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Review Article

Intraorbital ophthalmic artery aneurysm: A systematic review

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ABSTRACT

Background: Rare arterial abnormalities known as intraorbital ophthalmic artery aneurysms (IOOAAs) present considerable difficulties in diagnosis and treatment. To effectively manage these uncommon illnesses, sophisticated diagnostic methods and meticulous treatment planning are needed. The purpose of this study is to thoroughly examine the clinical manifestations, diagnostic techniques, therapeutic modalities, and results of IOOAAs.

Methods: A systematic review was conducted in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines, utilizing the PubMed and Scopus databases. The search terms included "ophthalmic artery," "aneurysm," "intraorbital," and "orbit." Studies were considered if they included patients with clinically confirmed IOOAAs and were published in English. The data collected encompassed patient demographics, aneurysm characteristics, clinical presentations, diagnostic imaging techniques, treatment methods, and outcomes.

Results: An analysis was conducted on fifteen studies involving fifteen patients with an average age of 46.4 years. Females constituted 40% of the study population. The most common symptom presented was loss of vision, which was seen in 93.3% of cases, while proptosis was found in 46.7% and pain in 40%. Aneurysms were found more on the right side, 66.7%, and varied in size and morphology; saccular aneurysms constituted 33.3% of the total number of aneurysms, while fusiform constituted 20%. The availability of advanced imaging techniques, especially angiography, magnetic resonance imaging (MRI), and computed tomography scans, considerably improved the diagnosis rate. Treatment approaches included surgery in 46.7% of the total number of patients, conservative treatment in 33.3%, and endovascular treatment in 20%. The outcome from these managements was variable, with complete resolution of symptoms seen in 33.3% of patients and partial improvement in 40%. The average time of follow-up and observation was 11.18 months.

Conclusion: Ophthalmic artery aneurysms within the intraorbital compartment are rare yet present formidable challenges for their diagnosis and management. Accurate localization of the aneurysm requires the use of very advanced imaging techniques. In this regard, high-resolution MRI and computed tomography angiography play a pivotal role in the detection of these anomalies within the complex structures of the eye. Proper planning of therapy that better suits the condition and proper follow-up care is important for the best outcomes. Future research should focus on guideline standardization in management and long-term outcome improvements.

Keywords: Diagnostic imaging, Endovascular, Intraorbital aneurysm, Ophthalmic artery aneurysm, Surgical intervention

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INTRODUCTION

Intraorbital ophthalmic artery aneurysms (IOOAAs) are uncommon vascular anomalies that pose significant challenges for both diagnosis and treatment. Effectively managing these rare conditions necessitates the use of sophisticated diagnostic tools and meticulous therapeutic planning. As these aneurysms occur within the orbit, they manifest as a range of symptoms, such as pulsating exophthalmos, visual disturbances, and orbital pain, depending on their size and location. Historically, diagnosing these aneurysms has been challenging without advanced imaging techniques, leading to their underreporting. Mortada (1961)[13] noted that IOOAAs were rarely documented, and many cases lacked anatomical verification. Early reports, such as those by Guthrie (1823) and others, often lacked the detailed diagnostic imaging necessary for confirmation.

The introduction of angiographic techniques in the mid-20th century greatly enhanced the diagnostic accuracy of intraorbital aneurysms. For example, Danziger (1974)[4] reported the radiographic identification of an IOOAA, highlighting the use of imaging to reveal the characteristic choroid blush.

Contemporary literature sheds light on the infrequency of intraorbital aneurysms and the increasing challenge in their treatment. Carter and Montgomery (1989)[2] discussed the appearance of intracanalicular as well as intraorbital aneurysms, often appearing like mass lesions within the orbit that are very difficult to diagnose and even more difficult to operate.

Furthermore, the case reports of Rahmat *et al*. (1984)[15] and Struckmeyer *et al*. (2012)[18] demonstrate the wide spectrum of clinical features in orbital aneurysms and emphasize that a management plan should be individualized according to each patient`s anatomical site involved as well as tailored to the specific clinical situation.

This systematic review aims to consolidate the clinical presentations, diagnostic methodologies, and management approaches of IOOAAs, thereby providing valuable insights into this rare condition.

MATERIALS AND METHODS

Literature search

The systematic review was conducted in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines.[14] The databases PubMed and Scopus were searched using a combination of Boolean operators "OR" and "AND" with the search terms: "ophthalmic artery," "aneurysm," "intraorbital," and "orbit." The identified studies were uploaded to Rayyan, where duplicates were removed.

Study selection

The study inclusion and exclusion criteria were clearly defined. Studies were included if they involved at least one patient with a clinically confirmed diagnosis, involved aneurysms exclusively in the intraorbital segment of the ophthalmic artery, and were written in English. Studies were excluded if they were reviews, book chapters, or animal or cadaver studies.

Two reviewers independently screened the titles and abstracts of collected articles and then assessed the full texts of studies that met the inclusion criteria. Eligible articles were included based on the predetermined criteria, and references were searched to include additional relevant studies.

Data extraction

Data were extracted by one reviewer and confirmed by two independent reviewers. The extracted data included authors, year, study design, location of the study, patient age, patient gender, aneurysm type, aneurysm size, aneurysm location, aneurysm rupture, risk factors, clinical presentation, imaging and diagnosis, preoperative treatment, intra-operative treatment, postoperative treatment, follow-up duration, and outcomes. Aneurysm sizes were classified based on their measurements in millimeters as follows: small if <5 mm, medium if ≥5 mm and <10 mm, large if ≥10 mm and ≤25 mm, and giant if >25 mm.

