Case Report

Eosinophilic Granuloma Syndrome with Conjunctival Involvement: A Case Report

Seyed-Hashem Daryabari [1,*], MD; Mohammad Ali Javadi [2], MD; Majid Shams [3], MD

1. Chemical Injures Research Center, Systems Biology and Poisonings Institutes, Baqiyatallah University of Medical, Tehran, Iran.
2. Professor of Ophthalmology, Shahid Beheshti University of Medical Sciences, Tehran, Iran.
3. Cornea Fellowship, Shahid Beheshti University of Medical Sciences, Tehran, Iran.

***Corresponding Author:** Seyed-Hashem Daryabari
E-mail: shdarya50@yahoo.com

Abstract

To report a rare case of eosinophilic granuloma with bilateral conjunctival involvement. A 27-year-old female patient was referred to our center due to granulomatous lesions in the upper conjunctiva of both eyes, which began about two years ago. Patient also had a history of lung disease (Asthma) from about 7 years ago. The patient was eventually diagnosed as a case of bilateral eosinophilic granuloma of eyes with pulmonary involvement. Treatment with systemic corticosteroid resulted in disappearance of the eye lesions after one month.Although eosinophilic granuloma of eye is a very rare condition it should be considered as a differential diagnosis for eye masses especially in patients with concurrent lung problems.

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Introduction

Histiocytosis X or Langerhans cell histiocytosis is a group of rare disorders with reticuloendothelial system involvement, which can appear in three ways: eosinophilic granuloma (a usually self-limiting condition appearing in children or young adults), Hand-Schuller-Christian (HSC) disease (chronic form of disease associated with bone involvement and internal organ involvements) and Letterer-Siwe disease (LSD) (fatal form of disease observed in young children) [1,2]. Here we describe a case of eosinophilic granuloma with bilateral eye involvement as a lesion in the upper conjunctiva of both eyes as well as pulmonary involvement, its diagnosis, treatment and follow up results.

Case Report

A 27-year-old woman was referred to our center complaining from granulomatous lesions in the upper conjunctiva of both eyes, which began about two years ago (Figure 1). She reported a pulmonary disease (Asthma), which began about seven years ago. There was no specific history of surgery, drug history and family history. In eye examinations the vision of both eyes was 10/10. In slit lamp exam a granulomatous lesion in the upper conjunctiva of both eyes was observed. Other examinations of the anterior and posterior segments were normal. Internal pressure of both eyes was 14 mm/ Hg which was in normal range.

Due to pulmonary involvement and the presence of conjunctival lesion, the differential diagnosis of the patient was divided into two categories: systemic or local disease involving eyes. The most important systemic differential diagnoses at this point were eosinophilic granuloma, Wagner granulomatous, and sarcoidosis.

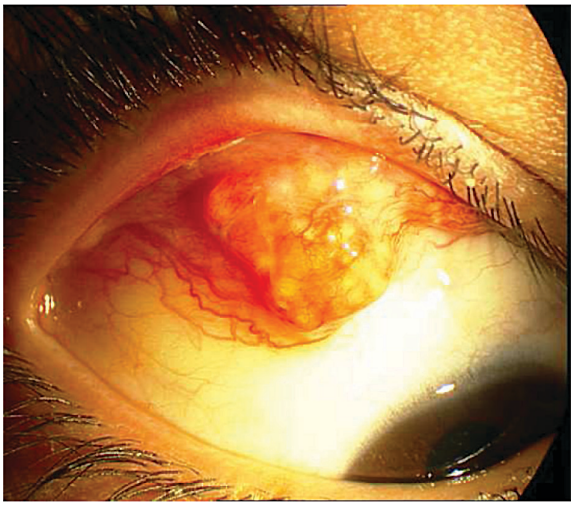

Figure 1: Upper conjunctival granulomatous lesions appearing in both eyes

The possible local differential diagnoses were squamous papilloma, conjunctival intraepithelial neoplasia, conjunctiva nevus, conjunctival melanoma, pyogenic granuloma, and Kaposi sarcoma.

Orbit CT-SCAN was performed which was normal. Perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA) and cytoplasmic antineutrophil cytoplasmic autoantibody (c-ANCA) tests were performed with normal results. A biopsy was performed from the patient's right eye lesion and allergic conjunctival granuloma was reported. The patient was asked to have a chest X-ray due to her history of pulmonary involvement and a reticulonodular diffuse lesion was observed. After trans-bronchial biopsy, it was confirmed that patient has eosinophilic granuloma.

After diagnosis of eosinophilic granuloma with eye and pulmonary involvement, systemic prednisolone 1 mg / kg was prescribed. About two weeks later the dosage was reduced to reach maintenance dose of five milligrams. The patient was examined after one month and symptoms of dyspnea were significantly improved. In slit lamp examination the

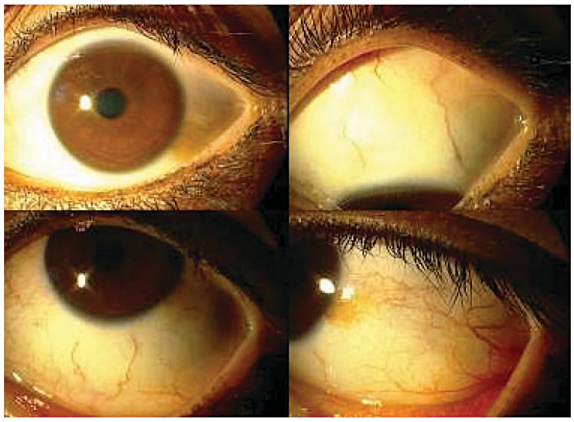

Figure 2: Complete recovery after treatment by corticosteroid

conjunctival lesions had been completely resolved (Figure 2).

Discussion

Histiocytosis X or Langerhans cell histiocytosis can cause single organ or multiple organ involvement. Single organ involvements might appear in lungs, pituitary gland, bone, skin, lymph nodes, thyroid, liver, spleen, and brain [3]. Multiple organ involvement itself can be divided into two categories: with pulmonary involvement and without pulmonary involvement. Eosinophilic granuloma is a form of disease which is usually self-limiting and appears in children or young adults [4]. Here we reported a case of eosinophilic granuloma with bilateral eye involvement as well as pulmonary involvement in a 27 year old patient. It should be noted that bilateral eye involvement, as seen in our case is a very rare finding.

Similar to our patient Demirci et al.,[4] in 2002 reported a five year old boy with bilateral eosinophilic granuloma involvement of the eyes. Kanazawa et al.,[5] reported a case of mono lateral eosinophilic granuloma eye involvement in a 32 years old patient, which was controlled with corticosteroid similar to our patient. Weissgold et al.,[6] reported a case of eosinophilic granuloma of the eyelid in a 15-year-old girl who responded well to incisional biopsy and required no additional treatment.

Conclusion

Although eosinophilic granuloma of eye is a very rare condition it should be considered as a differential diagnosis for eye masses especially in patients with concurrent lung problems.

Authors ORCIDs

Seyed-Hashem Daryabari:
https://orcid.org/0000-0002-7264-8637

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Footnotes and Financial Disclosures

Conflict of Interest

The authors have no conflict of interest with the subject matter of the present study.