

CHINESE MEDICAL SCIENCES JOURNAL

CASE REPORT

Conjunctival Langerhans Cell Histiocytosis: a Case Report

Di Chen and Han-yi Min*

Department of Ophthalmology, Peking Union Medical College Hospital,
Chinese Academy of Medical Sciences & Peking Union
Medical College, Beijing 100730, China

Key words: Langerhans cell histiocytosis; conjunctiva; immunohistochemistry

Chin Med Sci J 2015; 30(1):63-64

LANGERHANS cell histiocytosis (LCH) is a rare disease, mainly involving the bone, skin, lung, liver, spleen, and skin.¹ The heterogeneous nature of LCH makes it difficult to diagnose. Not only do the involved organs vary from case to case, but also its natural history. Herein, we describe a rare case of conjunctival LCH in an Asian woman.

CASE DESCRIPTION

A 47-year-old Asian female complained about conjunctival injection and red nodules of her both eyes for 4 months. On examination, the best-corrected visual acuity was 20/50 in the right eye and 20/30 in the left eye. Physical examination revealed diffuse conjunctival injection and smooth solid red nodules beneath the superior bulbar conjunctiva of her both eyes (Fig. 1A). The cornea was clear with medium-sized grey keratic precipitates (Fig. 1B). The erythrocyte sedimentation rate (ESR) and angiotensin-converting enzyme (ACE) value were all within normal limits. The rheumatoid factor (RF) and anti-nuclear antibody (ANA) were also negative. The nodule biopsy was performed to determine the diagnosis, which showed hyperplasia of histiocytes with massive plasma cells and

lymphocytes infiltration (Fig. 1C). Polymorphic giant cells could be seen (Fig. 1D). Immunohistochemical study revealed CD34 (–), S-100 (+) (Fig. 1E), and CD68 (++) (Fig. 1F). These findings were consistent with Langerhans cell histiocytosis (LCH). And the final diagnosis of conjunctival LCH was made.

Systemic therapy of prednisone 1 mg/kg along with cyclosporine A and cyclophosphamide was initiated. After one month's treatment, the conjunctival injection disappeared and the patient's visual acuity returned to 20/20 in the right eye and 20/15 in the left eye. The prednisone was gradually tapered down. The patient had been followed up for two years and there was no sign of recurrence.

DISCUSSION

LCH is characterized by the infiltration of pathogenic Langerhans cells.¹ The etiology of the rare disease is unknown. The ocular LCH mainly involves the orbit.² It has been occasionally reported LCH could induce uveitis, retinal vasculitis, and choroidal tumor,³ but the conjunctival LCH has never been reported before. The diagnosis of LCH is largely dependent upon immunohistochemical stains.⁴

The treatment of LCH remains controversial. The choice of treatment depends on the location of the lesions and the degree of local destruction.¹ As for this case, the

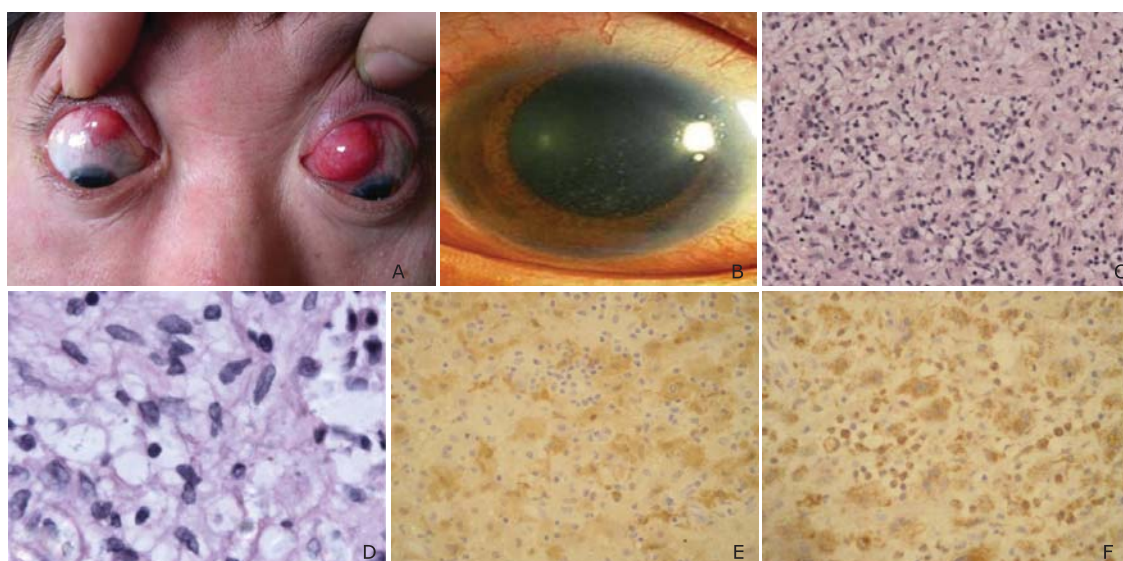


Figure 1. A 47-year-old woman with conjunctival Langerhans cell histiocytosis.

- A. Conjunctival injection and red nodules of both eyes.
- B. Keratic precipitates on the cornea.
- C. The biopsy of the nodules revealed hyperplasia of histiocytes with massive plasma cells and lymphocytes infiltration ($\times 100$).
- D. Histologically, polymorphic giant cells were seen ($\times 400$).
- E and F. Immunohistochemical study showed S-100 (+) and CD68 (++) ($\times 100$).

disease had affected the patient's visual acuity, which made systemic therapy necessary. After correct diagnosis and effective treatment, the patient had recovered well.

REFERENCES

1. Donadieu J, Chalard F, Jeziorski E, et al. Medical management of Langerhans cell histiocytosis from diagnosis to treatment. *Expert Opin Pharmacother* 2012; 106:1309-22.
2. Cheung N, Selva D, McNab AA, et al. Orbital Langerhans cell histiocytosis in adults. *Ophthalmology* 2007; 114: 1569-73.
3. Kim IT, Lee SM. Choroidal Langerhans' cell histiocytosis. *Acta Ophthalmol Scand* 2000; 78:97-100.
4. Crooks B, Grenier D. Langerhans cell histiocytosis: A complex recurrent disease. *Paediatr Child Health* 2010; 15:69-70.