Data synthesis and quality assessment

The primary outcomes of interest included the size of the aneurysm, any associated rupture, clinical presentation, and the outcomes following the management of the IOOAA. Each article's level of evidence was evaluated based on the 2011 Oxford Centre for Evidence-Based Medicine guidelines, and the risk of bias was assessed by two authors using the Joanna Briggs Institute checklists for case reports.[9]

RESULTS

Study selection

In this systematic review, a total of 15 studies were analyzed involving 15 patients diagnosed with IOOAAs, as detailed in Table 1.[1-6,8,9,10,12-13,16-18] The selection process is summarized in Figure 1.

Demographics

The demographic analysis indicated that the mean age of patients was 46.4 years, with a standard deviation of 14.04 years. Notably, 40% of the patients were female. This demographic data underscores the varied age range and

Table 1: An overview of intraorbital ophthalmic artery aneurysm studies: Demographics, aneurysm characteristics, clinical manifestation, treatment, and outcomes.

Author-Year	Study design	Patient age, years	Aneurysm type	Aneurysmal rupture	Risk factors	Imaging and Diagnosis
Mortada-1961[13]	Case Report	51	Fusiform	Not ruptured	Retinal arteriosclerosis	Lateral, antero-posterior, oblique skull angiogram
Rubinstein *et al*.-1968[16]	Case Report	36	Saccular	Not ruptured	Concussions, mild hypertension, obesity	Angiography
Meyerson and Lazar-1971[11]	Case Report	55	Saccular	Ruptured, hemorrhage spreading into the lids and periorbital tissues of the right eye.	Family history of intracranial aneurysms, previous head injury	Angiography
Danziger-1973[4]	Case Report	38	-	Not ruptured	-	Angiography
Rahmat *et al*.-1984[15]	Case Report	34	-	Not ruptured	Missile shell injury	X-ray, CT, Angiography
Carter and Montgomery-1989[2]	Case Report	44	Bilobed	Not ruptured	-	MRI, CT, arteriography
Ernemann-2002[6]	Case Report	64	Saccular	Not ruptured	-	MRI, angiography
Choi-2008[3]	Case Report	35	Fusiform	Not ruptured	Two minor traffic accidents	MRI, MRA, CT, Digital subtraction cerebral angiography
Struckmeyer *et al*.-2012[18]	Case Report	29	Saccular	Not ruptured	-	Contrast-enhanced CT
Zhao-2012[19]	Case Report	25	Pseudoaneurysm	Ruptured	Blunt head injury	CT, angiography, digital subtraction angiography
Della Pepa-2014[5]	Case Report	44	Saccular	Not ruptured	-	CTA, MRI, MRA, digital subtraction angiography
Hendryk-2017[9]	Case Report	75	-	Not ruptured	-	MRI, CT, CTA
Sattur *et al*.-2019[17]	Case Report	49	Fusiform	Not ruptured	-	MRI, angiography
Carey-2018[1]	Case Report	60	-	Not ruptured	Hypertension	MRI, MRA
Garala-2019[8]	Case Report	57	-	Not ruptured	Dilated cardiomyopathy, chronic kidney disease, hypertension, chronic obstructive pulmonary disease, and ulcerative colitis	Contrast-enhanced CT, angiography

(*Contd*...)

Table 1: (*Continued*).

Author-Year	Preoperative treatment	Intra-operative treatment	Postoperative treatment	Follow-up duration, months	Outcomes
Mortada-1961[13]	Compression of the left common carotid artery against Chassaignac's tubercle on the sixth cervical vertebra five times daily	Ligature of left common internal carotid artery	-	-	Symptoms resolution: Patient cured
Rubinstein *et al*.-1968[16]	-	Refused surgery	-	18	Symptoms persist
Meyerson and Lazar-1971[11]	Eye pressure bandages and sedatives	Right lateral orbitotomy using Berke's approach, necrotic mass was found and removed, bleeding controlled by pressure from a hemostat at the apex of the orbit, lateral rectus muscle resutured into position	Muscle movements full in all directions except for dextroversion, proptosis resolved within a week, postoperative angiography showed successful obliteration of the aneurysm	-	Successful obliteration of the aneurysm, preservation of the eye, though blind
Danziger-1973[4]	-	No treatment is recommended due to the absence of vision in the affected eye	-	-	With no surgical intervention, the patient remained blind in the affected eye.
Rahmat *et al*.-1984[15]	Conservative management with steroids, antibiotics, and antiepileptic drugs	Right frontotemporal craniotomy, clipping of the right ophthalmic artery at its origin from the internal carotid artery	Gradual subsidence of exophthalmos	3	Successful obliteration of the aneurysm and reduction of exophthalmos, the patient remained blind
Carter and Montgomery-1989[2]	Observation	No surgery due to the location beneath the optic nerve	-	14	Gradual improvement in visual acuity to 20/20; persistent defect in color vision; no surgical intervention
Ernemann-2002[6]	-	Surgical decompression through a pterional approach and orbital unroofing	Postoperative angiography showed the choroidal crescent supplied by external carotid collaterals	-	No recovery of visual functions

(*Contd...*)

Table 1: (*Continued*).

Author-Year	Preoperative treatment	Intra-operative treatment	Postoperative treatment	Follow-up duration, months	Outcomes
Choi-2008[3]	Observation	Endovascular treatment was chosen over surgical neck clipping due to the location near the optic nerve	Gradual recovery of visual acuity	24	Recovery of visual acuity to 20/20; successful occlusion of the aneurysm
Struckmeyer *et al*.-2012[18]	-	Lateral transcanthal approach, lateral canthotomy and preparation of the lateral orbital ligament, mobilization of the lateral rectus muscle, dissection, clipping, coagulation, and resection of the aneurysm, reattachment of the lateral palpebral ligament	Minor protrusion of the right eye receded, visual acuity improved to 20/40 after 2 days and 20/20 after 8 days	36	Complete resection of the aneurysm, full remission of visual function, no complications
Zhao-2012[19]	Conservative management initially transferred to a higher care facility due to the rapid deterioration.	Aneurysm clipping and resection through a right cranio-orbital approach under general anesthesia, fronto-temporal and superior lateral orbital rim bone flaps performed, removal of the posterior part of the orbital roof for decompression, temporary clipping of the proximal intraorbital segment ophthalmic artery to stop bleeding, resection of the pseudoaneurysm	Immediate relief of right eye luxation	1	Persistent blindness and total ophthalmoplegia in the right eye, successful relief of exophthalmos

(*Contd*...)

