• Clinical Research •

Bone destruction of orbital wall in idiopathic orbital inflammatory pseudotumor: does it always imply malignancy?

Qi-Han Guo[1], Xuan Zhang[2], An-Qi Huang[3], Ben-Tao Yang[4], Rui Liu[1], Nan Wang[1], Liang-Yuan Xu[1], Jian-Min Ma[1]

[1]Department of Ophthalmology, Beijing Tongren Hospital, Capital Medical University, Beijing 100730, China
[2]The Affiliated Tai'an City Central Hospital of Qingdao University, Tai'an 271000, Shandong Province, China
[3]Department of Pathology, Beijing Tongren Hospital, Capital Medical University, Beijing 100730, China
[4]Department of Radiology, Beijing Tongren Hospital, Capital Medical University, Beijing 100730, China
Co-first authors: Qi-Han Guo and Xuan Zhang
Correspondence to: Jian-Min Ma. Department of Ophthalmology, Beijing Tongren Hospital, Capital Medical University, Beijing 100730, China. jmma@sina.com
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Abstract

● **AIM:** To assess the clinical presentations and outcomes of idiopathic orbital inflammatory pseudotumor (IOIP) patients with orbital wall bone destruction (OWBD) and to propose an expanded classification system that includes bone destruction.

● **METHODS:** The study retrospectively reviewed clinical presentations, imaging findings, treatment modalities, and outcomes of six patients diagnosed histopathologically with IOIP and OWBD at the Beijing Tongren Hospital, Capital Medical University between October 2018 and June 2021.

● **RESULTS:** Over two years, 6 (10%) of 60 IOIP patients at our hospital exhibited OWBD, but this may overrepresent severe cases. The cohort consisted of three men and three women, aged 17 to 60y (mean 35.5±16.1y). Presenting symptoms included proptosis, eyelid swelling, decreased visual acuity with pain, and palpable mass. Imaging revealed multiple anatomical structures involved with the medial wall being the most common site of bone destruction. Histopathological examination showed classic type in five patients and sclerosing type in one patient. All patients underwent surgical resection followed by methylprednisolone treatment. Follow-up (mean 30.3±3.1mo) indicated three patients had no recurrence, while others had varying degrees of symptom persistence or recurrence.

● **CONCLUSION:** IOIP with bone destruction is a rare but significant subtype that mimics malignancy, leading to potential diagnostic and therapeutic challenges. Our findings suggest that complete surgical resection combined with adjunctive glucocorticoid therapy can yield favorable outcomes. However, larger-scale studies are needed to further optimize therapeutic approaches.

● **KEYWORDS:** idiopathic orbital inflammatory pseudotumor; bone destruction; orbital disease
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INTRODUCTION

Idiopathic orbital inflammatory pseudotumor (IOIP) refers to a benign orbital mass characterized by non-specific inflammation with no identifiable local or systemic cause. It is the most common cause of painful orbital mass in adults and the third most common orbital disease[1-3]. Rootman and Nugent's[4] classification system, widely recognized in the study of IOIP, offers a structured framework for categorizing IOIP based on anatomical involvement and clinical presentation. While invaluable for its clarity and diagnostic guidance, this classification, however, falls short in encompassing the full spectrum of IOIP manifestations, particularly when it comes to cases exhibiting bone destruction. Such occurrences, albeit rare, introduce complexities not adequately addressed by Rootman and Nugent's[4] criteria, leading to potential diagnostic oversights. The rarity of bone involvement in IOIP challenges clinicians and radiologists, often steering them towards a misinterpretation of these presentations as malignant, thereby impacting treatment decisions and patient outcomes[5]. Recognizing these limitations, we advocate for an

expanded classification system that integrates bone destruction as a critical variable, aiming to refine diagnostic accuracy and enhance therapeutic strategies for IOIP, thereby filling the gaps left by the current Rootman and Nugent's[4] classification.

SUBJECTS AND METHODS

Ethical Approval The study was conducted in accordance with the principles outlined in the Declaration of Helsinki. Informed consent for publication was obtained from all subjects.

