Idiopathic Orbital Inflammation: Distribution, Clinical Features, and Treatment Outcome

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Objective: To evaluate the distribution and clinical as well as treatment outcome characteristics of idiopathic orbital inflammation with the aim of delineating a more systematic approach to diagnosis and treatment.

Methods: A 10-year retrospective review of patients with idiopathic orbital inflammation treated at one institution.

Results: Ninety eyes in 65 patients (22 men and 43 women) were studied. Diagnoses were isolated dacryoadenitis (n=21), isolated myositis (n=19), concurrent dacryoadenitis and myositis (n=5), orbital apex syndrome (n=6), and idiopathic inflammation involving the preseptal region, supraorbital region, sclera, Tenon capsule, orbital fat, or optic nerve (n=14). The mean age at presentation was 45 years. Pain and periorbital swelling were the most common clinical features and were observed in 45 (69%) and 49 (75%) patients, respectively. Seventeen patients (26%) had bilateral involvement. Biopsy was performed in 19 patients (29%) with atypical presentations or who failed to respond to the initial therapy. Patients were treated with steroids alone (n=45), steroids and subsequent radiation therapy (n=8), steroids and nonsteroidal anti-inflammatory agents (n=6), nonsteroidal anti-inflammatory agents alone in mild cases (n=2), and, rarely, radiation therapy without steroids (n=1) or surgical debulking alone (n=1). Of 65 patients, 41 (63%) represented treatment successes, with complete symptom relief at the time of the last follow-up, and 24 (37%) represented treatment failures, with partial or no relief of symptoms. Treatment failures were often characterized by recurrence of inflammation after a period of quiescence (58%) and unremitting, recalcitrant inflammation (38%); 1 patient ultimately required an exenteration.

Conclusion: Systemic steroid with a slow taper has been the established first-line treatment for idiopathic orbital inflammation, but refractory cases accounted for a significant portion of treatment failures in our study, reflecting the need for a more systematic approach to the study of this multifaceted disease and for therapeutic alternatives to systemic steroids.

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Idiopathic orbital inflammation, also known as orbital pseudotumor, is defined as a benign, noninfective clinical syndrome characterized by features of non-specific inflammatory conditions of the orbit without identifiable local or systemic causes. It was first described in 1903 by Gleason and by Busse and Hochheim and characterized as a specific clinicopathological entity in 1905 by Birch-Hirschfeld, who described it as an orbital mass that simulated a neoplasm but was histologically inflammatory. Since the initial description, many classification schemes have been applied to idiopathic orbital inflammation based on the location of the inflammatory process, the histopathological characteristics, and the stage of inflammation. Idiopathic orbital inflammation is the third most common orbital disease, following Graves orbitopathy and lymphoproliferative diseases. It accounts for 4.7% to 6.3% of orbital disorders.

Idiopathic orbital inflammation has highly variable clinical features, ranging from a diffuse to very focal process targeting specific orbital tissues, such as the lacrimal gland, extraocular muscles, and orbital fat. This space-occupying infiltrating orbital process is typically characterized by an abrupt onset of pain, proptosis, and inflammatory signs and symptoms, such as swelling and erythema. Presentations vary according to the specific location and the degree of inflammation, fibrosis, and mass effect. Posis, chemosis, motility dysfunction, and optic neuropathy may also be found. Entrapment, compression, and destruction of orbital tissues may occur in patients with extensive sclerosis. Unilateral presentation is typical, but bilateral presentations are not uncommon. Symptoms most commonly de-
velop acutely (hours to days) with patients reporting a datable onset. In the minority of patients, presentation may occur over weeks (subacute) or may occur insidiously over a period of months (chronic). Pediatric idiopathic orbital inflammation is characterized by a number of features that differ from the adult presentation. Bilateral manifestation, as well as uveitis, disc edema, and eosinophilia, appear to be more common in the pediatric population.13-17

