Original Article

Orbital Vascular Malformations– Clinical Presentation and Management Strategies

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ABSTRACT

Purpose: To describe clinical features and management of Orbital Vascular Malformations (OVM) presenting to a tertiary care hospital of Lahore.

Study Design: Interventional case series.

Place and Duration of Study: Mughal eye hospital and Lahore General Hospital, Lahore, from January 2018 to December 2021.

Methods: A total of 26 patients with OVM were included in the study. All the patients underwent complete ocular examination. CT scan and digital subtraction angiography were performed for confirmation and extent of the lesions. The patients were photographed after informed consent to publish the photos. Management of three cases (representing three modalities) has been described in detail.

Results: Out of 26 patients, there were 16 males and 10 females. Mean age at presentation was 15 years (range 2 – 35 years). There were 14 cases with lymphatico-venous malformation (LVM) and 12 cases with arterio-venous malformation (AVM). One patient underwent surgical excision, second patient was managed with intralesional Bleomycin injection and in the third case, and intra-arterial embolization was performed by interventional radiologist. All three modalities proved successful in terms of function and cosmesis.

Conclusion: Orbital vascular malformations are uncommon lesions. Resection, sclerosing agent injection and intra-arterial embolization are successful treatments.

Key Words: Orbital Vascular Malformations, Lymphatic malformations, venous malformations, Lymphatico-venous malformations, Arterio-venous malformations.

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INTRODUCTION

International Society for the Study of Vascular Anomalies (ISSVA), the latest version 2018 is available online at: issva.org/classification.[1] It divides orbital vascular pathologies (anomalies) into tumors (haemangiomas) and vascular malformations. They usually manifest at birth or in infancy and grow in life. Exophthalmos, intralesional bleeding or cellulitis, long-term visual acuity impairment and amblyopia[2] are most common clinical features in children while chorioretinal folds have also been noted in a few cases.[2,3] Globe or optic nerve compression, or proptosis may result in reduced visual function.[4] Malformations have been divided into two categories, one is simple and the other is complex. Simple malformations consist of a single type of vessels. Complex malformations consist of multiple types of vessels.

Arteriovenous Malformations AVM have different flow characteristics and this requires different management options. Lymphatico-venous Malformations LVM are divided into two main types depending upon predominant components whether lymphatic or venous. Lymphatic malformations are

macrocysticormicrocystic depending upon the appearance. Functionally different types have different growth patterns. Treatment is difficult in both cases as the aim is to close large spaces in macrocystic and to decrease growth in microcystic variety. Venous malformations are characterized physiologically by flow as well as distensibility capacity because space in the orbit is limited.[5] The old terms e.g. lymphangioma, cavernous hemangioma, varix and arteriovenous fistula have been replaced now.

We describe three cases out of a total of 25 who presented to Mughal eye hospital, plus one patient from Lahore General Hospital. The three patients underwent three different treatments, which show a spectrum of management in these malformations.

Figure 1: Lymphatico-venous Malformations.

METHODS

Twenty five cases of Orbital venous malformations OVM presented in oculoplasty OPD of Mughal eye hospital during 2018 – 2021. These cases and one case which presented in Lahore General Hospital were included in the study. Clinical features of these patients were noted and photographs were taken. Management of three cases (representing three treatment modalities) has been described. Pre-treatment and post-treatment photographs were taken. Inclusion criteria was all vascular malformations (including lymphatic, venolymphatic and arteriovenous) presenting at any age. All orbital vascular tumours were excluded from the study.

Figure 2: Arterio-venous malformations.

RESULTS

Twenty-six patients with orbital vascular malformations were included in the study. Figure 1 and2 show different vascular malformations. There were 16 males and 10 females. Mean age at presentation was 15 years (range 2 – 35 years). All the patients were photographed and consent was taken to publish the photos. One patient was operated with the help of cryotherapy and minimum bleeding was encountered (Figure 4). Surgical excision markedly improved the cosmetic outcome. In another patient (Figure 5) intralesional bleomycin was injected which caused complete regression of the lesion. In yet another case (Figure 6), intraarterial embolization was performed by interventional radiologist, which caused regression of the vasculature and it was followed by surgical excision.

