CASE REPORT

GRANULAR CELL ASTROCYTOMA

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Granular cell astrocytoma (GCA) is a rare glial neoplasm composed of abundant granular cytoplasm gives immunoreactivity for GFAP and S100 stains. We report a case of GCA in a 64 years old male presented with history of fits, right sided weakness and loss of consciousness. The microscopy showed sheets of large cells with abundant eosinophilic granular cytoplasm. No high-grade features were seen. Its differential diagnosis includes most of the benign histiocytic conditions. Granular cell astrocytoma has an aggressive clinical course and its survival rate is less than 1 year. That's why early correct diagnosis is very essential.

Keywords: Granular cell Astrocytoma, Immunohistochemistry, GFAP

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INTRODUCTION

A rare entity which has not been yet incorporated in World Health Organization Classification of central nervous system (CNS) tumours 2021 is Granular Cell Astrocytoma (GCA).[1] It has different histo-morphological pattern and despite having morphology resembling to benign granular cell tumours, it has an aggressive outcome.[1] GCA lacks the typical features of conventional astrocytoma because of which it is at times misdiagnosed. Earlier it was diagnosed as granular cell tumour of central nervous system.[2] The new name of granular cell astrocytoma was given in 2002. At molecular level it shares the genetics with glioblastoma.

We report a case of 64 years old male patient diagnosed as granular cell astrocytoma.

CASE DESCRIPTION

A 64-year-old male patient presented in emergency department with history of fits, dysphasia, right sided weakness and sudden loss of consciousness. He was admitted and after his laboratory workup imaging study was done. 2 mm reconstructed images from scan performed on multiple sixteen (16) slice Computed tomography (CT) scan were reviewed which showed 58x45 mm intra-axial heterogeneous density lesion with internal haemorrhage causing mass effect on the left lateral ventricle with contralateral midline shift which raises the suspicious of malignant lesion. The differential diagnosis lies between high grade primary glial tumours vs. metastatic disease. Magnetic Resonance Imaging scan (MRI) of the brain was also performed which showed intra-axial, hyperintense, ring enhancing space occupying lesion in left parietal lobe as shown in figure-1.

Resection of the tumour was done. After detailed histopathological and immunohistochemical workup as shown in figure 2a & 2b, the diagnosis of GCA was confirmed.

Biopsy was received in 10% formalin; it was routinely processed. On initial haematoxylin and eosin (H&E) staining sections reveal brain parenchyma infiltrated by sheets of large polygonal cells with abundant granular eosinophilic cytoplasm with occasional prominent nucleoli as shown in figure-2. No marked nuclear atypia or mitosis was seen. Numerous macrophages and capillary sized blood vessel were evident. Focal areas of necrosis and microvascular proliferation were also seen. Periodic acid Schiff stain was applied which is positive in cytoplasmic granules as shown figure-3.

Immunohistochemical panel of glial fibrillary acidic protein (GFAP), oligodendrocyte transcription factor (Olig2), S100, CD68, IDH-1 and EMA were applied. The tumour cells were positive for GFAP (figure4), Olig2 and S100. CD68 was positive in intervening macrophages. IDH-1 and EMA were negative. Ki-67 proliferation index was <5% in highest proliferation areas. On the basis of histo-morphological features and positivity of PAS, GFAP and Olig2 diagnosis of GCA was made.

