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GRANULOMATOUS INFLAMMATION OF THE ORBIT: DIAGNOSIS, MANAGEMENT AND THERAPY

Slavica Konjević-Pernar, Renata Iveković, Katia Novak-Lauš and Zdravko Mandić

University Department of Ophthalmology, Sestre milosrdnice University Hospital, Zagreb, Croatia

SUMMARY – During the 2000-2005 period, we treated 32 patients with unilateral or bilateral granulomatous inflammation of the orbit. After careful exclusion of specific systemic and local diseases, the diagnosis was based on a combination of clinical and radiological findings. Examinations were performed to exclude the presence of a malignant or other form of tumor. Subsequently, we started treatment with high-dose steroids or a combination of steroids and surgical therapy. After therapy, we analyzed visual acuity, exophthalmometry, intraocular pressure and visual fields. There was no tumor recurrence during the follow up of 1-4 years. The findings suggested the treatment with high-dose steroids or a combination of steroids and surgical therapy to be an efficient therapeutic option for granulomatous inflammation.

Key words: *granulomatous inflammation, orbit, steroid therapy*

Introduction

Granulomatous inflammation, also known as orbital pseudotumor or idiopathic orbital inflammation is a disorder that was first described in 1903 by Busse and Hochheim[1]. It is a benign lesion of unknown origin that involves all or part of the fatty tissue within the orbit. The etiology, or cause, of the condition is unknown. The condition is rarely bilateral. Differential diagnosis includes orbital cellulitis, thyroid ophthalmopathy, sarcoidosis, lymphoid tumor, and metastatic carcinoma. Granulomatous inflammation can be subdivided into three different types: granulomatous, lymphoid and sclerosing. The diagnosis is usually a clinical one. Magnetic resonance imaging (MRI) is the most sophisticated radiological method of examination. The disorder is more prevalent in adult than in pediatric population.

Patients and Methods

During the 2000-2005 period, 32 patients with unilateral or bilateral granulomatous inflammation of the orbit were treated at University Department of Ophthalmology, Sestre milosrdnice University Hospital, with a combination of high-dose steroids and surgical therapy. Patients with the lesion localized in lacrimal gland were excluded from the study. Patients suspected of granulomatous inflammation of the orbit underwent complete ophthalmologic examination, which included visual acuity test according to Snellen, intraocular pressure (IOP) measurement, funduscopy, exophthalmometry according to Hertel, ultrasound orbit examination, visual field according to Goldmann, and MRI. The examinations were performed to exclude the presence of a malignant or other form of tumor. Treatment was initiated upon completion of these examinations.

First, surgical procedure of orbitotomy was performed at the site of intraorbital mass, with intraoperative exploration and removal of tumor tissue. Then, the patients were administered i.v. steroid therapy of 1000 mg Solu-Medrol in 1000 mL of Ringer's solution over three days, followed by *per os* steroid therapy over two months, with dose tapering. During the operative procedure and pulsed corticosteroid therapy, the patients were hospitalized at our Department, and subsequently they were followed-up at outpatient basis, i.e. on postoperative day 7 and 21, at 1 month, 3 months, 6 months, and then every 6 months.

Correspondence to: *Slavica Konjević-Pernar, MD*, University Department of Ophthalmology, Sestre milosrdnice University Hospital, Vinogradska c. 29, HR-10000 Zagreb, Croatia

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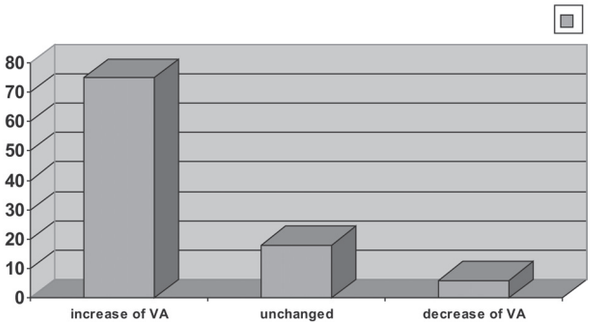

Fig. 1. Schematic presentation of postoperative visual acuity results.