We conducted a retrospective case series study involving patients who were diagnosed with IOIP at the Beijing Tongren Hospital, Capital Medical University between October 2018 and June 2021. Patient selection was based on the following inclusion and exclusion criteria: 1) Bone destruction of the orbit wall was detected by imaging investigations, including magnetic resonance imaging (MRI) and/or computed tomography (CT); 2) Diagnosis was established by histopathology; 3) The patients did not have any identifiable cause including thyroid-associated ophthalmopathy, granulomatosis with polyangiitis (Wegener's granulomatosis), lymphoma, or infection. During the study period, a total of 60 patients were pathologically diagnosed with IOIP, among which a total of six patients were eligible. The medical records of Beijing Tongren Hospital were used to collect the patients information. All the imaging and histopathological findings were interpreted by an experienced radiologist and pathologist respectively. Biopsy sections were visualized under a Nikon ECLIPSE Ni-U microscope. The acquisition software is OPLENIC CAMERA.

RESULTS

The six patients included three men and three women, ranging in age from 17 to 60y (mean 35.5±16.1y). One patient suffered from allergic rhinitis. One patient had a history that included orbitotomy of both eyes, and the pathological result was determined to be a lipoma. The remaining four patients had no special medical history or previous ocular diseases and surgeries (Table 1).

Presenting Symptoms Two of the six patients visited our clinic for proptosis, two for swelling of the eyelid, one for decreased visual acuity with pain, one for palpable mass discovered incidentally. The average duration from onset to presentation was 6.6±4.1mo, ranging from one month to 12mo.

Clinical Manifestations Two, three, and one case affected the right, left, and both eyes, respectively. The most common complaint was proptosis and eyelid swelling (five of the seven eyes respectively), followed by reduced visual acuity (four of the seven eyes), pain (three of the seven eyes) and tearing (two of the seven eyes). On physical examination, the most common sign was palpable mass (six of the seven eyes), followed by eye movement disorder and conjunctival congestion (three of the seven eyes respectively). The mean visual acuity was 0.7±0.4, ranging from no light perception (NLP) to 1.0 (NLP is considered as zero).

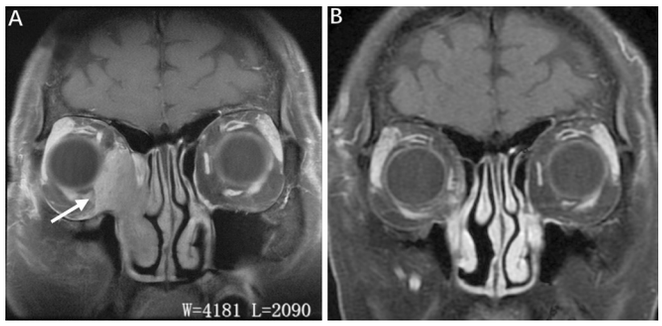

Figure 1 Coronal MRI A: A homogeneous soft tissue mass involving the lacrimal sac and nasolacrimal duct, extending to the right ethmoid sinus *via* lamina papyracea (white arrow). The medial rectus muscle, inferior oblique muscles and portions of the orbital fat were also involved. B: Almost complete resolution of the lesion one year after treatment (surgical resection and oral glucocorticoid). MRI: Magnetic resonance imaging.

Radiological Features Imaging studies including CT and/or MRI revealed six of seven eyes had more than one anatomical structure involved and one was a soft tissue mass of the lateral orbital wall. According to Rootman and Nugent's[4] classification, there were four cases of the diffuse type, two cases of the myositic type, and one case of the lacrimal type. The extraconal compartment was the most frequently involved site (five of the seven eyes) on imaging studies, with or without concurrent intraconal involvement. Imaging studies also revealed involvement of the orbital apex and extraocular muscles in three of the seven eyes, the lacrimal sac in two of the seven eyes, and the lacrimal gland in one of the seven eyes. The medial wall was the most common site of destruction (four of the seven eyes; Figure 1), followed by the lateral wall (three of the seven eyes), orbital floor and orbital roof (one of the seven eyes respectively). Given the bone destruction detected on the imaging, all the lesions were considered as probable malignancy.

Treatment All patients underwent surgical resection. The surgery aims to remove all tumors and fragmented bone pieces as thoroughly as possible. Postoperatively, five patients with more severe disease received intravenous methylprednisolone for 3 to 7d, followed by oral methylprednisolone treatment and the dose was then gradually reduced until withdrawal after 3 to 4mo. One patients was treated with oral methylprednisolone for 2wk, followed by dose tapering over 3 to 4mo.

