Journal of Pathology of Nepal (2024) Vol. **14**, 2236 -2238

Journal of
PATHOLOGY
of Nepal
www.acpnepal.com

Case Report

Lymphoplasmacyte-rich meningioma: A rare case report

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Keywords:

Meningioma; Immunohistochemistry; Lymphoplasmacyte-rich

ABSTRACT

We report a case of 51-year-old female with lymphoplasmacyte-rich meningioma, which is a rare histological subtype of meningioma. It often mimics intracranial inflammatory lesions during its initial course. Hence, early and accurate recognition is essential for proper management. Histopathology is the gold standard, and immunohistochemistry aids in final diagnosis of this tumor.

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Received : May 20, 2024; **Accepted :** June 18, 2024

Citation: Mishra P, Nayak GD, Patra Y, Pradhan D. Lymphoplasmacyte-rich meningioma: A rare case report. J Pathol Nep 2024;14(2):2236-8. DOI: 10.3126/jpn.v14i2.66010

INTRODUCTION

Meningioma, the most common intracranial tumor originates from the arachnoid cap cells, the outer lining of the arachnoid layer of the brain. They account for about 39% of all primary non-glial brain tumors.[1,2] There are 15 histologic subtypes of meningioma categorized by recent WHO classification 2021 that have been further divided into 3 different histologic grades based on varied biological behavior. Lymphoplasmacyte-rich meningioma (LPM) is rare, benign, and falls under WHO CNS grade 1 of intracranial meningiomas characterized by the presence of

dense lymphoplasmacytic inflammatory cell infiltrates.[2,3] LPM occurs most commonly over cerebral convexities.[4] Herein, we report a case of LPM in a 51-year-old female because of its rarity, hence recognition is important to give proper management and support.

CASE REPORT

A 51-year-old female presented to the neurosurgery OPD with complaints of headache and neck pain in sub-occipital region for 3 months. It was associated with a tingling sensation. Clinical examination showed exaggerated lower limb reflexes along with reduced sensation below the C3 level. Lumbar puncture showed lymphocyte predominance in cerebrospinal fluid. CECT Neck showed an enhancing lobulated hypodense lesion of size 32x31mm in the right foramen magnum with extension posteriorly to the arch of the atlas and right occipital condyle with anterior extension to the medulla. MRI brain showed an avidly enhancing isointense, fairly marginated CV junction lesion extending along the foramen magnum T1 and T2 suggestive of meningioma (fig.1). Occipital craniotomy was done. A single large well circumscribed fibrous, non-suckable tumor was found upto C2 region. Total tumor was removed along with adequate dural margin and occipital bone. The tissue was sent for histopathological examination. Postoperative CECT brain revealed enhancing soft tissue lesion at the foramen magnum with erosion of the right occipital condyle, suggestive of a small residual lesion. Post-operative contrast-enhanced MRI revealed occipito-cervical-parietal plane collection. However, the rest of the brain parenchyma was normal. Squash cytology revealed cohesive syncytial clusters of cells having ill-defined boundaries. Individual cells were round to oval shaped, having round to oval nuclei, eccentrically placed nucleus, and few showing prominent centrally placed nucleoli. Grossly, multiple greyish brown tissue altogether measuring 4x3x1.5cm was received. Microscopy revealed small clusters of meningothelial whorls with numerous lymphoplasmacytic infiltrations along with increased vascularity and numerous histiocytes. No area of hyper-cellularity, necrosis, mitoses, or any atypical cells were seen (fig.2). The diagnosis was further confirmed by immunohistochemistry where tumor cells showed positivity for EMA (fig.3a) and CD68 (fig.3b) along with CD3 positivity in infiltrating T-lymphocytes (fig.3c). MIB-1 index was 4% (fig.3d). A final diagnosis of LPM (WHO Grade I) was made.

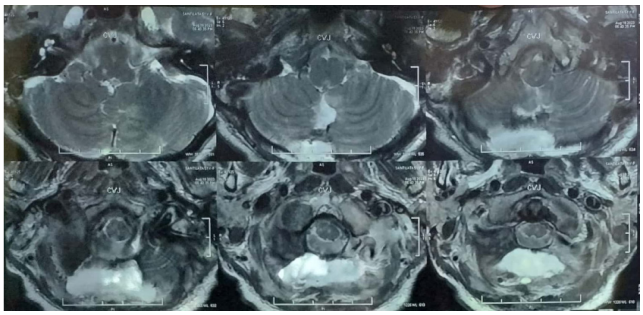

Figure 1: *Contrast MRI showing homogenously enhancing lesion with extension superiorly into the foramen magnum*

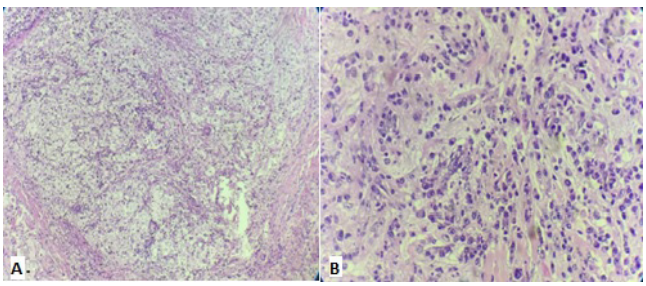

Figure 2: *A) Tumour cells forming whorls surrounded by numerous lymphocytes, plasma cells and histiocytes.(H&E stain 40x). B) Meningothelial cells in whorls (H & E stain 100x).*

