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Clinical and Surgical Insights into Primary CNS Lymphoma in Immunocompetent Patients

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ABSTRACT

Primary Central Nervous System Lymphoma (PCNSL) is a rare and aggressive extranodal non-Hodgkin lymphoma confined to the brain, eyes, and cerebrospinal fluid, with an incidence of 0.47 per 100,000 person-years. It accounts for 4%–6% of extranodal lymphomas and 4% of newly diagnosed CNS tumors. While commonly associated with immunocompromised states such as AIDS and organ transplantation, PCNSL in immunocompetent patients remains rare. The majority of cases are diffuse large B-cell lymphomas (DLBCLs), with other subtypes being extremely uncommon.

This case series presents five immunocompetent patients diagnosed with PCNSL at different intracranial locations, including atypical presentations in the cerebellum and cranial bones. Clinical symptoms ranged from focal neurological deficits, behavioral changes, and symptoms of increased intracranial pressure to localized swelling. Imaging revealed homogenously enhancing lesions, often in the supratentorial region, consistent with prior literature. All patients underwent surgical resection, primarily for diagnostic confirmation and decompression, followed by referral for oncological management. Histopathology confirmed high-grade B-cell lymphoma in all cases.

Our findings emphasize the variability in clinical and radiological presentations of PCNSL, necessitating a high index of suspicion for timely diagnosis. While the definitive treatment remains unclear, high-dose methotrexate combined with whole-brain irradiation is considered effective, and surgical resection can improve outcomes in select cases. This series highlights the need for multidisciplinary approaches to optimize management and improve prognosis in this challenging disease.

Keywords: *Primary CNS Lymphoma, Diffuse Large B-Cell Lymphoma, Immunocompetent Patients, Intracranial Tumors, Surgical Resection*

1. INTRODUCTION

Primary lymphoma of the central nervous system (CNS) is one of the rare causes of intracranial space occupying lesions. Primary CNS lymphoma (PCNSL) is a rare form of extranodal non-Hodgkin lymphoma that is typically confined to the brain, eyes, and cerebrospinal fluid without evidence of systemic spread. CNS lymphomas are commonly seen in immunocompromised patients. (1)

PCNSL occurs at an incidence of 0.47 per 100,000 person-years, accounting for 4%–6% of extranodal lymphomas and 4% of newly diagnosed CNS tumours. It is more common in males than females. (4)

PCNSL is an acquired immunodeficiency syndrome (AIDS)-defining illness. Organ transplantation is another risk factor, up to 2% of renal transplant patients and up to 7% of patients who have undergone cardiac, lung, or liver transplantation ultimately develop lymphoma in the CNS. EBV is also associated with 100% of PCNSL in patients living with AIDS.(1)

PCNSL in immunocompetent patients is rare and represents 4% of all intracranial neoplasms and 4% to 6% of all extranodal lymphomas. (4)

The majority of PCNSL cases consist of diffuse large B-cell lymphomas (DLBCLs). Other types include Burkitt's lymphoma, T-cell lymphoma, and low-grade malignant B-cell lymphoma but these are extremely uncommon. (2)

Patients with PCNSL develop neurologic signs over weeks, including focal neurologic deficits (56% to 70%), mental status and behavioural changes (32% to 43%), symptoms of increased intracranial pressure (headaches, nausea, vomiting, papilledema, 32% to 33%), and seizures (11% to 14%), depending on the site of CNS involvement. Imaging usually reveals a homogenously enhancing mass lesion, most often a single brain lesion (66%), with a supratentorial location (87%) and involvement of the frontoparietal lobes (39%).(3) Less frequently, eyes (15% to 25%),4 CSF (7% to 42%),5-7 and only in rare cases, the spinal cord are involved. (5)

The definitive treatment of PCNSL is unclear, but most studies have indicated that high-dose (≥3.5 g/m2) intravenous methotrexate (MTX) followed by whole-brain irradiation (WBI) can improve the outcome. The role of the monoclonal antibody rituximab remains controversial. (3)

Surgical resection might play a role in significantly improving overall survival (OS) and progressionfree survival (PFS) compared with stereotactic biopsy in a subset of patients. The type of surgery and tumour location are prognostic factors for PCNSL. (6)

Here we present 5 cases with different locations of PCNSL in immunocompetent patients. All were treated with surgical resection followed by referral to oncology department.

