Guest Editorial

Vascular lesions of the orbit: Conceptual approach and recent advances

Vascular lesions of the orbit have always been an enigma to the ophthalmologist. They comprise around 7% of all orbital pathology and even to the oculoplastic surgeon have been the holy grail of all orbital disorders. Consequences included dense amblyopia, blindness, globe dystopia, orbital and orbitofacial deformity, pain and ulceration, and nonrecognition of incidental, noncontiguous clinical intracranial vascular anomalies which may result in intracranial vascular incidents with residual severe morbidity.[1] Visual loss and presentation may also vary from chronic to acute and even catastrophic.[2] In fact, many a surgeon of the past may recall disappointing and sometimes even disastrous outcomes following attempted surgical management for these challenging conditions.[3]

Principal reasons for their poor management include the lack of clear understanding of the etiopathogenesis and their natural history, disparity between terminology and the underlying pathology, and inappropriate interventions. Examples of misnomers include common entities such as "cavernous hemangiomas" and "lymphangiomas" which are not "omas" (tumors) but in fact lesions arising from dysplastic vessels and thus "malformations." Another example of erratic management is the commonly diagnosed "capillary hemangioma," a vasoproliferative tumor. The lack of awareness of the difference between a congenital hemangioma (mature at birth and less likely to spontaneously regress) from an infantile hemangioma (appears after birth and has increased likelihood of spontaneous regression) may result in various forms of mismanagement, i.e., observation of noninvoluting lesions, medical management with intralesional and systemic agents when surgical excision is more appropriate.

Historically, the vascular lesions had been labeled purely by their clinical presentation, histopathology and were purely descriptive entities, for example, cavernous hemangioma, capillary hemangioma, and orbital varices. In the ophthalmological literature, an early attempt was made to classify vascular lesions into "static" and "dynamic" lesions with attempts to embolize dynamic lesions before surgical excision.[4] Wright *et al*. attempted to classify the host of lymphangiomas and orbital varices but lumped them all together into a seamless range of "orbital venous anomalies."[5] Subsequent clinical descriptions also led to characterizing lesions based on their "distensibility" (venous anomalies) and "pulsatility" (arteriovenous anomalies). Compared to the earlier era of orbital venograms of the 70s, the advent of modern radiological techniques with computed tomography scans, magnetic resonance imaging, and interventional radiographic techniques to describe flow characteristics, guided the Orbital Society to classify lesions based on the hemodynamic properties.[6] Harris classified lesions into no flow, venous flow, and arterial flow lesions. Similarly, Rootman in 2003 classified vascular lesions based on their angiographic characteristics into Type I (no flow e.g., lymphangiomas), Type II (low flow e.g., orbital varices), and Type III (high flow e.g., arteriovenous malformations) lesions.[7] The flow characteristics not only provided preoperative information to determine appropriateness and threshold to intervene but also facilitated preoperative embolization, intralesional glue application, etc., which made debulking and excisions of the orbital lesions safer and more predictable. More recently, the "map-glue-excision technique" for previously unresectable low flow lesions involves intraoperative angiography, occlusion of the outflow with injection of contrast laden glue followed by surgical excision.[8]

Despite these numerous and progressive advances, a conceptual approach to the diagnosis and management of vascular disorders have been lacking in most centers worldwide, often with disastrous consequences and mismanagement. Even though Mulliken and Glowacki had provided a scientific classification based on endothelial characteristics of vascular lesions in 1982,[9] it was not until 1992 with subsequent modification in 2014 by the International Society for the Study of Vascular Anomalies (ISSVA)[10] that there was greater understanding and awareness of the varied spectrum of vascular disorders with gradual and steady adoption by various specialties. An essential component of the ISSVA classification was not just the clinical presentation but also incorporation of the radiological (including interventional angiographic features), histological characteristics, and the natural history which in turn guided their management. Broadly, all vascular anomalies were classified into vasoproliferative tumors and vascular malformations. The latter are then classified into simple vascular malformations (venous, lymphatic, capillary, arteriovenous malformations, and arteriovenous fistulae), one of varied combined malformations (arterial, venous, capillary, and lymphatic), those arising from major arteries and finally syndromic malformations, for example, Sturge–Weber syndrome. A simplified adaptation for the ophthalmologist is shown in Table 1.[11]

