Desmoplastic infantile ganglioglioma / desmoplastic infantile astrocytoma

Desmoplastic infantile astrocytoma (DIA) and desmoplastic infantile ganglioglioma (DIG) are massive, enhancing, cystic and solid, superficially located tumors usually found in the cerebral hemisphere. The majority occur in infants less than 24 months of age, with a median of 6 month

Old

Desmoplastic infantile gangliogliomas are a form of desmoplastic infantile tumours, and despite their aggressive appearances tend to have a good prognosis.

Epidemiology

The vast majority occur in children with less than one year of age, and males are more commonly affected with a M:F ratio of approximately 2:1

Clinical presentation

A rapidly increasing head circumference is the most common presentation, with symptoms usually presenting in a short time (5 days to 3 months).

Seizure activity is uncommon.

Radiographic features

Desmoplastic infantile ganglioglioma manifests as an exceptionally large cerebral hemispheric mass composed of both cystic and solid portions. The frontal and parietal lobes are the most common sites.

CT

The solid portion of these large masses is typically slightly hyperattenuating and usually located along the cortical margin of the mass. Following administration of contrast, these masses usually enhance intensely, and may demonstrate a dural tail.

Calcification is not a feature of desmoplastic infantile ganglioglioma.

MRI

The solid portions typically have the following signal intensity:

T1: isointense to brain parenchyma

T2: isointense to brain parenchyma

T1 C+ (Gd)

intense enhancement

a dural tail may be seen

Treatment and prognosis

Surgical resection is the treatment of choice; however, because of the large size of these lesions and the firm attachment to the dura, complete resection is difficult.

In cases of partial resection, adjunctive chemotherapy may be considered and have been reported to produced some reduction in tumour volume

Overall prognosis is good for most patients.

Desmoplastic infantile astrocytoma (DIA) is a rare, supratentorial, dural-based, large cystic tumor that usually arises in the first 24 months of life. However, non-infantile cases were also reported in the literature. Desmoplastic infantile astrocytoma and desmoplastic infantile ganglioglioma (DIG) are both classified as grade I astrocytoma by the World Health Organization (WHO). Grossly, DIA/DIG are large tumors composed of solid and cystic portions. Although large in nature, they are slow-growing tumors, with good prognosis after complete surgical removal, and rarely require a chemotherapy or radiotherapy. However, there have been few cases of DIA that demonstrated malignant features and/or spontaneous recurrence or metastases which necessitates close-up monitoring after surgical intervention. Herein, we report a case of an 18-month-old boy who presented with progressive head enlargement that was discovered to be due to a large left frontal predominantly cystic tumor. The patient underwent subtotal resection (STR) and was diagnosed as DIA on histopathological examination. Over a duration of 18 months of follow-up, the patient's status deteriorated, and he eventually died [1)].

1)
Samkari A, Alzahrani F, Almehdar A, Algahtani H. Desmoplastic infantile astrocytoma and ganglioglioma: case report and review of the literature. Clin Neuropathol. 2017 Jan/Feb;36 (2017)(1):31-40. Review. PubMed PMID: 27668845.