The pathogenesis of idiopathic orbital inflammation has remained elusive. Idiopathic orbital inflammation has been associated with several infectious processes, including upper respiratory tract infections and flulike viral illness, but the exact nature of these associations is not clear.13,16-18,20 Several lines of evidence point to immune-mediated processes as the likely underlying ocular mechanism. Associations of idiopathic orbital inflammation with a number of systemic immunologic disorders, including Crohn disease, systemic lupus erythematosus, rheumatoid arthritis, diabetes mellitus, myasthenia gravis, and ankylosing spondylitis, have been reported in several studies.9,21-26 Mombaerts and Koornneef,27 for instance, found in their series that 10% of 58 patients with idiopathic orbital inflammation had concurrent autoimmune disease. In addition, idiopathic orbital inflammation typically responds favorably to systemic corticosteroid treatment and successful outcomes have been reported with other immunosuppressive agents, such as cyclophosphamide,28 methotrexate,29 and cyclosporine.30 Interestingly, 2 other disorders with a predilection for the orbit, namely Graves orbitopathy and ocular myasthenia gravis, are also immune-mediated.

An autoimmune process has been suggested as the ocular mechanism for idiopathic orbital inflammation by Atabay et al,31 who reported the presence of circulating autoantibodies against eye muscle antigens in patients with orbital myositis. Although autoimmunity is a plausible idea, it is not clear whether these autoantibodies are specific to idiopathic orbital inflammation or are also present in other forms of inflammation, such as scleritis and uveitis. In addition, the typical unilateral presentation of idiopathic orbital inflammation argues against the notion that autoimmunity is the primary mechanism. Alternatively, Mombaerts et al32 and Rootman et al33 proposed aberrant immune-mediated production of fibrogenic cytokines leading to aberrant wound healing as the ocular mechanism underlying the process of fibrosis in sclerosing orbital inflammation.

Although benign, idiopathic orbital inflammation may have a clinically malignant course, with severe vision loss and oculomotor dysfunction. Spontaneous remission may occur without any therapy, but systemic corticosteroids are the cornerstone of therapy in the acute phase. Despite the generally favorable response to steroid therapy, relapses and persistent inflammation complicate the clinical course and treatment. Therefore, idiopathic orbital inflammation poses a considerable diagnostic and therapeutic challenge.

METHODS

This study is a retrospective review of 79 patients with the diagnosis of idiopathic orbital inflammation who were treated at the Massachusetts Eye and Ear Infirmary (MEEI), Boston, from January 1991 to April 2001. The study protocol was approved by the institutional review board at MEEI. The diagnosis of idiopathic orbital inflammation was made based on the following clinical criteria: benign, noninfective, inflammatory condition of the orbit without identifiable local or systemic causes. Radiological workup was performed for many patients but was not a necessary diagnostic criterion for the study. Likewise, histopathological confirmation was not a diagnostic requirement for the study.

Of 79 patients, 6 with the eventual final diagnosis of Wegener granulomatosis or Graves orbitopathy were excluded from the analysis; an additional 8 patients with a follow-up period of less than 5 weeks were also excluded from the study. A lower limit for the follow-up period was set at 5 weeks because response to treatment is usually evident by this time. All the remaining 65 patients met the criteria for idiopathic orbital inflammation and had at least 5 weeks of follow-up.

These patients were managed according to the following treatment algorithm: at the initial manifestation, patients were typically treated with a high-dose oral steroid (1.0-1.5 mg/kg per day) for 1 to 2 weeks with taper during the ensuing 5 to 8 weeks. In the event of rebound of symptoms during steroid taper or recurrence of symptoms after a period of quiescence, steroid dosage was increased (with a slower taper) or restarted for typically no longer than 10 to 12 weeks. Alternatively, when the disease course was atypical or refractory to systemic steroids, biopsy was performed for definitive diagnosis. Radiation treatment was typically reserved for patients who did not respond to or were intolerant to systemic steroid therapy. Radiation therapy typically consisted of low-dose irradiation (usually 15 to 20 Gy fractioned over 10 days). Treatment outcome was considered a “success” if the patient had complete relief of symptoms at the time of the last follow-up and a “failure” if the patient had no or only partial relief of symptoms at the time of the last follow-up.

