Review Article

A Systemic Review on the Management Approaches in Non-Specific Orbital Inflammation

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Abstract

Background: Non-Specific Orbital Inflammation (NSOI) or Idiopathic Orbital Inflammation (IOI) or orbital pseudotumor is an uncommon, non-systemic, non-infectious, and non-neoplastic inflammatory disorder of the orbit with a broad spectrum of clinical features. Despite being the third most common orbital disease, its diagnosis and treatment are unreliable because no standardized protocols exist. This systematic review critically evaluates and consolidates current diagnostic methods and treatment plans for IOI to facilitate evidence-based clinical decision-making. **Methodology:** Systematic PubMed and Google Scholar searches were conducted to identify English-language, open-access studies. The PRISMA protocol was followed to select studies. Seventeen studies were included in the final analysis. **Result:** NSOI is diagnosed through clinical assessment, MRI or CT imaging, and selective biopsy. Systemic corticosteroids is first-line therapy, achieving >80% initial response but with 29–37% recurrence and frequent steroid-dependence. Steroid-sparing agents (methotrexate, azathioprine, mycophenolate) reduce relapse, while biologics (infliximab, rituximab) are effective in refractory cases. Radiotherapy serves as salvage for resistant disease, and surgery is mainly diagnostic or for selected structural indications. Prognosis depends on treatment timing and early response, with multidisciplinary care improving diagnostic accuracy and long-term outcomes. **Conclusion:** NSOI requires immediate diagnosis, tailored treatment, and multidisciplinary care to maximize results. Corticosteroids are the foundation, but immunosuppressants or biologics can be needed. Uniform diagnostic criteria and prospective studies are needed to optimize management algorithms and maximize long-term disease control.

Keywords: CT scan, MRI, Corticosteroid, Azathioprine, Rituximab, and Idiopathic Orbital Inflammation.

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Introduction

Non-Specific Orbital Inflammation (NSOI) or Idiopathic Orbital Inflammation (IOI) or orbital pseudotumor or idiopathic orbital inflammatory syndrome (IOIS) is an idiopathic, noninfectious, nonsystemic, and nonneoplastic type of inflammation limited to the orbital tissues.[1,2] Though classified as having an idiopathic status, NSOI is currently referred to as a heterogeneous spectrum of illnesses rather than as a homogenous pathological process with several clinical and histopathologic subtypes like orbital myositis, idiopathic dacryoadenitis, IgG4-related orbital inflammation, and sclerosing orbital inflammation.[3] NSOI is viewed as the third most common orbital pathology following thyroid eye disease and lymphoproliferative diseases, which account for approximately 6–16% of all orbital inflammatory diseases.[4] The disease implicates children as well as adults, but experiences peak incidence in the fourth and fifth decades of life, with mild female predominance being noted across various cohorts.[5,6] Clinically, NSOI presents a variety of characteristic signs like acute orbital pain, proptosis, periorbital edema,

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diplopia, and occasionally vision loss.[4,7] The signs generally overlap with orbital cellulitis, thyroid-associated orbitopathy, and orbital lymphoma, and thereby render the clinical diagnosis problematic and cause delayed treatment.[7] Radiologic assessment with CT scan and MRI is required to assess soft tissue extension and classically shows extraocular muscle hypertrophy with tendon involvement, useful but not pathognomonic for NSOI.[1,2] Laboratory investigations usually yield nondiagnostic findings and are used to exclude systemic autoimmune, infective, or neoplastic processes.[8] Histopathologic evaluation, while occasionally used in atypical or recalcitrant cases, is largely nonspecific, with most samples showing nonspecific lymphoplasmacytic infiltration and varying degrees of fibrosis.[9] Therapeutically, corticosteroids represent the primary stay for the treatment of NSOI due to their immediate effect; however, the recurrence rate is rather high at a rate of up to 50–70% of cases, especially in diffuse and sclerosing forms.[10,11] In steroid-refractory, relapsing, or intolerant cases, several second-line therapies, such as immunosuppressants (e.g., azathioprine, methotrexate, mycophenolate), biologic agents (e.g., rituximab), and low-dose radiotherapy, have been attempted. There are few clinical trials, and evidence regarding long-term efficacy and safety is limited.[11,12] Besides, diagnostic doubt persists due to the absence of universal biomarkers, the absence of a uniform diagnostic algorithm, and a lack of universal consensus regarding treatment sequencing and duration.[8,13] Clinical heterogeneity in presentation and histopathologic types, coupled with overlapping signs among orbital diseases, necessitates an evidence-based, holistic approach to diagnosis and management. In light of these limitations, the current study seeks to critically review and synthesize recent evidence over the past two decades on diagnostic modalities and therapeutic approaches in NSOI. By reviewing clinical outcomes, imaging features, histopathologic categories, and treatment approaches, this review aims to summarize the current state of knowledge, highlight key gaps, and inform the formulation of standardized diagnostic criteria and evidence-based treatment algorithms to guide clinical decision-making in NSOI.

