Indolent extranodal marginal zone lymphoma of the meninges producing chronic cerebral venous occlusion

Sir,

Primary meningeal lymphoma is very rare. We would like to report a case with some unusual features. A 65-year-old diabetic and hypertensive man presented to us with dressing difficulty. On examination, he had dressing and constructional apraxia and right parietal skull bossing. MRI showed a right temporo-parietal en-plaque meningeal lesion that was isointense to the parenchyma on T1WI, enhanced intensely with gadolinium and was hyperintense on T2WI [Figure 1]. MR venogram demonstrated near-occlusion of the right transverse and sigmoid sinuses [Figure 2]. CT scan (bone windows) showed parietal skull bossing without hyperostosis [Figure 3]. CSF, blood parameters, LFT, RFT, HIV (ELISA), PSA level, ESR [20 mm/hour], ultrasound abdomen, and colonoscopy were normal. The patient remained asymptomatic for the next 3 years after which left hemiparesis recurred. ESR was 60 mm/hour. Serum protein electrophoresis showed an M band. MRI brain showed a larger lesion with underlying mass effect.

An open biopsy revealed a tumor composed of sheets of small lymphoid cells with clear cytoplasm, round to irregular nuclei, clumped chromatin, and inconspicuous nucleoli. Many lymphocytes had a monocytoid appearance. There were scattered large lymphoid cells with scant cytoplasm, vesicular nuclei with prominent nucleoli and plasma cells with Russel bodies. Tumor cells
he was asymptomatic.

The CNS is more commonly involved with secondary lymphoma. 5-10% patients with disseminated NHL show leptomeningeal or parenchymal masses.\textsuperscript{[1]} Primary CNS lymphoma (PCNSL) is less common (~1% of all brain tumors). PCNSL involves the parenchyma more frequently than the meninges (12:1).\textsuperscript{[2]} The majority of PCNSL are high-grade B-cell NHL, with a poor prognosis. The MZL variant represents <1% of all PCNSL, but is associated with a good prognosis.

Our patient was unusual in three respects. This is the first case of primary meningeal lymphoma (PMeL) reported from India. Secondly, he developed parietal skull bossing overlying the tumor. Thirdly, our patient is one of the few men described with PMeL belying the overwhelming female predominance with PMeL tumor.\textsuperscript{[3]}

Extranodal MZL are a subgroup of MZL arising from mucosa associated lymphoid tissue (MALT). They display surface immunoglobulin (Ig) of the IgM subtype with only B-cell markers (CD19+, CD20+, CD22+). In the CNS, the arachnoidal meningoepithelial are analogous to MALT.\textsuperscript{[4]}

Clinical features of PMeL include headaches, visual field defects, hemiparesis, seizures, and cranial nerve palsies. PMeL is often indolent. Radiologically they present as en-plaque meningeal lesions, the differential diagnosis of which is vast. Treatment options include local resection, chemotherapy, or radiotherapy. In 30% of patients, disseminated disease is detected (such as the M protein in our patient) although prolonged remissions are still the norm.\textsuperscript{[5]}

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References


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