Desmoplastic infantile ganglioglioma: A questionably benign tumour

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SUMMARY
Desmoplastic infantile ganglioglioma is a rare intracranial tumour of childhood that involves the cerebral cortex and the leptomeninges. We report two patients with desmoplastic infantile gangliogliomas and multiple cerebrospinal metastases. To our knowledge, only two similar cases have been reported in the published literature. Pathologically, this rare intracranial tumour shows glial and ganglionic differentiation, accompanied by an extreme desmoplastic reaction. These are low-grade neoplasms that are questionably benign.

Key words: desmoplastic infantile ganglioglioma; magnetic resonance imaging.

INTRODUCTION
Desmoplastic infantile gangliogliomas (DIG) are rare primary neoplasms that comprise 0.5–1.0% of all the intracranial tumours.¹⁻³ They occur in infants. They are tumours commonly seen in the frontal or the parietal lobes with a large cystic and a small solid component representing the desmoplastic reaction located adjacent to the meninges and attached to the dura, demonstrating enhancement on the meningeal side of the mass.¹,⁴ We have encountered two children with features of a DIG with cerebral and spinal metastases.

CASE REPORTS
Case 1
A 3-month-old boy was brought to the emergency department of the hospital with a provisional diagnosis of meningoencephalitis. Examination revealed a full anterior fontanelle, irritability, one episode of clonic jerks of the left arm and left leg for a few seconds and evidence of a healing Herpes zoster rash.

The electroencephalogram showed bilateral abnormality, greater on the left side.

Magnetic resonance imaging of the brain showed a predominantly cystic mass lesion in the left cerebral hemisphere in the frontal, parietal and occipital regions. It had a small cortically based solid component that was centrolateral in relation to the cystic portions. The solid component enhanced intensely and homogeneously with gadolinium. Mass effect was noticed in the form of effacement of adjacent sulci, the left lateral ventricle and midline shift to the right side (Fig. 1). The preoperative diagnosis was that of DIG.

At surgery, the solid part of the mass was hard in consistency, cortically based and also adherent to the dura. It was completely resected.

A postoperative MRI showed a left-sided subdural hygroma that was treated with a subduroperitoneal shunt. He was readmitted about 8 weeks later with seizures and a repeat MRI showed enhancing tissue in the tumour bed and leptomeningeal enhancement in basal cisterns and in the supratentorium. These findings were suggestive of metastases (Fig. 2a). Progress MRI a week later revealed an increase in the number and size of metastatic foci along the basal cisterns, over the left cerebral hemisphere (Fig. 2b) and the spinal cord (Fig. 3). The boy is alive 8 months after the diagnosis.

Case 2
A healthy 4-month-old boy was admitted for the investigation of progressive increase in head circumference in addition to unusual head movements, irritability and lethargy. Examination revealed increased head circumference, a pulsatile anterior...
fontanelle, sunsetting of the eyes with inability to fix and follow, sluggish right pupil, antigravity of the limbs as well as equivocal plantar reflexes.

An ultrasound of the head showed significant dilatation of the lateral ventricles with a predominantly echogenic mass in the suprasellar cistern. The mass appeared to encase the adjacent vessels. It had a small cystic component. An MRI of the brain showed a solid mass in the suprasellar cistern extending superiorly into the base of the third ventricle and displacing the cerebral peduncles. There was dilatation of the lateral and the third ventricles with transependymal spread of cerebrospinal fluid (Fig. 4a). A T1-weighted low signal intensity lesion was also seen in the fourth ventricular outlet adherent to the posterior surface of the medulla in the mid sagittal plane. There was intense homogeneous enhancement of the solid component of the mass in the suprasellar cistern as well as the mass in the posterior fossa (Fig. 4b). The preoperative diagnoses were that of either a germ cell tumour or a primitive neuroectodermal tumour (PNET).

At surgery, a mass in the suprasellar cistern adherent to the neighbouring structures with encasement of the middle cerebral artery was found. The mass was described to be woody in consistency. A small segment of the mass was resected for histopathology. The suprasellar mass and that in the posterior fossa were not operated in view of their location. Two
ventriculoperitoneal shunts were placed in the frontal horns of the lateral ventricles.

Postoperatively, the child recovered well except for one episode of seizure after which he was admitted to the hospital and treated accordingly. Postoperative CT and MRI scans showed no significant change in the size of the residual mass, no reduction in the size of the ventricles as well as the presence of bilateral subdural collections with postoperative changes.

A follow-up MRI after 8 weeks revealed no change in the size of the suprasellar and posterior fossa mass lesions, bilateral subdural collections, prominence of the cerebral sulci and metastases along the left tentorium and the left lateral aspect of the brainstem (Fig. 5).