Table 1: (*Continued*).

Author-Year	Preoperative treatment	Intra-operative treatment	Postoperative treatment	Follow-up duration, months	Outcomes
Della Pepa-2014[5]	Conservative medical therapy with dexamethasone and heparin	No surgery	-	3	Visual acuity improved to 12/20, complete recovery of third cranial nerve palsy, partial sixth cranial nerve palsy persisted.
Hendryk-2017[9]	-	Clipping and total removal of the aneurysm through a left one-piece fronto-orbital approach	Full recovery in the left eye with good visual acuity, improved visual field, total remission of exophthalmos and ophthalmoplegia, and good cosmetic result	5	Complete resection of the aneurysm, full remission of exophthalmos and ophthalmoplegia, excellent cosmetic results
Sattur *et al*.-2019[17]	Awake balloon occlusion test (BOT) was performed for 30 minutes to assess collateral vascularity and intact visual function	Endovascular coil embolization of the aneurysm and occlusion of the parent artery performed under anesthesia, multiple Axium™ coils deployed to achieve complete occlusion	Vision and neurological function remained intact, and visual acuity in the left eye improved to 20/20, with no recurrence of the aneurysm on the CT angiogram	3	Successful occlusion of the aneurysm, preservation, and improvement of visual function
Carey-2019[1]	Conservative management with regular follow-up and serial neuroimaging studies	No surgery	-	12	Preservation of stable condition, no progression of the aneurysm, continued vision loss due to CRAO
Garala-2019[8]	Conservative management was chosen due to the low risk of rupture, low risk of intracranial hemorrhage, and the patient's comorbidities.	-	-	4	Improvement in vision and optic nerve function, stable condition with no further ocular symptoms

CT: Computed tomography, MRI: Magnetic resonance imaging, MRA: Magnetic resonance angiography, CTA: Computed tomography angiography, CRAO: Central retinal artery occlusion

gender distribution of individuals affected by this rare condition.

The distribution of study locations was as follows: the United States accounts for 4 studies at 26.67%, which is the highest among the countries. Similarly, in 4 studies, 26.67% did not specify the country of origin. The United Kingdom follows with 3 studies at 20.00%. India has 2 studies, 13.33%, while Ireland and Spain each have 1 study, 6.67%. These numbers are illustrated in Table 2.

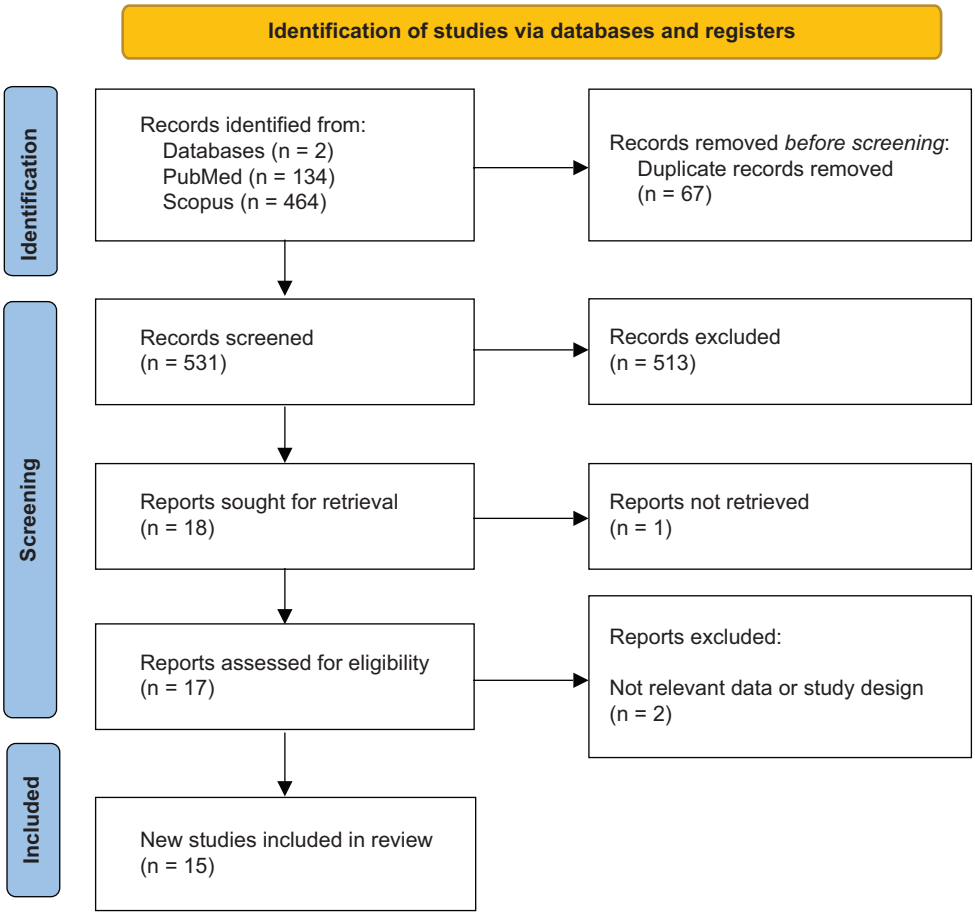

Figure 1: Preferred reporting items for systematic reviews and meta-analyses Flowchart of the included studies.

