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Desmoplastic infantile ganglioglioma; a rare supratentorial brain tumor of infancy

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Abstract

Desmoplastic infantile ganglioglioma (DIG) is rare uncommon intracranial brain tumor of infancy comprises of 0.5-1.0% of all the intracranial tumors. DIGs are commonly located in supratentorial region superficially with more affection to the frontal and parietal lobes and have voluminous size with both cystic and solid components. Histologically it is mixed glial and neuronal brain tumor and showed intense desmoplastic reaction at the periphery of the tumor and dural attachment. DIGs are most benign intracranial tumors and labeled as World Health Organization (WHO) grade-I. Data available from the literature suggest that surgical resection is the mainstay of treatment and no chemotherapy or radiotherapy is indicated if complete resection of the tumor has been achieved. In general, DIG has good prognosis and recurrence free intervals of up to 14 years have been reported. In our case report, a-12-month old girl presented to emergency department with history of first seizure of her life. Seizure duration was less than a minute, focal type and confined to mouth and left upper limb. Magnetic resonance imaging (MRI) brain with gadolinium showed diffuse right frontoparietal localized mass with well-defined right temporal cyst measuring 4×3×2.7 cm without mural nodule and gadolinium enhancement. There was extensive dural thickening and enhancement seen in the anterior temporal region on post contrast. Electroencephalogram was normal. Patient underwent right craniotomy and gross total excision of tumor done. Frozen section and final histology reported DIG (WHO grade-I).

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Introduction

Desmoplastic infantile gangliogliomas (DIGs) are rare primary tumors that comprise 0.5%-1.0% of all the intracranial tumors (1-4). DIG was first described by VandenBerg et al in 1987 (5). These tumors occur mostly with in the first 18 months of life and seen commonly in supratentorial region as a large cerebral mass with both cystic and a small solid component with intense desmoplastic reaction at the periphery to the tumor with attachment to the dura. The presence of atypical or immature cellular tissue is the hallmark of DIG (6). In this paper, we present a case of 12 months old girl was presented to the emergency department of the hospital with history of left sided focal seizure.

Key Point

Desmoplastic infantile ganglioglioma (DIG) is rare uncommon intracranial brain tumor of infancy comprises of 0.5%-1.0% of all the intracranial tumors. We present a case of 12 months old girl was presented to the emergency department of the hospital with history of left sided focal seizure. Our patient was regularly followed-up in outpatient clinic every 6 months and seen last time in December 2014 (approximately 22 months after surgery) with a recent magnetic resonance imaging (MRI) brain with gadolinium showed no evidence of tumor recurrence. She does not have any disability, thriving and seizure free.

Case Report

A-12-month old girl was presented to emergency department of the hospital with history of left sided focal seizure. Patient was full term and delivered by an uncomplicated spontaneous vaginal delivery. Apgar score was 9 and birth weight was 2745 g. Family history was unremarkable. Patient attained normal mile stones. Patient had first seizure of life that was confined to the mouth and left upper limb. Post seizure attack, she became drowsy for 30 minutes. No history of

vomiting or fever associated. General physical and neurological examination was normal. Anterior fontanelle was small and flat. Head circumference was 45 cm. Pediatric ophthalmologist consulted and no papilledema seen. Pediatric neurologist was consulted and phenobarbital was started empirically. Computerized tomography (CT)-scan brain plain showed large cystic lesion sized 4.1×2.7 cm occupying the right temporal lobe with surrounding edema causing effacement of right lateral ventricle. Electroencephalogram reported no epileptiform discharges noticed. MRI brain with gadolinium showed diffuse right frontoparietal localized mass with well-defined right temporal cyst measuring 4×3×2.7 cm without mural nodule and gadolinium enhancement. There was extensive dural thickening and enhancement seen in the anterior temporal region on post contrast (Figure 1).

Patient underwent right temporoparietal craniotomy with exposing the lateral part of frontal lobe, lateral part of temporal lobe and frontal operculum was done. The exposed cortex was grossly abnormal pale, flattened and tight. Tumor was encountered superficially on the temporal side while slightly deep in the frontal side. Right temporal cyst was drained and yielded clear amber colored fluid. Gross total excision of tumor was done including the part of frontal and parietal lobes and thickened anterior temporal dura. Post operatively patient developed left dense hemiplegia which had gradually but fully recovered. Phenobarbital was tapered and discontinued 3 months after surgery. No seizures seen during follow-up period.

Gross histopathology examination showed multiple fragments of soft grayish tissue measuring 4×3.5×0.5 cm and a second specimen consisting of large, soft, firm piece of irregular tissue with dural fragment measuring 5.5×5×2.5 cm. Microscopic examination showed dural attached tumor that was well delineated from normal brain. The tumor was composed of enlarged spindle shaped astrocytes and ganglion type cells with prominent desmoplastic stroma. No evidence of necrosis, vascular proliferation or mitotic activity seen (Figure 2).

Immunohistochemistry (IHC); the tumor was positive for reticulin stain, GFAP (in glial cells), NSE, neurofilament and synaptophysin (in neurons). A diagnosis of DIG grade-I (according to WHO Classification) was made.

