Primary Focal Intracranial Leptomeningeal Glioma- Diagnostic and Management Dilemma

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DISCLOSURE

• I do not have any financial relationships with any commercial interest.

• Conflict of interest- None.

• The approval for conducting the study has been obtained from the Institute Ethics committee board.
INTRODUCTION

• Primary focal intracranial leptomeningeal glioma [PFILG] is considered as a rare solitary glial tumor arising from the leptomeninges without brain attachment or evidence of primary neoplasm elsewhere within the neuraxis.

• It has two clinical variants- focal and diffuse; diffuse variant is relatively common but the focal variant is extremely rare.

• We report a case of PFILG in the left parietal location and its clinico-radiological, histo-pathological and management characteristics.
CASE DESCRIPTION

• A 75 year old lady presented with focal motor seizures involving right upper limb with associated headache.

• MR imaging of the brain revealed a small, focal lesion attached to left parietal dura [Fig A,B] with underlying brain parenchymal signal changes and contrast enhanced images showed heterogeneous enhancement of tumor as well as meninges.

• The repeat MRI brain images within a month [Fig C] of primary imaging revealed significant progression of the size of lesion along with invasion of underlying parietal lobe parenchyma.
• She underwent gross total resection of the lesion [Fig D] and the histo-pathological diagnosis was glioblastoma multiforme [GBM], WHO grade 4, IDH wild type.

• The patient recovered well from surgery without deficits, however she refused to take adjuvant treatment.

• The MR brain imaging repeated 3 months after surgery revealed significant progression of the GBM with mass effect [Fig E,F].

• Though adjuvant treatment was then started, she could not tolerate it and succumbed to death 4 months after surgery.
• Tumor sections showed strips or fragments of a glial neoplasm intertwined with meningotheelial proliferation, abutting the dura.

• Ki 67 showed a high proliferation index, at 17%.

• IHC with NeuN antibody highlighted rare scattered neurons in one small fragment of the neoplasm which did not have dysplastic features and other fragments of neoplasm devoid of neurons.

• It also depicted the neurons in a fragment of cortex that shows infiltration with neoplastic cells both by Virchow-Robin spaces and by single cell invasion.
DISCUSSION

• The diagnosis of this lesion is challenging as its radiological and histological appearance resembles other CNS tumors.

• Cooper and Kernohan introduced the diagnostic criteria for the first time in literature: the absence of attachment to the brain parenchyma, absence of intra-axial lesions, and the presence of leptomeningeal encapsulation.

• Surgery and adjuvant radiotherapy/chemotherapy was reported as the cornerstone of treatment to reduce the tumor load and progression of disease.

• The prognosis of PFILG varies and the progression free survival can extend from months to years which may depend on various factors such as age of the patient, size and location of the tumor, cystic changes, presence of systemic metastasis, extent of resection and adjuvant treatment status.
SUMMARY

• PFILG is a rare entity with varying patterns of clinical and histological presentations.

• The possibility of high grade PFILG should be considered in any case of an aggressive extra-axial focal lesion.

• The final diagnosis can only be established on histological examination.

• Surgery is the mainstay of treatment.

• The effectiveness of adjuvant treatment using radiotherapy, chemotherapy or monoclonal antibodies are still unclear due to the rarity of the tumor.