Table 2: Demographic data of patients with intraorbital ophthalmic artery aneurysm.

Number of cases	15
Age (years) mean (Standard deviation)	46.4±14.04
Gender (female) (%)	6 (40)
Location of the study (%)	
USA	4 (26.67)
Country not found	4 (26.67)
UK	3 (20.00)
India	2 (13.33)
Ireland	1 (6.67)
Spain	1 (6.67)

Risk factors and pathophysiology

IOOAAs result from a combination of systemic and local causes. The major systemic risk factors include hypertension, arteriosclerosis, and trauma. Thus, Mortada (1961)[13] reported the first case of retinal arteriosclerosis with two saccular aneurysms illustrating vascular fragility leading to aneurysm formation. Rubinstein *et al*. (1968)[16] related mild hypertension and obesity to the etiology of a saccular aneurysm, indicating systemic involvement in its emergence.

Other local factors would include trauma, and depending on the nature and force of the injury, it may cause ophthalmic artery structural weakness. Rahmat *et al*. in 1984[15] reported a case of IOOAA following a missile shell injury with a small aneurysm arising from the right ophthalmic artery. Moreover, the ophthalmic artery will have many turns and bifurcation points in its course; this tortuous anatomy may contribute to the susceptibility of these areas to aneurysmal transformation. For example, the case of blunt head trauma leading to ruptured pseudoaneurysm with rapid exophthalmos and luxation of the right eyeball was observed in Zhao's 2012[19] study. Pathophysiologically speaking, these aneurysms exert, but are not limited to, a mass effect on adjacent orbital structures due to their confined anatomical location. This was well reported in Struckmeyer *et al*. 2012[18] study, where a saccular aneurysm caused retrobulbar pain and progressive loss of visual fields, which eventually were all successfully treated.

Patient presentation

Patients exhibited a variety of symptoms associated with IOOAAs. The most prevalent symptom was reduced visual acuity, affecting 93.3% of patients and highlighting the significant impact on vision. Proptosis, characterized by abnormal eye protrusion, was observed in 46.7% of cases, while 40% of patients experienced pain. Ophthalmoplegia, characterized by paralysis or weakness of the eye muscles, was noted in 33.3% of the patients with such aneurysms. Additionally, headaches were reported by 20% of the patients, while scotoma (partial loss of vision or a blind spot) and diplopia (double vision) were each observed in only 13.3% of the cases. Ptosis, the drooping of the upper eyelid, was the least common symptom, occurring in 6.7% of the patients. These symptoms, summarized in Table 3, collectively highlight the diverse and often severe clinical manifestations of IOOAAs. IOOAAs are associated with various complications and side effects that significantly impact patient outcomes. Visual impairment was the most frequently observed complication, with 93.3% of patients reporting reduced visual acuity. Persistent visual disturbances, such as scotomas and diplopia, occurred in 13.3% of cases each, reflecting the critical impact of aneurysms on visual pathways. The striking features of proptosis were present in 46.7% and, most of the time presented an obviously cosmetic deformity with discomfort. The pain was variously described, from dull to severe, by the patients in 40% and was considerably accentuated by eye movements or palpation of the affected area. Also important were the neurological complications, especially ophthalmoplegia, in 33.3% of the patients. This condition may be dominated by paralysis/weakness of extraocular muscles, including restricted eye movements, which contribute to functional impairments such as diplopia and strabismus. Patients with ruptured aneurysms presented more serious complications: total ophthalmoplegia, blindness, and significant tissue damage, as was documented in the case reports of authors Meyerson and Lazar (1971)[11] and Zhao (2012).[19] Such observations show the pressing need for early diagnosis and timely intervention to avoid irreversible sequels.

Aneurysm location, size, type, and rupture status

The aneurysms were predominantly located on the right side, accounting for 66.7% of cases, with the remaining 33.3% on the left side. In terms of size, 33.3% of the aneurysms were classified as medium-sized (≥5 mm and <10 mm), while large (≥10 mm and ≤25 mm) and small (<5 mm) aneurysms each comprised 20% of the cases. In addition, 26.7% of the aneurysms did not have their size specified in the studies. These findings indicate that aneurysms can vary greatly in size and could tend to occur more frequently on the right side.

Table 3: Clinical manifestations of patients with intraorbital ophthalmic artery aneurysm.

Symptoms	No. (%)
Reduced visual acuity	14 (93.3)
Other	9 (60)
Proptosis	7 (46.7)
Pain	6 (40)
Ophthalmoplegia	5 (33.3)
Headache	3 (20)
Scotoma	2 (13.3)
Diplopia	2 (13.3)
Ptosis	1 (6.7)

The studies reported different types of aneurysms, with saccular aneurysms being the most prevalent, observed in 33.3% of the cases. Similarly, 33.3% of the aneurysms were not specified in the studies. Fusiform aneurysms were noted in 20% of the patients, while pseudoaneurysms and bilobed aneurysms each accounted for 6.7% of the cases.

Regarding the rupture status, the majority of the aneurysms, 86.7%, were not ruptured. Only two cases, 13.3%, and 86.7%, were not ruptured. Only two cases, 13.3%, reported ruptured aneurysms. Both cases in which ruptured aneurysms were reported had a history of head injury.

This distribution, summarized in Table 4, underscores the diversity in aneurysm morphology and the relatively low incidence of ruptures among the studied cases.