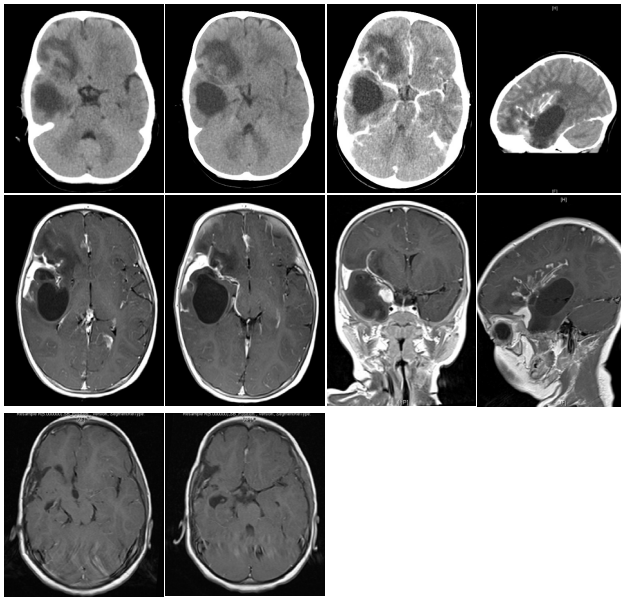

Figure 1. MRI brain with gadolinium showed diffuse right frontoparietal localized mass with well-defined right temporal cyst measuring 4×3×2.7 cm without mural nodule and gadolinium enhancement. There was extensive dural thickening and enhancement seen in the anterior temporal region on post contrast.

Discussion

DIG was first described as a rare intracranial tumor of infancy in 1987 by VandenBerg et al (5). The histopathological grading of the tumor was initially considered to be WHO grade-I until the year 2000, when an anaplastic variety was documented which was ultimately fatal (7). The most common presentation is under the age of 1 year (8), but a few exceptional cases are also reported in young adults (5). Males are more commonly affected with ratio of 2:1 and rapidly increasing head circumference in unclosed fontanelle is the most common symptoms (6). Patients may also present with variable localizing signs like seizures and paresis (9). DIGs are very rare tumors accounts for only 0.04% of all intracranial tumors (10). DIG have good prognosis with a survival of 8-20 years have been reported (5,7,10,11). Spontaneous regression of tumor after subtotal resection has also been documented (12,13).

Morphologically DIG present as exceptionally large cerebral hemispherical mass composed of both solid and cystic portions (6). The most common location of these tumors is in the frontal or parietal lobes and occasionally in the temporal lobes (5,6,14). Multi-lobar involvement is also frequent with over 60% presentation. The solid portion of the tumor frequently showed contrast enhancement and calcification in the tumors has not been documented (15). These tumors present with intense desmoplastic reaction at the periphery of the mass with attachment to the dura and have atypical cellular tissue distinguishing from con-

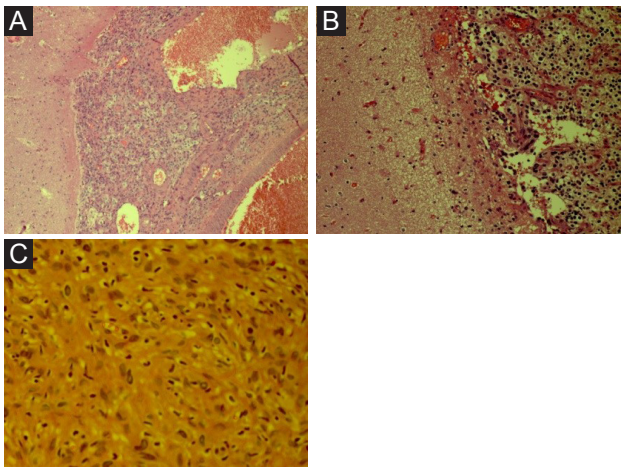

Figure 2. (A) Dural attached tumor that was well delineated from normal brain. (B) The tumor was composed of enlarged spindle shaped astrocytes and ganglion type cells with prominent desmoplastic stroma. (C) No evidence of necrosis, vascular proliferation or mitotic activity seen.

ventional ganglioglioma (6). Purely solid consistency of DIG has also been described in literature (6,16). Proliferative activity is very low in these tumors (17); however focal hypercellularity and mitotic activity in DIG may need to suspicion of high grade neoplasm. No CSF seeding or metastatic spread has been described (6).

Immunohistochemically DIG reacts diffusely with GFAP, NSE, synaptophysin and neurofilament elements (18). The differential diagnosis for DIG includes reticulin rich desmoplastic tumors such as pleomorphic xanthoastrocytoma (PXA) and gliofibromas (19,20). Also anaplastic large cell lymphoma (ALCL) may also be confused with DIG which is exceedingly rare and occurs in older age groups (21).

Surgery is the main stay of the treatment with complete excision of the tumor (12). The role of adjuvant therapy is very limited and chemotherapy is given to those candidates having high grade tumors showing brisk mitosis, aneuploidy and increased MIB labeling. It is also given to patients with tumors involving eloquent region of the brain, not safe for surgery (13). Our patient was regularly followed-up in out-patient clinic every 6 months and seen last time in December 2014 (approximately 22 months after surgery) with a recent MRI brain with gadolinium showed no evidence of tumor recurrence. She does not have any disability, thriving and seizure free.

Authors' contribution

All the authors wrote the first draft of the manuscript equally. AAK prepared the final paper. All authors read and signed the final manuscript.

Conflicts of interest

The authors declared no competing interests.

Ethical considerations

Ethical issues (including plagiarism, data fabrication, double publication) have been completely observed by the authors.

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