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Full Length Research Paper

Orbital Vascular Disorders; Clinical Aspects, Diagnostic techniques and Therapeutic Options

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Abstract

This research was undertaken to study the common presenting features, possible management options, and outcome results in a series of patients with intraorbital and extraorbital vascular disorders. Vascular lesions constitute up to 10–15 % of all orbital tumors. The shape, color, and invasiveness differ within each group: from bright red to deep blue and from a well-demarcated lesion to a more infiltrative tumor within the surrounding orbital tissue. Intraorbital vascular lesions can be categorized into arteriovenous, venous, and lymphatic disorders, each category have different clinical presentation, imaging characteristics, and management strategy. A retrospective non comparative case series study was conducted at ophthalmology department, Menoufia University Hospital, Egypt. Records of 10 patients diagnosed with orbital vascular disorders in the period from May 01, 2011 to December 31, 2013 were reviewed as regard to their diagnostic and therapeutic findings. Data of ten patients with unilateral vascular disorders were reviewed; the cohort included 4 women and 6 men with a mean age of 45 years (range, 5-66 years). Presenting findings included periocular mass (8 patients, 80%); periocular edema (6 patients, 60%); pulsation/bruit (3 patients, 30%); proptosis (5 patients, 50%); previous trauma (5 patients, 50%); elevated intraocular pressure (4 patients, 40%); pain and reduced visual acuity (2 patients each, 20%); and restriction of extraocular movements, and diplopia (1 patient, 10%). Orbital ultrasound, computed tomography (CT), magnetic resonance imaging (MRI), and computed angiography revealed specific findings according to type of vascular disorders. Treatment options ranged from conservative follow up till surgical excision. Two patients received embolization of feeder vessels; most of the patients had initial resolution of manifestations after treatment. Orbital vascular disorders can present with different symptoms and signs. Their diagnosis necessitates integrated cooperation between ophthalmological, interventional radiology, and vascular subspecialties. Computed tomography, magnetic resonance imaging and angiography are essential for both diagnosis and management. The treatment depends on patient-specific features and includes observation, embolization, and surgical excision or combined preoperative embolization/excision.

Key Words: *Orbital vascular disorders –Angiography – Orbital embolization*

Introduction

Vascular anomalies are classified into vascular tumors and vascular malformations based on the classification system of the International Society for the Study of Vascular Anomalies (ISSVA).Vascular tumors are characterized by proliferation and tumor enlargement, and the most common vascular tumor is infantile hemangioma, while a less common vascular tumor is hemangiopericytoma. Vascular malformations consist of non-proliferating vascular lesions in which blood flow is misdirected through anomalous channels. Malformations exist on a spectrum, ranging from high-flow arteriovenous malformation (AVM), low-flow venous (varix) and lymphovenous malformations, and capillary-venous malformations. Flow characteristics affect the natural history of vascular anomalies and are helpful in making the correct diagnosis.[1-2]

Patients with orbital vascular disorders seek medical advice for many reasons. Among these, exophthalmos, pain, and diplopia are most common. Patients presenting with these symptoms must undergo a comprehensive ophthalmic history and examination in order to document the type of the tumor and determine its clinical impact.[3-5]

Careful history taking is one of the most important tools available to the ophthalmologist. Main complaint usually includes periocular or retro-orbital pain, persistent or temporary visual loss, double vision or disfigurement. Appearance on initial examination is variable, as although the lesions are often congenital, they may not cause symptoms in childhood. Stimuli for growth include menarche,

pregnancy, and trauma. Common findings include periocular pain, dilated corkscrew vessels on the globe extending to the limbus, proptosis, pulsation, bruit, and raised intraocular pressure.[6]

Important ancillary tests include ultrasonography, computed tomography (CT), magnetic resonance imaging (MRI), CT angiogram and MR angiogram. Each of the three major orbital vascular malformations has its own pathognomonic characteristics.[7]

Management of orbital disorders may be difficult due to the risk of hemorrhage, vascular occlusion during treatment. The most critical concern is protection of the optic nerve and visual acuity, followed by preservation of periocular functions such as eyeball and eyelid movements.