Objective

The main objective of this review was to evaluate the diagnostic and therapeutic approaches in Non-Specific Orbital Inflammation (NSOI)

Methodology & Materials

Study Design:

This study was conducted in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines. The review process was implemented in five distinct and systematic steps: (1) formulation of a focused clinical question using the PICO framework, (2) comprehensive literature search across selected databases, (3) screening of identified studies based on predefined eligibility criteria, (4) data extraction and quality assessment and (5) synthesis and interpretation of results with attention to diagnostic performance and therapeutic outcomes.

Search Method:

To ensure the systematic retrieval of relevant literature on diagnostic and therapeutic strategies for Non-Specific Orbital Inflammation (NSOI) or Idiopathic Orbital Inflammation (IOI), a comprehensive search was conducted. The search was initiated by the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guide and navigated by the PICO (Population, Intervention, Comparison, Outcome) strategy to enhance clinical specificity and methodological transparency. The search targeted studies in human populations diagnosed with IOI that examined either diagnostic methods (e.g., imaging, biopsy, histopathology) or treatments (e.g., corticosteroids, immunosuppressants, biologics, radiotherapy) and reported measurable results on clinical efficacy, safety, recurrence, or diagnostic performance. A literature search was conducted across three primary databases: PubMed (MEDLINE) and Google Scholar, selected for biomedical, ophthalmologic, and immunologic literature coverage. Searches were limited to human-subject English-language studies to ensure transparency, availability, and clinical applicability of the findings.

Data Collection Procedure

After the first phase of literature search and screening, data collection was undertaken in accordance with PRISMA guidelines to ensure transparency, reproducibility, and methodological thoroughness. All studies fitting the inclusion criteria defined before these searches were reviewed in full text for systematic data extraction. Inclusion criteria were English -language peer-reviewed journals reporting on Non-Specific Orbital Inflammation (NSOI) or idiopathic orbital inflammation (IOI) in isolation, published before July 2025. Relevant study types were randomized controlled trials, observational cohort studies, retrospective and prospective case series, and systematic reviews with extractable primary data. Excluded were studies describing orbital inflammation secondary to infectious, neoplastic, traumatic, or systemic autoimmune disease to maintain the focus on idiopathic cases.

The data obtained were synthesized and tabulated for quantitative and qualitative analysis. The trials were categorized by diagnostic approach (e.g., imaging-first vs biopsy-first) and treatment type (e.g., corticosteroid-alone, combined immunosuppressive therapy, biologics, or radiotherapy). Such categorization enabled a comparative assessment of patient outcomes and recurrence rates. All information was safely stored and version-controlled for audit, with logs maintained in full for inclusion status monitoring and reviewer comments.

Systematic and rigorous selection was carried out to identify included studies on idiopathic orbital inflammation (IOI). Ultimately, 17 studies were included in the final qualitative synthesis.