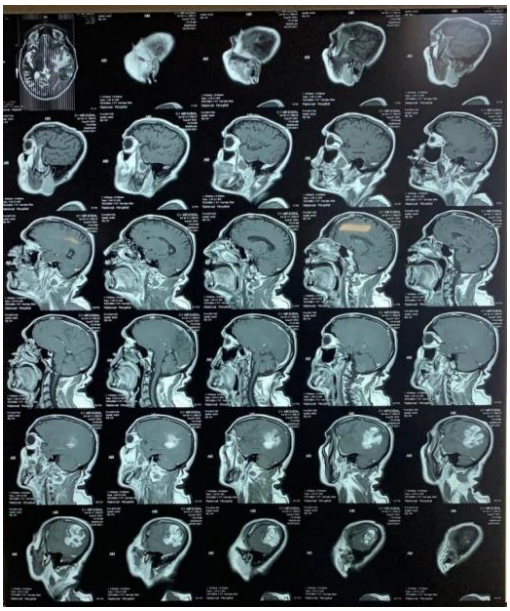

Figure-1: MRI Brain

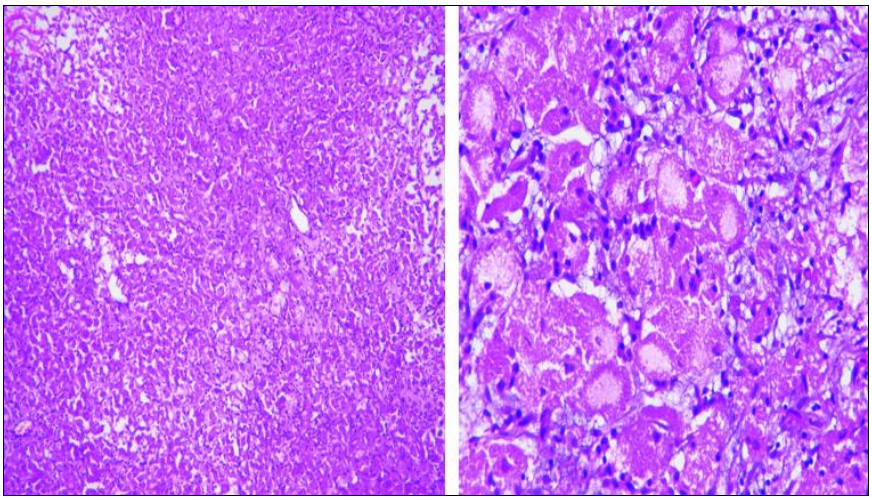

Figure-2a & 2b: H & E shows sheets of large cells with abundant cytoplasm 200x, 400x

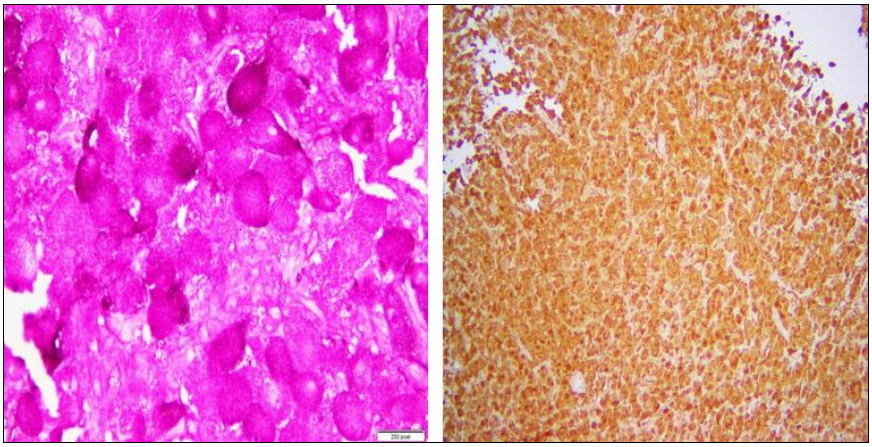

Figure-3 & 4: PAS stain highlights cytoplasmic granules and GFAP shows positive cytoplasmic staining

DISCUSSION

Granular cell astrocytoma is a rare infiltrating astrocytic neoplasm. So far no more than 70 cases have been documented in literarture.[1] Mean age of presentation is 50–60 years of life.[3] Some studies have shown predisposition for males.[1] The usual site of involvement is cerebral hemispheres.[4] GCA has aggressive behaviour despite absence of nuclear atypia or high-grade features. GCA has specific morphology characterized by large polygonal cells having abundant granular eosinophilic cytoplasm with bland nuclear morphology and PAS positive granules.[3] On basis of presence of granular cell component and abundant foamy macrophages, differentials also include inflammatory process, pleomorphic xanthoastrocytoma, histiocytic tumours. However, due to presence of typical infiltrating features of astrocytoma, larger cells than foamy macrophages with PAS positive granules and immunohistochemical markers help in reaching the final diagnosis.[2] GCAs are positive for GFAP, S100 and Olig2.Some studies has shown positivity of EMA[1] but in our case it was negative. Loss of IDH-1 was evident.In literature positivity of two stains olig2 and s100 with distinct morphological fearures aids in diagnosis of GCA.[5] Studies revealed that at genetics level GCA showed loss of 1P, 8p, 10q, TERT promoter mutations, EGFR amplification which are same as found in glioblastoma.[5,6] Electron microscopy show the presence of granules in GCA is due to intracytoplasmic lyosomes.[7] The pathogenesis is still not clear, it is assumed that GCA has taken its origin from astrocytes.[4] Treatment is same as for GBM which includes surgical resection, followed by radiotheraphy and chemotheraphy. Similarly, in our patient surgical resection of the tumour was done followed by radiotherapy. Two cycles of radiotherapy were done. Patient is followed at 3 months' interval.

In a nut shell, we reported a case of 64 years old male diagnosed with GCA which is a rare glial tumour with morphological features that can mimic granular cell tumor or histiocytic lesions. Therefore, detailed histological evaluation with immunohistochemical positivity of GFAP, S100 and negativity of IDH-1 mutation are important diagnostic points to confirm the diagnosis. GCA reveals the behaviour of WHO grade 2 to 4 and hence it is considered as tumour with aggressive behaviour and poor prognosis.[2,8]

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