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UNILATERAL INFILTRATION OF THE OPTIC NERVE AND ORBIT REVEALING RELAPSE OF AN ACUTE LYMPHOBLASTIC LEUKEMIA

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Abstract

Introduction. Ocular tissue remains a sanctuary for leukemic proliferation because of poor chemotherapy penetration of the blood-retina and blood-aqueous barriers. Thickening and enhancement of the optic nerve in patients with underlying leukaemia should be considered suspicious for leukaemic infiltration and should be differentiated from optic neuritis. The leukaemic infiltration of the optic nerve is more common in children. The chemotherapeutic drugs cannot penetrate the blood-brain barrier and the invasion of leukaemic cells in the small optic canal can interfere with the flow of CSF.

Purpose. To describe the clinical presentation and imaging features of a leukemic optic nerve and orbit infiltration. We present a case of unilateral infiltration of optic nerve and orbit revealing a relapse of an acute lymphoblastic leukemia.

Methods. A 10-year-old child with past history of treated acute lymphoblastic leukemia 5 years ago, presented to our clinic with decreased vision, eye pain during ocular motility and severe protrusion and exotropia of the left eye. At the moment of our examination, pediatrician confirmed that the child is healthy and no signs of leukemia.

Results. At presentation, we noticed very pronounced protrusion and exotropia of the left eye, lagophthalmos and conjunctival hyperemia. Ocular examination revealed normal visual acuity in his right eye and decreased visual acuity in his left eye 20/200. Pain

during ocular motility and limited elevation, abduction and adduction as well convergence insufficiency.

Ishihara test showed impaired red-green colour vision. Diminished light brightness and contrast sensitivity in his left eye and a relative afferent pupillary defect was present.

Fundus examination showed severe hyperaemia and oedema of the optic disc with blurring margins and whitish-yellowish peripapillary leukemic infiltrates, flame-shaped haemorrhages, few dot and blot retinal hemorrhages in the middle periphery of the retina and towards superior and inferior temporal retinal vein, as well as dilated and tortuous retinal veins.

MRI result showed the presence of a perineural lobulated expansive lesion that infiltrates the intraconal fat with loss of its signal as a sign of infiltration. The described mass displaces the eyeball ventral and causes axial proptosis while posteriorly extends to the prechiasmatic segment of the left ocular nerve with infiltration of the same; penetrates into the posterior third of the ethmoidal sinus with infiltration of the ethmoidal cells as well as towards the left cavernous sinus, para and suprasellar with dural involvement beside the left middle cranial fossa. The orbital apex is dilated and completely occupied by the described substrate. Size of this mass was about 5.3 cm and 4.5 cm.

We diagnosed as relapse of leukemic optic nerve and orbit infiltration and referred to the pediatrician-oncologist for further management. Bone marrow biopsy (myelogram), flow-cytometry and lumbar puncture was done and demonstrated blast infiltration and therefore was confirmed ocular relapse of the lymphoblastic leukemia.

Conclusion. Isolated optic nerve relapse of leukemic infiltration is of paramount importance to early diagnosis, as vision can be saved if treatment is initiated promptly. Optic nerve leukemic infiltration has a severe prognosis. Ophthalmic assessment is essential in patients with ALL in order to diagnose an early ocular involvement and the patient's vision can be preserved if treatment is initiated promptly.

Keywords: infiltration, optic, nerve, orbit, acute, lymphoblastic, leukemia.