Infiltrative optic neuropathy as an initial presentation of acute lymphoblastic leukemia

Sir,

We report a 20-year-old male with infiltrative optic neuropathy of the left eye, leading to irreversible blindness as an initial presentation of high risk T-cell acute lymphoblastic leukemia (ALL). Visual symptoms due to optic nerve infiltration by leukemic cells are rarely found in ALL. Blindness due to infiltrative optic neuropathy as the first presentation of ALL is extremely rare.

A 20-year old male presented with complaints of progressive decrease of vision in his left eye for last one month, leading to sudden blindness. There was no history of pain, redness, or watering. On examination, perception of light was absent in the left eye. Fundus examination of the left eye revealed temporal pallor of the optic disc with blurred margins [Figure 1]. Examination of the right eye revealed no abnormality. B-scan ultrasonography revealed left optic nerve thickening. Magnetic resonance imaging (MRI) of the head and face showed thickening of the extraocular portion of left optic nerve without any intracranial abnormality [Figure 2]. Peripheral blood picture and bone marrow examination revealed features consistent with ALL. Cerebrospinal fluid cytology revealed presence of leukemic blasts. Immunophenotyping showed CD3 and CD5 positivity. In view of positivity for chromosomal translocation t(4;11) (q21;q23), he was diagnosed as a case of high risk T-cell ALL with infiltrative optic neuropathy of the left eye due to leukemic cell infiltration. For optic neuropathy, the patient was recommended topical and systemic steroids. He was given chemotherapy as per ALL-BFM 95 protocol. At first remission, he received 18 Gray/10 fractions therapeutic cranial irradiation, followed by 6 Gray/3 fractions boost to left optic nerve with 3-Dimensional conformal radiotherapy (3D-CRT) technique. There was no improvement of the left eye vision, even after 3 months of treatment completion.

Figure 1: Fundoscopic image of the left eye showing temporal pallor of the optic disc with blurred margins

Figure 2: Magnetic resonance imaging (T1 sequence) showing thickening of the extraocular portion of left optic nerve without any intracranial abnormality
All can rarely present in adults as visual changes due to leukemic optic nerve infiltration.[1] In a prospective study of ocular manifestations in childhood acute leukaemia, Reddy et al.[2] found that although 3.6% of children presented with eye symptoms, on examination ocular changes were found in 17% cases, and these were 2.5 times more common in lymphoblastic leukemia than myeloid leukemia. In view of the high prevalence of asymptomatic ocular lesions in childhood acute leukaemia, the authors concluded that routine ophthalmic examination should be included as a part of evaluation at the time of diagnosis.[2]

The ocular lesions reported in All, were proptosis, intraretinal haemorrhage, white centered haemorrhage, cotton wool spots, macular haemorrhage, subhyaloid haemorrhage, vitreous haemorrhage, papilloedema, cortical blindness, sixth nerve palsy, exudative retinal detachment with choroidal infiltration;[1,2] peripheral ulcerative keratitis with bilateral optic nerve involvement;[3] central retinal artery occlusion associated with leukemic optic neuropathy.[4]

Ocular involvement in lymphoproliferative disorders other than All have also been reported e.g. in myeloid leukemia[2] and Hodgkin's disease.[5]

Acute-onset optic neuropathy in a patient with a history of a lymphoproliferative disorder may be the only sign of a re-emergence of the malignancy.[5,6]

In some reports, there was dramatic improvement of vision with urgent radiotherapy and high-dose systemic corticosteroids, in leukemic or lymphomatous optic nerve infiltration.[1,3] But, no visual gain could be achieved in some reports,[3] like in our patient, and the probable causes might be late presentation with irreversible optic nerve damage and delay in radiotherapy.

REFERENCES