

Recurrent giant chalazia in hyperimmunoglobulin E (Job's) syndrome

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Abstract Hyperimmunoglobulinemia E (Job's) syndrome is a rare autosomal dominant disorder appearing early in life with recurrent skin and pulmonary infections, characterized by markedly increased serum immunoglobulin E (IgE) levels. We describe a 50-year-old man with a 4-year history of recurrent, multiple giant chalazia in all eyelids. Medications and surgical intervention had produced only transient improvement. The patient had also had pulmonary and scalp infection. Laboratory tests disclosed elevated serum IgE (>1,000 IU/ml) and eosinophilia. As a result, based on the patient's history and clinical and laboratory findings, a diagnosis of Job's syndrome was made. Even though rarely, recurrent multiple giant chalazia may occur as an ophthalmic feature of Job's syndrome. Hyperimmunoglobulinemia E syndrome should be suspected in any case of recurrent giant chalazia, regardless of the patient's age. Measurement of serum IgE and eosinophils, along with internal evaluation, is essential to establish a proper diagnosis.

Keywords Hyperimmunoglobulinemia E · Job's syndrome · Recurrent giant chalazia · Middle-age

Introduction

Hyperimmunoglobulin E (Job's) syndrome is a rare, multisystem, autosomal dominant disorder characterized by high serum immunoglobulin E (IgE) levels (>1,000 IU/ml), eczematous skin, eosinophilic pustular folliculitis-like appearance, and recurrent pulmonary and skin infections [1]. Abnormal neutrophil chemotaxis due to decreased production of interferon gamma is believed to be the underlying mechanism of disease [2]. This syndrome was first reported in 1966 by Davis et al., who discussed two girls with red hair, chronic dermatitis, and recurrent staphylococcal abscesses and pneumonias [3]. Inspired by the biblical character whose body was smitten with boils by Satan, they referred to this condition as Job's syndrome. Reports of ocular involvement in patients with Job's syndrome are rare [4–8]. We describe herein a middle-aged patient with recurrent, multiple giant chalazia, who was later diagnosed as having hyperimmunoglobulin E syndrome.

Case report

A 50-year-old man was referred with a 4-year history of recurrent giant chalazia in all eyelids. Medications

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Fig. 1 Photograph of the right eye showing multiple giant chalazia in a 50-year-old man with Job's syndrome

and surgical intervention had produced only transient improvement. The patient also had a history of pneumonia and scalp abscesses. External examination disclosed facial asymmetry with hemihypertrophy, prominent forehead, broadened nasal bridge, deep-set eyes, increased alar and outer canthal distance, and multiple giant chalazia in all eyelids (Fig. 1). Best-corrected visual acuity was 20/20 OU. Examination of both eyes was unremarkable. Blood tests revealed elevated serum IgE (1,090 IU/ml; normal 4–100 IU/ml) and mild eosinophilia ($0.9 \times 10^3/\mu\text{l}$; normal $0\text{--}0.8 \times 10^3/\mu\text{l}$). As a result, based on the patient's history, characteristic facies, and laboratory findings, a diagnosis of hyperimmunoglobulin E syndrome was made. The patient underwent surgical incision and curettage and was postoperatively treated with oral doxycycline and tobramycin/dexamethasone ointment. Histology confirmed the diagnosis of chalazia. Microbial cultures of the chalazia contents were negative. Improvement was noted at week 3 postoperatively; however, new chalazia not necessitating further surgery arose after 3 months.

Discussion

Job's syndrome often appears early in life, but diagnosis up to the age of 63 years has been described [1, 9]. This condition is characterized by extremely elevated serum IgE and recurrent skin and

pulmonary infections, typically caused by *Staphylococcus aureus* and *Candida albicans* [1]. Many therapeutic approaches have been used to treat patients with Job's syndrome, including intermittent or long-term anti-staphylococcal antibiotics, cyclosporine, cimetidine and ascorbic acid, high-dose I.V. Ig and interferon gamma [1]. As more is understood about the immune defects in these patients, more treatment options with increased efficacy will become available.

Ocular infections in hyperimmunoglobulin E syndrome are rare, but they may be severe and lead to visual loss. *C. albicans* endophthalmitis and presumed *S. aureus* corneal ulceration and perforation despite aggressive systemic and topical antibiotic therapy have been reported [4–6]. Furthermore, De Stafeno et al. [7] described recurrent *S. aureus* chalazia despite appropriate medical and surgical therapy.

We are unaware of any previously reported cases of eyelid involvement in patients with Job's syndrome occurring in middle age. Even though rarely, recurrent giant chalazia may be an ophthalmic feature of Job's syndrome [8]. As a result, hyperimmunoglobulin E syndrome should be suspected in any case of recurrent giant chalazia, regardless of the patient's age. Measurement of serum IgE, along with internal evaluation, is essential to establish a proper diagnosis. Repeated surgical intervention and appropriate systemic and topical therapy may be required to obtain a satisfactory result in Job's syndrome with recurrent giant chalazia.

References

1. DeWitt CA, Bishop AB, Buescher LS, Stone SP (2006) Hyperimmunoglobulin E syndrome: two cases and a review of the literature. *J Am Acad Dermatol* 54:855–865
2. Borges WG, Augustine NH, Hill HR (2000) Defective interleukin 12/interferon-gamma pathway in patients with hyperimmunoglobulinemia E syndrome. *J Pediatr* 136:176–180
3. Davis SD, Schaller J, Wedgwood RJ (1966) Job's syndrome: recurrent, "cold", staphylococcal abscesses. *Lancet* 1:1013–1015
4. Frohn A, Weidle EG, Seelhorst A, Steuhl KP, Thiel HJ (1990) Eye involvement in hyperimmunoglobulin E syndrome. *Klin Monatsbl Augenheilkd* 197:432–435
5. Haslett RS, Moriarty AP, Vijayadurai P, McGalliard JN, Chandna A (1996) *Candida* endophthalmitis in Job syndrome. *Arch Ophthalmol* 114:617–618

6. Orhan M, Ozkan Y, Irkeç M (2001) Eye involvement in hyperimmunoglobulinemia E (Job's) syndrome. *J Pediatric Ophthalmol Strabismus* 38:313–314
7. DeStafeno JJ, Kodosi SR, Primack JD (2004) Recurrent *Staphylococcus aureus* chalazia in hyperimmunoglobulinemia E (Job's) syndrome. *Am J Ophthalmol* 138:1057–1058
8. Crama N, Toolens AM, van der Meer JW, Cruysberg JR (2004) Giant chalazia in the hyperimmunoglobulinemia E (hyper-IgE) syndrome. *Eur J Ophthalmol* 14:258–260
9. Rosenberg BE (2004) Hyperimmunoglobulin E syndrome. *Dermatol Online J* 10:4