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CASE REPORT

Lymphoplasmacyte-rich meningioma with invasion of bone: A case report and review of literature

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ABSTRACT

Lymphoplasmacyte-rich (LPR) meningioma is a rare variant of meningioma, which is characterized by conspicuous infiltration of plasma cells and lymphocytes and a variable proportion of meningothelial elements, and is classified as a grade I tumor in World Health Organization (WHO) classification of tumors of central nervous system. The origin and biological behavior of this rare variant of meningioma is still not clear. Till date, very few cases of LPR meningioma have been reported globally. Here, we are presenting a case of right parietal convexity LPR meningioma with invasion of bone in a 32-year-old male patient, who presented to us with complaints of focal seizures and weakness in left upper limb.

Key words: Grade I, invasive, lymphoplasmacyte-rich meningioma

Introduction

Meningiomas are common tumors of the central nervous system and originate from the meningeal coverings of the spinal cord and the brain. They account for about 13-26% of all primary brain tumors.[1,2] About 80% of all meningiomas are slow-growing tumors of World Health Organization (WHO) grade I, which can sometimes display aggressive behaviors such as invasion to brain, dura, and adjacent bone, and in 2.3-30% of histologically benign grade I tumors, meningiomas can recur following microscopically complete resection along with other involved structures.[3,4]

Lymphoplasmacyte-rich (LPR) meningioma is a rare, benign variant of meningioma, which is characterized by conspicuous infiltration of plasma cells and lymphocytes and a variable proportion of meningothelial tumorous elements, and is classified as grade I tumor in WHO classification of tumors of central nervous system. The origin and biological behavior of this rare variant of meningioma is still not clear. Till date, only 27 cases of LPR meningioma have been reported globally.[5] Most of the reported cases were benign in nature with only few case reports of invasion of brain parenchyma. We are presenting a case of LPR meningioma with invasion of bone in a 32-year-old male patient.

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Case Report

A 32-year-old patient presented with complaints of weakness and focal seizures involving left upper limb for 6 months duration and headache for 1 month duration. On clinical examination, the patient had weak elbow, wrist movement, and weak hand grip in the left side. Magnetic resonance imaging (MRI) brain was done which showed a large homogenous solid extra axial mass in the right parietal region with hyperostosis and sclerosis of adjacent calvarium and marked edema of underlying right fronto-parietal lobes and right lateral ventricle with midline shift of brain toward left-s/o meningioma [Figures 1-3]. Gross total removal of tumor along with adequate dural margin and parietal bone was done. Overlying parietal bone was found to be thickened and soft, and was invaded by the tumor. Macroscopically the tumor was firm, cauliflower shaped, moderately vascular with a broad dural base, with clear intervening plane between the tumor and cortical brain parenchyma, and preserved underlying sulcal and gyral architecture, although the underlying cortical surface was compressed [Figure 4]. Histopathologic examination revealed meningeal whorls with numerous lymphocytic infiltrations [Figure 5]. The inflammatory component was dominating the lesion and obscuring the neoplastic component. Several germinal centers and pink homogenous eosinophilic material with numerous foreign body type giant cells were seen. Immunohistochemistry revealed epithelial membrane antigen (EMA) positivity in the meningeal whorls [Figure 6].