Management principles of some of the common lesions are highlighted herewith. Clear recognition of congenital and infantile hemangioma guides the physician to educate parents regarding the natural history, define appropriateness of medical intervention (propranolol, corticosteroids, etc.,) or offer surgical excision [Fig. 1].[12] Lymphatic malformations are differentiated radiologically in to microcystic and macrocystic lesions. While microcystic lesions are amenable to doxycycline, sirolimus, or surgery, macrocystic lesions respond well to sensitization of the lining endothelium by sodium tetradecyl sulfate followed by one of the various sclerosant agents, for example, bleomycin, picibanil (OK-432), or alcohol [Fig. 2].[13] Venous malformations respond to sclerosant therapy and surgical excision with intraluminal glue application when indicated. Arteriovenous malformations typically follow the natural history proposed by Schobinger.[14] These lesions typically progress from bluish, discolored mass lesions to increased vascularity with pulsation, followed by extensive growth with pain and ulceration, and finally resulting in cardiac failure. While intervention at the earliest of stages with embolization alone may be successful, later stages may require mapping of feeders with embolization of the nidus followed by excision may be necessary. Acquired arteriovenous fistula, typically

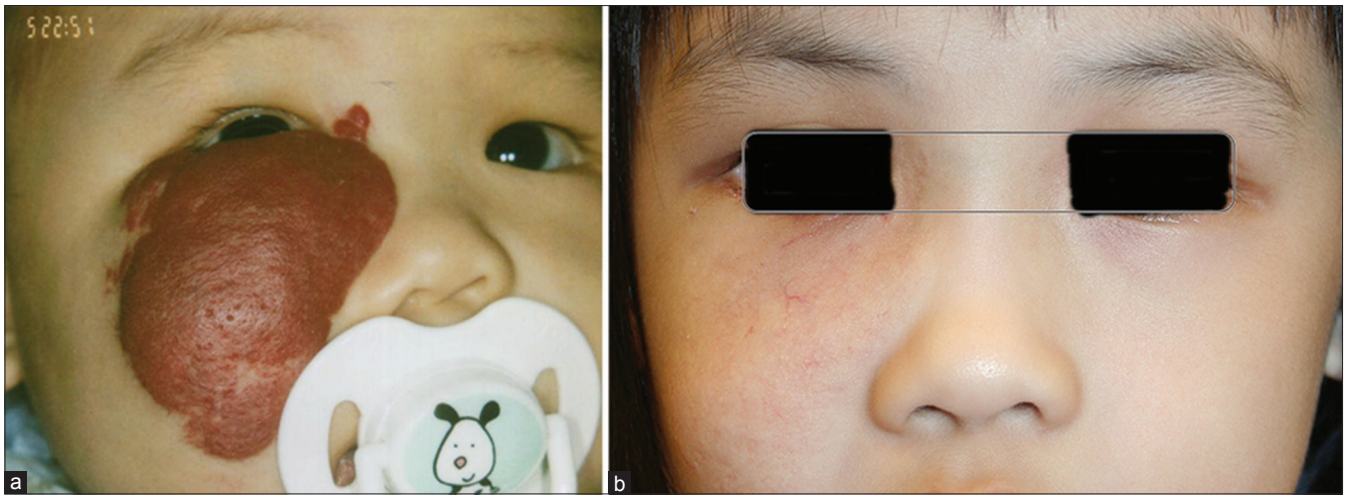


Figure 1: (a and b) Infantile hemangioma pre- and post-Rx

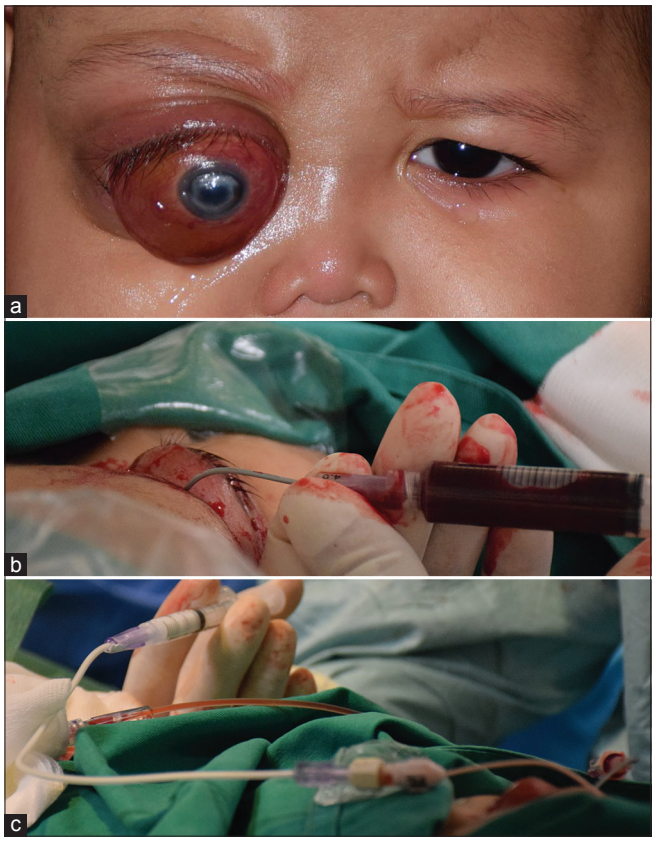


Figure 2: (a-c)Venolymphatic malformation with acute bleed, Decompression, Intralesioanl sclerotherapy

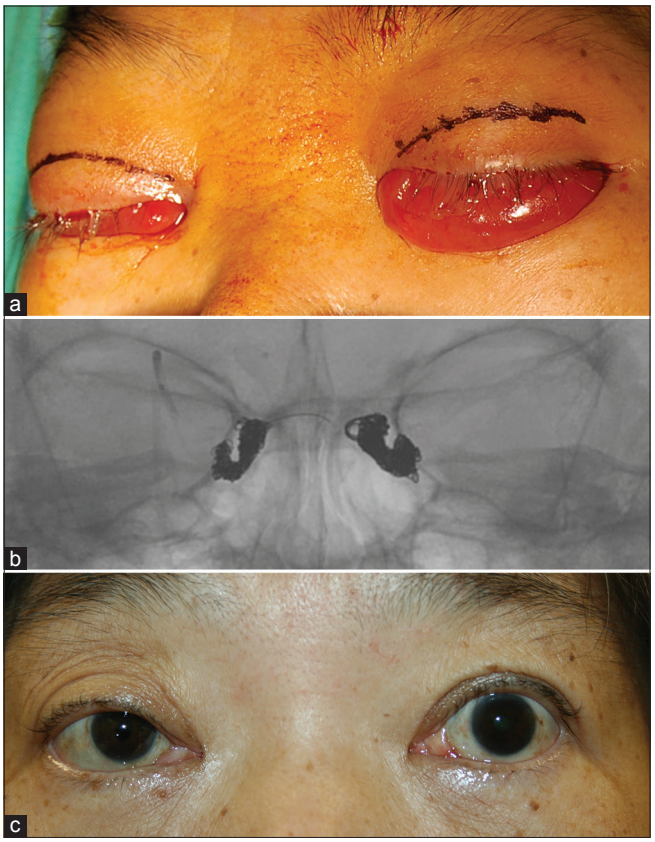


Figure 3: (a-c) Carotico-cavernous fistula preoperative view, postcoiling of bilateral cavernous sinus fistulae, and posttreatment