Result

Summary table on diagnosis approach, therapeutic approach, key findings, and conclusion of existing studies on Idiopathic Orbital Inflammation:

Article	Author (s)	Year published	Sample/ Participants	Diagnosis approach	Therapeutic approach	Key Findings	Conclusion
CT and MR imaging of orbital inflammation	Ferreira et al.14	2018	Review of imaging cases	MRI, CT, clinical correlation	Not applicable	MRI and CT reveal typical IOI patterns; MRI better for soft tissue	Imaging essential for diagnosis, especially MRI
Idiopathic orbital inflammation: distribution, clinical features, and treatment outcome	Yuen & Rubin4	2003	85 patients	Clinical features, imaging, biopsy in selected cases	Systemic corticosteroids; some radiotherapy	Pain, proptosis most common; 37% recurrence; steroid-dependence common	Steroids effective but relapse frequent
Idiopathic sclerosing orbital inflammation	Pemberton & Fay15	2012	Case series	Histopathology after biopsy	Steroids, immunosuppressants	Fibrotic subtype less responsive to steroids	Histologic subtype influences prognosis
Orbital inflammatory disease: Pictorial review and differential diagnosis	Pakdaman & Sepahdari16	2014	Review of imaging cases	MRI, CT, ultrasound	Not applicable	Different anatomic patterns seen on imaging	Radiology aids in differential diagnosis
Consensus on diagnostic criteria of IOI	Mombaerts et al.17	2017	Delphi panel	Consensus clinical + imaging + biopsy criteria	Not applicable	Established standardized IOI criteria	Consensus improves comparability across studies
Magnetic resonance imaging in orbital pathologies	Gokharman & Aydin18	2018	Review of MRI cases	MRI patterns in orbital disease	Not applicable	MRI shows distinct patterns for IOI vs other orbital disease	MRI key for differential diagnosis

Orbital inflammation: biopsy first	Mombaerts et al.19	2016	Expert review	Biopsy-based diagnosis	Not applicable	Early biopsy prevents misdiagnosis	Histology critical in atypical cases
The role of biopsy in diagnosing IOI	Bijlsma et al.20	2012	Clinical review	When to biopsy guidelines	Not applicable	Biopsy indicated in suspected malignancy or accessible lesions	Targeted biopsy improves accuracy
Successful treatment of IOI with infliximab	Miquel et al.21	2008	2 patients	Confirmed IOI via biopsy	Infliximab after steroid failure	Rapid symptom improvement	Infliximab viable in refractory IOI
Treatment Outcomes of Orbital Inflammatory Diseases	Al-Ghazzawi et al.11	2024	Multi-center cohort	Clinical diagnosis, imaging, biopsy	Steroids, DMARDs, biologics	Steroid-sparing reduced relapse	Biologics helpful in refractory cases
Long-term follow up of systemic rituximab therapy	Abou-Hanna et al.12	2022	Case series	Biopsy confirmed IOI	Rituximab as first-line or salvage	Good long-term control in selected cases	Rituximab can be effective as first-line in selected IOI
Radiotherapy for idiopathic inflammatory orbital pseudotumor	Sergott et al.22	1981	15 patients	Clinical, biopsy	EBRT after steroid failure	Short-term control; some relapses	Radiotherapy option when steroids fail
Radiotherapy with or without surgery for sclerosing IOI	Lee et al.23	2012	Series of refractory cases	Histopathology confirmed	Radiotherapy Â± surgery	Median long disease duration before RT; mixed success	RT can benefit selected refractory patients
Idiopathic orbital inflammatory syndrome: clinical features and treatment outcomes	Swamy et al.24	2007	73 patients	Clinical, imaging	Steroids, radiotherapy	Relapse rate 0.29/person-year	Early aggressive therapy reduces relapse
Association of clinical response and outcome with radiotherapy	Prabhu et al.25	2013	46 patients	Biopsy confirmed	Modern EBRT	Initial radiation response predicts long-term control	Good early RT response linked to better prognosis
Idiopathic orbital inflammation: review of literature and new advances	Yeşiltaş & Gündüz26	2018	Narrative review	Clinical features, imaging (MRI/CT), biopsy where needed	Steroids, immunosuppressants, biologics, radiotherapy	Summarizes classical and novel diagnostic and treatment options, including biologics and EBRT	Comprehensive reference for both diagnosis and therapy of IOI
Idiopathic orbital inflammatory disease: a multidisciplinary approach	Seyahi et al.27	2024	Case series and multidisciplinary clinical review	Clinical exam, imaging, biopsy, lab tests (autoimmune, inflammatory markers)	Steroids, immunosuppressants, biologics; coordinated rheumatology-ophthalmology care	Multidisciplinary management improves diagnostic accuracy and treatment outcomes	Interdisciplinary approach recommended for complex or refractory IOI

