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Primary intracranial meningeal marginal zone B cell lymphoma of malt type (PMML) with osseous infiltration

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Dear Editor,

We report the case of a 46-year-old immunocompetent woman with a 10-month-long history of progressive exophthalmia, right palpebral ptosis, and retro-orbital pain. The patient referred also a severe left ocular maculopathy since she was 17 years old. Physical examination revealed a right exophthalmia; blood cell count and other basic laboratory investigations were within normal ranges, with

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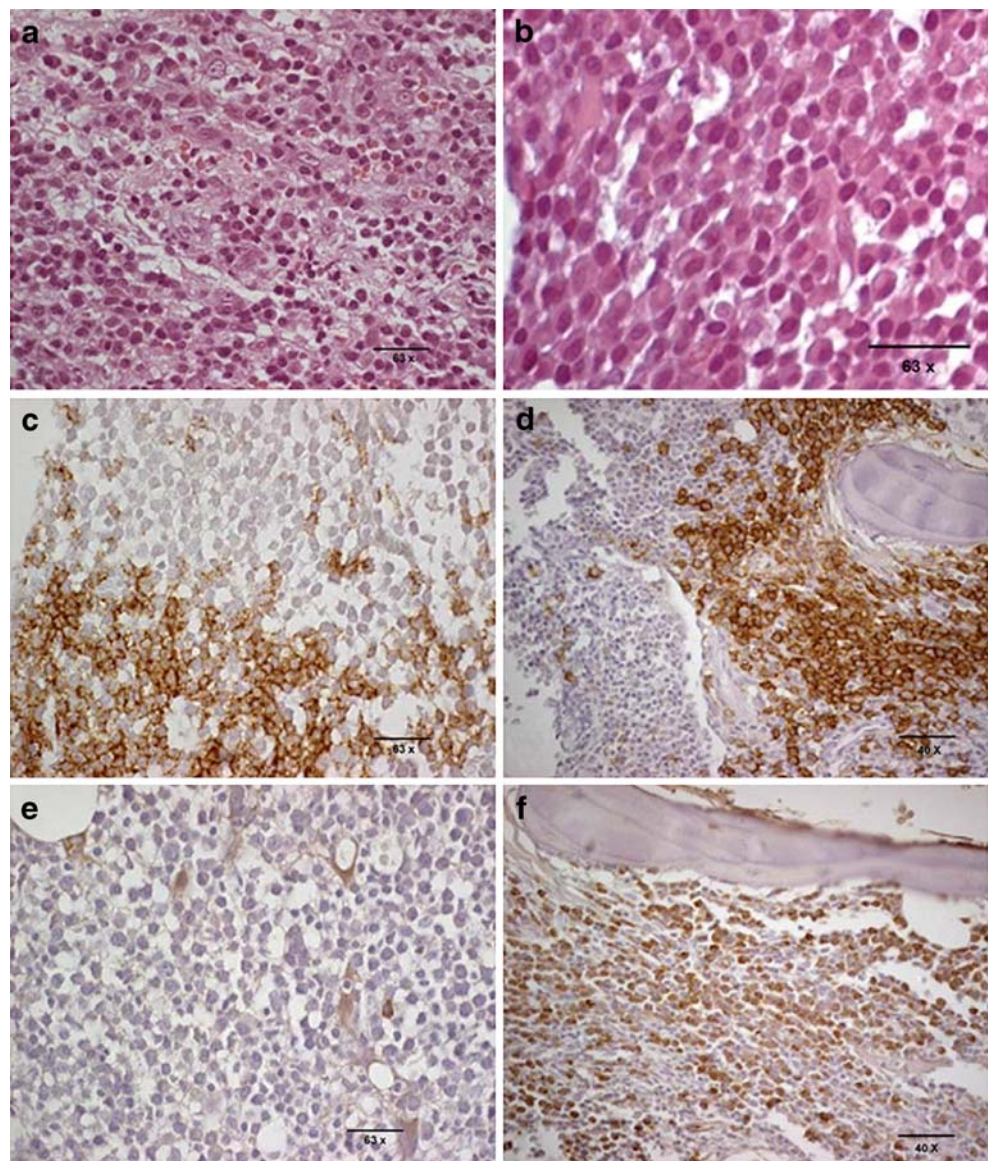
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the exception of a not quantifiable monoclonal component at serum electrophoresis. Magnetic resonance imaging (MRI) and computed tomography (CT) scan showed a right cavernous sinus mass with implant on the meninges associated with intrasellar and intraorbital extension and erosion of the right superior orbital fissure without parenchymal infiltration. These findings supported a neuroradiological diagnosis of meningioma. Cerebrospinal fluid examination was normal. A partial resection of the intraorbital mass was performed. The gross specimen consisted of small fragments (i.e., 0.5 cm in maximum diameter) including bone spiculae. Histopathology revealed a mucosa-associated lymphoid tissue non-Hodgkin's lymphoma (MALT-NHL) with an extensive plasma cell differentiation arising from the dura and infiltrating the underlying bone. Lymphoid cells were mainly represented by small- to intermediate-sized elements with somewhat regular nuclei and relatively abundant clear cytoplasm, forming loose aggregates in the intertrabecular areas of bone fragments (Fig. 1a). Lymphoid cells were immunoreactive for CD20 (Fig. 1c) and negative for CD3, CD21, CD138, IgM, IgD, cyclin D1, EMA, and myeloperoxidase. A significant proportion of tumor was represented by mature-appearing plasma cells, which, in some areas, were virtually the only recognizable elements (Fig. 1b) and showed immunoreactivity for CD138 (Fig. 1d) with lambda light chain restriction (Fig. 1e, f). The postoperative period was uneventful and a complete staging workup, including blood cell count, total-body CT scan, and bone marrow biopsy, ruled out any systemic lymphomatous involvement. *Helicobacter pylori* infection and gastric lymphoma were excluded by multiple biopsies. The patient's well-known long history of left ocular maculopathy prevented the adoption of radiotherapy to avoid further damage to both eyes. Accordingly, treatment with the anti-CD20 monoclonal

