Purely Cystic Desmoplastic Ganglioglioma: An Incidental Finding in a Giant Cyst

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Abstract

Desmoplastic infantile ganglioglioma (DIG) is a rare supratentorial brain tumor occurring mostly before the age of 2 years. It has a good prognosis and total excision of the tumor is curative, necessitating no further treatment. An accurate pathologic diagnosis is crucial. Until now, approximately 70 cases of this tumor type have been reported. Solid-cystic variety is mostly described with occasional purely solid type lesion also. We are presenting a 12-month-old male infant with developmental delay and a giant cyst on the right side of supratentorial compartment, which showed histological features of desmoplastic ganglioglioma. Hence, the current case is reporting the rare histologic finding of cystic change in desmoplastic ganglioglioma. Cystic form of desmoplastic ganglioglioma should be considered in differential diagnosis of supratentorial cystic lesion in an infant. Prognosis is excellent after complete excision.

Keywords
- cystic
- desmoplastic infantile ganglioglioma
- histopathology

Introduction

Desmoplastic infantile tumors (DITs) are rare benign lesions affecting infants, first described by Taratuto et al[1] in 1984. Neuroimaging typically shows a massive cystic and solid, well-defined supratentorial lesion. Two histologic forms have been described: desmoplastic infantile astrocytoma (DIA) and desmoplastic infantile ganglioglioma (DIG).[1,2] DIG was first described by VanderBerg et al[2] in 1987. Now this tumor is recognized as a distinct entity and is included in the World Health Organization (WHO) classification under the category of neural and mixed glioneuronal tumors, and is in the grade I of the WHO classification.[3] Until 2013, approximately 70 cases of DIG reported in literature.[4]

We are presenting a 12-month-old male infant, with developmental delay and a giant cyst on the right side of supratentorial compartment, which showed histologic features of desmoplastic ganglioglioma. Hence, the current case is reporting the rare histologic finding of cystic change in desmoplastic ganglioglioma.

Case Report

A 12-month-old infant was referred to neurosurgery out patient department from pediatrics department with history of developmental delay and a large cyst on the right side of supratentorial compartment. He was the first child of the family, born with full-term normal delivery. The family history was unremarkable. Physical examination revealed delayed developmental milestones, with head circumference of 44 cm; anterior fontanelle was open and tense. Magnetic resonance imaging (MRI) scan showed a large Hypointense area in the right frontotemporoparietal region, with midline shift. Computed tomography (CT) of the head showed large cystic lesion in right frontotemporoparietal region. The patient was planned for cyst decompression with provisional diagnosis of giant arachnoid cyst.

A right temporoparietal craniotomy followed by total excision of the cyst wall was done. Cyst contained crystal-clear fluid resembling cerebrospinal fluid (CSF). Excised cyst wall was sent for histopathology. The postoperative period was

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uneventful. Postoperative CT of the head showed adequate cyst decompression with reduced midline shift (►**Fig. 1**).

Histopathology showed strips of folded membranous structures composed of fibrocollagenous tissue containing glial and neuroepithelial elements with scattered ganglion cells. The stroma was fibroblastic with increase in reticulin fibers. There was no mitosis, necrosis, or any evidence of malignancy. On immunohistochemistry (IHC), cells in the membranous structure were positive for glial fibrillary acidic protein (GFAP) and synaptophysin. Features were suggestive of desmoplastic ganglioglioma with cystic changes (►**Figs. 2, 3**).

Until the last follow-up (6 months after surgery), the patient remained asymptomatic with lax anterior fontanelle with persistent delayed developmental milestones.

Discussion

DIGs are rare developmental neuroepithelial tumors, probably arising from neural progenitor cells, in the subcortical zone along with mature subpial astrocytes.[5] They are rare WHO grade I tumors of infancy, characterized by large-volume, superficial location, invariably supratentoriality, frontoparietal lobe predilection, and morphologically by an admixture of astroglial and neuroepithelial elements in a desmoplastic milieu.

DIGs are most likely diagnosed in the first 2 years of life.[3] Boys are affected more commonly than girls.[6] Symptoms of DIG include intracranial hypertension, sunset eye, enlarging head circumference, bulging fontanels, and variable localizing signs, including seizures or paresis.[7]

They are massive, firmly attached to the dura, and extensively infiltrate the subarachnoid space but do not involve the ventricular system.[8] Most commonly, CT scan and MRI show a large superficial large cerebral mass with solid and cystic areas.[6] The solid component of the tumor frequently shows contrast enhancement.

