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SHORT COMMUNICATION

Case report

Giant chalazia in the hyperimmunoglobulinemia E (hyper-IgE) syndrome

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PURPOSE. *To report the occurrence of recurrent multiple giant chalazia in the hyperimmunoglobulin E syndrome (hyper-IgE syndrome or Job syndrome).*
METHODS. *Two patients with hyperimmunoglobulinemia E (>500 IU/ml) had ophthalmologic examination and surgical treatment for chalazia of the eyelids.*
RESULTS. *The hyper-IgE syndrome is a rare immunodeficiency and multisystem disorder characterized by recurrent skin and pulmonary abscesses, connective tissue abnormalities, and elevated levels of serum IgE. In two patients with the hyper-IgE syndrome, multiple giant chalazia were seen in upper and lower eyelids. Despite surgical incision new giant chalazia arose.*
CONCLUSIONS. *Recurrent multiple giant chalazia may occur as an ophthalmic feature of the hyper-IgE syndrome. (Eur J Ophthalmol 2004; 14: 258-60)*

KEY WORDS. *Chalazion, Eyelids, Hyperimmunoglobulinemia, Hyper-IgE, Job syndrome, Ptosis*

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INTRODUCTION

The hyperimmunoglobulin E syndrome (hyper-IgE syndrome or Job syndrome) is a rare immunodeficiency disorder characterized by recurrent skin and pulmonary abscesses and extremely elevated levels of serum IgE (1).

The majority of patients also show connective tissue abnormalities, such as a lack of primary teeth root resorption, recurrent fractures, hyperextensible joints, and scoliosis. It has an autosomal dominant inheritance pattern with variable expressivity (1). The genetic defect of the syndrome has not been elucidated. In this report we describe two patients diagnosed with the hyper-IgE syndrome with recurrent chalazia, refractory to treatment.

Case reports

A 17-year-old woman with the diagnosis of hyper-IgE syndrome (IgE serum concentration 2500 IU/ml; normal reference level <500 IU/ml) was referred for ocular examination because of swelling of the eyelids. At the age of 1 month, she underwent surgery because of a perforation of the ileum. Since her first year of life,

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she has had eczema, pulmonary infections, and skin abscesses. Cultures taken from these infections usually yielded *Staphylococcus aureus*. A major problem was a progressive kyphoscoliosis, which is a recognized abnormality of this syndrome. Recently, she had developed painless, round, firm lesions in the upper and lower eyelids of both eyes, causing blurred vision and a secondary ptosis. The lesions were diagnosed as giant chalazia. Except for the chalazia, no ocular abnormalities were seen. The lesions were treated by surgical incision and complete curettage through conjunctiva and tarsus. The lesion showed a lipogranulomatous aspect with stagnated sebaceous secretion, typical for chalazion. A culture was taken, but was negative for bacteria. Despite strict eyelid hygiene, new lesions arose in both upper and lower eyelids. Only the larger lesions were again treated surgically (Fig. 1).

A 24-year-old man with an established diagnosis of hyper-IgE syndrome (IgE serum concentration 7000 IU/ml; normal reference level <500 IU/ml) was referred for ocular examination because of swelling of his left upper eyelid. In earlier years he had had eczematous skin lesions, sinusitis, otitis media, lymph node abscesses, and atopic conjunctivitis. The swelling of his eyelid was diagnosed as a giant chalazion, causing a severe ptosis. No other ophthalmic abnormalities were seen. The chalazion was treated by surgical incision and complete curettage through conjunctiva and tarsus, but despite the treatment new giant chalazia appeared in both upper eyelids (Fig. 2).

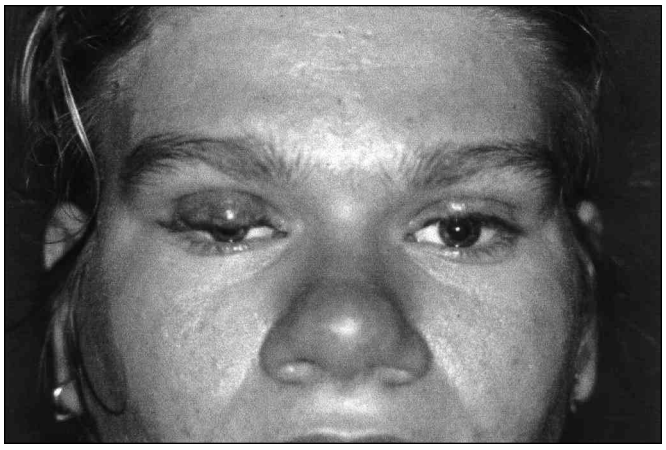

Fig. 1 - *A 17-year-old woman with hyper-IgE syndrome and giant chalazion of the right upper eyelid and a smaller chalazion of the left upper eyelid.*

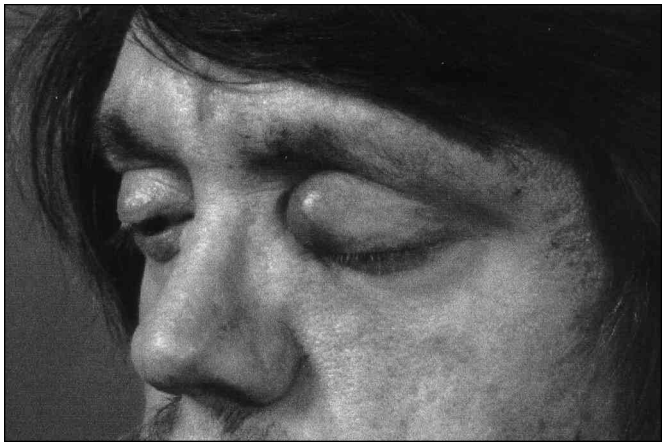

Fig. 2 - *A 24-year-old man with hyper-IgE syndrome and a giant chalazion of the left upper eyelid and a smaller chalazion of the right upper eyelid.*

DISCUSSION

Patients with the hyper-IgE syndrome have recurrent bacterial infections of the skin and sinopulmonary tract. Other ocular manifestations have been described in the literature, including conjunctivitis (2, 3), keratitis (3-5), and corneal perforation (4, 5). Endophthalmitis has been reported in two cases, one caused by streptococci (6) and the other caused by *Candida spp* (7).

We are unaware of previous reports concerning multiple chalazia in patients with the hyper-IgE syndrome and could find no references in a MEDLINE search. Even though contamination of the chalazia with staphylococci could not be confirmed, these two cases illustrate that recurrent chalazia can be an ophthalmic feature of the hyper-IgE syndrome.

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