Pachymeningeal involvement with blindness as presenting manifestation of non-Hodgkin lymphoma

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A 44 year old female presented with fever for 6 months and gradual onset progressive diminution of vision in both eyes for one month. On examination, she had enlarged cervical, axillary and inguinal lymph nodes, hepatomegaly (7 cm under right costal margin), splenomegaly (5 cm under left costal margin) and bilateral renomegaly. Examination of optic fundi (Figure 1A and 1B) showed bilateral disc edema (black arrowhead) with hemorrhages in the right eye (white arrowhead). Contrast enhanced magnetic resonance (Figure 2A) imaging of brain was done which showed pachymeningeal enhancement (white arrow). Histopathological examination of excised cervical lymph node showed infiltration by atypical lymphoid cells, with immunohistochemistry (IHC) suggesting diffuse large B cell lymphoma (DLBCL)-activated B cell like. Microscopic examination of cerebrospinal fluid showed infiltration by malignant lymphoid cells (Figure 2B). A diagnosis of non-Hodgkins Lymphoma- DLBCL with secondary Central Nervous System (CNS) involvement and bilateral grade 4 papilledema, likely due to pachymeningeal involvement was made. Patient was started on systemic and intrathecal chemotherapy.

CNS involvement with aggressive lymphomas is uncommon at initial presentation and usually occurs during relapse after primary therapy (1). Ophthalmological abnormalities are usually attributed to the direct invasion of the optic nerve and ocular structures by the lymphoma (2) which was not seen in our case.

REFERENCES
Figure 1A - Right fundus photograph showing optic disc edema with multiple hemorrhages
1B- Left fundus photograph showing large optic disc with blurred margins suggestive of papilledema

Figure 2A- CEMRI Brain showing patchy meningeal enhancement and thickening, suggestive of pachymeningitis
Figure 2B- CSF Cytology showing atypical lymphoid cells 2-3 times the size of normal lymphoid cells with prominent nucleoli