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Primary CNS lymphoma- A case report

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Abstract
Primary CNS lymphoma is an uncommon form of extranodal Non Hodgkins lymphoma and consists of 2.4 to 3% of all brain tumours and 4 to 6 % of all extranodal lymphomas. The incidence has increased in past two decades in patients aged above 60 years. It can occur in both immunocompetent as well as immunocompromised individuals. The occurence in immunocompetent individuals are associated with diagnostic and therapeutic issues and the management is also different from other extranodal NHL. Primary CNS lymphoma can involve eye and CSF and so diagnostic assessment of these regions are also important.

Keywords: primary CNS lymphoma

Introduction
The primary CNS lymphoma is a rare extranodal Non Hodgkins lymphoma which is particularly confined within brain, eyes and cerebrospinal fluid without evidence of systemic spread. It differs from other lymphoma entities in several aspects. The clinical, neuroradiological and pathological features are important in diagnosing the disease as well as in its therapeutic facet.

Case Presentation
We would like to report a case of primary CNS lymphoma
A 46 years old man with systemic hypertension, presented with complaints of swaying while walking to either sides of 2 weeks duration. He also had associated recurrent vomiting and dull aching continuous holocranial headache. He has a history of blurring of vision in left eye 2 months before the onset of symptoms which was resolved completely. On clinical examination, his Montreal cognition score was 17/30. Impaired tandem walking and gait ataxia was noticed. Cranial nerves, motor system examinations were within normal limits. All hematological and biochemical investigations were within normal limits. Imaging revelaed evidence of extensive corpus callosal hyperintensity as well and T2 hyperintensity around the periventricular area with relative enlargement of the corpus callosum with only minimal contrast enhancement (Refer MRI images).In view of past history of possible optic neuritis, workup for autoimmune neurological conditions were done and found to be negative. CSF study was negative for malignant cells. In view of the contrast enhancement and the corpus callosal enlargement with MRS findings, possible CNS lymphoma was considered as the most possible etiology
Multiple soft grey brown tissue bits were received and histopathology of which revealed a malignant neoplasm of monotonous population of lymphoid cells with diffuse and perivascular arrangemet and having angiocentric infiltration, Tumour cells were positive for LCA, CD20 and BCL2 & BCL6 overexpression noted, histomorphologically suggestive of CNS lymphoma, B cell type.
Patient was started on chemotherapy and symptoms improved and is kept under follow up

Discussion
Primary CNS lymphoma is a form of extranodal Non Hodgkin lymphoma. It remains particularly confined to the CNS. Affects brain, leptomeninges and eyes. About 90% of primary CNS lymphoma are Diffuse large B cell lymphoma [1]. It is found in both immune compromised and immune competent individuals. The incidence has increased in immune competent patients [2]. Acquired Immunodeficiency syndrome is an important association with primary CNS lymphoma considering the immunocom promised patients, but the incidence in these patients has decreased due to development of newer antiviral drugs [3] It has a peak incidence in fifth to seventh decade in immunocompetent patients. Diffuse infiltrative growth pattern is associated with cognitive dys function, psychomotor slowing, personality changes and disorientation. This can present as single or multiple masses, more commonly at the periventricular brain tissue. MRI is the neuroradiological gold standard investigation which reveals hypointense enhancing lesions on T1 W images and hyperintense lesions and edema in T2W

images [4] The lesions may be circumscribed or infiltrative, pale with a homogenous cut surface sometimes with areas of hemorrhage and necrosis. [5] Histopathological diagnosis using a stereotactic biopsy is required for confirming the disease before starting treatment [6]. Histopathologically, the tumour is diffusely growing, cellular with geographical necrosis, angiocentric invasion [7]. Major classification systems include Revised European Amerian Lymphoma system, Kiel classification and Working Formulation of non-Hodgkin's Lymphomas for Clinical Usage. The Kiel subtypes include immunoblastic, centroblastic, lymphoblastic and unclassified. The molecular pathology include deletion of CDKN2A/2B and TP53 mutation [8]. Immunohisto chemically, tumour cells are positive for B cell markers, CD10 is only rarely positive. [9] This disease shows a favourable prognosis to both chemotherapy and radiation unlike other CNS tumours, but has a less overall survival compared to lymphomas of other sites [10].