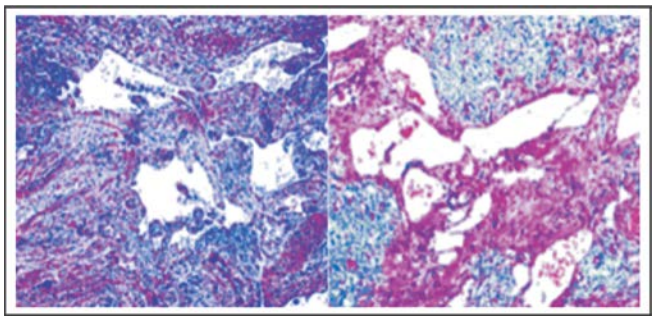

Figure 3: Pure LMs are serum- filled channels lined by flat endothelial cells that have immunostaining patterns consistent with lymphatic capillaries. Scattered follicles of lymphoid tissues are found in the interstitium. These have Infiltrative pattern, not encapsulated.

Figure 4: Arterio-venous Malformation; Left, pre-operative; Right, after resection

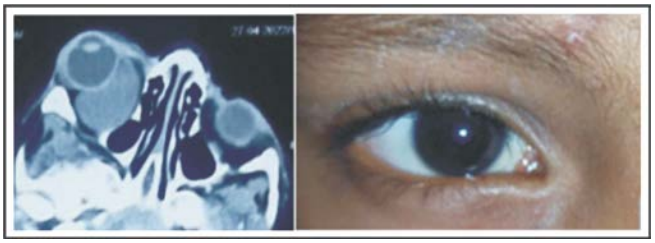

Figure 5: Left Photo: AVM pre injection. Right: Resolution of the lesion after injection Bleomycin.

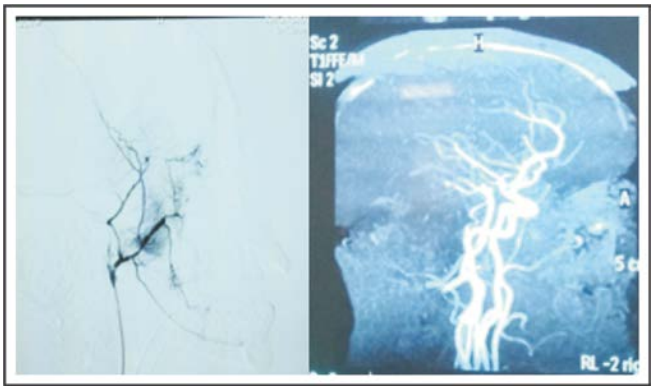

Figure 6: Left Digital subtraction angiography (DSA) showing preoperative tumor blush/vascular malformation in the branches of Internal Maxillary Artery. Interventional neuroradiologist embolized the feeder vessel using N-butyl cyanoacrylate (NBCA).

DISCUSSION

PubMed search "Orbital Vascular Malformations" yielded 958 results from 1953 to 2022. Out of these, 41 articles were from year 2022 and 55 articles were of the year 2021. All (41+ 55 = 96) of these articles were studied to collect latest knowledge about orbital vascular malformations.

ISSVA[5] enables meaningful classification for orbital vascular malformations. Four types of vascular malformations have been identified: OVM1 (cavernous), OVM2 (varix), OVM3 (infiltrating) and AVM.[6] Valsalva maneuver-augmented computed tomography angiography has been used to determine outflow in orbital vascular malformations.[7] A simplified version of ISSVA (2018) has been proposed. Imaging findings in pediatric vascular malformations divide these lesions into "simple", "combined" and syndromic or "low flow" and "high flow" (both entities can arise in isolation or as part of syndromes).[8] Analysis of blood flow by plane-wave ultrasound (compound coherent) has been found useful for imaging of orbital vascular malformations.[9]These OVM may be associated with intraocular vascular malformations.[10]

Lymphatic malformations (LM) can be macrocystic (> 1 cm^3) which generally present like encapsulated mass and produce proptosis.[10] Lymphatic endothelial cells line large spaces and stromal tissue is scanty. MRI and ultrasound may reveal cystic nature of the lesion. Intralesional bleed may cause rapid proptosis and may be due to respiratory tract infection. Resulting "chocolate cyst" may require urgent drainage.

Sclerosing agents e.g. Bleomycin, Sodium Tetradecyl Sulfate, Doxycycline, Picibanil (OK-432), Sodium Morrhuate, and Ethanol have been used for treatment of these malformations. Percutaneous injection can be given unaided or under ultrasound or fluoroscopic guidance.[12]

Microcystic lesions present as infiltrative process leading to motility reduction, squint, vision loss and pain. These have predominant cellular proliferation as compared to macrocystic in which fluid filled cavities predominate. Many lesions have both types. Observation, periodic debulking, Sirolimus and combined Bleomycin and Bevacizumab have all been used successfully.[13] Transnasal endoscopic sclerotherapy has also been found effective for LM atorbital apex.[1]

LVM are dealt with combination procedures depending on predominant portion type. Sodium tetradecyl sulfate,[12] oral (an mTOR inhibitor) Sirolimus,[13] Bleomycin [14,15] and combination of Sirolimus and Rivaroxaban (a factor Xa inhibitor)[16] have been used to treat LVM.