Treatment and outcomes

Various treatment approaches were employed to manage IOOAAs. Surgical intervention was the most common, used in 46.7% of cases. Conservative management, which involves close monitoring and noninvasive measures, was adopted in 33.3% of cases. Endovascular treatment, a minimally invasive procedure, was utilized in 20% of cases. Treatment outcomes varied: surgical intervention led to complete resolution of symptoms in 20% of studies (3 cases), partial improvement in 13.3% (2 cases), no improvement in 6.7% (1 case), and unspecified outcomes in another 6.7% (1 case). Conservative management resulted in complete resolution of symptoms in 1 study (6.7%), partial improvement in 2 studies (13.3%), no improvement in 1 study (6.7%), and was not specified in 1 study (6.7%). Endovascular treatment outcomes showed complete resolution of symptoms in 1 study (6.7%) and partial improvement in 2 studies (13.3%), with no studies reporting no improvement or unspecified outcomes.

Across all treatment modalities, 40% of the patients experienced partial improvement in their symptoms, 33.3% achieved complete resolution, 13.3% did not experience any

Table 4: Aneurysm characteristics among patients with intraorbital ophthalmic artery aneurysm.

Aneurysm characteristics	No. (%)
Location	
Right	10 (66.7)
Left	5 (33.3)
Size	
Medium	5 (33.3)
Not specified	4 (26.7)
Large	3 (20)
Small	3 (20)
Giant	0 (0)
Type	
Saccular	5 (33.3)
Not specified	5 (33.3)
Fusiform	3 (20)
Pseudoaneurysm	1 (6.7)
Bilobed	1 (6.7)
Rupture Status	
Not ruptured	13 (86.7)
Ruptured	2 (13.3)

improvement, and outcomes were not specified for 13.3% of the cases. These findings, detailed in Tables 5 and 6, highlight the varied efficacy of different treatment approaches and the potential for significant symptom resolution in many patients.

Postoperative outcomes

This was effective with the treatment modalities; however, the characteristics of the aneurysm, patient comorbidities, and choice of intervention also played a role, as will be demonstrated below. Regarding overall treatment modalities employed, surgical intervention was applied most frequently, at 46.7%. Of these, 20% had a complete resolution of symptoms, while 13.3% showed partial improvement. Even with successful aneurysm obliteration, complications such as persistent blindness occur. Conservative management was utilized in 33.3% of cases, particularly for patients with small-sized or asymptomatic aneurysms or in situations where surgical intervention was deemed unsuitable. The conservative modality of treatment produced complete resolution of symptoms in 6.7% and partial symptomatic improvement in another 13.3%. However, in cases such as Carey *et al*.,[1] limited success due to the persistence of visual disturbances resulting from Central Retinal Artery occlusion was seen.

The endovascular approach, utilized in 20% of cases, demonstrated outcomes comparable to those of surgical intervention and conservative management, achieving complete resolution in 6.7% of cases and partial improvement

Table 5: Treatment and outcomes in patients with intraorbital ophthalmic artery aneurysm.

Treatment Type	No. (%)
Surgical intervention	7 (46.7)
Conservative management	5 (33.3)
Endovascular treatment	3 (20)
Outcome	
Partial improvement	6 (40)
Complete resolution of symptoms	5 (33.3)
No improvement	2 (13.3)
Not specified	2 (13.3)

Table 6: Correlation between treatment modality and its corresponding outcomes.

Treatment type	Outcome	No. (%)
Surgical intervention (7 studies)		
	Complete resolution	3 (20)
	Partial improvement	2 (13.3)
	No improvement	1 (6.7)
	Not specified	1 (6.7)
Conservative management (5 studies)		
	Complete resolution	1 (6.7)
	Partial improvement	2 (13.3)
	No improvement	1 (6.7)
	Not specified	1 (6.7)
Endovascular treatment (3 studies)		
	Complete resolution	1 (6.7)
	Partial improvement	2 (13.3)
	No improvement	0 (0)
	Not specified	0 (0)

in 13.3%. This minimally invasive alternative was very useful in those with aneurysms that were located at anatomically dangerous locations or for those with high surgical risks. For illustration, Cho[3] reported a remarkable recovery of visual acuity following endovascular occlusion near the optic nerve.

Recurrence and long-term monitoring

Although none of the reviewed cases reported recurrence of the aneurysms, long-term follow-up is still essential. The average follow-up period of related studies was 11.18 months, with remarkable observations noted about recovery and residual effects. Some late complications, such as progressive visual decline or enlargement of an aneurysm, may require further intervention, with regular imaging studies such as magnetic resonance imaging (MRI) and computed tomography angiography (CTA) during follow-ups being important.

Quality of life implications

Outcomes following IOOAA management were very varied.33.3% achieved complete resolution of symptoms, 40% demonstrated partial improvement, and 13.3% remained unimproved. These findings point to the extreme complexity of IOOAA management and the necessity for highly individualized management strategies. Aneurysm type, size, location, and rupture status were all important factors in determining outcomes. Such was the case with saccular aneurysms, which were more likely to rupture and, therefore, required aggressive management. In such cases, a distinctly tailored therapeutic approach is often required.

Follow-up duration

The follow-up duration across the studies averaged 11.18 months, with a standard deviation of 11.09 months. Such an extended observation period enabled a comprehensive evaluation of both immediate and long-term results of the treatment interventions. Consequently, this led to substantial revelations on the therapy techniques and prognosis for individuals with these aneurysms.

DISCUSSION

IOOAA are rare but significant vascular anomalies that provide unique challenges during both diagnosis and management. This systematic review focuses on the clinical spectrum of presentation, features of aneurysms, approaches in treatment, and outcomes with these aneurysms. The very rare occurrence of an IOOAA in this region with complex anatomical arrangements calls for a very sophisticated and nuanced approach regarding their diagnosis and management.

