Corneal Involvement In A Case Of Mycosis Fungoides - Case Report

A 42 year old gentleman, who was diagnosed as Mycosis fungoides in 2006, currently undergoing radiotherapy, presented to our OPD with complaints of pain, watering and defective vision in left eye of 1 month duration. He was under the care of a local ophthalmologist and was on systemic and topical acyclovir, topical antibiotics, cycloplegics and preservative free lubricants for 1 week.

On examination, he had generalized hyperpigmented plaques over the body and face including the periorbital region. His visual acuity was: OD: 6/6, OS: 6/18. OD: upper lid showed an ulcerative plaque like lesion 2 mm from the anterior lid margin with localized keratinization of the skin. Anterior segment and fundus were otherwise unremarkable.

OS: There was a linear epithelial defect in the inferior paracentral cornea simulating a dendrite. Stroma surrounding the epithelial lesion was edematous, with cellular reaction and Descemet’s level folds. There was stromal scarring nasally with punctate keratitis There was no vascularization.

Fundus examination in both eyes were within normal limits.

Our diagnosis was OD: Infiltration of upper lid secondary to Mycosis fungoides, OS: Herpes Simplex Virus epitheliopathy. Corneal infiltration secondary to Mycosis fungoides was also considered as a differential diagnosis.

He was continued on Acyclovir eye ointment for 2 weeks, Prednisolone acetate 1% eye drops QID and preservative free lubricant eye drops. Since his was a biopsy proven case of Mycosis fungoides, futher biopsy from the lid lesion was deferred.

At one week review, the corneal lesion was resolving with mid stromal scar and mild punctate keratitis. Acyclovir eye ointment was stopped after 3 weeks and topical steroids tapered. At one month follow up, the lesion healed completely with mid stromal scaring.

Discussion

Mycosis fungoides is a malignant cutaneous T-cell lymphoma characterized by clonal expansion of T helper cells and rarely T-suppressor or natural killer cells, that usually present as a widespread chronic cutaneous eruption, which progresses to form plaques and tumours. Eventually non-cutaneous involvement supervenes with infiltration of lymph nodes and visceral organs[1].

Ocular involvement may occur in up to a third of advanced cases of Mycosis fungoides and most frequently presents as tumours or infiltration of the eyelids [2]. Less frequent manifestations include involvement of the caruncle[3] the cornea[2] the sclera[4] the optic disc[5] and optic nerve[6].
and the orbit[7]. Intraocular involvement has been reported only rarely[5,8].

Corneal involvement is a rare occurrence in Mycosis fungoides. This was first reported by Besnier and Hallopeau in (1892) and histologically confirmed by Herman in 1951. Occasionally this may be the first evidence of the disease, but usually it is seen in well developed cases and occasionally as a terminal event.

Corneal manifestations vary. They may appear as a regular linear infiltrate, somewhat resembling a dendrite, which progresses to an ulcerative keratitis involving the stroma and Descemets membrane. Alternatively progressive and usually heavily vascularised stromal infiltrates appear which may suffer acute exacerbations and may breakdown to ulceration or undergo spontaneous resolution.

Pathological examinations have demonstrated the typical infiltration characterized by large mononuclear cells of the primitive lymphoblastic type.

This case was reported because of the rare corneal presentation of Mycosis fungoides. Antivirals were used in the treatment keeping in view of the more common herpetic keratitis. But the lesion lacked the typical dendritic borders and terminal bulbs. Reference to the older literature pointed more towards a self resolving variant of corneal involvement in Mycosis fungoides. Therefore a high index of clinical suspicion is needed in the diagnosis of the unusual presentation of this rare disease.

References