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Orbital Pseudotumor: Clinical Features and Outcomes

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SUMMARY To describe the clinical characteristics of orbital pseudotumor, a retrospective analysis was performed on patients with orbital pseudotumor at Siriraj Hospital for ten years. Forty-nine patients (24 males and 25 females; 62 eyes) with a mean age of 43.75 years were included (a mean follow-up of 25 months). Thirty-six patients (73.5%) had unilateral disease. The clinical features were proptosis (79.6%), ocular motor deficit (61.2%), pain (51%), lid swelling or lid mass (44.9%), ptosis (24.5%), and chemosis (18.4%). The most common presenting sign was proptosis (49%). All were treated with corticosteroids with clinical improvement in 40 (81.6%) patients. Ten (83.3%) of 12 patients with visual loss improved with mean recovery time of 10.3 days. Ocular motility recovered in 24 (80%) patients, occurring an average of 17.8 days after initiation of therapy. It is concluded that the clinical features of orbital pseudotumor are varied. Most patients were improved with corticosteroids treatment.

Orbital pseudotumor or idiopathic orbital inflammation syndrome are used to describe a nonspecific benign orbital inflammation without evidence of specific local or systemic cause. It is an uncommon disorder diagnosed by exclusion.[1] The clinical features of orbital pseudotumor vary widely, with the presentation of orbital pain, proptosis, eyelid swelling, chemosis, double vision, and visual loss.[1,2] Corticosteroids are the mainstay of therapy and can produce dramatic results.[1-3] Although the etiology of orbital pseudotumor is unknown, immune process has been suggested as a mechanism for orbital pseudotumor.[1,3-7] The association of orbital pseudotumor with some systemic diseases and successful treatment of corticosteroids or other immunosuppressive agents also suggest that an autoimmunologic process is the mechanism.[1,5-7] The present study was undertaken to evaluate clinical characteristics, orbital-imaging findings, treatment, and outcome in patients with orbital pseudotumor.

PATIENTS AND METHODS

We reviewed medical records of patients with a diagnosis of orbital pseudotumor treated at the Department of Ophthalmology, Siriraj Hospital, between August 1997 and May 2006. These records were identified from the central database of the hospital. This study was approved by the ethical committee of the hospital. The diagnosis of orbital pseudotumor was made clinically on the basis of orbital pain, proptosis, lid swelling, chemosis, conjunctival injection, ophthalmoplegia, and visual loss. All patients have no other identifiable cause such as orbital tumor, thyroid orbitopathy, orbital fracture, sinusitis, dacryocystitis, orbital cellulites, or other infection and inflammation. Neuro-imaging and pathologic re-

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sults may have been used to support the diagnosis.

We collected the following information of each patient: age at onset, sex, best-corrected initial visual acuity, eye examination, laboratory findings, neuro-imaging, treatment, visual and extraocular motility recovery time and follow-up data. Proptosis was defined with a Hertel exophthalmometer as greater than 2 millimeters (mm) compared with the noninvolved eye or greater than 21 mm in both eyes. Recurrent orbital pseudotumor was defined as repeated attacks affecting one or both eyes over an interval greater than one month. If the patients had complete relief of symptoms, they were defined as improved.

RESULTS

A total of 49 patients (62 eyes) with orbital pseudotumor were identified. Twenty-four (49%) were male and 25 (51%) were female. The mean age was 43.75 years (range 4-84 years). Patients underwent a follow-up with a mean of 25 months (range 1 month-8 years). Thirteen patients (26.5%) had bilateral disease whereas 36 patients (73.5%) had unilateral involvement. Right eyes and left eyes were 17 (34.7%) and 19 (38.8%), respectively (Table 1). The clinical features of orbital pseudotumor varied. The most common chief complaint that patients were concerned about, and came to the hospital with, was proptosis (24 patients, 49%). Six patients (12.2%) came with the chief complaint of orbital pain and three (6.1%) came with visual loss. Other chief complaints were lid swelling or lid mass (22.4%), diplopia (6.1%), and ptosis (4.1%). We had summarized the symptoms and signs of the chief complaints in Table 2. Most patients had proptosis (79.6%). Twenty-five patients (51.0%) had orbital pain and 12 patients (24.5%) complained of visual loss. Other clinical features were limitation of eye movement (61.2%), lid swelling or lid mass (44.9%), ptosis (24.5%), and chemosis (18.4%). Clinical features are summarized in Table 3.

Disc examination at presentation revealed normal in 58 of 62 eyes (93.5%), optic disc swelling in 3 of 62 eyes (4.8%), and optic atrophy 1 of 62 eyes (1.6%).

All patients were treated with corticosteroids with clinical improvement in 40 patients (81.6%).

Table 1 Clinical characteristics of patients with orbital pseudotumor

	N (%)
Total	49
- Male	24 (49)
- Female	25 (51)
Mean age (years)	43.75
Unilateral	36 (73.5)
- Right eye	17 (34.7)
- Left eye	19 (38.8)
Bilateral	13 (26.5)
Recurrence	10 (20.4)
Mean time of follow up (months)	25

Table 2 Chief complaints of patients with orbital pseudotumor

	N (%)
Proptosis	24 (49)
Lid swelling or lid mass	11 (22.4)
Orbital pain	6 (12.2)
Diplopia	3 (6.1)
Visual loss	3 (6.1)
Ptosis	2 (4.1)

Forty-two of 49 patients (85.7%) underwent orbital-imaging evaluation. Magnetic resonance imaging and computed topography were performed in 34 and 8 patients, respectively. All orbital images demonstrated an infiltrative mass or extraocular muscle enlargement. Five patients showed enlargement of lacrimal glands.