Patients and methods

This is a retrospective non comparative case series study for all patients diagnosed with orbital vascular disorders at ophthalmology department, Menoufia university hospital, Egypt from May 1, 2011 to December 31, 2013. The study was designed according to the principles outlined in the Declaration of Helsinki and after obtaining the approval of the institutional review board at the Department of Ophthalmology, Menoufia University, Egypt, also a written consent for surgery, photography, and to be included in the study was obtained from all cases. Medical records were reviewed as regard to: age; sex; duration of symptoms and signs; clinical presentation; site involved; imaging findings on ultrasonography , angiography, computed tomography, or magnetic resonance imaging angiography were reviewed (Fig. 1 A-F); treatment modalities; and outcomes. Response to treatment was evaluated by resolution of the signs and symptoms noted at the initial examination and patient satisfaction.

Results

This study included 10 patients (4 women and 6 men) with a mean age of 45 years (range, 5-66 years). All of the cases were unilateral. The mean duration from initial clinical signs and symptoms to diagnosis and treatment was 10 years (median, 8 years; range, 6 months to 20 years). The presenting signs and symptoms are summarized in Table 1.The most common finding was periocular mass (8 patients, 80%) and periocular edema (6 patients, 60%). Computed tomography or magnetic resonance imaging localized the lesions to the superomedial quadrant of the orbit in 2 patients, the superolateral quadrant in 2 patients, posterior orbit in 1 patient and intracranial in 1 patient. Seven cases needed surgical interferences as shown in table 2 which were aspiration, embolization, surgical excision / debulking or combined embolization and debulking. Substantial symptomatic improvement was noted regarding proptosis, raised intraocular pressure, reduced visual acuity, and periocular swelling in most of the patients. Pain was persistent in 2 patients (20%), with 1 requiring further surgical intervention. Diplopia was reported in 1 patient (10%) postoperatively, which required strabismus surgery to attain binocular vision. Patients' demographics and management are outlined in Table 2. We encountered in neurophthalmology unit 3 cases of indirect carotid cavernous fistula whose Visual acuity was good, pupillary reaction was normal in both eyes. Difference in exophthalmometric readings between both eyes was 2-3 mm. Ocular motility was normal in both eyes. No bruits were noted on auscultation and the proptosis was not accentuated by Valsalva maneuver. Ophthalmologic examination of the affected eye revealed dilatation of conjunctival vessels especially in the inferior fornices and chemosis. Ophthalmoscopy was unremarkable. Neurologic examination revealed no deficits. Brain computed tomography (CT) was normal, multiplanar MRI revealed ectatic & dilated left superior ophthalmic veins. After few months, 2 cases showed raised IOP = 30-40 mm Hg (even uncontrolled by antiglaucoma eye drops).

Angiography revealed an indirect carotid cavernous fistula, feeder vessels from meningeal branches of both internal & external carotid arteries. Embolization of feeder vessels by embolic agent 6% ethylene vinyl alcohol copolymer, soluted in dimethyl-sulfoxide (onyx18) was performed in 2 patients. One month after embolization, there's improvement of symptoms, manifestations and IOP was less than 24 mmHg without any treatment .Feeder vessels were ligated in another patient, with careful excision of the malformations ensuring hemostasis but failed. Histologic analysis in cases underwent surgical debulking revealed lesions with thickened arterial and venous components, consistent with AVMs.

Illustrative case (Patient no. 3)

A 53 years old female patient presented with an one-year history of left sided headache, pain, redness and congestive symptoms together with protrusion of her left eye. She had no history of orbital trauma or family history of vascular disorders. Visual acuity was 20/30 and 20/20 in right and left eyes, respectively. Pupillary reaction was normal in both eyes. Exophthalmometric readings were 20 mm in the right and 22 mm in the left eye. Ocular motility was normal in both eyes. No bruits were noted on auscultation and the proptosis was not accentuated by Valsalva maneuver. Ophthalmologic examination of the affected eye revealed dilatation of conjunctival vessels especially in the inferior fornices and chemosis (Fig. 2-A). Ophthalmoscopy was unremarkable. Neurologic examination revealed no deficits. Brain computed tomography (CT) was normal, multiplanar MRI revealed ectatic & dilated left superior ophthalmic veins (Fig. 2-B). After 2 months, worsening of symptoms & manifestations IOP = 30-40 mm Hg (even uncontrolled by antiglaucoma eye drops). Angiography revealed a left indirect carotid cavernous fistula, feeder vessels from meningeal branches of both left internal & external carotid arteries (Fig.2-C). Endovascular treatment of left dural CCF by

embolization using Onyx 18 was done. One month after embolization, there's improvement of symptoms, manifestations and IOP was 23 mmHg without any treatment (Fig. 2-D).