2. CASE SERIES

Case 1:

A 63-year-old male presented with complaints of giddiness for 2 months and left upper and lower limb weakness for 1 month. MRI Brain revealed an ill-defined, heterogeneously enhancing lesion with significant vasogenic edema involving the right corona radiata, right periventricular parietal region, and splenium of the corpus callosum on the right side, extending into the contralateral side. The lesion caused a mass effect and midline shift towards the left side with subfalcine herniation. The patient underwent a right parietal craniotomy with tumor excision.

Histopathology revealed diffuse large B-cell lymphoma.

Immunohistochemistry:

- CD20: Positive
- CD3: Highlighted reactive T cells in the background
- Ki67 labelling: 60-70%
- ALK, CD30: Negative

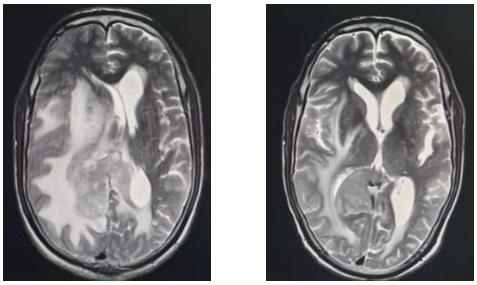

Fig. 1a Fig. 1b

Case 2:

A 23-year-old male presented with a painless swelling over the right parietal region for 3 months. The swelling was hard in consistency, had distinct borders, and was not associated with local temperature rise or tenderness. MRI Brain revealed a lytic mass lesion of the right parietal skull bone with a soft tissue component causing mild mass effect on the adjacent brain parenchyma (suspected Ewing's sarcoma).

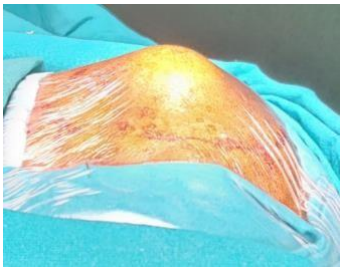
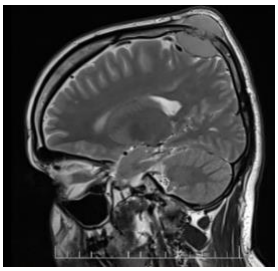
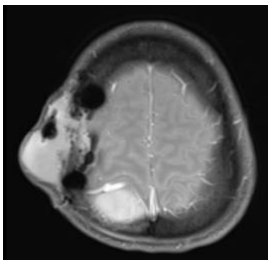

Fig. 2 Fig. 3a Fig. 3b

The patient underwent a right parietal craniotomy with tumour excision and bony reconstruction.

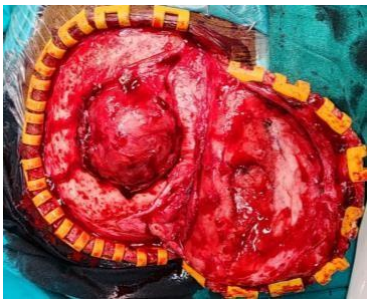
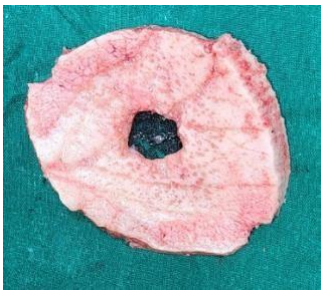
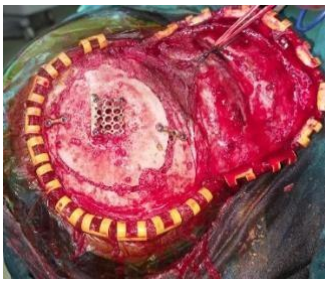

Fig. 4a Fig. 4b Fig. 4c

Histopathology showed fibrocollagenous tissue with a cellular high-grade lymphoid neoplasm composed of sheets of atypical lymphoid cells. These cells displayed moderately pleomorphic vesicular nuclei, clumped chromatin, visible nucleoli, moderate eosinophilic to clear cytoplasm, brisk mitosis, and bone trabeculae infiltration without necrosis.