Pathological Examination All patients were diagnosed by pathological examination. According to the histopathological classification, five patients had classic type, and one had sclerosing type[6]. Infiltration of lymphocytes and plasma cells within reactive fibrous connective tissue was seen in the classic form (Figure 2), while noticeable sclerosis with a sparse mixed

Table 1 Characteristics, clinical findings, and outcomes of IOIP patients with bone destruction

Patient	Sex	Age (y)	Side	Time to presentation (mo)	VA	Clinical manifestations	Structure involved	Bone destruction	Pathology	Treatment	Outcome	Follow up (mo)
1	F	48	OD	6	0.6	Eyelid swelling, tearing, conjunctival injection, palpable mass	Lacrimal sac, nasolacrimal duct, medial rectus muscle, lower eyelid	Medial orbital wall, orbital floor, bony nasolacrimal duct	Chronic inflammation with fibrosis	Surgical resection, oral methylprednisolone	Relief of symptoms, no recurrence	32
2	M	17	OD	6	1.0	Pain, proptosis, bluish eyelid, fluctuated visual acuity, palpable mass	A soft tissue mass of the lateral orbital wall	Lateral orbital wall	Infiltration of lymphocytes and plasma cells in fibrous connective tissue	Surgical resection, intravenous and oral methylprednisolone	Relief of symptoms, no recurrence	34
3	M	35	OS	5	1.0	Proptosis, tearing, palpable mass	Lacrimal sac, extraconal fat, medial rectus muscle	Medial orbital wall, left ethmoid sinus	Dense infiltrate of lymphocytes and plasma cells in fibrous connective tissue, with lymphoid follicle formation	Surgical resection, intravenous and oral methylprednisolone	Relief of symptoms, no recurrence	27
4	F	22	OS	4	NLP	Decreased vision, proptosis, pain, ocular movement disorder, palpable mass	Intra/extraconal mass, medial, inferior and lateral rectus muscle, orbital apex	Medial orbital wall, orbital floor, orbital roof, left ethmoid sinus, left maxillary sinus	Chronic inflammatory with fibrosis	Surgical resection, intravenous and oral methylprednisolone	Visual acuity remained NLP and experienced bilateral eyelid swelling 3mo after the surgery which improved by glucocorticoids	35
5	F	31	OS	1	1.0	Eyelid swelling, pain, conjunctival injection, palpable mass	Extraconal mass, lacrimal gland	Lateral orbital wall, the greater wing of sphenoid	Acute and chronic inflammation in fibrous connective tissue and adipose tissue, fat necrosis, histiocytes, multinucleated giant cells in certain area	Surgical resection, intravenous and oral methylprednisolone	Lost to follow-up	NA
6	M	60	OD	12	0.5	Proptosis, eyelid swelling, decreased vision, ocular movement disorder	Intra/extraconal mass extending to pterygopalatine fossa, inferior orbital fissure and infratemporal fossa, the optic nerve, the right temporal bone and surrounding soft tissue	Right temporal bone	Chronic inflammation with fibrosis	Surgical resection, intravenous and oral methylprednisolone	Mild proptosis of the right eye 22mo after the surgery	28
			OS	12	0.5	Proptosis, eyelid swelling, decreased vision, conjunctival injection, ocular movement disorder, palpable mass		Left medial orbital wall	Sparse lymphocytes and plasma cells with fibrosis, lymphoid follicle formation		No recurrence	28

IOIP: Idiopathic orbital inflammatory pseudotumor; F: Female; M: Male; VA: Visual acuity; NLP: No light perception; OD: Right eye; OS: Left eye; NA: Not available.

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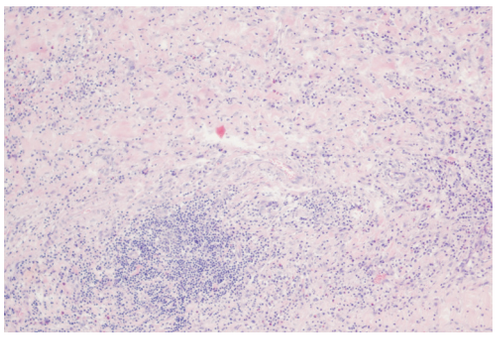

Figure 2 Hematoxylin-eosin staining shows noticeable fibrous hyperplasia accompanied by prominent foci of lymphocytic infiltration (×10).

inflammatory cell replacing normal anatomical structures was observed in the sclerosing type. Besides, lymphoid follicle formation was observed in two patients. Immunostaining showed more than 50 immunoglobulin G4-positive (IgG4+) plasma cells per high power field (HPF) in one patient, 10-15 in two patients, 2-4 in one patient and zero in two patients. The ratio of IgG4+/IgG+ plasma cells was more than 10% in one patient, 10% in one patient and 2%-4% in one patient.