Table 1: Initial Clinical Signs and Symptoms in 10 Patients with Orbital vascular Malformations

Number (%)	Manifestations
8 (80)	Periocular mass
6 (60)	Periocular swelling
3 (30)	Pulsation/bruit
6 (60)	Proptosis
5 (50)	Previous trauma
4 (40)	Elevated intraocular pressure
2 (20)	Pain
2 (20)	Reduced visual acuity
1 (10)	restriction of extraocular movements, and diplopia

Table 2: Patient Demographics, Diagnosis and Management of Orbital vascular disorders

Patient No./ sex/age	Duration of symptoms	Symptoms	Side	ultrasound CT MRI Angiography	Treatment	Progress	comments
1/M/27	7 y	Periocular mass, slowly increasing size, pulsation positive	R	AVM anterior superolateral orbit; multiple ICA feeder vessels	Refused treatment	Lost to follow-up after 3M	Boxing trauma to left side
2/M/26	5 y	Periocular mass, intermittent eyelid swelling (fig.1A)	L	CT show varices with calcification (phleboliths) posterior orbit (fig.1B)	Conservative treatment & Follow up	remission & exacerbation	Idiopathic
3/F/53	1.5 y	Pain, eyelid lesion, and bulbar conjunctival hyperemia (Fig. 2)	L	Indirect carotid cavernous fistula, feeder vessels from meningeal branches of both external & internal carotid arteries to cavernous sinus (barrow grade IV)	Embolization by onyx 18	Resolution of symptoms	Hypertensive & menopause
4/F/43	27 y	Both upper & lower lid ,Episcleral mass and swelling, increase in size with valsalva (anterior varices)	R	CT show no posterior lesion	Surgical debulking	Incomplete excision	Idiopathic
5/M/62	10 y	Periocular mass, pulsation, episcleral congestion and reduced VA (fig.1 C)	L	Giant AVM filling entire lateral orbit & upper lid ; feeder vessels identified from ophthalmic artery (fig.1 D)	Ligation of the left superficial temporal artery	Failed surgery & patient refused another treatment	Hypertensive
6/M/6	3 months	Sudden painful proptosis	R	Isolated orbital lesion without arterial or venous connection (posterior lymphangioma)	Drainage of the chocolate cyst by maxillofacial surgeon	Resolution of symptoms; follow-up 48 M	None
7/M/32	10 y	Periocular mass and swelling, 5-mm	R	AVM anterior inferolateral orbit;	Preoperative	Resolution of signs and symptoms;	None

		proptosis, decreased VA, increased IOP, edema, and congestion of episclera		feeder vessels from maxillary, facial, lingual, and ophthalmic arteries	embolization using onyx 18;surgical excision	follow-up, 12 M	
8/F/9	2 y	Several soft bluish masses in upper nasal quadrant of epibulbar surface not increase in size with valsalva (anterior lymphangioma)	R	No posterior lesion	surgical debulking	recurrence; follow-up, 12 M	Upper respiratory tract infection
9/F/70	3 months	Periocular mass, pulsation positive	R	AVM anterior superomedial orbit; multiple feeder vessels from supraorbital vessels	Surgical excision	resolution of symptoms;diplopia temporary supraorbital nerve hypoesthesia follow-up, 48 months	None
10/M/53	2 Y	Mild proptosis, periocular edema & congestion (Fig.1 E)	L	Dilated superior ophthalmic vein by orbital ultrasound (fig.1 F) AVM posterior orbit; Feeder vessels from ophthalmic artery and Internal maxillary artery	No treatment	Spontaneous thrombosis of superior ophthalmic vein; disappearance of AVM	none