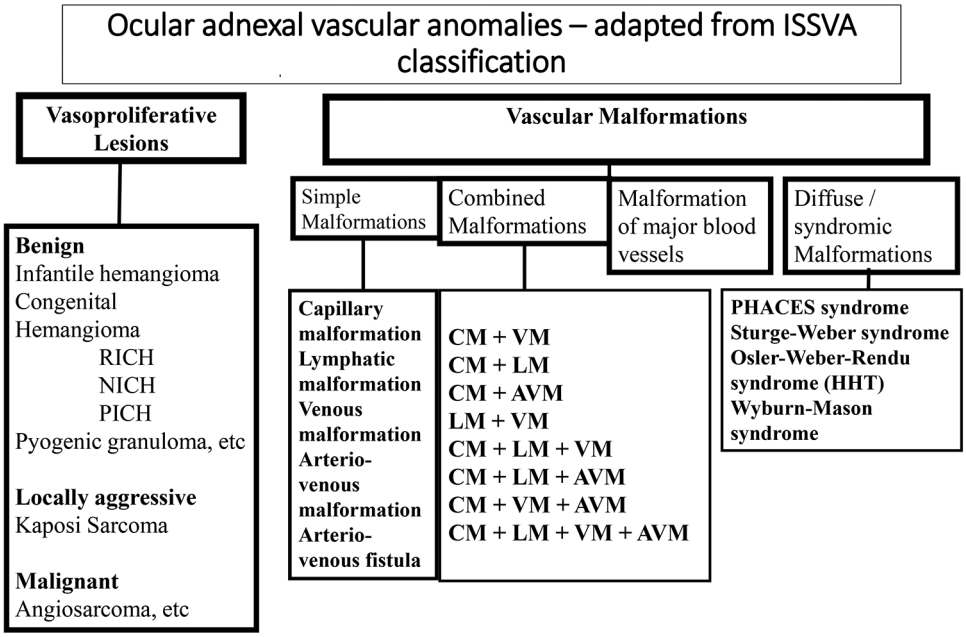
intracranial, but with ophthalmic consequences, may be managed purely by interventional neuroradiological techniques with balloons or platinum coils, occasionally through the transorbital approach [Fig. 3].[15]

To summarize, the following are some of the major contributions to the efficient management of complex vascular disorders of the orbit are listed below.

1. Conceptual approach to the diagnosis and management of vascular tumors and malformations guided by the ISSVA classification.
2. The advent of selective and superselective contrast-enhanced digital subtraction angiograms using fine catheters with application of intravascular platinum coils and contrast-laden endovascular glue, for example, ethylene-vinyl alcohol polymer (Onyx).
3. Collaborative approaches between multidisciplinary teams comprised by ophthalmologists/orbital surgeons, the

Figure 4: Hybrid operating theater

Table 1: Ocular adnexal vascular anomalies adapted from the International Society for the Study of Vacular Anomalies classification



diagnostic and interventional neuroradiologist, the pediatrician, the head-and-neck surgeon, and occasionally the neurosurgeon.

4. The introduction of hybrid operating theaters[16] [Fig. 4] which have facilitated seamless, simultaneous angiogram followed either by angioembolization and excision or direct interventional procedures to place coils or glues to the communication between the arteries and veins.

In this issue of the Indian Journal of Ophthalmology, Mukherjee *et al*. have showcased a conceptual diagnostic and surgical approach to the treatment of one such malformation of the periorbital region.[17] The case report highlights the collaboration between the clinician and the radiologist, the value of digital subtraction angiography to identify the feeder vessels and the flow characteristics, injection of contrast-laden glue into the lesion followed by an excision. This case report also highlights the often seen misdiagnosis and mismanagement, delay in appropriate treatment resulting in dense amblyopia with disfiguration, and the role of the orbital surgeon working in a tertiary care center as a member of a multidisciplinary team.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Access this article online

Quick Response Code:

Website: www.ijo.in

DOI: 10.4103/ijo.IJO_1272_17

Cite this article as: Sundar G. Vascular lesions of the orbit: Conceptual approach and recent advances. Indian J Ophthalmol 2018;66:3-6.

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Dr Gangadhara Sundar is currently the Head of Orbit and Oculofacial Surgery, with the Department of Ophthalmology, National University Hospital, Singapore. Having had his basic training in Chennai at the Madras Medical College and the Regional Institute of Ophthalmology/Government Ophthalmic Hospital and advanced subspecialty training in Oculoplastic Surgery and Ocular Oncology at Henry Ford Hospital, Detroit, he has since furthered the cause of advancing Oculoplastic Surgery and Ophthalmic Oncology in Singapore and Southeast Asia. Having trained residents and fellows from South Asia, Asia Pacific region and the West, his special interests include Ophthalmic Oncology, Thyroid Eye Disease, Orbitofacial Trauma, and minimally invasive orbitofacial and lacrimal surgery. A strong believer of multidisciplinary, multimodality approach to complex orbitofacial conditions, he serves in leadership roles in various national and international ophthalmic and oculoplastic societies.