Patients with IOOAAs present with variable symptoms depending on the effects of aneurysms on the ocular structures. Reduced visual acuity is the most common first symptom and is present in 93.3% of cases, reflecting the ominous nature of these aneurysms. Initially, patients may experience subtle blurring or a loss of acuity of vision, which gradually progresses as the aneurysm increases in size or exerts more pressure on the surrounding structures.

Proptosis, found in 46.7% of the patients, usually starts as an insidious bulging of the eyes [Figure 2]. It progressively increases in protrusion and is usually associated with significant discomfort, eventually leading to obvious facial asymmetry. The pain was a presenting complaint in 40% of cases, ranging from dull aching or throbbing to a severe excruciating one, and it may be present continuously or intermittently. The pain generally increases on moving the eyes or touching the affected area.

Ophthalmoplegia is a condition present in 33.3% of patients, characterized by paralysis or weakness of the muscles of the eye, which gives rise to defects or impaired movement of one or both eyes. This can also start as a limitation in the range of extraocular muscle movement and later progress to partial or complete paralysis of the muscles of the eye. As the problem worsens, the patient may experience diplopia or strabismus.

Other less common symptoms include headache reported in 20% of the patients, which may present as a dull, and persistent ache or more acute pain episodes. Scotoma observed in 13.3% of cases, is characterized by partial loss of vision or blind spots in the visual field. Diplopia, found in 13.3% of the patients, or double vision, can occur due to misalignment of the eyes caused by the growing IOOAAs, while ptosis, the drooping of the upper eyelid, reported by 6.7% of cases, often indicates significant nerve involvement and is typically a later manifestation.

Additional symptoms not previously highlighted include visual hallucinations described by Carter and Montgomery

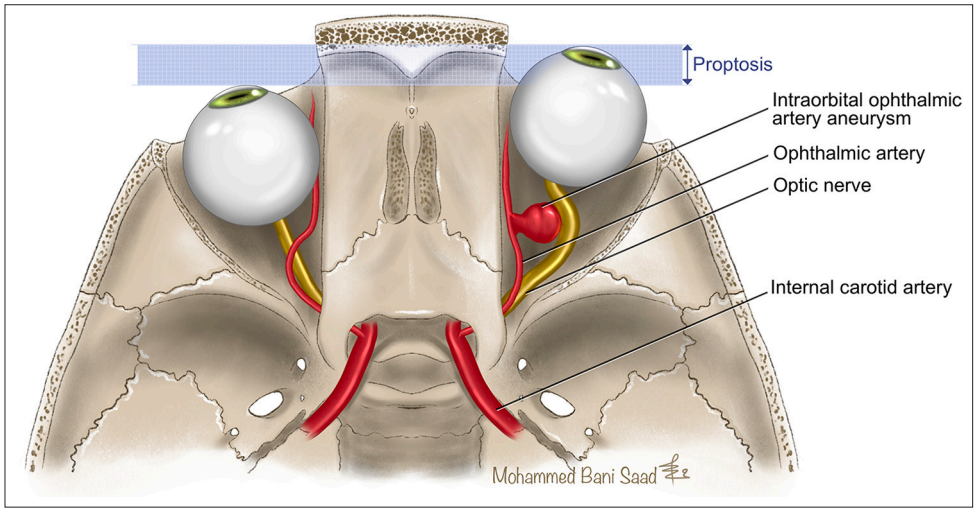

Figure 2: Illustrative view of intraorbital ophthalmic artery aneurysm.

(1989),[2] where patients saw spots of yellow with a pink center floating down from the top of their vision on either side of fixation. Rubinstein *et al*. (1968)[16] reported lacrimation (excessive tearing). Patients may also experience nystagmus (involuntary eye movement) and afferent pupillary defects, which indicate impaired optic nerve function.

Aneurysm rupture, though less common, is a critical emergency requiring immediate intervention. This review identified two cases of ruptured aneurysms. In Meyerson's and Lazar's (1971)[11] study, a 55-year-old male presented with a ruptured saccular aneurysm. The rupture caused hemorrhage into the lids and periorbital tissues, leading to acute proptosis, severe pain, and a fixed, dilated pupil with no light perception. Immediate surgical intervention was necessary to control the bleeding and remove the necrotic aneurysmal mass. Postoperatively, the aneurysm was successfully obliterated, and the eye was preserved, although the patient remained blind in the affected eye.

Another case described by Zhao (2012)[19] involved a 25-year-old male who suffered a ruptured pseudoaneurysm following blunt head trauma. This rupture led to severe headache, vomiting, rapid development of exophthalmos, and luxation (dislocation) of the right eyeball. The patient underwent urgent surgical clipping and resection of the aneurysm. Postoperatively, the patient experienced immediate relief from exophthalmos; however, he suffered persistent blindness and total ophthalmoplegia in the right eye, underscoring the severe consequences of aneurysm rupture.

Regarding the location of the aneurysms, right-sided aneurysms were predominant, accounting for 66.7% of the 15 cases, suggesting a probable anatomical predisposition. The reasons for this lateral preference are not fully understood but may relate to the vascular anatomy of the ophthalmic artery and its branches. This would be better understood through further research into anatomical variations. The sizes were varied with medium being the most common at 33.3%. The distribution also incorporated large and small, each accounting for 20% of cases. None of the aneurysms were giant. Individualized treatment plans should be made and tailored in such a way as to fit the specific characteristics of each aneurysm. Larger aneurysms, on the other hand, are of greater concern with a higher propensity to rupture and, therefore, may require more aggressive treatment plans.

