Management Strategies for Orbital Lymphatic-Venous Malformations: Insights from a Case Series

Estratégias de Tratamento das Malformações Linfático-Venosas da Órbita: Experiência de uma Série de Casos

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ABSTRACT

INTRODUCTION: Orbital venous-lymphatic malformations (VLMs) are complex vascular anomalies with diverse clinical manifestations that pose significant therapeutic challenges due to their proximity to critical orbital structures and risk of visual loss. Surgery carries high risk and laser therapy is limited to superficial lesions, whereas imaging-guided sclerotherapy with bleomycin offers a minimally invasive, effective, and repeatable treatment, particularly for deep or retrobulbar VLMs. However, microcystic subtypes often respond poorly, with recent studies suggesting oral sirolimus as a promising alternative in refractory cases.

We aimed to describe the clinical presentation, management and outcomes of orbital VLM cases across different age groups.

METHODS: A retrospective study was conducted on selected patients with orbital VLMs treated at Unidade Local de Saúde de Coimbra between July 2010 and September 2025 to represent the spectrum of management approaches available. All patients underwent ophthalmic examination and orbital imaging with computed tomography and/or magnetic resonance. Clinical photographs, treatment response, number of sessions, and procedure-related complications were systematically documented and analyzed.

RESULTS: Five patients (ages 9–52 years) with orbital VLMs were analyzed. Treatment modalities included intralesional bleomycin, oral sirolimus and surgical excision, with most patients undergoing multimodal therapy. Intralesional bleomycin achieved significant lesion regression and symptomatic improvement in all treated cases, although one patient required ongoing therapy for residual maxillary involvement. Sirolimus was used in two refractory cases, showing lesion stabilization, while surgical excision achieved complete resolution in two patients with well-circumscribed lesions. Visual acuity was preserved at 20/20 in four patients. No major ophthalmic or systemic complications were observed across treatment modalities.

CONCLUSION: Management of orbital VLMs remains challenging due to their proximity to critical structures, risk of recurrence and hemorrhagic complications. In this series, multimodal therapy—including intralesional bleomycin, sirolimus and surgery—was well tolerated, with no complications. Surgical excision and embolization achieved favorable outcomes in selected cases with well-defined lesions. Intralesional bleomycin provided effective lesion control and symptom

relief, while sirolimus demonstrated promising results in refractory microcystic lesions. Overall, individualized, multimodal treatment strategies of orbital VLMs appear safe and effective, supporting their use across a wide range of patient ages, lesion types, and disease severities.

KEYWORDS: Bleomycin; Lymphatic Abnormalities; Orbital Disease; Sclerotherapy; Sirolimus; Vascular Malformations.

RESUMO

INTRODUÇÃO: As malformações venosas-linfáticas orbitárias (MVLs) são anomalias vasculares complexas devido à sua proximidade de estruturas críticas e risco de perda visual. A cirurgia apresenta elevado risco e a terapia a laser é limitada a lesões superficiais, enquanto a escleroterapia com bleomicina é uma opção minimamente invasiva e eficaz, particularmente para MVLs profundas ou retrobulbares. No entanto, os subtipos microcísticos respondem de forma insuficiente, com estudos recentes a sugerirem a administração oral de sirolimus como alternativa promissora em casos refratários.

O nosso objetivo foi escrever a apresentação clínica, a gestão e os resultados de casos de MVL orbitária em diferentes faixas etárias.

MÉTODOS: Estudo retrospetivo de doentes selecionados com MVL orbitária tratados na Unidade Local de Saúde de Coimbra entre julho de 2010 e setembro de 2025, para representar as diferentes estratégias de tratamento disponíveis. Todos os doentes foram submetidos a exame oftalmológico e imagiologia por tomografia computorizada e/ou ressonância magnética. Fotografias clínicas, resposta terapêutica, número de sessões e complicações foram registadas e analisadas.

RESULTADOS: Cinco doentes (9-52 anos) com MVL orbitárias foram analisados. O tratamento incluiu bleomicina intralesional, sirolimus oral e excisão cirúrgica, sendo a maioria dos doentes submetida a terapêutica multimodal. A bleomicina intralesional obteve regressão significativa das lesões e melhoria sintomática em todos os casos tratados, embora um doente mantenha terapêutica por envolvimento residual maxilar. O sirolimus foi utilizado em dois casos refratários, mostrando estabilização das lesões, enquanto a excisão cirúrgica obteve resolução completa em dois doentes com lesões circunscritas. A acuidade visual manteve-se em 20/20 em quatro doentes. Não foram observadas complicações oftalmológicas ou sistémicas.

CONCLUSÃO: A gestão das MVLs orbitárias continua desafiante devido à proximidade de estruturas críticas, ao risco de recidiva e complicações hemorrágicas. Nesta série, a terapêutica multimodal, bleomicina intralesional, sirolimus e cirurgia, foi bem tolerada, sem complicações. A excisão cirúrgica e a embolização obtiveram resultados favoráveis em lesões circunscritas. A bleomicina intralesional proporcionou controlo eficaz das lesões e alívio sintomático, enquanto o sirolimus demonstrou resultados promissores em lesões microcísticas refratárias. No geral, o tratamento multimodal e individualizado das MVLs orbitárias é seguro e eficaz, apoiando a sua utilização em diferentes idades, tipos de lesão e gravidade da doença.