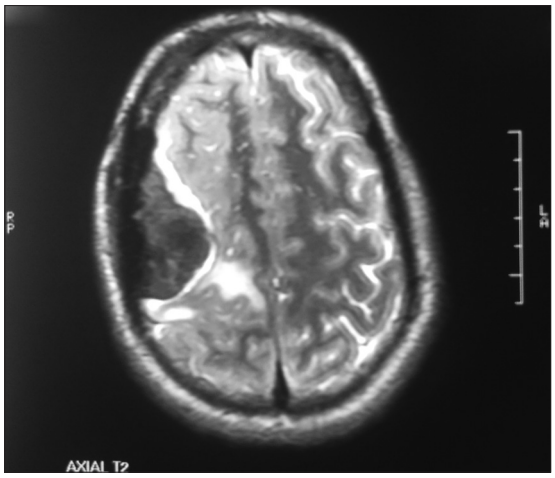

Figure 1: Axial T2 image of MRI brain

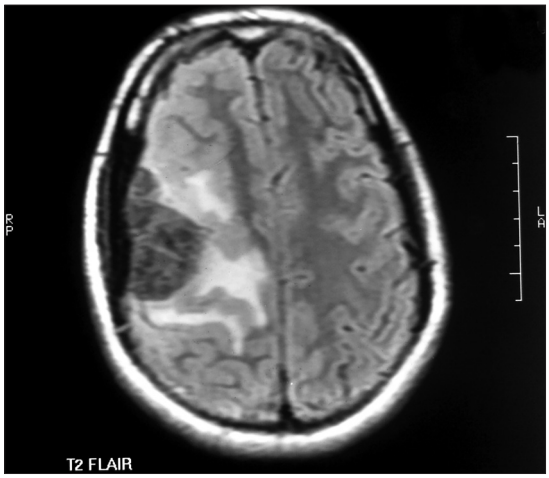

Figure 2: Axial T2 FLIAR image of MRI

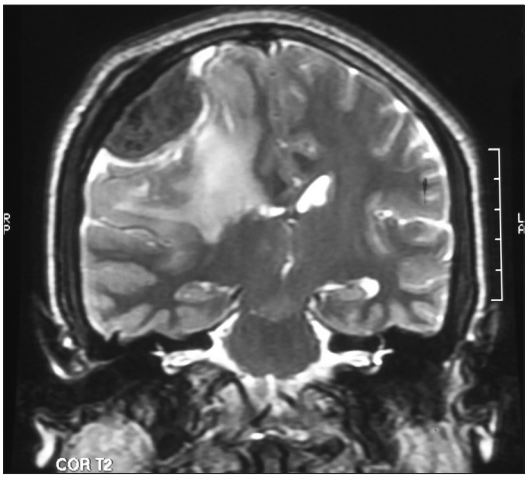

Figure 3: Coronal T2 image of MRI

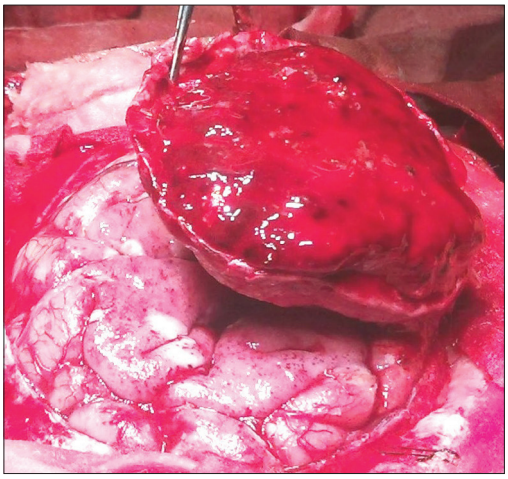

Figure 4: Intraoperative image showing cauliflower-shaped tumor with broad dural base, with clear intervening plane between tumor and cortical brain parenchyma and preserved underlying sulcal and gyral architecture

The meningothelial component was also strongly positive for vimentin [Figure 7] and negative for CD138 and glial fibrillary acidic protein (GFAP). The Ki-67 antigen/MIB-1 monoclonal antibody index was approximately 0-1%. A final diagnosis of LPR meningioma WHO grade I was made.

Discussion

The definition of LPR meningioma was established by the WHO in their new histological classification.[6] First described by Benerjee and Blackwood in 1971, LPR meningioma is one of the rarest variants of meningioma and was thought of as a collision tumor between a meningioma and a plasmacytoma.[7] Rubinstein (1977) studied three examples of this peculiar type of meningioma and favored the interpretation that the infiltration of plasma cell is a secondary character. In 1979, Horten *et al.* described five cases of this type of meningioma and noted various histological features and observed that the plasma cell component is not neoplastic. Thus, they chose to consider these cases as meningiomas with prominent plasma cell–lymphocytic infiltration, which may be a mechanism of host resistance to the tumor. They also noted its occasional association with hypergammaglobulinemia.[8] In 1980, Stam *et al.* performed immunohistochemical analysis which revealed the polyclonal nature of plasma cell component and regarded them not to be neoplastic and described the plasma cell infiltration is secondary in nature.[9] Immunohistochemical assays indicated that the lymphoplasmacellular proliferation probably represents an immune reaction of the host.[10] However, the relationship between inflammatory lesions and meningiomas remains uncertain.

LPR meningioma can occur at any age, but more often has been described in children and young adults. This type of meningioma is usually accompanied by prominent peripheral blood abnormalities, anemia, and/or polyclonal gammopathy, which usually disappear after surgical removal of the tumor. LPR meningioma has been described to occur in various locations that include cerebral convexity, falx, cerebellopontine (CP) angle, foramen magnum, and spinal cord.

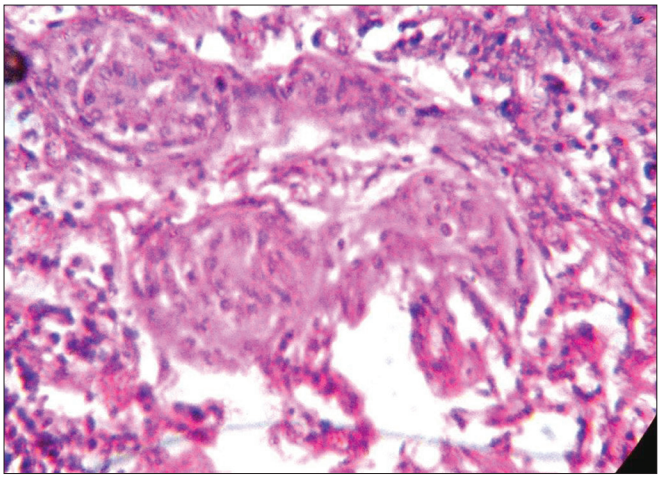

Figure 5: Meningothelial cells forming whorls with massive plasma cells and lymphocytic infiltration (high power, H and E, x40)