Although Tseng et al[9] described cystic changes in a purely solid DIG on follow-up over a period of 18 months. Duffner et al[10] and Sperner et al[11] have published two cases of DIG that have been purely solid. The present case is purely cystic form of desmoplastic ganglioglioma. Differential diagnosis of supratentorial cystic lesion in an infant includes primitive neuroectodermal tumor, ependymoma, ganglioglioma, and dysembryoplastic neuroepithelial tumor.[12]

Histologically, the most prominent feature of DIG is desmoplasia and spindle cells with a storiform pattern of arrangement.[7] There is also a ganglion cell component, which is present as single cells or clusters.[13] The first component can be shown to be GFAP positive, but the latter component is of neuroepithelial origin and reactive with markers such as synaptophysin.[13]

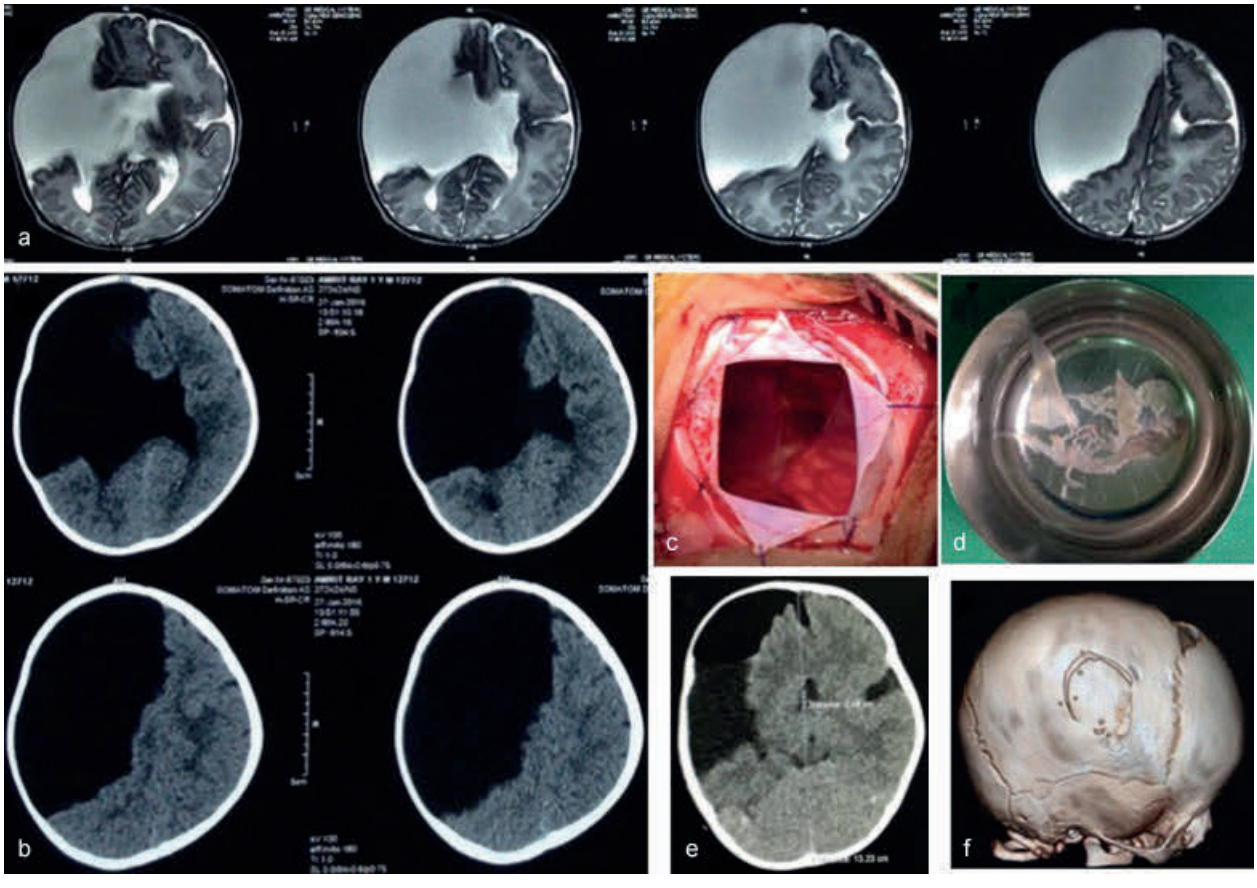

Fig. 1 (**a**) MRI of the brain and (**b**) CT of the head showing giant right frontotemporoparietal cyst with midline shift. (**c**) Intraoperative photograph is showing giant cyst with clear fluid and (**d**) excised cyst wall. (**e**) Postoperative CT of the head showing adequate cyst decompression with reduced midline shift and (**f**) right parietal bone flap. CT, computed tomography; MRI, magnetic resonance imaging.