PALAVRAS-CHAVE: Anomalias Congénitas Linfáticas; Bleomicina; Doenças Orbitárias; Escleroterapia; Malformações Vasculares; Sirolimus.

INTRODUCTION

Orbital venous-lymphatic malformations (VLMs), formerly termed lymphangiomas, are rare, benign vascular anomalies characterized by the coexistence of venous and lymphatic components within the orbit.[1-3] They account for approximately 0.3%–4% of all orbital tumors and typically are congenital and manifest in childhood, with equal distribution between males and females.[3] According to the updated International Society for the Study of Vascular Anomalies (ISSVA) classification,[4] these lesions are considered slow-flow combined vascular malformations, consisting of abnormal vascular spaces lined by endothelial cells, including venous (internal venous flow) and lymphatic components (low flow). The ISSVA Classification is being continuously updated reflecting the evolving understanding of these entities. The orbital location is particularly challenging due to the absence of native lymphatic channels in this region and the close relationship of VLMs to vital ocular and periocular structures.

Clinically, patients often present with gradual, painless proptosis, dystopia, or ocular motility restriction. Acute hemorrhage within the lesion, sometimes triggered by minor trauma or infection, may result in rapid, painful proptosis, placing the patient at risk of compressive optic neuropathy and corneal exposure.[1,2,5] Recurrent swelling related to infection or systemic illness is also a common feature. Over time, untreated lesions can cause progressive visual impairment, cosmetic deformity, and functional disability.[5]

Diagnosis is based on clinical presentation and neuroimaging including magnetic resonance imaging (MRI) and/or computed tomography (CT) of the brain and orbits. VLM typically present in intra- and extraconal orbital spaces and can extend to the oropharynx and sinuses.[1] Non-contiguous intracranial vascular malformations can be associated. Dynamic neuroimaging assessment (CT or MRI angiography and angiography) is indicated to determine the best treatment approach.[1]

Management of orbital VLMs remains complex. Natural history of VLM is highly variable and despite a conservative approach being reasonable in cases with slow progression, other vison-threatening cases demand complex treatment options. Complete surgical excision is frequently unattainable due to their infiltrative, unencapsulated and vascular nature of the lesion, and high risk of recurrence and morbidity.[1,2,6] Intralesional sclerotherapy has emerged as a minimally invasive, imaging-guided treatment option, with favorable results particularly in macrocystic lesions. Sclerotherapy can be used alone or in combination with surgical debulking. Newer modalities such as foam sclerotherapy and systemic agents like sirolimus are increasingly being explored for microcystic or refractory cases.[1,2,7,8] Given their heterogeneous behavior, treatment often requires a tailored, multidisciplinary approach.

In this article, we describe the clinical spectrum, radiologic characteristics, therapeutic approach, and outcomes of a 15-year retrospective series of orbital VLMs treated at a tertiary care center.

METHODS

STUDY DESIGN AND ETHICAL APPROVAL

This retrospective case series included selected patients diagnosed with orbital VLMs and treated at Unidade Local de Saúde de Coimbra between July 2010 and September 2025. The study was conducted in accordance with institutional ethical standards and the Declaration of Helsinki. Written informed consent for publication of clinical details and accompanying images was obtained from all patients or their legal guardians in the case of minors.

INCLUSION CRITERIA

Inclusion criteria consisted of a selected subset of treated patients with a confirmed diagnosis of orbital VLM and adequate clinical and imaging documentation, chosen to illustrate the spectrum of management approaches used in our center.

DATA COLLECTION

All patients' records were reviewed for demographic data, clinical presentation, ophthalmic examination, radiologic features, treatment modality, number of treatment sessions, and procedure-related complications. Ocular characteristics comprised: best-corrected visual acuity (BCVA); ocular motility; globe position by Hertel exophthalmometer; biomicroscopy and fundoscopy. Ocular and systemic comorbidities were also collected. Orbital imaging with CT and/or MRI was performed. Clinical photographs were available for documentation in all cases.

TREATMENT PROTOCOLS

Management strategies included observation, sclerotherapy, and/or surgery, selected according to lesion type, location, and clinical impact. For patients undergoing sclerotherapy, bleomycin was prepared at a concentration of 1 mg/mL and administered at a dose of 0.5–1 mg/kg per session, with a maximum of 15 mg. Injections were performed percutaneously either under ultrasound guidance using a 23G needle by the ophthalmology team or under fluoroscopic guidance by the interventional radiology team.

FOLLOW-UP AND OUTCOME MEASURES

Patients continued to be followed at regular intervals, and outcomes were assessed based on changes in lesion size, degree of proptosis, ocular motility, visual function, pain, cosmetic appearance, and recurrence.