Follow-up All the six patients were contacted by phone calls, and one of them was lost to follow-up. Mean follow-up time was 30.3±3.1mo. Three patients reported no recurrence and a total relief of symptoms. One patient's visual acuity remained NLP and experienced bilateral eyelid swelling 3mo after the surgery. After corticosteroid treatment, her symptom alleviated. One patient with binocular lesions experienced mild proptosis of the right eye 22mo after the surgery and did not visit any clinic.

DISCUSSION

IOIP is a diagnosis of exclusion and lacks characteristic clinical and imaging features. Although it is a benign lesion, it has the potential to locally invade the surrounding tissues[7]. Indeed, pseudotumor is named for its ability to mimic invasive malignancies on clinical presentation, imaging, and histological findings. The bony orbit is generally unaffected by IOIP, with no erosive or compressive changes in the adjacent bones on CT/MRI scan. Bone destruction in IOIP refers to the rare occurrence where the inflammatory process extends beyond soft tissue, causing erosion or damage to the orbital bones, identified through advanced imaging techniques and posing significant diagnostic and therapeutic challenges. When bone destruction is observed, it is usually considered malignant and hence further investigation, including biopsy, should be performed to exclude aggressive diseases[8].

From October 2018 to June 2021, we observed a 10% incidence rate of bone defects in patients with IOIP at our hospital. However, this incidence rate needs further investigation because the duration of this study was short, and our hospital, being specialized in ophthalmology, included a higher proportion of patients with severe conditions.

The first documented case of IOIP with bone destruction was reported by Eshaghian and Anderson[9] in 1981. Echography showed an irregular mass with bony erosion of the right orbital floor and thickening of the mucous membranes of the right maxillary sinus that was interpreted as a carcinoma of the right maxillary sinus. Therefore, the patient underwent a right Caldwell-Luc antrostomy which is considered radical and kept as a last resort[10]. The biopsy specimen is compatible with a diagnosis of pseudotumor so the patient received a right orbitotomy six days later. This case have pointed out that IOIP with bone destruction is under-recognised and this lack of awareness may cause unnecessary medical trauma to the patients, as well as psychological distress.

The most commonly used classification is based on the orbital structures involved. IOIP was first classified in 1982 by Rootman and Nugent[4] as anterior, diffuse, posterior, lacrimal, and myositic. In recent years, more classifications of IOIP had been gradually reported. Some classified "focal mass" as a single subtype while some considered "periscleritic" and "perineuritis" as subtypes[11-12], but IOIP with bone destruction have not been proposed as a classification. To date, only a few cases of this type of IOIP have been reported (Table 2)[9,13-28]. Although our series presents the largest number of cases so far, it is still a small number. However, this six patients may still reflect several characteristics of this type. Typically, dacryoadenitis and myositis are the most frequent manifestations of IOIP[29-30]. However, IOIP with bone destruction tends to damage a wide range of surrounding tissues, mimicking the aggressive growth of malignancy. Six of seven eyes had more than one anatomical structure involved including the extraocular muscles, lacrimal gland, eyeball, eyelid and so on, showing an extensive invasion. In addition, this type often damage the orbital apex. Normally, orbital apex involvement was noted in 5.3% on imaging studies[30], but our imaging investigations revealed nearly half of the affected eyes have orbital apex and/or optic nerve involvement, causing visual impairment. One patient presented with NLP and did not improve after treatment, probably due to the persistent mass effect on the tissues surrounding the optic nerve within the narrow orbital apex, despite decongestion by surgery and glucocorticoids. Posterior IOIP tends to have a serious clinical presentation with severe orbital pain, loss of visual acuity, and poor visual outcome after treatment. Studies have showed that although there was initial pain relief with high-dose steroids, visual acuity did not improve in almost all cases[31].