The literature on Non-Specific Orbital Inflammation (NSOI) spans a wide range of diagnostic and therapeutic approaches, from imaging reviews to clinical series and interventional studies. Imaging-based diagnostic characteristics took center stage in the work of Ferreira et al.[14], Pakdaman & Sepahdari[16], and Gokharman & Aydin[18], who all highlighted MRI and CT as critical tools, with MRI superior for soft-tissue resolution and pattern differentiation between IOI and other orbital pathologies. These imaging findings were augmented by Mombaerts et al.[17], who established consensus diagnostic criteria based on clinical, imaging, and biopsy parameters to standardize IOI diagnosis.

Histopathology and biopsy-driven studies, such as those of Pemberton & Fay[15], Mombaerts et al.[19], and Bijlsma et al.20, emphasized the value of directed biopsy, especially in atypical or refractory cases, for confirmation of diagnosis, guiding classification, and avoiding misdiagnosis.

Therapeutically, systemic corticosteroids remain the cornerstone of treatment, as demonstrated in large series by Yuen & Rub'n4 and Swamy et al.[24], with excellent initial response rates but a high relapse rate, necessitating adjunctive therapy. Oral prednisone is usually initiated with doses of 1–1.5 mg/kg/day for one to two weeks, followed by tapering slowly over six to twelve weeks, depending on clinical response.[4,26] In advanced or vision-threatening disease, intravenous pulses of methylprednisolone (1 g/day for three consecutive days) may be employed initially before oral therapy is resumed. Early response rates are similarly high, typically greater than 80% but the frequency of relapse remains high, particularly if tapering is rapid. Steroid-dependence, by which recurrence occurs when doses are tapered, is present in approximately 30–40% of patients. In steroid-refractory, steroid-intolerant, or steroid-dependent patients, second-line and steroid-sparing drugs are implemented to maintain remission and minimize corticosteroid use. Drugs such as methotrexate (10–25 mg/week, oral or subcutaneous), azathioprine (1–2 mg/kg/day), and mycophenolate mofetil (1–3 g/day) were attempted by Al-Ghazzawi et al.11 and Yeşiltaş & Gündüz26, all of which have been found effective in lowering recurrence rates and facilitating steroid tapering.

Biologic agents, particularly infliximab, as described by Miquel et al.[21] and rituximab in the long-term follow-up study by Abou-Hanna et al.[12], were effective in steroid-refractory or intolerant patients, with rapid symptom control and long-term remission in well-selected cases. Rituximab, an anti-CD[20] monoclonal antibody, was also successfully used as first-line therapy in carefully selected patients and as salvage therapy, with excellent long-term control.[12] These biologics are usually reserved for those unresponsive to standard immunosuppressants or for patients with the presence of concurrent systemic inflammatory disorders.

For radiotherapy, early research by Sergott et al.[22] and later reports by Lee et al.[23] and Prabhu et al.[25] demonstrated its value as a salvage therapy for steroid-resistant disease, with variable recurrence rates, and the importance of response at onset as a prognostic indicator. It is typically given at 20–30 Gy in fractionated schedules and achieves partial or complete remission in a significant proportion of patients.[22,23] Recurrence does, nonetheless, remain possible, and long-term sequelae such as radiation cataract, retinopathy, and optic neuropathy must be considered.