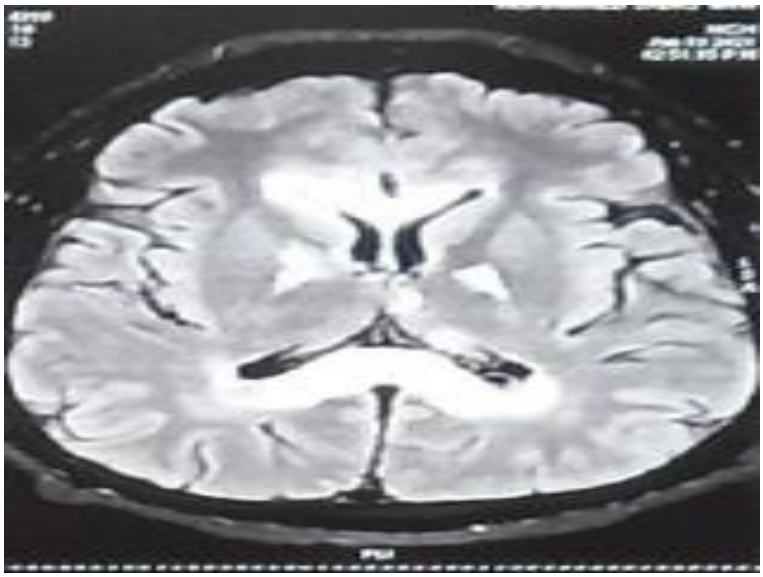

Fig 1: MRI showing extensive corpus callosal hyperintensity

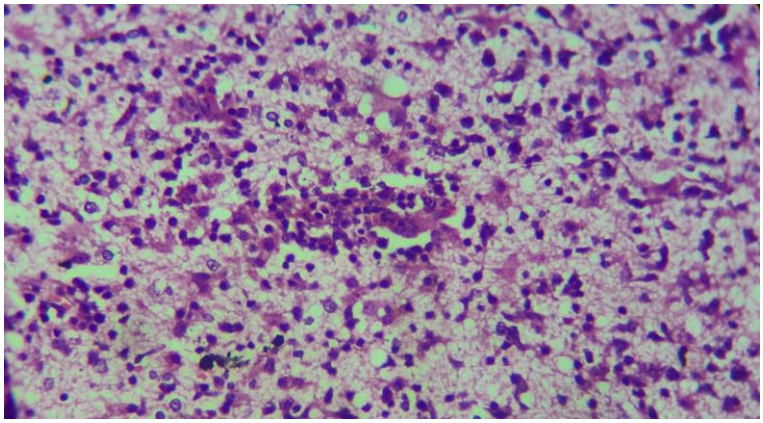

Fig 2: lymphoid cells in sheets

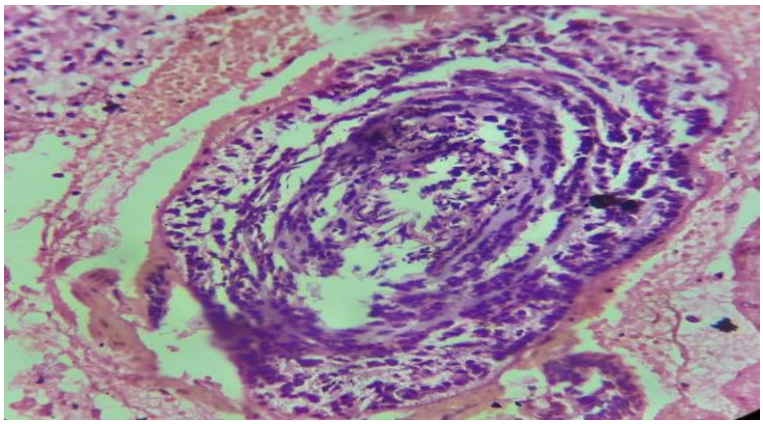

Fig 3: angiocentric infiltration

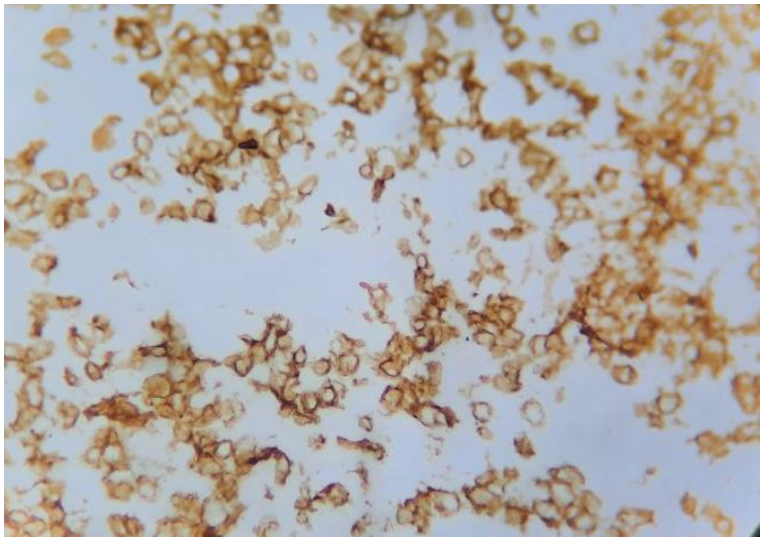

Fig 4: CD20 positivity

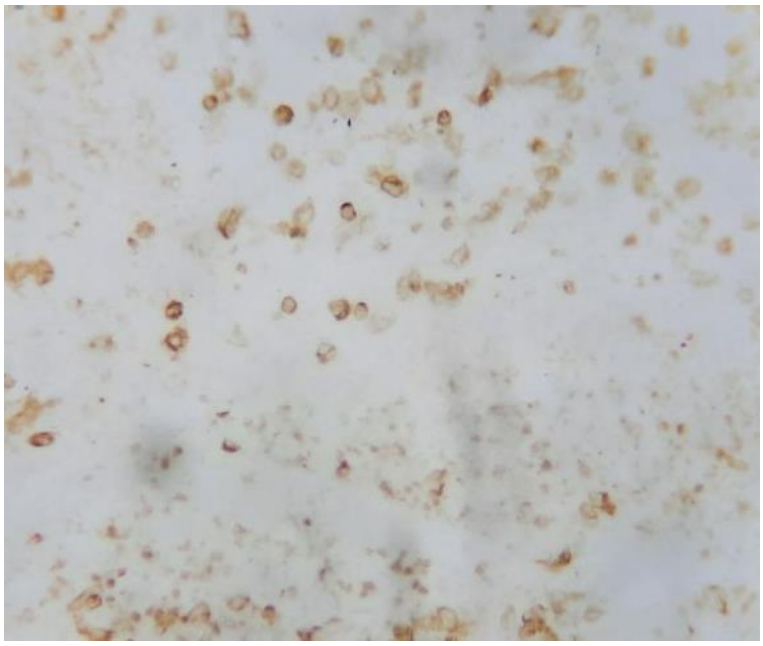

Fig 5: BCL2 overexpression

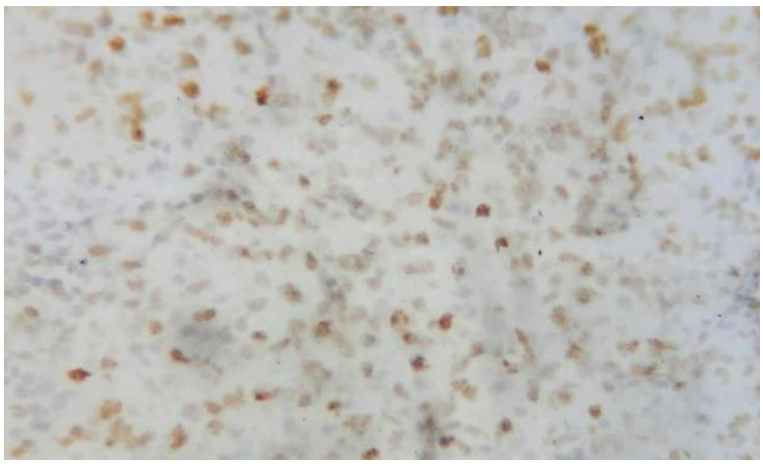

Fig 6: Bcl6 overexpression

Conclusion

In conclusion, we describe a case of primary CNS lymphoma in the periventricular region with relative enlargement of the corpus callosum. Clinicoradiologically, Possibility of Primary CNS lymphoma was considered because of the enhancement on radiology even though minimal. The pathologist should be aware of

the typical angiocentric infiltration pattern as well as the immunohistochemistry inorder to differentiate from systemic lymphoma

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