Gangliocytoma

Tumors with hypothalamic neuronal differentiation are classified as gangliocytomas or neurocytomas based on large and small cell sizes, respectively [1)]

Gangliocytomas are rare benign (WHO grade I) CNS tumours which differ from gangliogliomas by the absence of neoplastic glial cells. Both tumours are defined by the presence of displaced ganglion cells (large mature neurons that show cytological or architectural abnormalities).

On imaging these tumours are usually characterised by cortical solid lesions with little associated mass effect and minimal or no surrounding vasogenic edema. Calcification and cyst formation can occur, and contrast enhancement is generally present.

Terminology

It should not be confused with dysplastic cerebellar gangliocytoma, which is also known as Lhermitte-Duclos disease.

Epidemiology

They account for 0.1-0.5% of all brain tumours and occur in children and young adults.

Clinical presentation

Tumours in the cerebral cortex present most commonly with epilepsy.

Pathology

The key feature is a lack of glial cells. The tumour is composed of abnormal mature ganglion cells.

Markers

Lack immunoreactivity to glial fibrillary acidic protein (GFAP).

Associations

Frequently associated with a dysplastic and malformed brain to varying degrees.

Location

This tumour may arise anywhere within the neuroaxis. Occurs most frequently at the floor of the 3rd ventricle followed by the temporal lobe, cerebellum, parieto-occipital region, frontal lobe, and spinal cord.

Radiographic features

CT

Typically appears hyperattenuating on non contrast imaging.

Has little associated mass effect and minimal or no surrounding vasogenic oedema. Calcification and cyst formation can occur.

MRI

T1: solid components typically hypointense T2: solid components are typically mildly hypointense 2; cystic areas are hyperintense; calcification if present can be hypointense T1 C+ (Gd): solid components enhance Treatment and prognosis

They tend to grow slowly and do not undergo anaplastic change.

Differential diagnosis

Ganglioglioma

Cortical dysplasia

Dysembryoplastic neuroepithelial tumor (DNET).

Case reports

Mullarkey et al. reported 2 cases of medulloblastoma maturing into gangliocytoma after receiving multimodal therapy.

The first patient, an 11-year-old boy diagnosed with high-risk (non-WNT, non-SHH) medulloblastoma, was treated with near-total surgical resection followed by craniospinal irradiation therapy with weekly vincristine. He then received maintenance chemotherapy with vincristine, cyclophosphamide, and cisplatin. On surveillance MR imaging studies residual tumor in the lateral aspect of the tumor bed was noted to be slowly growing, eliciting gross-total resection of the residual tumor. Histopathology showed benign gangliocytoma without residual medulloblastoma. The second patient, a 3-year-old girl, was diagnosed with medulloblastoma, desmoplastic nodular variant. She was initially treated with gross total resection and chemotherapy with etoposide, carboplatin, and high-dose methotrexate. At 4 months off therapy, she was noted to have local recurrence along the resection cavity. Second-line therapy was started with irinotecan and temozolomide, but MRI assessment during treatment showed further disease progression. She then received craniospinal radiation. Eleven months off therapy, further radiographic progression was noted, and the patient underwent second-look surgery, with pathology showing gangliocytoma and treatment-related gliosis.

The maturation of medulloblastoma into a ganglion cell-rich lesion is very rare, with few well-characterized previous reports. Given the rare nature of this entity, it would be of great value to understand the process of posttreatment maturation and the genetic and treatment factors which contribute to this phenomenon [2)].

1)
Asa SL, Mete O, Perry A, Osamura RY. Overview of the 2022 WHO Classification of Pituitary Tumors. Endocr Pathol. 2022 Mar;33(1):6-26. doi: 10.1007/s12022-022-09703-7. Epub 2022 Mar 15. PMID: 35291028.
2)
Mullarkey MP, Nehme G, Mohiuddin S, et al. Posttreatment Maturation of Medulloblastoma into Gangliocytoma: Report of 2 Cases [published online ahead of print, 2020 Sep 3]. Pediatr Neurosurg. 2020;1-10. doi:10.1159/000509520