In terms of the diagnosis of IOIP, the most divergent views are held on the discriminative value of a corticosteroid response trial and biopsy[32-33]. A corticosteroid response is defined as a

Table 2 Previous cases of IOIP with bone destruction

Author, year	Sex	Age,y	Side	Onset time	VA	Clinical findings	Anatomic location	Bone erosion	Extra-orbital extension	Pathology	Treatment	Prognosis/follow-up
Proia[13], 2019	F	36	OD	4mo	1.0	Tearing, periorbital swelling, pain, diplopia, proptosis	Orbital apex, globe, muscles, fat	Orbital floor and lamina papyracea	Maxillary sinus and ethmoid sinus	Dense lymphoplasmacytic infiltrate, marked fibrosis	Surgical excision, oral prednisone, injection of triamcinolone acetonide	Symptoms were resolved/2mo
Pimpha[14], 2018	F	44	OD, OS	4mo	1.0	Pain, periorbital swelling and redness, diplopia	Bilateral lacrimal glands	Lateral orbital wall	None	Chronic inflammation of the lacrimal gland with multifocal lymphoid follicles, no fibrosis	Incisional biopsy, oral prednisolone	Asymptomatic/3y
Yan[15], 2016	M	56	OS	6mo	25/20	Proptosis, ocular motility limitation	Lateral recti muscle	Lateral and inferior orbital walls	Maxillary sinus	Infiltration of lymphocytes and plasma cells	Surgical excision, intravenous dexamethasone, oral prednisone	Improvement in proptosis and ocular movements/6y
Işık Balcı[16], 2015	M	4	OD	2mo	20/20	Periorbital swelling, ptosis, hypersensitivity	Mass at the lateral orbit	Zygomatic bone	Temporal fossa	Chronic inflammation with eosinophil dominant inflammatory cell infiltration	Fine needle biopsy, oral prednisone	All the problems was completely solved/9mo
Lauwers[17], 2014	M	71	OS	NA	0.4	Proptosis, periorbital swelling, ocular motility limitation, afferent pupillary reflex defect	Extraconal enhancing mass	Lamina papyracea	Ethmoid sinus, superior orbital fissure and middle cranial fossa	Fibrous stromal changes, dense infiltration of inflammatory cells, and myofibroblasts	Endoscopic biopsies, surgical excision, oral methylprednisolone	Only a mild left mechanical abduction restriction remained/1y
Ginat[18], 2012	F	89	OS	NA	NA	Proptosis	Lateral rectus muscle	Orbital floor	Maxillary sinus	NA	Surgical excision	Immediate relief of visual symptoms/NA
Shinder[19], 2012	M	18	OD	1.5mo	20/70	Proptosis, pain, decreased visual acuity, limited eye movements, diplopia, right facial hypesthesia	Lacrimal fossa, orbital apex	Orbital floor, medial wall of the maxillary sinus	Maxillary sinus	A mixture of chronic inflammatory cells with a large fibrotic component	Incisional biopsy, prednisone, methotrexate	Visual acuity improved to 20/25, visual symptoms improved except proptosis/ 15mo
Shinder[19], 2012	F	5	OD	3mo	20/30	Pain, periorbital swelling, proptosis, limited eye movements, diplopia	Lacrimal fossa, mass in the superolateral orbit	Lateral orbital wall, zygomatic bone	Temporal fossa, temporalis muscle	Chronic inflammatory cells along with fibrotic bands	Surgical excision, intralesional injection of triamcinolone, systemic prednisone	Almost complete resolution of the lesion/16mo
Shinder[19], 2012	F	35	OS	5mo	20/20	Pain, proptosis, periorbital erythema	Inferior and medial rectus muscles	Orbital floor	Maxillary sinus	Chronic inflammation and fibrosis	Biopsy, oral prednisone	Asymptomatic/26mo
Orgaz[20], 2010	M	37	OD	6mo	20/20	Epiphora, ptosis, eyelid swelling, proptosis, pain	Medial and inferior recti muscles	Turbinate bone	Right inferior nasal cavity, maxillary and ethmoid sinuses	Dense collagenous tissue with sparse infiltration of mixed inflammatory cells, occasional eosinophils and histiocytes	Surgical excision, oral prednisolone	Visual symptoms improved/1y
Zborowska[21], 2006	M	32	OS	NA	6/60	Pain, rapid deterioration of vision, proptosis	Orbital floor mass	Orbital floor	Pterygopalatine fossa, middle cranial fossa, sphenoid sinus, temporal lobe	Chronic inflammatory process with prominent fibrosis	Biopsy, intravenous methylprednisolone, intravenous cyclophosphamide and dexamethasone	Left eye visual acuity improved to 6/9, 70% regression of the mass size, symptom-free/2y
Zborowska[21], 2006	F	48	OD	4mo	6/12	Diplopia, periorbital swelling, pain, ptosis, proptosis, right relative afferent pupillary defect	Lateral and superior recti muscles, lacrimal gland, fat	Posterior portion of the greater wing of sphenoid, orbital roof	Superior orbital fissure, middle cranial fossa and anterior cavernous sinus	Dense fibrosis and a paucicellular inflammatory infiltrate of lymphocytes and histiocytes	Biopsy, pulsed steroids, cyclosporine, cyclophosphamide	Complete resolution of all orbital symptoms/2y
Cruz[22], 2003	M	35	OD	1mo	20/20	Proptosis, diplopia, pain	The lateral, inferior, and medial rectus muscles, intraconal fat	Lateral orbital wall, sphenoid bone	Pterygopalatine fossa and inferior orbital fissure	Chronic inflammatory cells with dense collagen deposition	Biopsy, radiotherapy, oral prednisone	Gradual improvement on ocular motility and relief of symptoms/8wk