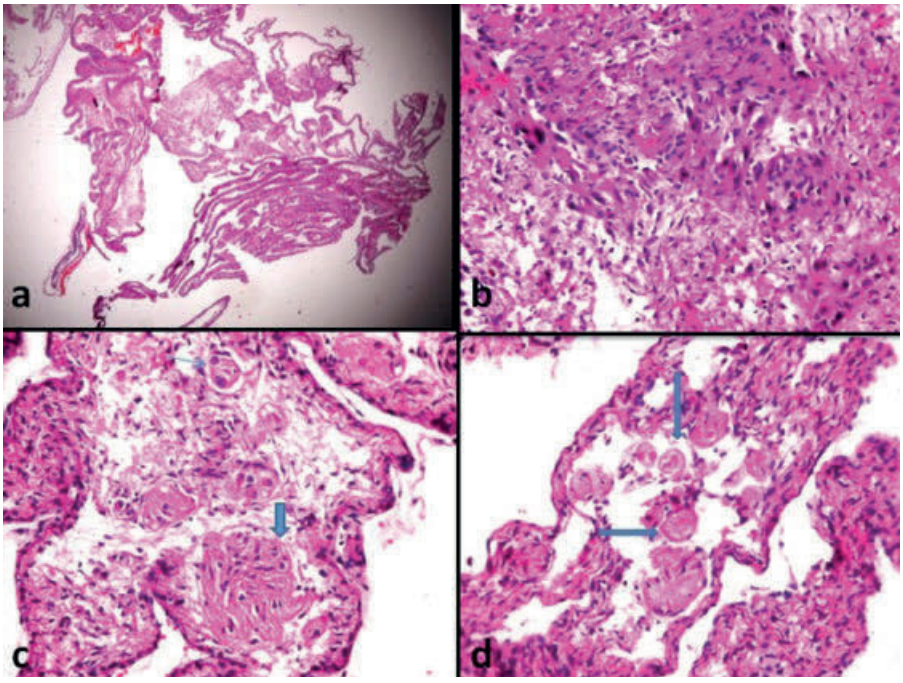

Fig. 2 (**a**) Scanner view of cyst wall consisting of fibroblast-like cells (H&E stain ×40). (**b**) Neoplastic astrocytes and atypical ganglionic cells (H&E stain ×400). (**c**) Nerve plexuses (thick arrow) and scattered ganglion cells (thin arrow, H&E stain ×400). (**d**) Ganglion cells (arrows) in higher magnification (H&E stain ×400). H&E stain, hematoxylin and eosin stain.

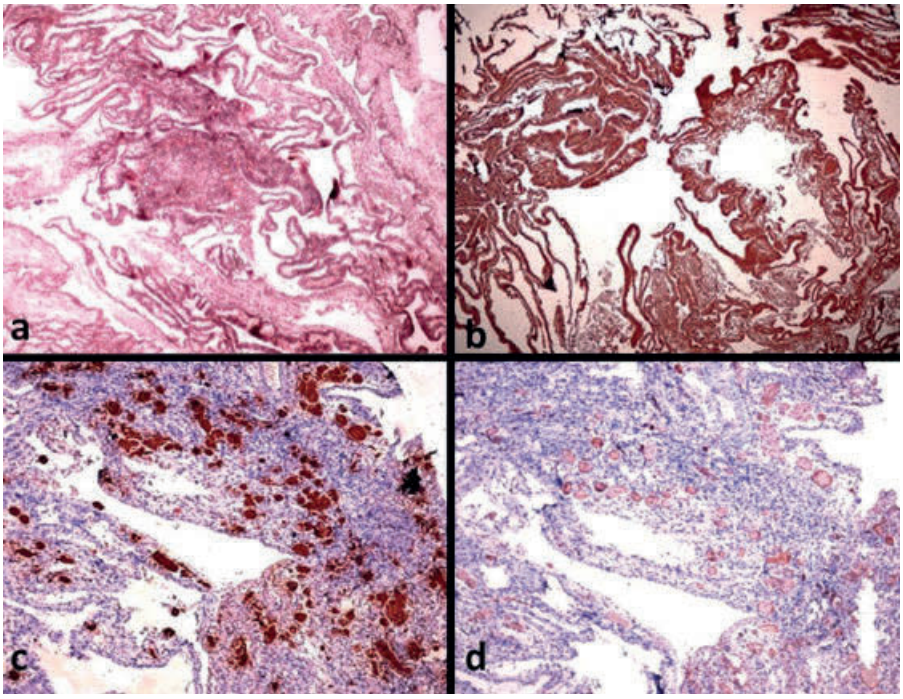

Fig. 3 (**a**) Reticulin stain highlights the reticulin fibers (reticulin stain ×40). (**b**) Diffuse vimentin positivity (IHC stain ×40). (**c**) Ganglion cells highlighted by GFAP (IHC stain ×400). (**d**) Synaptophysin staining the ganglion cells (IHC stain ×400). GFAP, glial fibrillary acidic protein; IHC stain, immunohistochemical stain.

The present case was a 12-month-old baby with large head. CT showed a large cyst in right side of the head. Being benign slow-growing tumor, it is likely that the lesion started in intrauterine in the fetus. Pathology of the cyst wall revealed histologic and IHC features consistent with desmoplastic ganglioglioma.

Complete excision is usually curative with no further additional therapy.[14] Although DIGs are considered to be

benign tumors, deeply located DIGs rarely can be aggressive. The best choice of treatment is complete surgical excision. The use of adjuvant therapy is still controversial, particularly in incompletely resected cases. However, these tumors are common at a young age; therefore, in partially resected cases, regular neuroimaging is recommended in follow-up. There may be a need for adjuvant chemotherapy in deep-seated tumors with malignant histologic features; however, there is no consensus in this regard.

In conclusion, cystic form of desmoplastic ganglioglioma should be considered in differential diagnosis of supratentorial cystic lesion in an infant. Prognosis is excellent after complete excision.

Funding
None.

Conflict of Interest
None.

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