Desmoplastic Non-Infantile Ganglioglioma - A Case Report

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Abstract

Desmoplastic ganglioglioma is a rare markedly desmoplastic variant of ganglioglioma that usually presents in the first year of life. It is a mixed glial and cerebral neuronal tumour. A few cases of desmoplastic ganglioglioma have been reported in non infantile patients. We report a case of a 17 year old girl who presented with a right temporal lobe space occupying lesion in the outer third of right sphenoid wing enhancing well on contrast, attached to pia. Histological examination revealed a cellular glioneuronal tumour showing nuclear atypia. Atypical ganglion cells were present in the tumour. Immunohistochemistry revealed GFAP positive glial component and synaptophysin and chromogranin positive ganglion cells. This case confirms that desmoplastic ganglioglioma is an entity that can be seen even in young adults.

Introduction

Desmoplastic infantile gangliogliomas are rare, superficial, supratentorial tumours of early childhood i.e. they occur within the first two year of life.[1] Tumours with similar characteristics are exceedingly rare in the non infantile population.[1] We report a case of desmoplastic non-infantile ganglioglioma in a 17 year old girl. This case adds to the limited data available for desmoplastic ganglioglioma in the non infantile population.[1]

Case Report

A 17 year old girl presented with history of headache since one year and generalized tonic clonic seizures since 3 months. MRI demonstrated a right temporal lobe space occupying lesion in the outer third of right sphenoid wing enhancing well on contrast, attached to pia, causing mass effect and cerebral oedema (Fig. 1). On gross examination , the tumor was globular, grayish white measuring 3 x 2.5 x 2.5 cms (Fig. 2). Cut surface of the tumour was grayish white with haemorrhagic areas. Light microscopy revealed a cellular glioneuronal tumour showing lobules of dysplastic neuronal cells and multinucleate giant cells. Perivascular lymphocytic infiltrate and occasional mitosis were noted. Necrosis was not seen (Fig. 3). Reticulin staining demonstrated the reticulin rich glial component and desmoplastic stroma (Fig. 4). Immunohistochemistry demonstrated GFAP positive glial component and synaptophysin and chromogranin positive ganglion cells.The above mentioned morphological and immunohistochemical features are compatible with a desmoplastic noninfantile ganglioglioma .

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Discussion

Eleven examples of a distinctive paediatric tumour designated desmoplastic supratentorial neuroepithelial tumours of infancy (also known as desmoplastic infantile ganglioglioma) were originally described by Vandenberg *et al* in 1987.[2]

Desmoplastic infantile gangliogliomas are a distinct form of developmental neuroepithelial tumours probably arising from neural progenitor cells in subcortical zone along with mature subpial astrocytes.[3] They are rare WHO Grade I tumours of infancy characterized by large volume, superficial location, invariable supratentoriality, fronto-parietal lobe

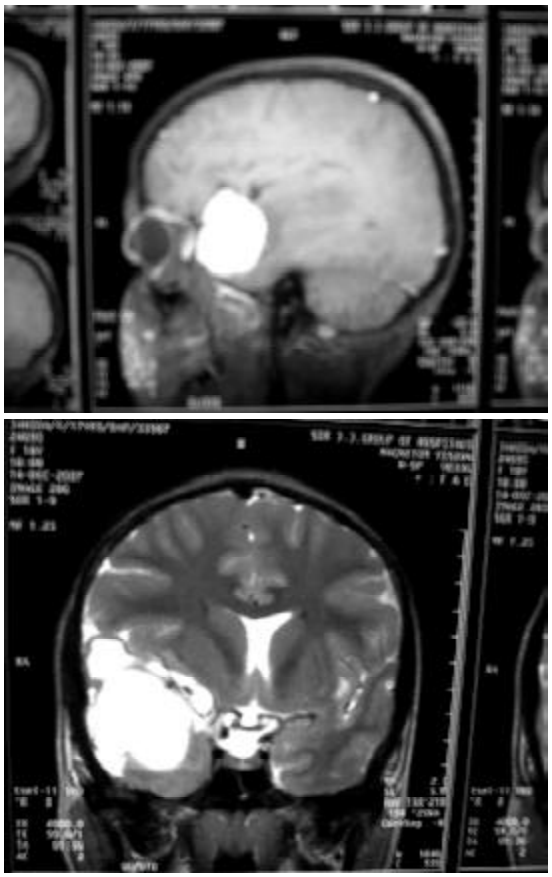

Fig. 1 : Radiologic images showing a contrast enhancing mass attached to the pia.