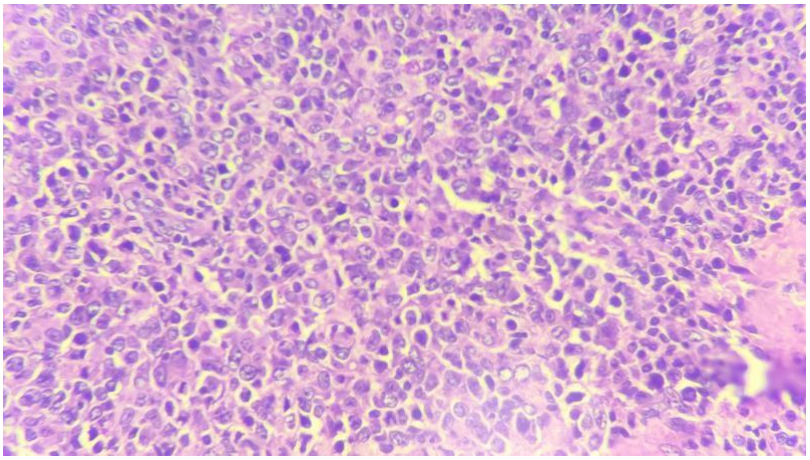

Fig. 5

Immunohistochemistry:

- CD20: Positive
- CD99, CD68: Positive in few perivascular cells
- CD3: Highlights reactive T cells
- FLI, S100, CD1a: Negative
- MIB-1 labeling index: >90%

Final Impression: High-grade B-cell Non-Hodgkin lymphoma.

Case 3:

A 67-year-old male, known diabetic, presented with giddiness, vomiting, and headache for 20 days. MRI revealed intensely enhancing mass lesions with significant perilesional edema likely arising from the choroid plexus of the left lateral ventricle and left thalamus. The lesion infiltrated adjacent brain parenchyma, causing mass effect and midline shift, suggestive of a

choroid plexus tumor.

The patient underwent left temporo-parietal craniotomy with transcortical decompression of the lesion under neuronavigation guidance. Histopathology revealed diffuse large B-cell lymphoma.

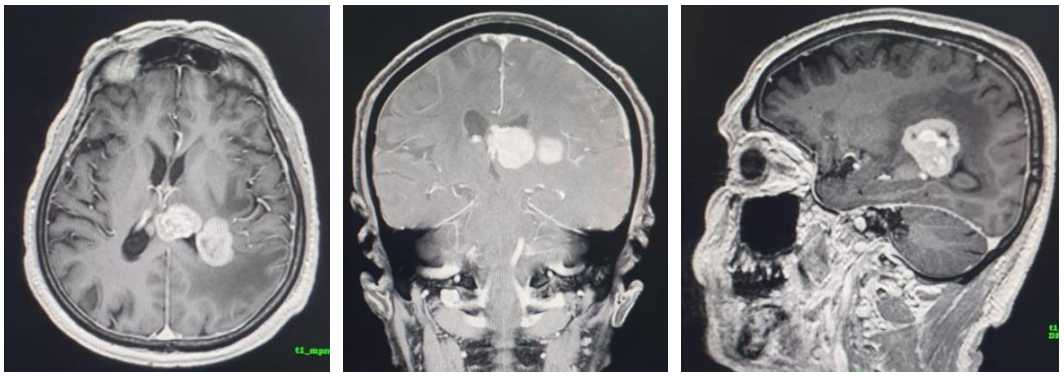

Fig 6a, 6b, 6c

Case 4

55-year-old male presented with headache, vomiting, and giddiness. MRI showed an avidly enhancing, well-defined mass lesion with a few central necrotic areas and surrounding perilesional edema in the right cerebellar hemisphere. The lesion caused significant mass effect. The patient underwent suboccipital craniotomy and microscopic subtotal excision of the right cerebellar lesion. Histopathology revealed diffuse large B-cell lymphoma.

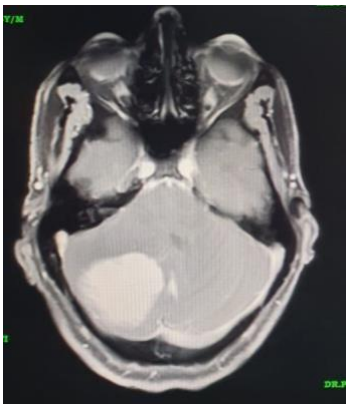

Fig 7

Case 5

A 40-year-old female presented with complaints of left upper limb numbness, headache, vomiting, and imbalance while walking. MRI showed intracranial lesions in the right frontal and thalamocapsular regions. The patient underwent right anterior frontal lobectomy. The sample was sent for biopsy.

Histopathology revealed high-grade Non-Hodgkin B-cell lymphoma.