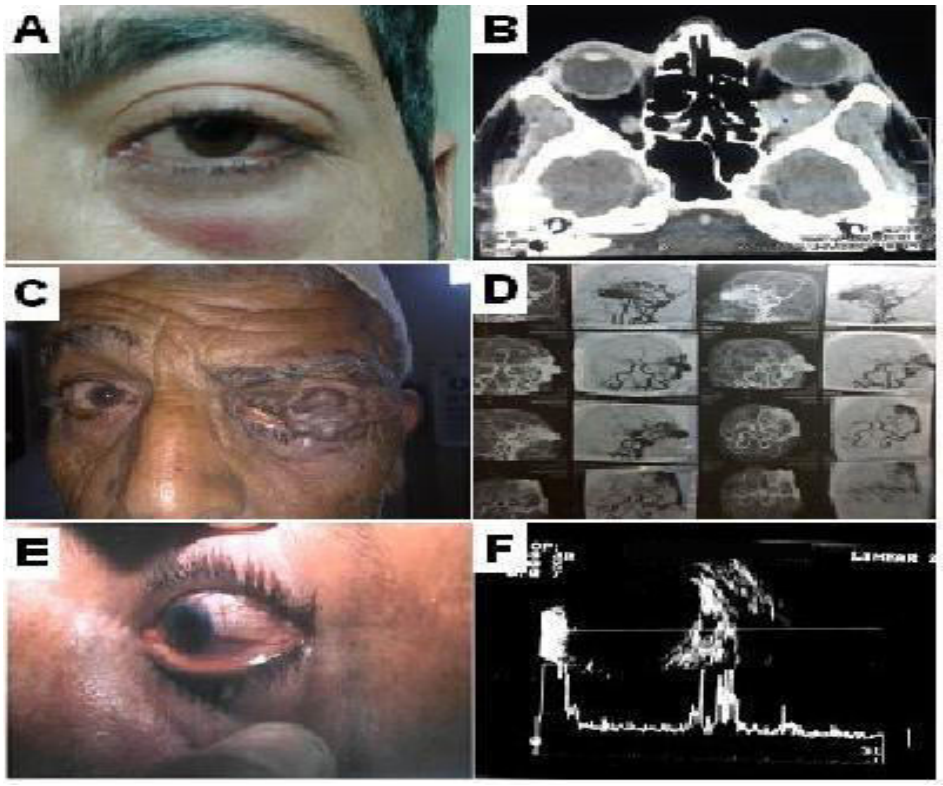

Fig.1: (A) patient no. 2 with LT lower lid congestion, (B) CT show orbital varix in posterior orbit with phlebolith , (C) patient no. 5 with giant vascular malformation in left eye, (D) angiography identify feeder vessels from ophthalmic artery, (E) patient no. 10 with low flow fistulae that mimic chronic conjunctivitis, (F) orbital ultrasound showing dilated superior ophthalmic vein of patient with low flow arteriovenous malformations

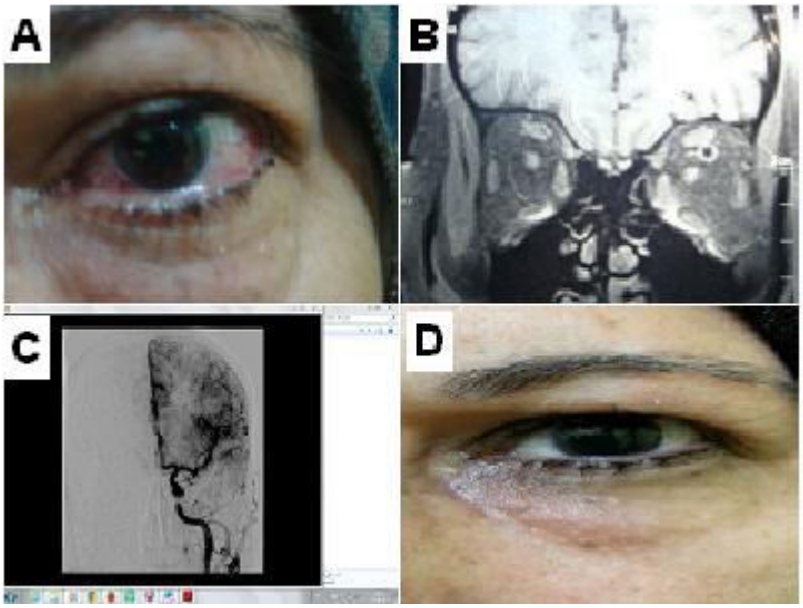

Fig.2: Patient no. 3 (A) dilatation of left conjunctival blood vessels, chemosis and lid puffiness, (B) coronal MRI show dilated left superior ophthalmic vein, (C) external carotid angiography demonstrated left cavernous sinus filling during early arterial phase, and (D) patient 1 month after embolization by onyx 18.