Surgical treatment is seldom the primary treatment for IOI. Still, it may be indicated for diagnostic purposes (incisional biopsy for suspicious lesions), debulking of fibrotic tumors in sclerosing disease, or decompression of the orbit in cases of optic nerve involvement not responsive to medical therapy. Surgical outcomes are unpredictable and are based on the associated histologic subtype.[15]

In multiple studies, response to early symptom resolution with corticosteroids is consistently high, but recurrence rates are 29% to 37%.[4,24] Median remission time is extremely variable, from several weeks in responsive cases to several months in chronic or fibrotic disease.

Finally, multidisciplinary care, as advocated by Seyahi et al.[27], bringing together expertise in ophthalmology, rheumatology, and radiology,

was associated with improved diagnostic accuracy and therapeutic outcomes. Overall, the evidence indicates that optimal IOI practice entails early diagnosis, a personalized therapeutic approach, and the willingness to escalate therapy based on disease severity, subtype, and response to initial treatment.

Clinical presentations, CT scans, and MRI images of some patients are shown in Figures 1 and 2 below.

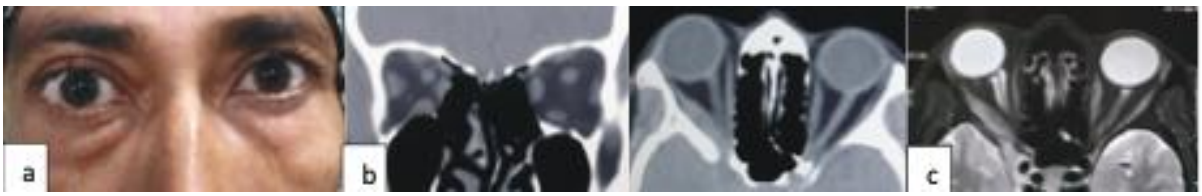

Fig 1: a) A 49-year-old female presented with right proptosis and ophthalmoplegia due to IOI, b) CT scan orbit coronal and axial view demonstrates rectus muscle hypertrophy with tendon involvement, c) T2 weighted MRI- Hyperintense enlarged rectus

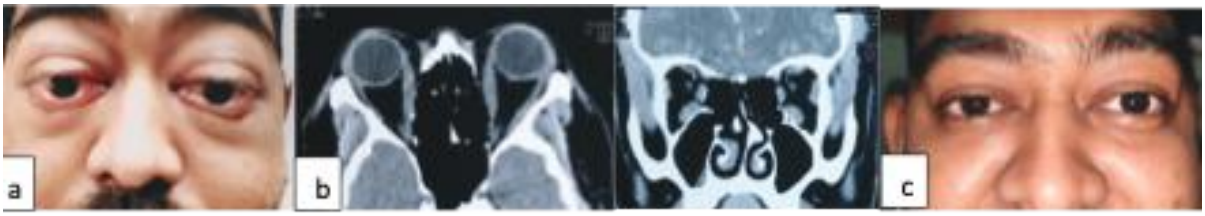

Fig 2: a) A 30-year-old male presented with bilateral proptosis, ophthalmoplegia, periocular swelling, and conjunctival prolapse due to IOI, b) CT scan orbit axial and coronal view demonstrate rectus muscle hypertrophy with tendon involvement, c) Improvement after corticosteroid therapy.

Discussion

This systematic review synthesizes current evidence on diagnostic and therapeutic strategies for Idiopathic Orbital Inflammation (IOI) from imaging-based reviews, clinical cohorts, histopathological studies, and therapeutic trials spanning more than 4 decades. The literature collectively underscores the heterogeneity of IOI in presentation, course, and response to treatment and underlines the necessity of individualized and multidisciplinary therapy.