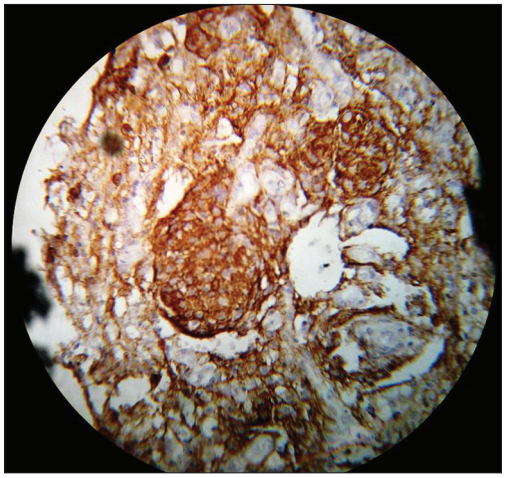

Figure 6: EMA-positive meningothelial cells (high power, x40)

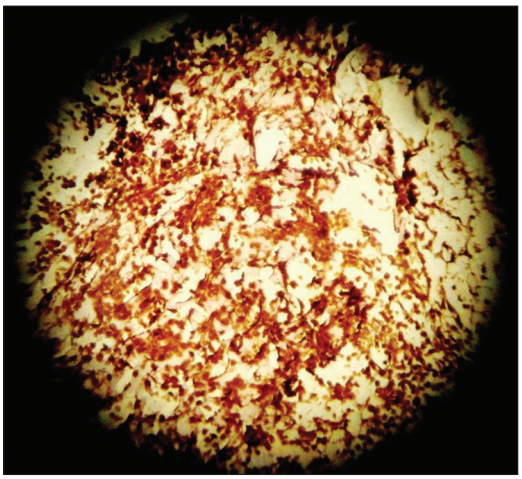

Figure 7: Strongly vimentin-positive meningothelial cells (low power, x10)

Clinical manifestations of LPR meningioma are usually related to the site of occurrence, which include seizures, paresis, and cranial nerve palsies; nonspecific symptoms like memory disturbances and headache have also been described.

Although peritumoral edema is said to be characteristic of LPR meningiomas, the diagnosis of LPR meningioma would be hard to establish preoperatively because peritumoral edema is not so rare in other types of meningiomas.[11] Brain infiltration may be essential for the development of peritumoral brain edema.[12]

Although there are reports of LPR meningioma with invasion to brain parenchyma,[8,13] till date, no report has been found in literature citing invasion of this tumor to the bone.

Histologically, LPR meningioma is characterized by the presence of a prominent polyclonal infiltrate of histologically benign lymphocytes and plasma cells. Germinal centers, Russell bodies, and amyloid deposits may also be present. The proportion of the tumor which is recognizable as meningothelial or transitional meningioma varies from case to case, and may be hard to detect. The most commonly used marker in meningioma diagnostics is EMA which yields at least patchy positivity in most meningiomas. Vimentin staining can also be helpful, as all meningiomas strongly express this intermediate filament. The major phenotypic changes in the transformation of meningiomas from the classic to the anaplastic type are loss of meningioma architecture, decreased expression of EMA, increased expression of vimentin, and metaplastic expression of α-internexin and neurofilament proteins.[14] Proliferative index as measured by Ki-67/MIB index indicates the risk of recurrence.

Treatment options advocated are the same as for grade I meningioma. Simpsons grade I resection is ideal where possible.

Prognosis of LPR meningioma is not clear, although recurrence after complete resection has not been reported till date. The significance of plasma cells and lymphocytic infiltration is still not clear. No protective action afforded by this lymphoplasmacytic infiltration has been noted. The biological behavior of these meningiomas seems to depend not upon the infiltrates, but rather upon the meningiomatous component.[8] Factors considered to have prognostic significance include sheeting, hypercellularity, cytologic atypia, increased mitotic index, necrosis, small cell change, brain invasion, and elevated proliferative index of MIB-l. Extent of resection is also a powerful prognostic indicator.

In our case, diagnosis of LPR meningioma was done on histopathologic examination of the resected specimen. A search for associated hematological abnormality was done. Our patient did not have anemia, and protein electrophoresis did not show M band or hypergammaglobulinemia, although albumin and gammaglobulin were toward the higher side of normal reference value. Postoperative MRI scan was done 2 months after surgery, which showed only postoperative changes and no residual/recurrent mass. Patient remained

clinically well and seizure free during 3 months of follow-up period.

Conclusions

LPR meningioma is a rare variant of meningioma, which is classified as WHO grade I meningioma. As the significance of lymphoplasmacytic infiltration of this type of meningioma is still not clear and invasive nature of these tumors has been noted, a close watch and awareness of this condition is needed.

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