Unusual ganglioglioma with extensive calcification and ossification

Vikas Shashikant Kavishwar, Kirti G. Chadha, Shaikhali Moiz Barodawala, Anuradha Krishna Murthy

Department of Histopathology, Metropolis Healthcare Limited, Mumbai, Maharashtra, India

Address for correspondence:
Dr. Vikas Shashikant Kavishwar, Department of Histopathology, Metropolis Healthcare Limited, Central Laboratory, 4th Floor, Commercial Building - 1A, Kohinoor Mall, Vikhroli (West), Mumbai - 400 070, Maharashtra, India. E-mail: kavishwarvikas@gmail.com

ABSTRACT

Ganglioglioma is a slow-growing relatively low-grade mixed glioneuronal tumor with most cases corresponding to the WHO Grade I category. It frequently presents with seizures. The temporal lobe is the most common location followed by frontal, parietal, and occipital lobes. These generally behave in a benign fashion and have a favorable prognosis. We describe a case of a 24-year-old male presenting with convulsions and a calcified parieto-occipital mass. This mass removed from the parietal lobe showed neoplastic glial and dysplastic neuronal tissue amidst extensive areas of calcification and foci of ossification. On immunohistochemistry, the glial component expressed glial fibrillary acidic protein whereas the dysplastic neuronal component expressed synaptophysin and CD34. Epithelial membrane antigen was negative and Ki-67 showed a low proliferative index. After the surgery, the patient is free of neurological symptoms. Widespread calcification and ossification are very unusual in ganglioglioma, which prompted us to report this case.

KEY WORDS: Calcification, ganglioglioma, ossification

INTRODUCTION

Ganglioglioma corresponds to the WHO Grade I and the WHO classifies it in mixed glioneuronal tumors. It is a well-differentiated, slow-growing glioneuronal tumor composed of neoplastic mature ganglion cells and neoplastic glial cells. It may occur in any age group, temporal lobe being the most common location. In this patient, the calcified mass removed from the parietal lobe showed neoplastic glial and dysplastic neuronal tissue amidst extensive areas of calcification and foci of ossification. This type of widespread calcification and ossification is very unusual in ganglioglioma, which prompted us to report this case.

CASE REPORT

A 24-year-old Indian male working in the Middle East complained of repeated attacks of abnormal movements of the right side of the body for 1 week with an episode of loss of consciousness. He had a fall during these convulsions and injured his face and skull. There was no previous history of trauma. Speech and ophthalmologic examination was within normal limits. The clinical impression was tuberculoma.

Computed tomography and magnetic resonance imaging showed a calcified left parieto-occipital mass measuring 16 mm × 16 mm, which was hypointense on T1- and T2-weighted images and nonenhancing on postcontrast with mild perifocal vasogenic edema and erosion of the left inner table. There were no other lesions in the brain.

The differential diagnoses were (1) encephalomalacia/dystrophic calcification (2) low-grade nonenhancing neoplastic space occupying lesion. A spectroscopic examination of the aforementioned area was advised.

The surgeon noted a very unusual appearance of the lesion having three distinct components - a solid chunk of bony hard mass, an amorphous component, and typical soft glial element [Figure 1a]. The tissue was received as excised in three parts: tissue from thickened arachnoid, a calcified subcortical mass, and amorphous subcortical tissue.

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

How to cite this article: Kavishwar VS, Chadha KG, Barodawala SM, Murthy AK. Unusual ganglioglioma with extensive calcification and ossification. Indian J Pathol Microbiol 2016;59:395-7.
Sections showed lamellated calcospherites [Figure 1b] amidst cortical tissue. There was neuronal disorganization with occasional cytomegaly [Figure 1c]. Extensive calcification was also noted along the vessel wall. A distinct glial component was identified. Eosinophilic granular bodies were also seen [Figure 2a]. Admixed bony trabeculae were identified [Figure 1d]. No necrosis, abnormal mitoses, or anaplastic features were noted in any of the sections. Widespread calcification and formation of bony trabeculae are very rare findings.

Glial fibrillary acidic protein (GFAP), synaptophysin, CD34, epithelial membrane antigen (EMA), and Ki-67 (Labvision) immunohistochemical stains were performed using the UltraVision Quanto HRP detection system on the Thermo Scientific Lab Vision 480S Autostainer.

The glial component expressed GFAP [Figure 2d] whereas the dysplastic neuronal component expressed synaptophysin and CD34 [Figure 2b and c]. EMA was negative. Ki-67 showed a low proliferation index.

Hence, a diagnosis of ganglioglioma of WHO Grade I was made.

After the surgery, the patient is free of neurological symptoms.

**DISCUSSION**

Gangliogliomas are uncommon neoplastic lesions which represent 1.3–2% of all brain tumors.[1‑3] Nair et al. reported an incidence of 0.32% in their 5-year study of 1560 cases.[3] The age of patients is variable ranging from 2 months to 70 years. Data from a large series with a total of 626 patients indicate an average age at diagnosis ranging from 8.5 to 25 years, with only a slight male preponderance.[1] Our patient was a 24-year-old young male.

Majority of cases occur in the temporal lobe (>70%) and present with seizures.[1,2] These also occur in the frontal, parietal, and occipital lobes.[1‑3] In our case, the tumor was located in the parieto-occipital lobe and the patient presented with convulsions. Nair et al. also reported a case in the parietal lobe in their series of five cases.[3]

Intracranial calcification is a common finding in neuro-radiology, associated with a broad spectrum of disorders.[4,5] However, the morphology of a mixture of neuronal and glial cell elements as seen in our case is characteristic of ganglioglioma. In addition, abundant lamellated calcospherites and metaplastic ossification were also observed, which is unusual and rare. Makariou and Patsalides and Kiroglu et al. report that 35–50% of all gangliogliomas show calcification.[4,5] In the study by Nair et al., all cases showed calcification and perivascular lymphocytes.[3] Like us, Hori et al. also reported a case of ganglioglioma containing osseous tissue.[6]

On immunohistochemistry (IHC), neuronal proteins such as microtubule-associated protein 2, NeuN, neurofilaments, and synaptophysin are useful to demonstrate the neuronal component, and GFAP highlights the glial component. CD34 positivity is also observed in the neuronal component.[3] Our case showed a similar immunophenotype.

Heavily calcified localized tumefactive lesions in the brain have relatively limited differential diagnoses. Among the non-neoplastic lesions, meningiomatosis and vascular malformation calcifying pseudotumor of neuraxis can be considered, but display characteristic pathological features, which were not present in our case.[7]

Among the neoplastic lesions, psammomatous and rarely metaplastic meningioma are important differentials. The concentric lamellated calcification may coalesce and dense calcification can almost completely replace meningotheial...
cells.[8] Here, concentric lamellated calcification was seen, but there were no meningothelial cells, and tumor cells were negative for EMA.

Other primary intra-axial tumors known for extensive calcification include oligodendroglioma and less frequently subependymal giant cell tumor, ependymoma, choroid plexus papilloma, craniopharyngioma, neurocytoma, supratentorial primitive neuroectodermal tumor, and metastatic tumors.[4] However, histomorphology and clinicoradiological findings were not supportive of any of these.

Thus, based on the gross, histomorphology, and IHC, the diagnosis of ganglioglioma of parieto-occipital region of WHO Grade I was established. Because of its uncommon location, typical morphology but with abnormal ossification and calcification, and rarity of reports in world literature, we are eager to share this unusual case.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES