ISSN: 0975-8585

Research Journal of Pharmaceutical, Biological and Chemical Sciences

Clinical profile of idiopathic orbital inflammatory syndrome: An Observational study from a South Indian tertiary center

Umarani Thangapandi[1]*, Ponmani R[2], Shakila A[2], and Hemapriya AS[3].

[1]Assistant Professor, Dept of Ophthalmology, Government Krishnagiri medical college, Tamil Nadu, India.
[2]Senior Resident, Dept of Ophthalmology, Government Krishnagiri medical college, Tamil Nadu, India.
[3]Master of Public Health, Independent Researcher, Chennai, Tamil Nadu, India.

ABSTRACT

Idiopathic orbital inflammatory syndrome (IOIS) is a rare, non-infectious orbital disorder characterized by diverse clinical and radiological manifestations, often posing a diagnostic challenge. This study aimed to evaluate the demographic profile, clinical features of IOIS in patients presenting to a tertiary center in South India. In this Observational Study, both newly diagnosed cases of idiopathic orbital inflammatory syndrome (IOIS) confirmed radiologically by computed tomography (CT) and patients with recurrent or follow-up presentations were included. Demographic details, clinical presentation, ocular findings, imaging characteristics, and subtype classification were collected and analyzed. The primary outcomes assessed were age and gender distribution, laterality, clinical features, radiological involvement, and frequency of IOIS subtypes. A total of 29 patients were included. The majority were aged 21–40 years (44%), with a slight female predominance (56%). Unilateral disease occurred in 92% of cases. Pain was the most frequent symptom (80%), followed by proptosis (24%) and diplopia (16%). Limited extraocular movements were seen in 55%, and globe displacement in 41%. Visual function was preserved in most, with only one showing relative afferent pupillary defect and optic disc pallor. Imaging revealed predominant involvement of the lateral rectus (52%) and inferior rectus (48%), with lacrimal gland disease in 16.7% and orbital fat infiltration in 24%. Myositis was the most common subtype (50%), followed by diffuse disease (25%) and dacryoadenitis (16.7%). IOIS in this cohort primarily affected young to middle-aged adults, typically presenting as unilateral painful myositis with rectus involvement. Recognition of these patterns facilitates accurate diagnosis and timely management.

Keywords: Orbital Diseases, Orbital Pseudotumor, Orbital Inflammatory Syndrome, Tamil Nadu, South India.

Corresponding author

https://doi.org/10.33887/rjpbcs/2025.16.6.10

ISSN: 0975-8585

INTRODUCTION

Idiopathic Orbital Inflammatory Syndrome (IOIS) is a benign, non-specific inflammatory condition representing the third most common orbital disease in adults [1, 2]. In India, IOIS demonstrates a striking prevalence of approximately 23%, substantially higher than global averages of 8–11% [1,3]. Despite this significant burden—where inflammatory conditions account for over one-third of orbital diseases [1].

IOIS remains underdiagnosed due to variable clinical presentations and diagnostic challenges. [2, 4]. Understanding regional clinical profiles is crucial for improving diagnostic accuracy and treatment outcomes in Indian tertiary care settings [1, 5].

METHODS

This hospital-based retrospective study was conducted from January 2022 to April 2025. All patients who presented to the orbit clinic with orbital pseudotumor during the study period and met the inclusion criteria were recruited. In this retrospective study, both newly diagnosed cases of orbital pseudotumor confirmed radiologically by computed tomography (CT) and patients with recurrent or follow-up presentations were included. Cases with other identifiable conditions such as orbital tumors, thyroid orbitopathy, rheumatoid arthritis, Wegener's granulomatosis, sarcoidosis, and orbital cellulitis were excluded. This study was approved by the Institutional Ethics Committee and individual patient consent was obtained. The study adhered to the principles of the Declaration of Helsinki.

Each patient underwent a detailed ophthalmic and medical history assessment, general physical examination, and evaluation of the ear, nose, throat (ENT), and paranasal sinuses (PNS). Comprehensive ophthalmic evaluation included assessment of visual acuity, refraction, intraocular pressure, anterior segment using slit-lamp biomicroscopy, proptosis using Hertel's exophthalmometer, fundus examination with a 90D lens, colour vision testing with Ishihara's chart, and visual field analysis using HFA screen.

All patients underwent thyroid function tests and ultrasonography B-scan. Selected cases were investigated further with hematological tests including p-ANCA, c-ANCA, rheumatoid factor, and serum angiotensin-converting enzyme (ACE) levels to rule out systemic inflammatory conditions. Orbital imaging was performed using CT scan, and MRI was used when indicated. Histopathological examination was done in cases where biopsy was required for diagnostic confirmation.

RESULTS

In Table 1, the demographic and clinical characteristics of the study participants are presented. The majority of participants were between 21–40 years of age (44%), followed by those above 41 years (32%), while only 20% were aged 20 years or below. Females (56%) were slightly more represented than males (40%). Regarding the eye involved, 60% of cases affected the right eye, while 40% involved the left eye. A small proportion of participants had associated systemic illnesses such as diabetes mellitus (8%), hypertension (8%), or a combination of diabetes, hypertension, and cardiac disease (4%). Most cases were unilateral (92%), whereas only 4% presented with bilateral involvement. In terms of presenting complaints, pain was the most common symptom, reported by 80% of patients. Other notable complaints included protrusion (24%), double vision (16%), drooping (12%), and defective vision (8%), while redness was reported by 20% of participants.

Table 1: Demographic Characteristics of Study Participants

Characteristics	Frequency (n)	Percentage (%)
Age		
≤20 years	6	20%
21-40 years	13	44%
>41 years	10	32%
Gender		
Male	12	40%
Female	17	56%
Eye		

ISSN: 0975-8585

Right Eye	17	60%
Left Eye	12	40%
Systemic Illness		
Diabetes Mellitus	2	8%
Hypertension	2	8%
Diabetes/Hypertension/Cardiac	1	4%
Laterality		
Unilateral	28	92%
Bilateral	1	4%
Presenting Complaints		
Defective Vision	2	8%
Double Vision	5	16%
Droop	4	12%
Protrusion	7	24%
Pain	24	80%
Redness	6	20%

**n = number of participants*

Table 2: Clinical Examination among study participants

Examination	Frequency	Percentage
Extraocular Movements		
Full	13	45
Limited	16	55
Orbital Mass		
Not Palpable	19	65.5
Palpable	10	34.5
Pre-Orbital Rim		
Continuous	29	100
Interrupted	0	0
Globe Displacement		
Present [Axial (8), Eccentric (4)]	12	41
Absent	17	59
Globe Retropulsion		
Present	1	3.4
Absent	28	96.5
Thrill/Pulsations		
Present	0	0
Absent	29	100
Valsalva		
Positive	0	0
Negative	29	100
Conjunctiva		
Normal	18	60
Congested	12	40
Diffuse	5	16
Localized	7	24

In Table 2, the clinical examination findings of the study participants are described. More than half of the participants (55%) showed limited extraocular movements, while 45% had full ocular motility. An orbital mass was not palpable in most patients (65.5%), though it was palpable in 34.5% of cases. On assessment of the pre-orbital rim, continuity was maintained in all cases (100%). Globe displacement was observed in 41% of participants, with 8 presenting axial displacement and 4 showing eccentric displacement, whereas 59% exhibited no displacement. Globe retropulsion was positive in only one patient (3.4%) and absent in the rest (96.5%). None of the participants demonstrated orbital thrill or pulsations, nor was there any positive Valsalva response (both 100% negative). Evaluation of the conjunctiva revealed that 60% appeared normal, while 40% showed congestion. Among those congested, 16% had diffuse congestion and 24% demonstrated localized congestion.

Table 3: Ocular Examination among study participants

Examination	Frequency	Percentage
Visual Acuity		
UCVA		
6/6	18	60
6/9 to 6/36	4	12
6/60	1	4
BCVA		
6/6	7	24
6/9	1	4
6/18	1	4
6/24	2	8
Anterior Segment		
Cornea		
Clear	29	100
ACD		
Normal Depth	29	100
ACD Reaction		
None	29	100
Iris		
Normal	29	100
Pupil		
Normal	28	96.5
RAPD	1	3.4
Lens		
Clear	29	100
Posterior Segment		
Normal	28	96.5
Abnormal (Pale Disc)	1	3.4
Colour Vision, Central Fields		
Colour Vision	28	92
Central Fields	1	4
TFT	29	96
IOP		
Intra-ocular pressure	19.0±5.3mm Hg	

**UCVA = uncorrected visual acuity; BCVA = best corrected visual acuity; RAPD = relative afferent pupillary defect; IOP = intraocular pressure*

ISSN: 0975-8585

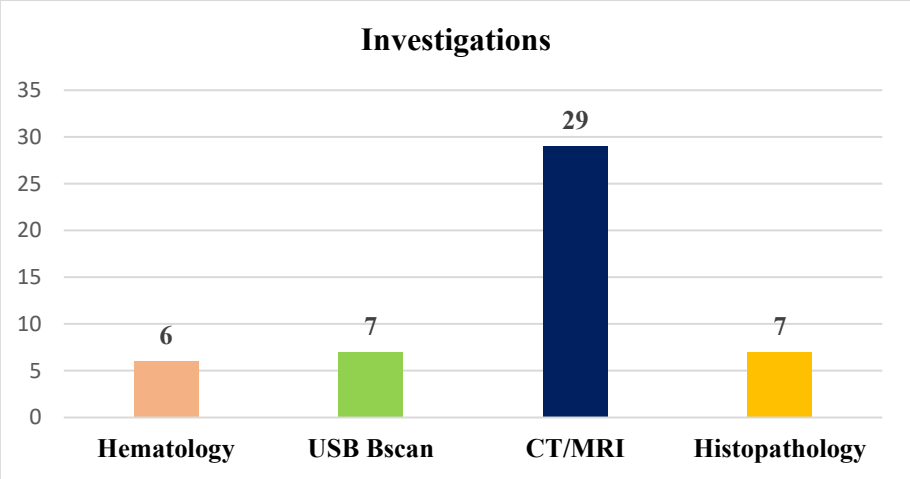
In Table 3, ocular examination findings among the study participants are summarized. Regarding visual acuity, 60% of participants had an uncorrected visual acuity (UCVA) of 6/6, while smaller proportions had reduced vision ranging from 6/9 to 6/36 (12%) and 6/60 (4%). Best corrected visual acuity (BCVA) was 6/6 in 24% of cases, with various decreases to levels such as 6/9 (4%), 6/18 (4%), and 6/24 (8%). The anterior segment examination revealed normal findings across all participants, with clear corneas, normal anterior chamber depth (ACD) and no reactions detected. The iris and lens were also normal in all cases. The pupil appeared normal in 96.5% of patients, but one patient (3.4%) exhibited relative afferent pupillary defect (RAPD). For the posterior segment, 96.5% of participants had normal findings; however, one participant (3.4%) had an abnormal pale optic disc. Color vision was intact in 92% of patients, while one patient (4%) showed central field changes. The thyroid function test (TFT) was normal in 96% of participants. The mean Intraocular pressure was 19.0±5.3mm Hg

Table 4: Lid Abnormalities among study participants

Lids	Present	Absent	Total
Lids signs	13	16	29
Ptosis	7	22	29
Lid lag	0	29	29
Lagopthalmus	0	29	29
Lid edema	12	17	29

In Table 4, the prevalence of lid abnormalities among the study participants is presented. Lid signs were observed in 13 out of 29 participants, while 16 participants showed no lid signs. Ptosis was present in 7 participants, whereas the majority (22) did not exhibit this condition. There were no cases of lid lag or lagophthalmos among the participants, as all 29 had these signs absent. Lid edema was found in 12 participants, while 17 showed no eyelid swelling.

Figure 1: Investigations done among Study Participants



In Figure 1, the usage of various diagnostic investigations among the study participants is displayed. Histopathology and ultrasound B-scan (USB Bscan) were performed in 7 participants each. Hematology tests were conducted in 6 participants. Computed tomography (CT) or magnetic resonance imaging (MRI) scans were the most commonly used diagnostic modalities, performed in all 29 participants.

Table 5: CT scan findings among study participants

CT Scan Finding	N	%
Lateral rectus	16	52
Inferior rectus	14	48
Medial Rectus	11	36

SR+LPS	12	40
Oblique Muscle	5	16
Lacrimal gland only	5	16
Lacrimal gland + EOM	5	16
Lateral rectus +SR+ IR+ Optic nerve	1	4
Orbital Fat involvement	7	24

**SR+LPS = superior rectus and levator palpebrae superioris complex;*
EOM = extraocular muscles

Table 5 summarizes the CT scan findings among the study participants. The lateral rectus muscle was the most frequently involved extraocular muscle, affected in 52% of cases, followed closely by the inferior rectus at 48%. The superior rectus and levator palpebrae superioris (SR+LPS) complex were involved in 40% of participants, while the medial rectus was affected in 36%. Involvement of the oblique muscles and the lacrimal gland alone was seen in 16% of cases each, with an additional 16% showing combined involvement of the lacrimal gland and extraocular muscles. One participant (4%) demonstrated extensive involvement including the lateral rectus, SR, inferior rectus, and optic nerve. Orbital fat involvement was observed in 24% of cases.

Table 6: Diagnosis and Classification among Study participants

Diagnosis	n	%
Diffuse	7	25
Myositis	15	50
Apical	1	4.2
Anterior	1	4.2
Lacrimal gland adenitis	5	16.7
Total	29	100

Table 6 presents the diagnosis and classification of the study participants. Myositis was the most common diagnosis, affecting 50% of participants. Diffuse disease was diagnosed in 25% of cases. Lacrimal gland adenitis accounted for 16.7% of the diagnoses. Both apical and anterior lesions were rare, each observed in 4.2% of the participants. Overall, these classifications cover the entire cohort of 29 patients.

DISCUSSION

This retrospective study contributes to the understanding of idiopathic orbital inflammatory syndrome (IOIS) by analyzing the clinical and radiological features of both newly diagnosed cases of orbital pseudotumor confirmed radiologically by computed tomography (CT) and patients with recurrent or follow-up presentations in a tertiary care center in South India.

Demographic profile: The majority of our patients were young to middle-aged adults, with the highest prevalence between 21–40 years (44%), and a slight female preponderance (56%). Similar trends have been reported in multiple series, including Yuen and Rubin [6] and Swamy et al., [7] who documented IOIS most frequently in middle-aged adults with a modest female predominance. However, Yeşiltaş and Gündüz et al. [8] reported that although IOIS can occur at any age, pediatric and elderly presentations are less frequent, supporting our findings.

Laterality: Unilateral disease was the predominant presentation (92%), with bilateral involvement in only one case. This finding is consistent with Shields et al. [9], who reported unilateral involvement in 85–90% of cases. Yuen and Rubin [6] also found unilateral disease in approximately three-quarters of their cohort.

Presenting complaints: Pain was the most common presenting symptom in our series (80%), followed by proptosis (24%), diplopia (16%), ptosis (12%), redness (20%), and defective vision (8%). These results

are consistent with prior studies, where pain and periorbital swelling were the most frequently reported complaints, often accompanied by diplopia and motility restriction [6, 10]. The slightly higher prevalence of pain in our series may reflect the predominance of the myositic subtype.

Clinical examination: Limitation of extraocular movements was observed in 55% of patients, correlating with muscle involvement on imaging. This parallels findings by Kang et al.,[10] who noted that ocular motility restriction is strongly associated with the specific rectus muscle affected. Globe displacement was seen in 41% of cases, which is within the range described by Weber et al. [11] Ptosis (24%) and lid edema (41%) were also observed, consistent with earlier descriptions of lid abnormalities as part of IOIS manifestations.[4]

Ocular examination: Most patients maintained good visual acuity, with 60% recording 6/6 unaided vision, while a minority presented with reduced acuity (16% <6/18). Best-corrected visual acuity was preserved in the majority, suggesting that visual impairment is uncommon unless the optic nerve is involved. In our study, one patient (3.4%) demonstrated a relative afferent pupillary defect (RAPD) and optic disc pallor, indicative of apical extension or optic nerve compression. Similar low frequencies of optic nerve involvement have been reported by Chaudhry et al.,[4] who emphasized that optic neuropathy is a rare but sight-threatening manifestation of IOIS. Color vision was intact in most patients (92%), with only one case showing central field involvement, again underscoring that visual pathway compromise is uncommon. The mean intraocular pressure (19.0 ± 5.3 mmHg) was within the normal range, comparable to prior series where raised IOP has not been a consistent feature of IOIS.[10,12]

Imaging features: The lateral rectus (52%) and inferior rectus (48%) were the most commonly involved muscles in our series, followed by the superior rectus/levator palpebrae superioris complex (40%) and medial rectus (36%). These findings agree with Ferreira et al., [12] who noted that the rectus muscles are the most frequently affected, particularly the lateral and superior rectus. Kang et al.[10] similarly reported preferential involvement of lateral and superior rectus. In contrast, Lakerveld et al. [13] emphasized that the inferior rectus may be the most commonly affected in certain populations. Orbital fat involvement (24%) in our study reflects the diffuse nature of disease in some cases, consistent with Weber et al. [11] Lacrimal gland involvement was present in 16.7% of cases, in line with EyeWiki [14] who reported dacryoadenitis as a frequent subtype of IOIS.

Classification: Myositis was the most common subtype (50%), followed by diffuse disease (25%) and dacryoadenitis (16.7%). This distribution is consistent with previous studies. Kubota et al.[15] identified myositis as the predominant subtype in their Japanese cohort, while Chaudhry et al. [4] also documented myositis as the most frequent manifestation. The recognition of diffuse disease and dacryoadenitis in our series underscores the heterogeneity of IOIS presentations.

Diagnostic approach: IOIS remains a diagnosis of exclusion, requiring careful differentiation from thyroid orbitopathy, orbital cellulitis, and neoplastic conditions. Our diagnostic process incorporated thyroid function testing, autoimmune serology, radiological evaluation, and selective biopsy, which is consistent with the consensus criteria established by Mombaerts et al. through a modified Delphi approach [2].

The present study confirms the predominance of unilateral, painful, myositic IOIS in young to middle-aged adults, with lateral and inferior rectus muscles most frequently involved. Most patients had preserved vision, with rare optic nerve involvement. These findings are in close agreement with global literature and emphasize the importance of early recognition. Long-term multicentric studies are required to evaluate recurrence, treatment outcomes, and the role of emerging immunomodulatory therapies.

The present study demonstrates that IOIS predominantly affects young to middle-aged adults with a slight female preponderance and is characterized mainly by unilateral, painful presentations. Myositis emerged as the most frequent subtype, with preferential involvement of the lateral and inferior rectus muscles, while visual function was generally preserved and optic nerve compromise remained rare. These findings underscore the importance of early clinical recognition of IOIS, systematic exclusion of differential diagnoses, and timely initiation of therapy. Larger multicentric studies with extended follow-up are warranted to further elucidate prognostic determinants and optimize management strategies in IOIS.

Limitation

This study has certain limitations. First, the sample size was relatively small (n = 29), reflecting the rarity of IOIS presentations in a single tertiary care center, which may limit the generalizability of the findings. Second, the study was conducted in a single institution over a limited period of time, and therefore regional referral patterns may have influenced the clinical spectrum observed. Third, long-term treatment outcomes and recurrence rates were not evaluated, restricting insights into disease prognosis and therapeutic efficacy. Finally, although imaging and selected laboratory investigations were used to exclude systemic causes, biopsy confirmation was not performed in all cases, which may pose a diagnostic challenge in atypical presentations. Future multicentric studies with larger cohorts and long-term follow-up are warranted to validate these findings and provide more robust conclusions.

ACKNOWLEDGEMENT

The authors sincerely thank the Department of Orbit and Oculoplasty, for providing access to patient records and imaging data essential for this study. We express our gratitude to all the faculty members, medical officers, and staff for their valuable support during data collection and analysis.

REFERENCES

[1] Kim UR, Khazaei H, Stewart WB, Shah AD. Spectrum of orbital disease in South India: an aravind study of 6328 consecutive patients. Ophthal Plast Reconstr Surg. 2010;26(5):315–22.

[2] Mombaerts I, Bilyk JR, Rose GE, McNab AA, Fay A, Dolman PJ, et al. Consensus on Diagnostic Criteria of Idiopathic Orbital Inflammation Using a Modified Delphi Approach. JAMA Ophthalmol. 2017 July;135(7):769–76.

[3] Rootman J. Diseases of the orbit : a multidisciplinary approach [Internet]. Philadelphia : Lippincott; 1988 [cited 2025 Sept 25]. 666 p. Available from: http://archive.org/details/diseasesoforbitm0000root

[4] Chaudhry IA, Shamsi FA, Arat YO, Riley FC. Orbital Pseudotumor: Distinct Diagnostic Features and Management. Middle East Afr J Ophthalmol. 2008;15(1):17–27.

[5] Ramdoss J, Jain A, Thejesvi G, Biswas J. Prevalence, clinical profile, investigations, and visual outcome of sarcoid intermediate uveitis in a tertiary eye care center in South India. Indian J Ophthalmol. 2022 July;70(7):2454–7.

[6] Yuen SJA, Rubin PAD. Idiopathic orbital inflammation: distribution, clinical features, and treatment outcome. Arch Ophthalmol Chic Ill 1960. 2003 Apr;121(4):491–9.

[7] Swamy BN, McCluskey P, Nemet A, Crouch R, Martin P, Benger R, et al. Idiopathic orbital inflammatory syndrome: Clinical features and treatment outcomes. Br J Ophthalmol. 2007;91(12):1667–70.

[8] Yeşiltaş YS, Gündüz AK. Idiopathic Orbital Inflammation: Review of Literature and New Advances. Middle East Afr J Ophthalmol. 2018 June;25(2):71.

[9] Shields JA, Shields CL, Scartozzi R. Survey of 1264 patients with orbital tumors and simulating lesions: The 2002 Montgomery Lecture, part 1. Ophthalmology. 2004 May 1;111(5):997–1008.

[10] Kang MS, Yang HK, Kim N, Hwang JM. Clinical Features of Ocular Motility in Idiopathic Orbital Myositis. J Clin Med. 2020 Apr;9(4):1165.

[11] Weber AL, Romo LV, Sabates NR. Pseudotumor of the orbit. Clinical, pathologic, and radiologic evaluation. Radiol Clin North Am. 1999 Jan;37(1):151–68, xi.

[12] Ferreira TA, Saraiva P, Genders SW, Buchem MV, Luyten GPM, Beenakker JW. CT and MR imaging of orbital inflammation. Neuroradiology. 2018;60(12):1253–66.

[13] Lakerveld M, van der Gijp A. Orbital Muscle Enlargement: What if It's Not Graves' Disease? Curr Radiol Rep. 2022 Feb 1;10(2):9–19.

[14] Eissa L, Abdel Razek AAK, Helmy E. Arterial spin labeling and diffusion-weighted MR imaging: Utility in differentiating idiopathic orbital inflammatory pseudotumor from orbital lymphoma. Clin Imaging. 2021 Mar;71:63–8.

[15] Kubota T, Iwakoshi A. Clinical heterogeneity between two subgroups of patients with idiopathic orbital inflammation. BMJ Open Ophthalmol [Internet]. 2022 June 6 [cited 2025 Aug 17];7(1). Available from: https://bmjophth.bmj.com/content/7/1/e001005