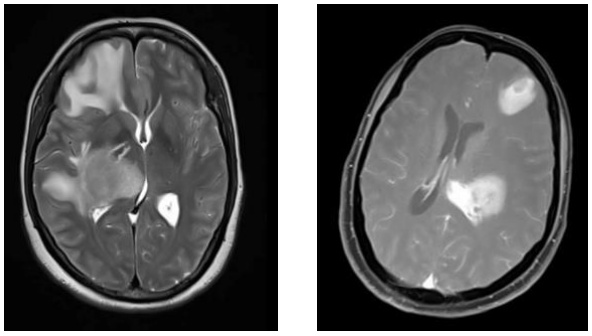

Fig. 8a, Fig. 8b

3. DISCUSSION

Primary central nervous system lymphoma (PCNSL) has an incidence rate of 0.47 per 100,000 person-years and accounts for 4%–6% of extranodal lymphomas and 4% of newly diagnosed central nervous system (CNS) tumors. It is more common in males than females. (4) PCNSL is considered an acquired immunodeficiency syndrome (AIDS)-defining illness, with a strong association to Epstein–Barr virus (EBV) in AIDS patients. Nearly 100% of PCNSL cases in AIDS patients are linked to EBV. Organ transplantation is another significant risk factor, as up to 2% of renal transplant recipients and 7% of patients who have undergone cardiac, lung, or liver transplants may eventually develop CNS lymphoma. (1)

In immunocompetent patients, PCNSL is rare, comprising approximately 4% of all intracranial neoplasms and 4%–6% of all extranodal lymphomas. (4) Interestingly, in our case series, none of the patients were immunocompromised. Additionally, literature specific to the Indian population suggests that immunocompromised status is less common among PCNSL patients. (7)

Histologically, the majority of PCNSL cases are diffuse large B-cell lymphomas (DLBCLs). Other subtypes, such as Burkitt's lymphoma, T-cell lymphoma, and low-grade malignant B-cell lymphoma, are extremely uncommon. (2)

Patients with PCNSL typically present with neurological symptoms that develop over several weeks. Common manifestations include focal neurological deficits (56%–70%), mental status and behavioral changes (32%–43%), signs of increased intracranial pressure such as headaches, nausea, vomiting, and papilledema (32%–33%), and seizures (11%–14%), depending on the site of CNS involvement. Imaging studies often reveal a homogenously enhancing mass lesion, with a single brain lesion being most common (66%), predominantly located in the supratentorial region (87%), particularly in the frontoparietal lobes (39%). (3) Other sites of involvement include the eyes (15%–25%), cerebrospinal fluid (7%–42%), and, rarely, the spinal cord. (5)

Most PCNSL lesions occur in the supratentorial region, commonly affecting the frontal lobe and periventricular areas. However, in our series, one patient had a lesion in the cerebellum and another in the cranium, which are relatively rare presentations. (8)

The optimal treatment for PCNSL remains uncertain. However, most studies suggest that high-dose intravenous methotrexate ($\geq$3.5 g/m^2) followed by whole-brain irradiation (WBRT) can improve outcomes. (3, 9) The role of rituximab, a monoclonal antibody, remains controversial. Surgical resection may play a role in improving overall survival and progression-free survival in a subset of patients, depending on tumor location and the type of surgery performed. In our case series, surgeries were primarily performed either due to a suspicion of glioma or other brain tumors or for decompression to improve the sensorium. (6) All the patients in our series were referred to the oncology department for further management following surgical intervention.

4. CONCLUSION

This case series highlights the clinical, radiological, and pathological features of Primary Central Nervous System Lymphoma (PCNSL), emphasizing its rarity and aggressive nature. Early diagnosis is critical, as timely intervention with treatments such as corticosteroids, chemotherapy, and radiotherapy can significantly improve patient outcomes. The wide variability in clinical presentations necessitates a high degree of clinical suspicion, particularly in patients with unexplained neurological symptoms or imaging findings suggestive of lymphoma.

Our findings also underscore the importance of a multidisciplinary approach, integrating advanced imaging techniques and histopathological evaluation to establish an accurate diagnosis. Future research should focus on refining diagnostic algorithms, exploring novel therapeutic options, and identifying prognostic biomarkers to optimize management strategies. For clinical practice, raising awareness about PCNSL and ensuring prompt referral to specialized centers can play a pivotal role in improving prognosis and survival rates.

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