IOI diagnosis remains a test of elimination, relying on clinical assessment, selective imaging, and selective biopsy. MRI emerged in some studies as the modality of choice for delineating soft-tissue extension, particularly for distinguishing IOI from neoplastic, infectious, or thyroid orbitopathy.[14,16,18] T2 hyperintensity, increased muscle belly size with tendon involvement (excluding thyroid eye disease), and contrast enhancement are the characteristic MRI features. CT remains relevant for acute and skeletal involvement, while ultrasound is useful for anterior or superficial lesions. The application of the consensus criteria of Mombaerts et al.[17] has increased uniformity in diagnosis but with residual variation, particularly where institution-based criteria or limited imaging availability shape practice. CT provides satisfactory delineation of osseous structures and can detect calcifications, sinusitis, or bone destruction. CT is particularly useful in emergency cases and for patients who cannot be imaged with an MRI. CT imaging in IOI will be

similar to MRI findings, but with reduced soft-tissue differentiation.[16] Biopsy emerges as an emergent procedure in atypical, bilateral, or drug-resistant instances where histopathology not just diagnoses but also enables identification of IgG4-related disease, an extension with prognostic-specific significance.[19-20] B-scan ultrasonography is an inexpensive, widely available imaging tool for evaluating anterior orbital mass and scleral thickening. It may be used in communities without advanced imaging or follow-up for superficial disease.[4]

Therapeutically, systemic corticosteroids are the foundation of first-line treatment, with the majority of patients achieving rapid symptomatic relief as outlined in studies such as Yuen & Rubin4 and Swamy et al.[24] However, the very high rates of relapse, ranging from 29% to 37% and steroid-dependence in a third of cases, necessitate early consideration of steroid-sparing interventions. Immunosuppressants (methotrexate, azathioprine, mycophenolate) have been successful in maintaining remission and reducing cumulative steroid exposure.[11,26] Biologic therapy has added to the toolkit for refractory disease. Anti–TNF agents such as infliximab achieved long-term remission in chronic myositis variants, while rituximab was both first-line and salvage effective.[12,21] These are generally reserved for severe, relapsing, or multi-system disease, often in conjunction with a rheumatologist. Radiotherapy continues to play a certain role in steroid-resistant IOI, particularly in fibrotic or sclerosing forms. Initial reports and recent EBRT regimens showed partial or complete responses, with early radiation response as a predictor of long-term control.[22,23,25] Surgery, although not as a first-line therapeutic procedure, is required for tissue diagnosis and can provide decompression or debulking in some fibrotic cases.[15]

Prognostically, outcomes vary by histologic subtype, timing of the therapy, and initial response to therapy. Fibrotic subtypes are at higher risk for relapses and might require longer immunomodulation. Delayed therapy and incomplete early response are good predictors of poor long-term outcome. Multidisciplinary care, as described by Seyahi et al.[27] has been associated with increased diagnostic accuracy, better coordination of escalating therapy, and improved long-term disease control.

Overall, though corticosteroids remain first-line therapy, the therapeutic approach to IOI has increasingly shifted toward early incorporation of steroid-sparing agents, biologics, and adjunctive modalities in judiciously selected patients. Prospective, multicenter studies grounded in uniform diagnostic criteria should guide future research to further define optimal treatment regimens and markers of outcome, particularly with regard to biologic therapy.

Limitations

This systematic review is limited by its heterogeneity, as most of the included studies are retrospective in design, use non-standardized diagnostic criteria, or have small sample sizes. Heterogeneity in treatment protocols and outcome measures made direct comparisons and meta-analyses difficult. Exclusion of non-English literature and unknown effects of publication bias may have led to an underreporting of some therapeutic methods. The use of open-access databases may therefore have limited the publication of relevant evidence behind paywalls, thereby limiting the scope of evidence reviewed.

Conclusion

This systematic review highlights that Non-Specific Orbital Inflammation is a heterogeneous, often relapsing condition requiring timely diagnosis and individualized management. MRI remains the imaging modality of choice, with biopsy reserved for atypical or refractory cases. Systemic corticosteroids remain first-line therapy, but high relapse and steroid-dependence rates necessitate early use of immunosuppressants or biologics in selected patients. Radiotherapy and surgery have defined roles in the management of resistant or structurally compromising disease. Prognosis is influenced by histologic subtype, treatment timing, and early response. Multidisciplinary collaboration optimizes outcomes, and future research should focus on standardized diagnostic criteria and prospective evaluation of advanced therapeutic strategies.

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