RESULTS

Ninety eyes in 65 patients (22 men and 43 women) with a final diagnosis of idiopathic orbital inflammation and at least 5 weeks of follow-up were included in the data analysis. Patients were followed for a mean of 20 months (range, 1 month to 9 years). Each patient was evaluated at least twice.

DISTRIBUTION OF SUBTYPES

The 65 patients had isolated dacryoadenitis (n=21), isolated myositis (n=19), concurrent dacryoadenitis and myositis (n=5), orbital apex syndrome (n=6), and idiopathic inflammation involving the preseptal region, supraorbital region, sclera, Tenon capsule, orbital fat, or the optic nerve (n=14). The frequency of these subtypes is given in the Table. The mean age at manifestation was 45 years (range, 2.5 weeks to 89 years).

CLINICAL FEATURES

Pain was the most common symptom and was evident in at least 45 patients (69%). Diplopia was less frequent but present in at least 20 patients (31%). Periorbital edema was the most common sign and was present in at least 49 patients (75%), followed by red eye, proptosis, and chemosis occurring in at least 31 (48%), 21 (32%), and 19 (29%) of patients, respectively (Figure 1). Seven-
ten patients (26%) had bilateral involvement, either concurrently or sequentially. Twenty-six patients (40%) showed involvement of the extraocular muscles, either as the primary presentation, as in myositis, or as a less prominent part of a more diffuse presentation. The most frequently involved muscle was the medial rectus (n=18; 31%), followed by the superior rectus (n=15; 25%), lateral rectus (n=14; 24%), and inferior rectus (n=12; 20%).

NEUROIMAGING AND BIOPSY

Neuroimaging studies were obtained for 63 patients (97%). The most common neuroimaging study was the computed tomographic (CT) scan, which was obtained for 59 patients (91%). Magnetic resonance images (MRIs) were obtained for 10 patients, 7 in combination with a CT scan. Ultrasonograms were obtained only for 3 patients, 2 in combination with a CT scan.

Radiological findings allowed subtypes of idiopathic orbital inflammation to be more precisely classified according to the specific orbital tissues involved as follows:

- **Lacrimal gland:** diffuse, oblong enlargement of the lacrimal gland with preservation of the shape of the gland that may be accompanied by an inflammatory reaction in the periglandular tissue, blurring the gland margin.
- **Extraocular muscles:** enlargement of the extraocular muscles (single or multiple; with or without the involvement of the associated tendons) accompanied by some spillover of the inflammatory process into orbital fat bordering the muscle, blurring the margin of the muscle.
- **Orbital fat:** diffuse infiltration in the orbital fat, enveloping the globe, that may involve the optic nerve sheath complex.
- **Preseptum, sclera, episclera, Tenon capsule, and uvea:** inflammatory and enlargement of the tissues.
- **Optic nerve:** inflammation of the optic nerve sheath with thickening of the margin of the nerve and streaky densities in the contiguous orbital fat.
- **Orbital mass:** orbital mass of heterogeneous composition, occasionally invading extraorbital structures, extending along the optic nerve sheath from the globe to the optic canal.
- **Orbital apex and cavernous sinus:** inflammatory process at the apex may compress, obliterate, or displace the optic nerve and may have intracranial extension into the cavernous sinus.

Biopsy was performed in 19 patients (29%) with atypical presentations or who failed to respond to the initial therapy. Of the biopsy specimens, 14 showed a nonspecific inflammatory pattern, and the remaining 5 showed a sclerosing pattern, with dense fibrosis as the primary feature.

