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Desmoplastic ganglioglioma

Report of a non-infantile case

Flávio Ramalho Romero[1], Sérgio Listik[1], Paula Annunciato Fabris[1]

Desmoplastic infantile gangliogliomas (DIGs) are rare, superficial, supratentorial tumours of early childhood i.e. they occur within the first two years of life representing 1.25% of all intracranial tumors in children[1,2]. Tumours with similar characteristics are exceedingly rare in the non infantile population[1]. These tumours, which are composed of a mixture of glial and neuronal cells and a fibrous stroma, affect mainly young patients and arise at the surface of the cerebral hemispheres[1,2]. Despite its histologically malignant appearance, DIGs are associated with excellent prognoses[1-3].

A 19 year old young man presented with history of a year old headache and generalized tonic clonic seizures 3 days before consultation. MRI showed a right temporal lobe solid-cystic lesion, causing mass effect and cerebral edema (Fig 1A). The patient was operated by craniotomy and micro neurosurgery and the lesion totally removed. Light microscopy revealed a cellular glioneuronal tumour showing lobules of dysplastic neuronal cells and multinucleated giant cells. Immunohistochemistry demonstrated GFAP positive glial component and synaptophysin and chromogranin positive ganglion cells.

Post operative MRI showed no evidence of residual lesion (Fig 1B). The patient and his parents agree with this report.

In 1982 Taratuto et al.[2] defined desmoplastic infantile astrocytomas as meningocerebral astrocytomas attached to the dura mater with a desmoplastic reaction. Five years later VandenBerg et al.[2,3] recognized a ganglion cell component in part of these tumours, and such tumours were called "desmoplastic infantile gangliogliomas".

Desmoplastic infantile gangliogliomas are a distinct form of developmental neuroepithelial tumours probably arising from neural progenitor cells in the 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 predilection and morphologically by an admixture of astroglial and neuroepithelial elements in

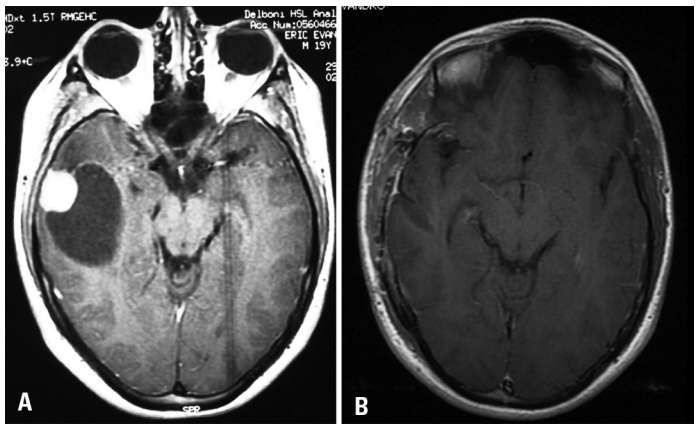

Fig 1. [A] MRI showing a right temporal solid-cystic lesion in Axial view. [B] Pos operative MRI in axial view.

Correspondence
Flávio Ramalho Romero
Rua Pascoal Vita 366 / apto 94
05445-000 São Paulo SP - Brasil
E-mail: frromero@ig.com.br
romeroncr@gmail.com

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GANGLIOMA DESMOPLÁSICO: RELATO DE UM CASO NÃO INFANTIL

[1]Neurosurgeon, Serviço de Neurocirurgia do Hospital São Luiz, Unidade Anália Franco, São Paulo SP, Brazil.

a desmoplastic millieu. 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 found in the parietal or temporal lobes, and present as a large cystic mass with peripheral contrast enhancement[4].

Histopathological examination reveals a well demarcated low grade glial tumour with prominent desmoplasia. Ganglion cells with dysplastic features, clustered focally are also present. Perivascular lymphocytic cuffs and low mitotic activity are also observed[5].

Immunohistochemically, the glial components are GFAP positive while the ganglion like neuronal cells are positive for NSE, neurofilaments and synaptophysin[4,5]. Like infantile cases, noninfantile desmoplastic gangliogliomas seem to have good prognosis without additional therapy, if a total surgical resection can be performed[5].

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. In this case, total tumor resection was made and no adjuvant therapy was necessary.

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