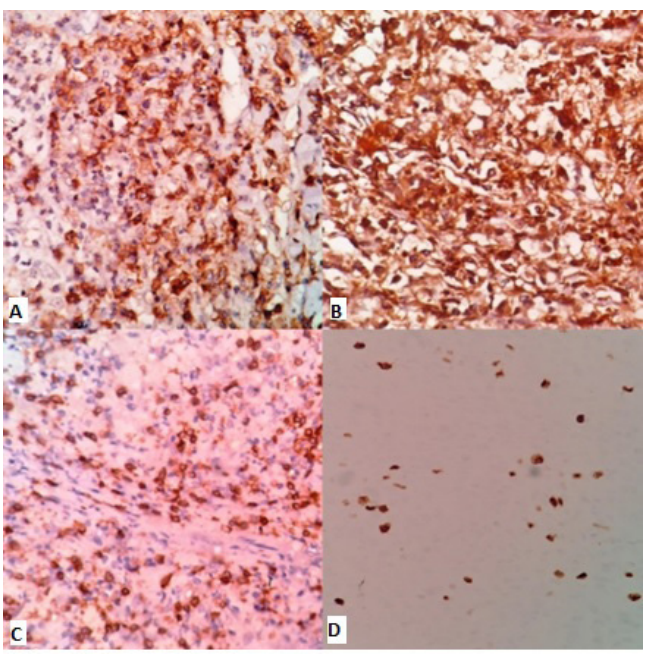

Figure 3: *A)Tumor cells showing strong cytoplasmic staining for EMA (IHC: 400x). B) Strong cytoplasmic staining for CD68 in the histiocytic component of the tumor (IHC: 400x). C) Cytoplasmic staining of CD3 in infiltrating T-lymphocytes (IHC: 400x). D) Nuclear staining for MIB-1 with labeling index 4% in highest proliferating area of tumor (IHC: x 400).*

DOI: 10.3126/jpn.v14i2.66010

DISCUSSION

LPM is categorized under WHO CNS grade 1 meningiomas. It is a rare pathologic variant with an estimated incidence of less than 1% of all meningiomas.[5] LPM can occur at any age with higher incidence seen amongst young and middle-aged adults, predominantly affecting females.[6,7] LPM as a separate entity was added to the WHO classification in 1993 however it was first originally reported by Banerjee and Blackwood in 1971.[6-8] Majority of LPM are localized along the cerebral convexities, skull base, sphenoid ridges, olfactory grooves, parasellar region, tentorium, falx and posterior fossa. It usually presents with headache, hemiparesis, seizure, and vomiting, visual disturbance. The origin, whether neoplastic or inflammatory is still not known; and it is usually confused as an intracranial inflammatory mass rather than meningioma in resource-poor settings. LPM may show prominent peripheral blood abnormalities like anemia, and/or polyclonal gammopathies, which usually do not persist after surgical removal of the tumor. MRI imaging usually shows multiple linear irregular enhancements extending to adjacent brain parenchyma along with the presence of peritumoral brain edema suggesting an inflammatory origin, which explains the clinical presentation not usually seen in other variants of WHO grade 1 meningiomas.[9] These tumors present as hyperdense lesions on non-contrast CT scans and may show restricted diffusion on MRI due to the high cellularity because of dense lymphoplasmacytic infiltration. Cyst formation in adjacent brain parenchyma is seen in about 30% of cases due to inflammatory infiltrates.[10] Histopathologically, LPMs are characterized by excess infiltration of lymphocytes, plasma cells and numerous histiocytes.[9,10] LPMs are benign in nature, and hence do not show any clinical or pathological signs of invasion with only one third of diagnosed cases showing a proliferation index higher than 3%.[6] The differential diagnoses include granulomatous disease, histiocytic tumors, inflammatory pseudotumor, hypertrophic pachymeningitis, and internal hemorrhagic pachymeningiosis (IHP). On immunohistochemistry, tumor cells show strong immunoreactivity for EMA, hence ruling out other intracranial lesions and narrowing down to meningothelial tumors. Intracranial plasma cell granulomas and dural plasmacytomas are also the considered differentials and are difficult to differentiate at histopathological level. Inflammatory pseudotumor is characterized by the presence of myofibroblastic spindle cells along with lymphoplasmacytic infiltrate. IHP shows diffuse lamellar thickenings rather than a localized nodular involvement. Moreover, inflammatory fibrous histiocytic tumors, and sinus histiocytosis with massive lymphadenopathy (Rosaidorfman's disease, RDD) are also considered as differentials. RDD shows large histiocytes with abundant faintly eosinophilic cytoplasm with prominent nucleoli and also shows emperipolesis, hence ruling out LPMs. Therefore, comprehensive evaluation along with radiologic and histopathological findings are necessary to rule out LPM. Gross total resection of the tumor is the treatment of choice, followed by long-term clinical, and radiological follow-up, since the prognosis of these tumors is still uncertain despite its benign nature.

CONCLUSIONS

Lymphoplasmacyte rich meningioma is a rare variant of meningioma. It is necessary to differentiate it from all intracranial inflammatory conditions which tend to mimic its initial presentation. Pathologists need to be well aware of this entity. Recognition of this rare variant important to ensure proper management.

Conflict of interest: None

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