TREATMENT OUTCOME

Of 65 patients included in the study, 45 (69%) were treated with steroids alone, 8 (12%) with steroids and subsequent radiation therapy, 6 (9%) with steroids and non-steroidal anti-inflammatory agents, 1 atypical case (2%) with radiation therapy and nonsteroidal anti-inflammatory agents, 2 mild cases (3%) with nonsteroidal anti-inflammatory agents alone, and 1 rare case (2%) with surgical debulking alone; treatment was deferred in the remaining 2 cases (per patient request or pending completion of workup). Of 65 patients studied, 41 (63%) represented treatment successes and had complete symptom relief; 24 (37%) represented treatment failures, with 23 patients experiencing only partial relief and 1 patient experiencing no relief. These treatment outcome results may reflect an overall higher rate of steroid failures than that generally observed in the community because MEEI serves as a referral center, which would introduce some degree of selection bias toward more complicated and recalcitrant disease. The stringent definition for treatment success used in our study, ie, complete relief of symptoms may also inflate the overall higher failure rate. Treatment outcomes are shown for different subtypes of idiopathic orbital inflammation (Figure 2) and treatment modes (Figure 3).

TREATMENT FAILURE ANALYSIS

The clinical course for many patients in our study was complicated by incomplete or no resolution of inflammation, ie, treatment failure. For 14 (58%) of 24 pa-
tients who failed to respond to treatment, inflammation recurred after a seemingly favorable response to treatment with a period of quiescence. For 9 (38%) of 24 patients, the disease course was characterized by unremitting, recalcitrant inflammation, with no real relief provided by treatment; 1 of these patients ultimately required an exenteration.

Systemic steroid therapy has traditionally been the mainstay of treatment for idiopathic orbital inflammation. However, many patients in our study, particularly those who ultimately failed to respond to treatment, had unsatisfactory outcomes from systemic steroids. Rebound of symptoms during steroid taper (ie, steroid dependence) and adverse reaction to steroid therapy (ie, steroid intolerance) occurred in 8 (33%) and 3 (13%), respectively, of the 24 patients who failed to respond to treatment. Steroid dependence and steroid intolerance also occurred in 12% and 2%, respectively, at some point in the clinical course of patients who ultimately had successful outcomes (Figure 4).

Age was not a predictor of treatment failure because the mean age of patients successfully treated (42 years) was comparable to that of patients who failed to respond to treatment (45 years). Likewise, sex was not significantly associated with treatment failure because the male-female ratio remained comparable for the treatment success group (13:28) and the treatment failure group (9:15). Inflammation involving the orbital apex was relatively uncommon in our study but was associated with the poorest treatment outcome; 4 (66%) of 6 patients with this subtype failed to respond to treatment. Sclerosing orbital inflammation also appeared to be associated with poor outcome because all 4 cases in our study failed to respond to treatment and 1 patient ultimately required an exenteration.

A more detailed characterization of treatment failure was beyond the primary scope of this study and remains to be further explored. Our study did not systematically address several questions that are important in defining more fully the pattern of recalcitrant disease and thereby improving treatment outcome, such as: Is there a particular window period of peak recurrence after the initial presentation, and would an aggressive treatment targeted at this window ultimately improve outcome? Does a more prompt initiation of treatment reduce the rate of recurrence and treatment failure? Is there an optimal course of steroid treatment or tapering for more difficult cases that would ultimately reduce the rate of recurrence?

COMMENT

In our study, idiopathic orbital inflammation occurred more frequently in middle-aged women. The true incidence of
idiopathic orbital inflammation in the literature is difficult to assess, given the wide spectrum of manifestations and the lack of a universally accepted definition of the disease entity. However, the peak incidence reported in the literature appears to be predominantly in the adult population, typically in middle-aged persons, with pediatric cases accounting for only 6% to 17% of the total incidence.36 No strong sex predilection has been reported for idiopathic orbital inflammation. However, a 2:1 predominance in women,35 as well as a rare instance of familial occurrence,36 has been reported for orbital myositis. In our study, an overall 1.8:1 predominance in women was observed across the different subtypes; the highest predominance in women (5:1) was observed for the orbital apex syndrome group. Dacryoadenitis and myositis were the most common subtypes observed in our study.