The child died a month later of respiratory insufficiency and dysfunction of the hypothalamus.

Histopathological examination confirmed the mass lesions in both cases to be DIG. The first case showed a component of small primitive cells with a focally high mitotic rate of 4–5 per high-power field and a proliferative index of over 5% focally. The second case showed a mitotic rate of less than 1 per 10 high-power fields and a proliferative index of 2%. There was no focus of endothelial proliferation or tumour necrosis in either case.

**DISCUSSION**

Desmoplasic infantile ganglioglioma was recognized as a distinct clinicopathological entity in 1987, following a report of 11 cases by VandenBerg et al.\(^5\) They are rare tumours with only 60 cases being reported in the published literature.\(^5,6\) Desmoplasic infantile gangliogliomas are closely related to desmoplasic infantile astrocytomas (DIA), which were described by Taratuto et al. in 1984.\(^7\) Desmoplasic infantile gangliogliomas and DIA are classified by the World Health Organization on the category of low-grade superficially located desmoplasic neuroepithelial tumours of infancy.\(^6,8\) The DIA differs from the DIG in that the former does not have a neuronal component.\(^9\)

Desmoplasic infantile gangliogliomas occur in infants within the first 18 months of life, usually the first 4 months, and their large size at presentation suggests a fetal or perinatal origin according to VandenBerg et al.\(^5\) This tumour is uncommon and affects males more often than females with clinical features of a rapid increase in head size, seizures and/or paresis.\(^10\) They have a good prognosis with a survival of 8–20 years.\(^5,11–13\)

The majority of these are located in the frontal or parietal lobes, occasionally in the temporal lobe.\(^4,5,14\) Whilst VandenBerg

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**Fig. 3.** Case 1. Postoperative follow-up MRI: post-contrast T1-weighted sagittal image of the spine showing metastatic foci along the spinal cord.
A TARANATH ET AL. has reported a DIG with suprasellar involvement. Setty et al. have reported a DIA with suprasellar and hypothalamic involvement.\textsuperscript{11,15} They also suggest that multilobar involvement is frequent, with over 60% of reported cases showing involvement of more than one lobe.\textsuperscript{15}

The DIG are typically neuroepithelial tumours characterized by a voluminous size, intense desmoplasia, astrocytic and ganglionic differentiation. They have two distinct components: a solid component with intense desmoplasia that is always located close to the meninges and a cystic component, which is dominant and an integral part of the tumour.\textsuperscript{4,7,9,16}

Although Tseng et al. have described cystic changes in a purely solid DIG on follow up over a period of 18 months,\textsuperscript{17} Duffner et al. as well as Sperner et al. have published two cases of DIG that have been purely solid.\textsuperscript{14,18}

One should consider a number of pathologies such as PNET, typical gangliogliomas, supratentorial ependymomas, and cerebral astrocytomas in the differential diagnosis. Malignant teratomas and sarcomas can be considered in infants than in children.\textsuperscript{1} Primitive neuroectodermal tumours are deep seated, with calcification and cystic changes. Ependymomas
can involve the supratentorial neuroparenchyma, enhance heterogeneously and calcify. Rare meningoencephal astrocytomas attached to the dura and involving the superficial cortex have been reported by Taratuto et al. A published literature search reveals two cases: one a case of a hemispheric DIG with multilobar involvement/metastases at presentation and another one that was a suprasellar and hypothalamic DIA with posterior fossa and spinal canal metastases. In summary, our first case had typical features of a DIG with the mass in the left cerebral frontal, parietal and occipital regions showing a large cystic and a small cortically based, intensely enhancing solid component. The solid component was found to be adherent to the dura in the left frontal and parietal region. It was completely excised. Histology showed a few primitive cells, a focally high mitotic rate and proliferative index which could explain the metastases in the progress scans. The second case had a central, deep-seated mass, located in the suprasellar cistern, extending and adherent to the neighbouring structures with a dominant solid and a small cystic component. A nodule was seen adherent to the posterior surface of the medulla. Differential diagnoses of either a germ cell tumour or a PNET were considered. It is quite possible that the suprasellar mass in the second case could have been related to the dura. The nodule in the posterior fossa could represent a multifocal presentation of the DIG or a secondary deposit. Although it is possible that the inability to resect the tumour completely was the cause of the morbidity, the fact that there was a deposit at the fourth ventricular outlet and that the child developed metastatic deposits in the next few months suggest that this was a mass with inherently aggressive behaviour.

In conclusion, we have reported two cases of DIG that support the possibility that these tumours can have a varied biological behaviour and their benign nature needs to be questioned.

REFERENCES