Twelve eyes (11 patients) of 62 eyes (49 patients) had visual loss. Ten of 12 eyes (83.3%) were improved with mean recovery time of 10.3 days. Two patients with unilateral visual loss who were not improved with the treatment had visual acuity only as light perception at the presentation, and one of these two patients showed disc pallor at the presentation.

Ocular motility deficit recovered in 26 of 32 eyes (81.3%) within an average of 17.8 days after

Table 3 Clinical features of patients with orbital pseudotumor

	N (%)
Proptosis	39 (79.6)
Ophthalmoplegia	30 (61.2)
Orbital pain	25 (51.0)
Lid swelling or lid mass	22 (44.9)
Visual loss	12 (24.5)
Ptosis	12 (24.5)
Chemosis	9 (18.4)

initiation of the therapy. Five patients with unilateral ocular motility impairment who had not improved with corticosteroids treatment demonstrated extraocular muscle enlargement on orbital image. Another patient revealed an infiltrative lesion at the superior part of the orbit and had vertical diplopia with limitation of upgaze greater than downgaze.

Ten patients (20.4%) had recurrence. Seven of these patients were female. Furthermore, eight of these patients had developed recurrence within two years.

Twenty-five patients underwent tissue biopsy. All showed pleomorphic cellular infiltration with lymphocytic predominate. Histopathologic subtypes were not classified.

DISCUSSION

In our study, the results corresponded to the clinical features reported in the literature. There was no gender predilection.[1] It may occur in any age groups,[1,3] ranging from 4-80 years[3] with mean age of 45 years. The clinical features varied with proptosis, ocular motor impairment, orbital pain, eyelid swelling, ptosis, chemosis, and visual loss; all responded quite well to systemic corticosteroids.[1,2]

Our study showed no gender predilection, while some studies reported female predominance.[2,3] However, this predominance was not significant; the highest predominance in females (5:1) was in the orbital apex syndrome group.[2] Our study also showed that the most common clinical features were proptosis and opthalmoplegia whereas pain and periorbital swelling were the most common clinical features in Western patients.[2] The higher incidence of periorbital swelling in the Western study[2] than in our study (75% *vs*. 10%) may be from the higher frequency of dacryoadenitis (40% *vs.* 10%). However, the higher incidence of proptosis and ophthalmoplegia in our study than in the Western study[2] (61% *vs.* 31%) may be due to the higher incidence of myositis and inflammation of retrobulbar region, (86% *vs.* up to 59%). There might be also a wide and variable spectrum of orbital pseudotumor. It is possible that the characteristics and natural history of pseudotumor are different between Asian and Western patients. This warrants further study.

The mainstay of therapy for orbital pseudotumor is corticosteroids.[1,2] The response is typically rapid. Although good response to corticosteroids was reported in most patients, the advantage is sometimes temporary and partial, and symptoms occasionally recurred as treatment was tapered off. All of our patients were treated with systemic corticosteroids with clinical improvement in 85% and a recurrence rate of 20%. Patients with visual loss or ocular motor impairment had good prognosis for recovery after treatment with corticosteroids. Many previous studies demonstrated varied success and recurrence rates in different subtypes of orbital pseudotumor. One reported that 78% of orbital pseudotumor, which excluded myositis and lacrimal gland pseudotumor, responded well with corticosteroids initially but only 37% were cured, with a 52% recurrence rate.[8] Another study showed a 63% success rate and 58% recurrence rate in idiopathic orbital inflammation.[2] Additionally, some studies reported 15-23% recurrence rate in orbital myositis with corticosteroids.[3,9] The differences of response and recurrence rate might be due to the differences in subtypes of orbital pseudotumor, diagnostic criteria, degree of inflammation, dose and duration of corticosteroids treatment, and the natural history of the disease. Previous studies reported good response to corticosteroids in patients with isolated dacryoadenitis or myositis and orbital pseudotumor with optic nerve involvement.[8,10] However, unfavorable outcomes were showed in patients with myositis and inflammatory mass, posterior, sclerosing and vasculitic orbital pseudotumor.[4,10]

Our result supported that studies which indicate good response to corticosteroids is not a pathognomonic sign and is not reliable as a diagnostic test for identifying orbital pseudotumor.[4,8]

In this study, 70% of recurrence of orbital pseudotumor was found in females. Most of recurrence (80%) developed within two years. This is the same as a previous reports that indicate patients who reduce or stop corticosteroids had recurrence within one year.[4] The recurrence rate in females (7/25 = 28%) was higher than in males (3/24 = 13%). This finding is in contrast to a previous Western report which found a higher recurrence rate in males (4/11 = 36%) than females (2/15 = 13%) in patients with orbital myositis.[9]

However, further study with a large number of patients and long term follow-up is needed to understand the clinical manifestations of orbital pseudotumor that might help the physician to approach and differentiate this condition from other common and uncommon causes of proptosis leading to proper and early management.

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