Results

Bilateral inflammation of the orbit was diagnosed in three, and unilateral inflammation of the orbit in 29 of 32 study patients. There were 19 female and 13 male patients, mean age 52 (range 39-82) years. The follow up period was 1-4 years. Visual acuity according to Snellen was measured both before and after therapy. After treatment, visual acuity increased by one or more lines according to Snellen in 24 (75%), remained unchanged in six (18.75%), and decreased in two (6.25%) patients (Fig. 1). We also measured protrusion of the bulb using exophthalmometer according to Hertel. Before therapy, protrusion amounted to 21-32 mm, whereas the values after therapy ranged from 16 to 26 mm (Fig. 2). During the follow up period, we recorded no signs of reactivation of inflammation or postoperative complications in any of our patients.

Discussion and Conclusion

We report on a retrospective study conducted from 2001 to 2005 at University Department of Ophthalmology, Sestre milosrdnice University Hospital in Zagreb, which included 32 patients diagnosed with granulomatous inflammation of the orbit, manifesting as an orbital mass that infiltrates the soft tissue structures within the orbit. The origin and etiology of the disease remain unknown.

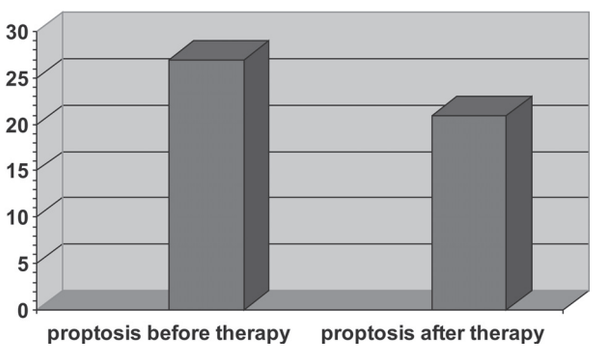

Fig. 2. Schematic presentation of results on pretherapeutic and post-therapeutic proptosis.

We decided on a therapeutic option with a combination of steroids and surgical intervention, based on literature data that indicate favorable inflammation response to steroid therapy (over 40%), and by far better results when combined with orbitotomy and tumor extirpation (97%)[2]. We followed the patients' visual acuity both before and after the treatment, along with measuring protrusion of the bulb. After the treatment, 75% of our patients showed an increase of visual acuity by one or more lines according to Snellen, and an even greater percentage showed a decrease of eyeball protrusion by $^{3}2$ mm. No signs of reactivation of inflammation were noticed during the follow up period.

Accordingly, it is concluded that good therapeutic results can be achieved in patients suffering from granulomatous inflammation of the orbit by thorough clinical examination and accurate diagnosis, followed by combined treatment with steroids and surgical intervention.

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Sažetak

GRANULOMATOZNA UPALA ORBITE: DIJAGNOZA, PRAĆENJE I TERAPIJA

S. Konjević-Pernar, R. Iveković i *Z. Mandić*

U razdoblju od 2000. do 2005. godine na Klinici za očne bolesti Kliničke bolnice „Sestre milosrdnice“ liječili smo 32 bolesnika s jednostranom ili obostranom granulomatoznom upalom orbite. Nakon pažljivo učinjenih pretraga kako bi se isključilo postojanje specifičnih sustavnih ili lokalnih bolesti postavili smo dijagnozu koja se temeljila na kombinaciji kliničkih i radioloških nalaza. Kod svih bolesnika prije i nakon liječenja analizirana je vidna oštrina, mjeren je očni tlak, učinjena egzoftalmometrija po Hertelu te vidno polje po Goldmannu. Svi bolesnici najprije su bili podvrgnuti kirurškom zahvatu orbitotomije, eksploracije i uklanjanja tumorskog tkiva, potom su svi primili pulsnu dozu kortikosteroida kroz tri dana, a nakon toga se nastavilo s liječenjem *per os* kroz dva mjeseca. Kod 75% bolesnika nakon završetka liječenja došlo je do porasta vidne oštrine za jedan ili više redova po Snellenu, a kod 96.88% bolesnika došlo je do smanjenja protruzije očne jabučice za 2 ili više mm. Tijekom razdoblja praćenja nismo zabilježili znakove reaktiviranja upale. Zaključuje se kako se pažljivim kliničkim pretraživanjem i primjereno postavljenom dijagnozom te kombiniranim liječenjem steroidima i kirurškim zahvatom postižu dobri rezultati kod bolesnika s granulomatoznom upalom orbite.

Ključne riječi: *granulomatozna upala, orbita*