Fig. 1 PMML with plasma cell differentiation. Two main components are appreciable: in some areas, the tumor displays a prevalence of monocytoid cells, scattered large cells, and few plasma cells intermingled (**a** hematoxylin and eosin stain); in other areas, particularly close to bone trabeculae, plasma cells organized are the only recognizable elements (**b** hematoxylin and eosin stain, enlarged vision of a 63× original magnification). Monocytoid and large cells display immunoreactivity for CD20 (**c**); plasma cells are CD138 positive (**d**) with a lambda chain restriction (**e** k light chain)



antibody rituximab at a dose of 375 mg/m², once a week for 4 weeks, was chosen as first-line therapy, and it was able to obtain a partial clinical response. Ten months later, due to progressive disease, rituximab therapy was repeated, allowing a further partial regression of the lymphoma. Presently, at 30 months from diagnosis, the patient is alive and well, showing a small residual disease at the brain MRI.

Since the initial description in 1997 by Kumar et al., 56 cases of primary meningeal MALT-NHL (PMML) have been reported so far [1–5], mostly under the form of case reports [2–5], but one large study [1]. Review of literature shows that PMML has somewhat common clinical characteristics and there are not well-established therapeutic guidelines. Presently, local treatments, including surgery followed by radiotherapy or radiotherapy alone, are the most commonly used approaches, but some patients additionally receiving systemic chemotherapy have also

been reported; rarely, chemotherapy was the sole treatment (Table 1). In addition to the intrinsic rarity of PMML, the present case adds further insights to the pathologic and therapeutic features of this lymphoma. In particular, the possibility of bone infiltration by PMML, until now previously reported in only one case [4], should be taken into account at diagnosis. When this eventuality occurs and/or diagnostic material is fragmented and scarce, the monocytoid-looking cells of PMML have to be differentiated from B cell neoplasms with higher propensity for bone involvement, such as hairy cell leukemia, which actually displays much different natural history, prognosis, and treatment. Furthermore, plasma cell differentiation, which could be prominent and unevenly distributed, such as in our case, may mimic a plasma cell myeloma and lead to the erroneous prognostic and therapeutic implications. These arguments suggest that the diagnostic accuracy of PMML

Table 1 Frequency of more commonly reported data in primary dural MALT type lymphoma cases [1–5]

Cases	56
Sex	9 M and 47 F
Age (range)	47–66 years for M 29–75 years for F
Symptoms	Headache, seizure, visual deficit, dizziness
Suspected diagnosis	Meningioma 56/56
Morphology	Small lymphocytes 16/56 Monocytoid cells 21/56 Centrocytic-like cells 4/56 Plasma cell differentiation 2/56
Therapy	22/56 k and 8/56 λ PR and RXT 14/56 PR, RTX and CHT 4/56 CR and CHT 3/56 RXT alone 12/56 CHT alone 3/56 CR alone 3/56
Follow-up (range)	3–100 months

M male, *F* female, *k*, λ type of restricted immunoglobulin light chain, *PR* partial resection, *CR* complete resection, *RXT* radiotherapy, *CHT* chemotherapy

could be implemented by a wide excision of the tumor in order to prevent inappropriate therapy. Finally, this case would suggest rituximab as a good resource to control those PMML cases in which a local treatment is not recommended.

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