Our patients typically had unilateral periorbital pain and edema at the initial visit, which is consistent with the clinical features reported in the literature. Other features such as diplopia, redness, chemosis, and proptosis occurred less frequently in our study. Restriction, compression, and destruction of orbital tissues occurred rarely in our study and only in patients with extensive sclerosing and poor treatment outcomes. Medial rectus was the most commonly involved extraocular muscle in our study, followed by superior rectus and lateral rectus; inferior rectus was the least frequently involved. In their studies of orbital myositis, Mombaerts et al27 and Siatkowski et al37 reported that medial and lateral rectus were most commonly involved.

Radiological imaging studies allow tissue characterization and localization without surgical intervention and thereby have become invaluable diagnostic tools. Computed tomography is the preferred imaging mode for idiopathic orbital inflammation and was by far the most common imaging mode used in our study. Idiopathic orbital inflammation is typically seen on CT scans as a focal or diffuse mass, usually poorly demarcated and enhancing with contrast. Common CT findings include enhancement with contrast medium, infiltration of retrolubular fat, proptosis, extraocular muscle enlargement, muscle tendon or sheath enlargement, apical fat edema, optic nerve thickening, uveal-scleral thickening, edema of the Tenon capsule, and lacrimal gland infiltration.33 Tendons of the extraocular muscles may be involved or spared.38 Magnetic resonance imaging is generally used, either alone or in combination with CT, in patients with extraorbital or intracranial extensions. Intracranial extension and bone destruction have been reported but are rare findings, usually occurring with the sclerosing process.39–41 Echography may be useful as an alternative if CT or MRI is not readily available.37

The histopathological spectrum of idiopathic orbital inflammation is typically nondiagnostic, wide, and diverse, ranging from the typical diffuse polymorphous infiltrate to the atypical granulomatous inflammation, tissue eosinophilia, and infiltrative sclerosis. In the absence of systemic fibroinflammatory, granulomatous, and vasculitic disease, these atypical presentations are considered to be subclasses of idiopathic orbital inflammation.42 Histopathological confirmation was sought in our study for patients who had atypical presentations or who failed to respond to the initial therapy. A benign, non-specific inflammatory pattern was the general finding in our study, with the exception of 4 cases, which showed dense fibrous connective tissue as the predominating component with little inflammatory infiltrate (ie, the sclerosing form). The dense collagenization seen in the sclerosing form is considered by some authors to be the end stage of the histological continuum of idiopathic orbital inflammation, with the earlier stage characterized by greater lymphocytic component that then progresses toward fibrosis in the later chronic stage.43–46 Others, however, support the notion that idiopathic sclerosing inflammation of the orbit is a unique clinicopathological entity distinct from idiopathic orbital inflammation.31,47 In either case, the sclerosing form runs an insidious, frequently progressive course that replaces and damages orbital structures through cicatricial entrapment. It tends to be more aggressive than the nonsclerosing forms and appears to have a poor therapeutic outcome, consistent with the findings in our study.

Systemic steroid therapy with a slow taper has been the established first-line treatment, but refractory cases accounted for a significant portion of treatment failures in our study. Steroid dependence and intolerance were prominent features of the patients who failed to respond to treatment. Steroid therapy, in general, hastens clinical resolution in the acute phase. However, as observed in our study, there are serious shortcomings to steroid therapy. Although many patients with idiopathic orbital inflammation do have favorable responses to steroid therapy, incomplete resolution is common. Steroid resistance and dependence as well as potential adverse reactions, such as mood changes, hyperglycemia, dyspepsia, and weight gain, further complicate the clinical course and therapy. Mombaerts et al27,32,46,47 report a low cure rate (37%) and a high recurrence rate (52%) obtained with steroid therapy in their retrospective studies and question the value of steroids as the primary treatment modality. They propose that systemic steroid therapy not be used as the initial step but be reserved as therapy in selected patients who have associated optic neuropathy or who may benefit from rapid though possibly transient symptomatic relief. They point out the need for a controlled prospective study that compares the efficacy of different therapeutic modalities. However, until more information on the efficacy of these alternative treatment modalities becomes available, systemic steroid therapy appears to be the best-accepted first-line treatment for idiopathic orbital inflammation. Nonsteroidal anti-inflammatory drugs, such as ibuprofen, may be used as an alternative to or in combination with steroid therapy. Mannor et al48 recommend a trial of nonsteroidal anti-inflammatory drugs for up to 3 weeks or until clinical resolution, with steroids reserved for refractory cases.