Types of aneurysms identified were saccular in 33.3% of cases, fusiform in 20%, pseudoaneurysm in 6.7%, and bilobed in 6.7%. Saccular aneurysms, particularly those that are spherical or rounded with a small neck size, have an increased potential for rupture. They will require close monitoring and possibly intervention at shorter intervals because they are more prone to rupture. Fusiform aneurysms are spindle-shaped with a greater length of the artery involved, which may require different treatment measures. Pseudoaneurysms occur due to trauma and usually have some defect in the arterial wall, causing the contained hematoma. Bilobed aneurysms have two interconnecting lobes, and the management of these is more complex.

Diagnosis of IOOAAs was quite difficult during the last century. With the development of advanced imaging techniques, diagnosis has now become very accurate. Angiography, MRI, and CT scans are most important in determining the location, size, and type of aneurysms.

Angiography has been described as the gold standard by which IOOAA can be diagnosed with detail. Danziger (1974)[4] reported that angiography usually demonstrates a characteristic choroid blush related to intraorbital aneurysms. This procedure utilizes a contrast medium injected into the vascular system, followed by visualization of the vascular structures and identification of any abnormality. Angiography gives details of high-resolution images that can allow an exact evaluation of shape and location in relation to surrounding structures.

MRI and magnetic resonance angiography (MRA) are both noninvasive means of visualization without any ionizing radiation. MRI is of particular importance in the study of the soft tissues of the orbit, and it enables the surgeon to obtain images pertinent to conditions such as compression or edema of the optic nerve. MRA, as described previously, is a special MRI method that provides an operator with quality images of blood vessels accompanied by abnormalities in them. Choi (2008)[3] used both MRI and MRA to localize an aneurysm and for presurgical planning. These investigations are essential for comprehensive preoperative planning, particularly for patients who cannot undergo standard angiography.

Similarly, computed tomography and CTA offer rapid and detailed evaluation of both hard and soft tissues. CTA enhances CT imaging because the contrast used gives a clear definition of the blood vessels. This strategy works very well with the fast detection and evaluation of aneurysms, especially in case of an emergency when early diagnosis is key. The works by Rahmat *et al*. (1984)[15] and Garala (2019)[8] made use of CTA to confirm the diagnosis and determine the course of treatment in cases of IOOAAs.

Digital subtraction angiography (DSA), on the other hand, provides clearer visualization of the vessels by subtracting precontrast images from postcontrast images. DSA is highly effective in detecting aneurysms and aiding in procedural planning for endovascular interventions. Zhao (2012)[15] used DSA in the diagnosis and treatment of a ruptured pseudoaneurysm. This allowed adequate surgical planning with an excellent result.

IOOAAs are much less frequently diagnosed using ultrasound. Doppler ultrasound can be used for the evaluation of blood flow inside the aneurysm and helps

to define the hemodynamic status. There are also some drawbacks to its use. Its resolution is inferior to the one of MRI and CT scans; it indeed depends to a great extent upon the operator's skill and experience. Still, because of all these drawbacks, ultrasound can, at the same time, be an effective supplementary tool in those cases where more information concerning blood flow is important.

In terms of IOOAAs treatment, plans should be individualized according to the features of the aneurysm and the general health of the patient with the aneurysm. In this review, three phases of treatment are described: preoperative, intra-operative, and postoperative. A crucial preoperative evaluation is essential before deciding the most appropriate intervention for aneurysms. This phase includes the imaging studies detailed earlier. Such imaging techniques can then be used to define the size and location of the aneurysm with respect to neighboring structures. Such a process helps in the formulation of a precise surgical plan, which in turn assures that the intervention is effective and safe. For instance, Choi (2008)[3] used the MRI as well as DSA in precisely mapping the aneurysm anatomy before opting for an endovascular approach. Patients also receive health assessments to identify comorbidities that may influence surgical outcomes.

At times, preoperative management usually involves the administration of medication as a stabilizer of the patient's condition. For instance, in a report by Rahmat *et al*. (1984),[15] steroids, antiepileptics, and antibiotics have been used to control inflammation and prevent complications in a patient with a traumatic aneurysm. Similarly, Sattur *et al*. (2019)[17] used awake balloon occlusion tests in the study of collateral circulation and prediction of aneurysm occlusion results.

The choice of surgical approach, on the other hand, is determined by the properties and location of the aneurysm in question. Most often, the mainstream primary mode of treatment is surgical, particularly if the associated risk of rupture linked to the aneurysm is high or if the symptoms are highly severe. Techniques can range from open surgery to minimally invasive procedures performed endovascularly.

The more traditional open surgical approaches generally involve a craniotomy to access the aneurysm directly. Meyerson and Lazar (1971)[11] outlined an approach in which he needed to perform a right lateral orbitotomy with Berke's approach to remove a necrotic aneurysmal mass. Struckmeyer *et al*. (2012)[18] similarly outlined using a lateral transcanthal approach to clip and resect a saccular aneurysm. These surgical procedures almost always involve delicate dissection and handling of the muscles around the eye. The aneurysm itself has to be clipped or resected with great caution since rupture has to be avoided, and surrounding tissues have to be protected from injury.

Endovascular coil embolization and stenting are less invasive than open surgery. Choi (2008)[3] has reported a successful case of endovascular occlusion for the treatment of a large aneurysm adjacent to the optic nerve with minimal complication. The patient was treated with endovascular coil embolization and underwent an awake balloon occlusion test, resulting in improved visual function without recurrence. These techniques are particularly beneficial for aneurysms located in anatomical locations that are otherwise difficult to access or for patients presenting at a higher surgical risk. On the other hand, postoperative care is extremely important because it will allow the monitoring of recovery and also help in complication management. Patients often receive follow-up imaging to confirm whether the operation was successful and identify any remaining or repeated aneurysms. Follow-up imaging may be required over a period ranging from a few months to several years after surgery to monitor for any residual or recurrent aneurysms.