Bleomycin is generally safe but occasionally it has been found to be associated with severe orbital inflammation and hemorrhage.[17] In our case, use of bleomycin was safe and effective. It did not initiate any inflammatory reaction or haemorrhage and resulted in complete regression of the lesions.

Percutaneous embolization of orbital LVMs has been found effective before surgical resection. It is

done with the Onyx Liquid Embolic System (Medtronic, Ireland).[18] In our case, percutaneous embolization was performed by interventional radiologist. However, ophthalmologist was present also to prevent any unnecessary damage to ocular structures.

There are some latest techniques for treating vascular malformations. Rapamycin has been used to treat successfully orbital vascular malformations.[19] Symptomatic orbital cavernous malformations (OCVMs) are usually found a decade later than symptomatic lesions. Endonasal endoscopy has been used to remove OCVMs.[20]

Venous malformations increase in size during Valsalva's maneuver and bending. Surgical excision is easy as it does not invade local structures. Bleeding is a possibility. In our cases, surgical excision was uneventful. Initially it was done with cryotherapy. Cryo probe was placed right at the malformations and after freezing, surgical excision was completed with scissors. No bleeding was encountered.

Intralesional diode laser pretreatment has been found to facilitate surgery for orbital venous malformations.[21] Sclerotherapy may provide temporary improvement in some distensible venous-dominant moderate-flow orbital LVM cases, however in the long run in one study, recurrence occurred in all cases. Embolization with excision prevented recurrence in all cases for three years.[22] Endovascular embolization by glue (distends vessels) or onyx can be done percutaneously with the help of an interventional radiologist. For large and fast outflow systems, inflating an endovascular balloon and injecting embolic material through central microcatheter can be a better option. In enophthalmos, glue can be left in orbit to increase volume.

AVM are bunch of vessels where blood flows from arteries to veins without passing through capillaries. AVM can be localized or diffuse. These are high flow, rare and congenital (remain asymptomatic in infancy) but enlarge to become symptomatic in early adulthood. These usually present as mass with colour change and are associated with palpable pulsations and audible bruit. Arteriography shows anastomotic network of vessels and rapid filling of veins in the arterial phase. Doppler shows high flow and MRI shows flow void. Many can be observed while a few require treatment if cause proptosis or cosmetic disfigurement.

Embolization alone or followed by resection is the usual treatment. Selective embolization of arterial system aims to close arterial flow of the lesion without interfering the blood supply of surrounding healthy tissue. Complete embolization is often difficult as arterial flow has multiple sources. Partial embolization often results in recurrence. Resection can be tried alone or following embolization to improve hemostasis during resection. Anti VEGF have been used successfully in Hereditary Hemorrhagic Telangiectasia (HHT).[23]

Orbital veno-lymphatic anomalies have been found to be associated with giant cavernous malformation of the posterior fossa (with lymphangiomatous phenotype).[24]Both cavernous and lymphatic malformations express vascular endothelial growth factor receptors (VEGFR) to a variable degree and thus treatment with anti VEGF can be explored.[25]

In our series surgical resection, injection bleomycin and intraarterial embolization followed by resection, all produced satisfactory results.

CONCLUSION

AVM are treated by embolization of vessels with or without surgical excision. AVMs have a tendency to recur and antiangiogenic treatment may be required. LMs, macrocystic have a large lumen and are minimally invasive. These can be managed with sclerotherapy. Microcystic have more cellular proportions and can be approached with sclerosing and antiangiogenic substances. Excision may be required if there is rapid increase in size due to hemorrhage or thrombosis. VMs are distensible. Endovascular embolic techniques make excision easy.

Conflict of Interest: Authors declared no conflict of interest.

Ethical Approval

The study was approved by the Institutional review board/Ethical review board **(OSP-IRB/004-2023).**

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Authors Designation and Contribution

Khawaja Khalid Shoaib; Professor: *Concepts, Design, Literature Search, Data Acquisition, Data Analysis, Statistical Analysis, Manuscript Preparation, Manuscript Editing, Manuscript Review.*

Ahsan Mehmood; Consultant Ophthalmologist: *Concepts, Design, Data Acquisition, Data analysis, Manuscript Preparation, Manuscript Review.*