predilection and morphologically by an admixture of astroglial and neuroepithelial elements in a desmoplastic milieu.With over 50 cases described, the histologic and radiologic spectrum has been well characterized.[4]

Rare tumours with the same morphologic and radiologic features have been described in older subjects.[5] The patients present with an array of symptoms e.g. seizures,weakness and unsteady gait.[1,5] These tumours are generally localized in parietal or temporal lobes, present as a large cystic mass with peripheral contrast enhancement.[6] Histopathological examination reveals a well demarcated low grade glial tumour with

Fig. 2 : Gross photographs showing a globular gray white tumour.

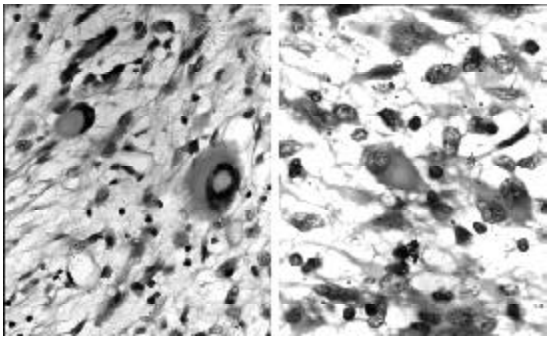

Fig. 3 : Microphotograph showing a cellular tumour with dysplastic neuronal cells (H and E 40X).

prominent desmoplasia. Ganglion cells with dysplastic features, clustered focally are also present. Perivascular lymphocytic cuffs and low mitotic activity are also observed.[7] Immunohistochemically, the glial components are GFAP positive while the ganglion like neuronal cells are positive for NSE, neurofilaments and synaptophysin.[6,7] Like infantile cases, noninfantile desmoplastic gangliogliomas seem to have a

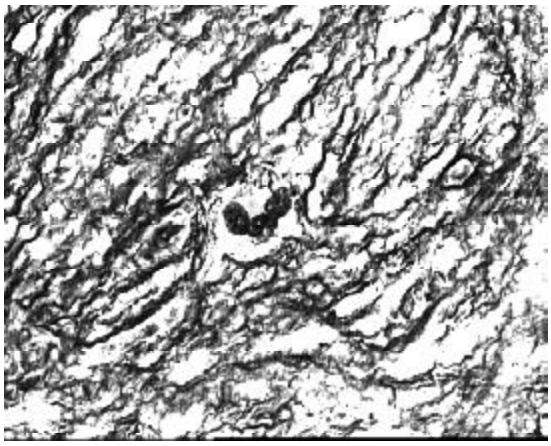

Fig. 4 : Microphotograph showing reticulin rich glial component and desmoplastic stroma (Retic 20X).

favourable prognosis without additional therapy, if a total surgical resection can be performed.[5] The identical radiologic and light microscopic findings in infantile and non infantile cases emphasizes on the fact that desmoplastic ganglioglioma can no more be considered a specific entity of infancy and must be well recognized even in young adults because it may be misdiagnosed as malignant glioma.[6]

In conclusion, although accepted as a tumour of infancy, desmoplastic ganglioglioma can also be encountered in older patients. Careful diagnosis and differentiation with other tumours particularly malignant gliomas is important since the therapeutic strategies may differ.

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RISK OF MYOCARDIAL INFARCTION AND NUCLEOSIDE ANALOGUES

In today's Lancet, the D:A:D Study Group analyses whether the nucleoside reverse transcriptase inhibitors zidovudine, didanosine, stavudine, lamivudine, and abacavir are associated with increased risk of myocardial infarction.

Participants in the D:A:D study who were receiving abacavir and didanosine had greater risk of coronary heart disease than those prescribed other nucleoside reverse-transcriptase inhibitors.

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