Reports of alternatives to steroid therapy, such as immunosuppressive chemotherapy, are fairly limited in the idiopathic orbital inflammation literature. Pulsed chemotherapy consisting of either cyclophosphamide or chlorambucil combined with prednisone has been reported to be effective in the treatment of idiopathic orbital inflammation refractory to both steroid and radiation
therapy. Methotrexate and intravenous immunoglobulin have similarly been found to be effective in treating idiopathic orbital inflammation that did not respond to steroids. The role of chemotherapy in the treatment of idiopathic orbital inflammation remains to be further explored.

Radiation therapy was used in our study to treat patients with idiopathic orbital inflammation who were unresponsive to steroid therapy, became steroid dependent, or had intolerable adverse reactions to steroids. Radiation therapy was used as the initial treatment in only 1 atypical case in which Graves orbitopathy and atypical lymphoma were strongly considered in the differential diagnosis. Radiation therapy did not significantly affect the treatment of idiopathic orbital inflammation in our study, because 8 of 9 patients who underwent radiation therapy ultimately failed to respond to treatment. This is not surprising given that the patients who ultimately undergo radiation therapy tend to have more complicated or atypical disease. However, several studies have reported favorable outcomes with radiation therapy, indicating that radiation therapy remains a viable treatment option for idiopathic orbital inflammation. Radiation therapy is an attractive alternative to steroid therapy, particularly given the relatively few potential adverse effects of low-dose external-beam radiation treat-
orbital fat volume. These findings may occur in the euthyroid setting in the absence of any objective thyroid dysfunction as the initial presentation of the disease process or after adequate control. A number of clinical and radiological features distinguish idiopathic orbital inflammation from thyroid orbitopathy. Abrupt onset of pain and inflammatory signs, such as periorbital erythema and swelling, are typical early manifestations of idiopathic orbital inflammation. In contrast, Graves disease has a slower, more insidious course, and extraocular motility dysfunction and visual disabilities tend to occur later in the disease process. In addition, pain is not a prominent feature of Graves orbitopathy. Radiological findings for idiopathic orbital inflammation are typically unilateral and may involve any of the orbital structures, including the extraocular muscles, muscle tendons, orbital fat, perineural connective tissues, Tenon capsule, and sclera. Graves orbitopathy, in contrast, has findings that are typically bilateral, and the primary focus is on the enlargement of extraocular muscles and increased orbital fat volume.

Wegener granulomatosis is a necrotizing, granulomatous inflammation and vasculitis that affects the respiratory and renal systems; there is ocular involvement in about 50% of cases. Wegener granulomatosis is a rare disease but an important differential diagnosis for idiopathic orbital inflammation because of its association with high morbidity and mortality. Bilateral eye pain, proptosis, redness, and ocular motility dysfunction are common clinical features. Ocular and orbital manifestations of Wegener granulomatosis include conjunctivitis, marginal ulcerative keratitis, scleritis, uveitis, retinal vasculitis and optic neuropathy, dacryoadenitis, and nasolacrimal duct obstruction. Histological findings consist of necrotizing, granulomatous inflammation and vasculitis. Serum levels of antineutrophil cytoplasmic antibodies that display a cytoplasmic immunofluorescent staining pattern (c-ANCA) are elevated in many cases having orbital extension. Thyroid orbitopathy is exceedingly rare in children and is much lower in the differential diagnosis than it is for idiopathic orbital inflammation. Retinoblastoma is also an important consideration because it is the most common intraocular tumor of children, with 10% of cases having orbital extension. Thyroid orbitopathy can provoke a significant granulomatous inflammatory reaction in the surrounding orbital tissue. Lymphangioma is usually slow growing but can also have a fulminating inflammatory presentation on rupture and hemorrhage.

