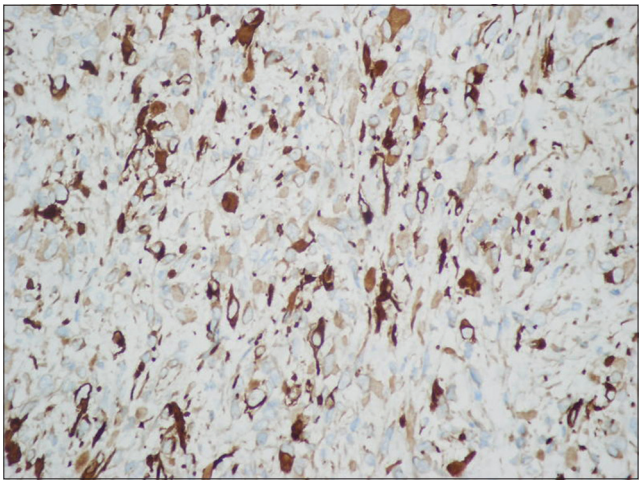

Figure 5: Randomly distributed intermingled GFAP reacting astrocytes in tumor are seen. (Streptavidin–biotin complement, GFAP, ×200)

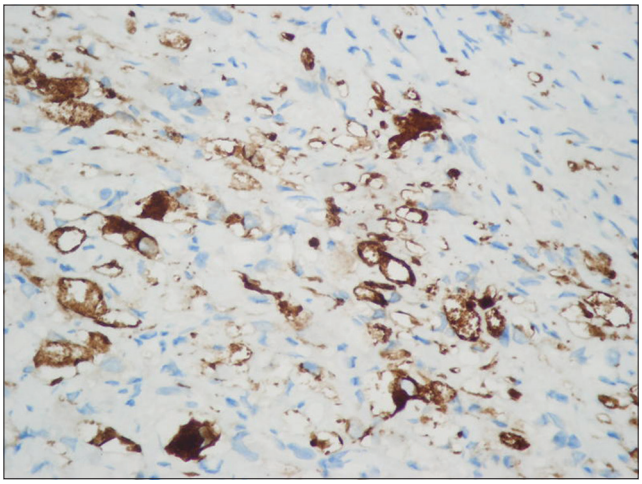

Figure 6: Scattered synaptophysin reacting ganglion cells are a major component of tumor. (Streptavidin-biotin complement, synaptophysin, ×200)

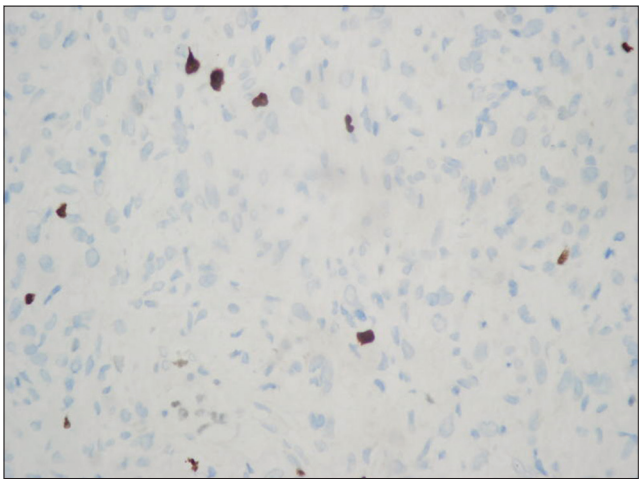

Figure 7: MIB-1 index is 2% (Streptavidin–biotin complement, anti-Ki-67 [MIB-1], ×200)

physician because of swelling under the skin. It was found that the bone tissue adjacent to the tumor was almost eroded, and the overlying dura was herniated into the skin.

Although DIGs are considered to be benign tumors, deeply located DIGs present aggressive behavior. The best choice of treatment is complete surgical resection. The use of adjuvant therapy is still controversial, particularly in incompletely resected tumors. However, these tumors are common at a young age; therefore, in partially resected cases, only neuroimaging is recommended for follow-up. There may be a need for adjuvant chemotherapy in deep-seated tumors with malignant histological features.

In conclusion, a cranial defect is not a common clinical finding of DIGs. A progressive increase in intracranial pressure might lead to head circumference enlargement accompanied by a cranial defect and bulging of the dura over the tumor. Therefore, DIG should be considered among differential diagnoses when there are any signs or symptoms reflecting cranial bone defects.

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