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Table 2 Previous cases of IOIP with bone destruction (continued)

Author, year	Sex	Age,y	Side	Onset time	VA	Clinical findings	Anatomic location	Bone erosion	Extra-orbital extension	Pathology	Treatment	Prognosis/follow-up
Whyte[23], 1992	F	75	OD	3mo	6/24	Pain, diplopia, visual blurring, proptosis, eye movement limitation	Mass in the inferior orbit	Superior lateral antral wall	Maxillary antrum, temporal fossa	Orbital fat infiltrated by a granulomatous inflammatory lesion	Biopsy, prednisolone, radiotherapy	Visual acuity improved to 6/9 after steroid but reduced to 6/36 after radiotherapy; relief of other symptoms/NA
McNicholas[24], 1991	M	67	OD	NA	NA	Pain, proptosis, diplopia, markedly reduced vision	Orbital apex	Anterior clinoid process, optic canal, superior orbital fissure	Ethmoid air cells, the cavernous sinus, the middle cranial fossa	Inflammatory pseudotumor	Biopsy, steroid, irradiation	Complete regression of the lesion but no recovery of vision/2y
Noble[25], 1986	F	46	OS	1mo	NLP	Decreased visual acuity, ptosis	Optic foramen, sphenoid sinus mass	Floor of the sella turcica	Sphenoid sinus	Granulation tissue with acute and chronic inflammation	Surgical excision, dexamethasone, radiotherapy	Regained light perception, but intradural extension after 2y/2.5y
Frohman[26], 1986	M	72	OD	1mo	Blind	Loss of vision, pain, decreased sensation of ophthalmic nerve, ptosis, absent corneal reflex, weight loss	Orbital apex	Anterior clinoid	Superior orbital fissure, sphenoid sinus	Connective tissue with fibroblasts, histiocytes, acute and chronic inflammatory cells, and plasma cells	Biopsy, methylprednisolone sodium succinate, radiotherapy	Symptomatic relief of his pain/6wk
Frohman[26], 1986	M	48	OS	A few weeks	NLP	Loss of vision, eye movement limitation	Lateral rectus muscle, optic canal	Lateral wall of the sphenoid sinus	Sphenoid sinus	Dense collagenous connective tissue focally infiltrated with lymphocytic and plasma cells	Methylprednisolone sodium succinate	Visual acuity returned to 20/40 within 48h/NA
Frohman[26], 1986	F	48	OD	29y	NLP	Loss of vision, proptosis, eye movement limitation, absent corneal reflex	Complete orbit	Right sphenoid bone, right lamina papyracea	Right cavernous sinus, ethmoid sinus, middle cranial fossa	Sclerosing type of inflammatory pseudotumor	Prednisone, radiation therapy	Symptomatic relief was attained/NA
Kaye[27], 1984	M	71	OD	12mo	20/30	Eyelid swelling, proptosis,	Mass in the supero-medial orbit	Right and left lamina papyracea, cribriform plate	Both ethmoid sinuses, left orbit, anterior cranial fossa	Small lymphocytes, plasma cells, and some macrophages embedded in a fibrous stroma	Biopsy, prednisone, nitrogen mustard, methotrexate, x-ray therapy, intracranial excision	Temporary slight decrease in the size of the Tumor after steroid, cytotoxic agent and radiotherapy, but developed a intracranial mass/4y
Edwards[28], 1982	F	32	OS	4y	NA	Proptosis	Medial rectus muscle	Medial wall of the orbit	Ethmoid sinus	Inflammatory granulomatous process characteristic of orbital pseudotumor	Biopsy, steroid	The only residual finding was loss of the normal thin fat stripe medial to the medial rectus muscle on the left/12wk
Eshaghian[9], 1981	M	41	OD	2wk	6/6	Diplopia, hypertropia, discomfort	Mass in the right inferior orbit	Orbital floor	Maxillary sinus	Inflammatory pseudotumor, mainly histiocytes	Right caldwell-luc maxillary antrostomy, surgical excision, prednisolone	Done well with no recurrence/18mo
Eshaghian[9], 1981	F	40	OS	NA	6/6	Proptosis, intermittent sharp pain	Medial rectus muscle	Left lamina papyracea and frontal process of left maxilla	Ethmoid sinus	Inflammatory pseudotumor of mainly lymphocytes and plasma cells	Surgical excision, prednisone	Done well with no evidence of recurrence/16mo