After the local and systemic causes have been ruled out, treatment for presumed idiopathic orbital inflammation can be initiated. In patients with a mild clinical presentation, clinical progression may be monitored while awaiting test results, or nonsteroidal anti-inflammatory drug therapy may be initiated. In patients with a moderate to severe clinical presentation and a strong possibility of idiopathic orbital inflammation, systemic corticosteroid therapy may be initiated as long as suspicion for an infectious etiology is low and there is no other contraindication for steroid therapy. Systemic prednisone can be started with an initial dosage of 60 mg to 100 mg per day for 1 to 2 weeks and a taper typically over 5 to 6 weeks. Clinical progression and potential adverse reactions need to be closely monitored once systemic corticosteroid therapy has been initiated. In the event of persistent or recurrent episodes that are refractory to systemic steroid therapy, biopsy (open biopsy or, if applicable, fine needle aspiration) should be strongly considered for definitive diagnosis, particularly if the lesion is easily accessible (eg, lacrimal gland). If the biopsy findings are consistent with idiopathic orbital inflammation, consider the following options in patients with a disease course that is refractory to systemic steroid therapy: (1) if the patient had a favorable response to steroid therapy
initially, oral steroid therapy may be restarted or, if the patient is in the steroid taper phase, the dosage may be increased with a slower taper and (2) if the patient is steroid intolerant, nonresponsive, or dependent, low-dose external beam irradiation (typically 15 to 20 Gy fractioned over 10 days) may be considered.

For idiopathic orbital inflammation refractory to both systemic corticosteroid and radiation therapy, treatment options are limited. It would be reasonable to step back and repeat or expand the radiological evaluation, laboratory workup, and biopsy (of a different site if multiple sites are involved) to reassess the possibility of other causes. Alternatively, chemotherapeutic agents, such as cyclophosphamide, may be considered. Surgical debulking may be a consideration if the lesion is easily approachable or in patients with a severely progressive and disabling clinical course (eg, orbital apex syndrome with optic nerve compression).

CONCLUSIONS

Idiopathic orbital inflammation is a multifaceted disease with a wide spectrum of clinical, radiological, and histopathological presentations. In our study, idiopathic orbital inflammation occurred more frequently in middle-aged persons, affected more women than men, and typically manifested with unilateral periocular pain and edema. Dacryoadenitis and myositis were most common. Response to steroid therapy was generally favorable initially in our study, but persistent and recurrent cases accounted for a significant portion of the treatment failures. Steroid dependence and intolerance were prominent features of patients who failed to respond to treatment in our study.

Idiopathic orbital inflammation encompasses a broad spectrum of disease entities with a high degree of variability in clinical presentation and outcome. Many aspects of this disorder remain unknown. The pathogenesis of idiopathic orbital inflammation has remained elusive despite several lines of evidence for an immune-mediated etiology. Moreover, no satisfactory animal model has been developed to date to aid in the understanding of the disease process or to assist in determining optimal treatment protocols to limit sequelae from inflammation. Relapses are exceedingly frustrating to both patients and physicians because there are few treatment alternatives in these refractory cases. Factors predictive of these recurrent and refractory cases need to be further delineated. There is a great need for a more systematic and comprehensive characterization of this disease. This would provide a framework for a controlled prospective study comparing the efficacy of different treatment modalities for idiopathic orbital inflammation and thereby aid in further exploration of therapeutic alternatives to systemic corticosteroids.

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REFERENCES


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A look at the past . . .

I t is now certain that accommodation does not affect intraocular tension, so increased accommodative efforts due to wearing the correcting glass for near cannot tend to increase the myopia. This is rather due to the pressure of the extrinsic muscles, whose deleterious action is favored when objects are held close to the eyes.