During the postoperative phase, some medications may be advised to the patients to reduce pain, infections, or inflammations. For instance, Meyerson and Lazar (1971)[11] administered sedative medicine along with pressure bandages on his patient to reduce pain and help with healing. The extent of success can be gauged through the measures of symptom resolution and functional improvement of the visual function.

The outcomes after treatment varied significantly. About 40% of patients saw partial improvement, while 33.3% experienced complete resolution of their symptoms. These results highlight the potential for significant clinical improvement with appropriate management. However, the persistence of symptoms in 13.3% of patients and the variation in follow-up durations highlight the necessity of long-term monitoring.

The correlation between treatment modalities and outcomes shows varying results. Surgical intervention, reported in seven studies, led to complete resolution in 3 cases (20%), partial improvement in 2 cases (13.3%), no improvement in 1 case (6.7%), and unspecified in 1 case (6.7%). Conservative management, discussed in five studies, resulted in complete resolution in 1 case (6.7%), partial improvement in 2 cases (13.3%), no improvement in 1 case (6.7%), and an unspecified outcome in 1 case (6.7%). Endovascular treatment in three studies showed a case fully recovered in 1 (6.7%) and partial improvement seen in 2 cases (13.3%). No improvement or unspecified result was observed.

Recurrence or residual aneurysm must be noted carefully at follow-up assessments as outcomes can vary. For example, Hendryk (2017)[9] reported that at the end of a 5-month follow-up, the patient fully recovered with excellent cosmetic results. In comparison, Ernemann (2002)[6] reported that there was even no improvement in visual functions with

surgical decompression performed, which testifies to the complexity and variableness of patient outcomes.

Long-term follow-up is essential to monitor potential late complications. Della Pepa (2014)[5] repeated the need for regular follow-up imaging studies to detect changes in aneurysm conditions to guide further management. This, with an average follow-up duration of 11.18 months, contributed majorly to enormous inferences on both the immediate and long-term outcomes. A full-time frame allows health practitioners to understand more about the course of illness and recovery of the patients to enable intervention tailoring for general well-being.

One major limitation of our study is the small sample size, reflecting the rarity of IOOAA. Our limited sample, therefore, restricts the generalizability of our findings to broader populations. However, the rarity of these aneurysms supports the importance of a systematic analysis of available cases to consolidate clinical knowledge. This limitation highlights, more than ever, the need for setting up a multicenter registry with prospective data collection and collaborative studies. The result of this would be to help gather more data on IOOAA regarding their clinical behavior and long-term outcomes. These avenues will further be used to refine diagnostic protocols, treatment guidelines, and prognostic indicators for this rare condition by expanding the evidence base.

Furst *et al*.[7] described a true ophthalmic aneurysm treated with a novel combined balloon test occlusion technique of flow diversion and adjunctive coiling. This was an occlusive test of the adequacy of collateral flow/retinal perfusion, as also confirmed before deploying an assist flow-diverting stent (Surpass Evolve) and subsequent coilings within the aneurysm dome. The treatment had very satisfying results: complete obliteration of the aneurysm while maintaining the vision as well as the neurological status of this patient intact.

The management of IOOAA involves fully informed consent regarding the risks and benefits, including discussion of alternative treatments. Complications of blindness to other deformities may result; ethical considerations regarding the treatment option to be considered are very applicable. Treatments must be in concert with the priorities of the patients and should weigh invasiveness against the conservative approach to surgical or endovascular treatment. Treatment options are most often based on the size of the aneurysm, risk of rupture, and concerns of the individual about quality of life. IOOAAs significantly impinge on the quality of life, with a loss of vision occurring in 93.3% of the patients. Thus, the management should be performed with symptomatic therapy, functional recovery, and follow-up to enable patients to achieve an optimal outcome.

The results of this review have underlined unique clinical and diagnostic challenges related to IOOAAs. The very few cases of such aneurysms necessitate high clinical suspicion and dependence on various advanced imaging modalities, including angiography and MRI/CT, for accurate diagnosis. Treatment strategies, such as surgical, endovascular, and conservative treatments, have to be tailored in a very individual way according to the size, location, and rupture condition of an aneurysm, together with the general condition and risk profile of the patient. We acknowledge the limitations of our study, including the small sample size and the heterogeneity across the included cases, which precluded the use of meta-analytical techniques. Despite these constraints, we believe the qualitative synthesis provides valuable insights into the clinical and therapeutic aspects of IOOAAs. Future studies, in this regard, should be focused on bridging the gaps that exist in knowledge by pursuing multicenter collaborations that establish registries for IOOAAs. Larger data accrual would thus be possible with such studies, shedding light on many aspects of their natural history, best management options, and long-term outcomes. Further prospective studies are also needed to elucidate the role of emerging imaging techniques, minimally invasive procedures, and advanced therapeutic tools in the care of these patients. Standardized algorithms of treatment and follow-up would further enhance uniformity in quality patient care.

CONCLUSION

IOOAAs are rare but emergent conditions that call for high suspicion for diagnosis and a highly advanced approach for their management. This review brings out their different clinical presentations in relation to the presence of decreased visual acuity, proptosis (bulging of eyes), and ophthalmoplegia (restricted eye movements). The aneurysms themselves vary in size and type; therefore, advanced imaging, in the form of angiography, MRI, or CT scan, is necessary to ascertain the correct nature of the problem. Management can range from surgical intervention to conservative care and endovascular techniques, all individualized according to the characteristics of the aneurysm and the overall health of the patient. The outcomes also vary, but for many of the patients, the symptoms show improvement; thus, this requires management with personalized treatment modalities and follow-ups. Future research has to focus on multicenter studies to have a better understanding of the IOOAA and to formulate guidelines for the management so that the outcomes for these patients can be better.

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