IOIP: Idiopathic orbital inflammatory pseudotumor; NA: Not available in the literature; F: Female; M: Male; VA: Visual acuity; NLP: No light perception; OS: Left eye; OD: Right eye.

dramatic improvement of signs and symptoms within 48h after administration of systemic prednisolone, 1 mg/kg·d[34]. As for IOIP with bone destruction, we believe the diagnosis should be based on biopsy and surgical resection is warranted. Possible reasons include the following. First, surgery may remove the damaged bone pieces since spontaneous absorption on its own is very slow and sometimes even no absorption. Subsequent steroid treatment might further facilitate the inflammatory resorption during the healing process after surgery. Second, removal of the lesion may alleviate symptoms such as proptosis, diplopia, and restricted eye movement. Moreover, in instances where vision is compromised due to the mass effect, surgical resection can lead to the improvement of visual function. Besides, corticosteroid is a strong, nonspecific anti-inflammatory drug, so even lymphoma can respond to the corticosteroid treatment. Misdiagnosis causes delays in its cure which may even be life-threatening[35]. Therefore, biopsy is essential to exclude other diseases and make a definitive diagnosis. Third, corticosteroid only has a fair therapeutic effect on myositic IOIP, in contrast, with nonmyositic IOIP, the diagnostic corticosteroid trial has poor specificity and low positive predictive value. Studies have found that dacryoadenitis and sclerosing IOIP have a poor corticosteroid response of only 21% and 45% respectively[30,32]. Moreover, 16 percent of our patients have posterior involvement of the orbit, much higher than the occurrence in patients with IOIP in general. From our experience and review of the literature, complete surgical resection is the most effective option for patients with bone destruction[36]. However, as this type is often diffuse, surgery may not achieve complete removal, and thus postoperative corticosteroid adjuvant therapy is required due to their anti-inflammatory effects, which can help manage residual disease and reduce the risk of recurrence[1]. Nonetheless, the decision to proceed with surgical resection should be made after a thorough evaluation of the individual patient's condition. The specific benefits of surgery over other treatment modalities would depend on the size, location, and effects of the pseudotumor, as well as the overall health and preferences of the patient.

In conclusion, while IOIP account for approximately 8% to 11% of all orbital tumors, the specific presentation involving bone destruction is less frequent[3]. In our hospital, over a period of more than two years, 10% of patients diagnosed with IOIP exhibited bone destruction. However, as we are a tertiary hospital specializing in ophthalmology, our patient population tends to have more severe conditions. Therefore, the true frequency of bone destruction in orbital pseudotumors may be a lot lower than 10%.

IOIP with bone destruction is predominantly of the diffuse type, involving multiple orbital structures. The inner wall of the orbit is the most frequent site of bone invasion. These characteristics make it difficult to distinguish from malignant tumors, often leading to clinical misdiagnosis and incorrect treatment. We propose a new classification to enhance awareness and understanding of this condition. For this type, we believe most patients could benefit from complete surgical resection, complemented by corticosteroid therapy.

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