Discussion

Management of orbital vascular disorders is a real challenge. As regard malformations can occur at any point during the developmental sequence of the vascular system, including the arterial, venous, or lymphatic vessels, and can consist of these elements alone or in combination. Vascular malformations of the head and orbit are derived from the vessels of the brain and therefore, not infrequently, have intracranial vascular and structural components. Intraorbital vascular disorders are rare lesions.[8]

Wright[9] reported only 3 cases of arteriovenous shunts among 627 patients with various orbital diseases. In this study, we reported 5 cases with orbital AVMs, 2 cases were high flow shunts and 3 cases were low flow. Intraorbital AV disorders are most often related to an intracranial or maxillofacial arteriovenous shunts.[8] Management of intraorbital AV disorders may be difficult due to the risk of hemorrhage, vascular occlusion during treatment, and collateral damage to surrounding organs.[8] Identification of all arterial feeders from both internal and external carotid systems is critical in developing a therapeutic plan. AV disorders may be treated by surgical excision or embolization alone. However, in the hands of an experienced interventional neuroradiologist and surgical team, most AV disorders may be treated by a combined approach of preoperative embolization followed by surgical excision of the vascular mass. The goal of therapy is closure of the low-resistance shunt. In our cases, the AV disorders had different feeders' vessels; from the external carotid system, the internal carotid system or ophthalmic artery. Embolization of the external carotid system feeder vessel was performed and seems to be easier, but intervention through the ophthalmic artery was not possible without iatrogenic risks.

Chakrabortty et al [10] recorded a case report of complete removal of the intraorbital contents after unsuccessful endovascular and surgical treatment of AVM. Kim and Kosmorsky[11] reported a case of arteriovenous communication between branches of the internal and external carotid systems and the ophthalmic veins located within the orbit. Embolization therapy of the lesion resulted in a branch retinal artery occlusion. These show how much such cases are rare and difficult in their management. The therapeutic options are very limited in cases of AVM in the posterior orbit, particularly when the dura of the optic nerve is involved.[12] In our cases, because of the posterior extension of the lesion, it was not possible to remove the nidus surgically and debulking was limited to the anterior vascular loops. Significant decrease in symptoms, chemosis and exophthalmos in our patients after embolization of the feeding artery suggests that this method is useful for similar cases and may be considered as an appropriate treatment option for intraorbital AVMs.

Rootman et al [13] described their clinical and radiologic experience with orbital vascular malformations using the International Society for the Study of Vascular Anomalies (ISSVA) classification and the preferred radiologic techniques. They concluded that evolving and future treatment strategies for orbital lesions should be based on a better understanding of the pathophysiology and use of a common classification. The standard of care and reporting should include dynamic assessment before, during, and after therapy. There are possibilities for new glue agents, sclerosants, or gels that could be left in the lesions, thereby curing them via minimally invasive

image-guided methods. Also, as noted, serial injection of newer sclerosants may be useful in ablating and controlling progression of the lesions and may be combined with surgical interventions with and without gluing techniques. In this case series, we recorded 2 cases with orbital varices, one in the posterior orbit which is followed up without any interference while the other is at anterior orbit and underwent surgical debulking. Also, 2 cases of lymphangioma were recorded. One of them complicated by chocolate cyst and was aspirated by maxillofacial surgeon, the other one was excised by us.

In our case series, CT was helpful in detecting calcification (phleboliths) of orbital vairx while MRI was more diagnostic in cases of AVMs. Although, angiography is an invasive technique, it's very important when decision of surgery or sclerotherapy is taken to localize the feeder vessels. This agrees with Warrier et al, [14] who record 8 cases of orbital AVMs. The risk-benefit ratio must be evaluated on a case-by case basis before interventional management is undertaken in orbital vascular disorders. Their natural history must be understood and considered, alongside the risks of neuroradiologic and surgical interventions. Visual compromise and persistent or progressive patient discomfort are the main indicators for intervention.

Conclusion

Orbital Vascular disorders form a major multidisciplinary problem that needs comprehensive understanding of many anatomical and pathological aspects. Computed tomography, magnetic resonance imaging and angiography are essential for diagnosis and for planning the management of different orbital vascular lesions. The management of theses cases necessitates integration between ophthalmological, radiological and vascular subspecialties, this cooperation should continue throughout the management process in order to achieve the best results.

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