CASE 1

A 10-year-old girl presented to the emergency department with spontaneous subconjunctival hemorrhage, inferior eyelid ecchymosis, and mild proptosis of the right eye (OD), over two days. Ophthalmologic examination confirmed proptosis with dystopia (Hertel OD 21 mm, OS 18 mm) (Figs. 1A and 1B) and BCVA was 20/20 in both eyes (OU). Imaging studies were performed, which revealed a right intraconal lesion extending in both intraconal and extraconal spaces, predominantly between the medial rectus muscle and the optic nerve (Figs. 1C and 1D).

Initial management included oral methylprednisolone (1 mg/kg/day), however, in the following week, progressive proptosis and the onset of vertical diplopia prompted surgical intervention. A superomedial anterior orbitotomy via the upper eyelid crease allowed complete excision of the lesion. A negative pressure drain and compressive dressing were maintained for 48 hours to prevent pos surgical hemorrhage. Histopathological analysis confirmed the clinical diagnosis.

The postoperative course was uneventful. At monthly follow-up, the patient demonstrated good clinical and cosmetic results and BCVA of 20/20, which was preserved at 1 year (Fig. 1E). Imaging at 3 months with MRI postoperatively showed minimal residual lesion (Fig. 1F). Seven years after surgery, the patient remains asymptomatic without clinical or radiologic recurrence.

CASE 2

An 8-year-old boy presented to the emergency depart-

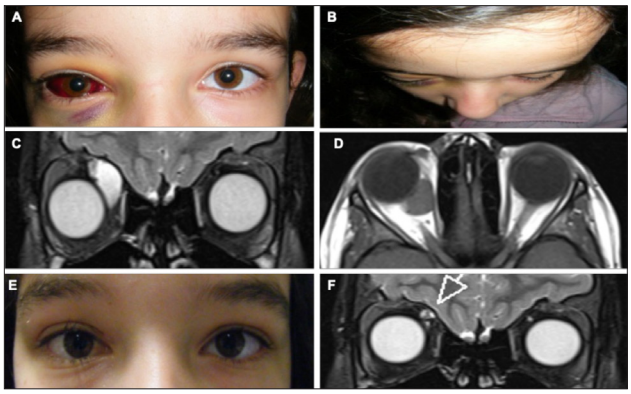


Figure 1. A) Subconjunctival hemorrhage and inferior eyelid ecchymosis OD; B) proptosis OD; C) Right intraconal lesion extending in both intraconal and extraconal spaces, predominantly between the medial rectus muscle and the optic nerve, showing hyperintense signal on T2-weighted MRI; D) Right intraconal lesion slightly heterogeneous hypointense signal on T1-weighted MRI; E) 1 year postoperatively; F) Residual right intraconal lesion slightly heterogeneous hyperintense signal on T2-weighted MRI.

ment with acute-onset proptosis OS for three days, associated with an upper respiratory infection. Ophthalmologic examination revealed proptosis (Hertel: OD 12 mm, OS 17 mm) and limitation of abduction and elevation OS. BCVA was 20/20 OU and the remainder of the ophthalmic examination was unremarkable. Orbital imaging demonstrated an expansive intraconal retrobulbar lesion in the left orbit. Initial management included aspiration followed by partial surgical excision of the lesion, with histopathological confirmation of an orbital VLM. Postoperatively, the patient achieved complete resolution of symptoms and remained stable for fourteen years.

In 2019 and 2023, at the ages of 22 and 26, respectively, the patient presented with recurrent episodes of acute left-sided proptosis (Fig. 2A). On both occasions, ultrasound-guided intralesional bleomycin sclerotherapy was performed (2 mL and 1.3 mL), resulting in significant improvement of symptoms (Figs. 2B and 2C).



Figure 2. A) Proptosis and inferior conjunctival chemosis OS; B) Ultrasound-guided intralesional bleomycin sclerotherapy; C) Postoperative clinical improvement at one-month after sclerotherapy; D) Conjunctival lesion reminiscent conditioning ectropion; E) and F) Postoperative clinical improvement at two-weeks after surgical excision.

Following the second sclerotherapy session, the patient experienced residual ectropion and a small conjunctival remnant OS, prompting reconstructive surgery to address both functional and aesthetic concerns (Fig. 2D). Follow-up assessments demonstrated a sustained recovery, with visual acuity stability and ocular motility improved with minimal restriction. MRI of the orbits demonstrated residual venous malformation with marked reduction in lesion size (Fig. 3). The patient expressed satisfaction with functional and aesthetic outcomes and continued with biannual follow-ups to monitor potential recurrence (Figs. 2E and 2F).

CASE 3

A 1-week-old male newborn was referred to pediatric ophthalmology for proptosis OS since birth. Pregnancy and delivery were unremarkable. Ophthalmologic examination revealed proptosis (Hertel: OD 11 mm, OS 14 mm) that increased with Valsalva maneuver, lagophthalmos, and a soft mass involving the ipsilateral hemiface (Fig. 4A). Reflexes and fundus examination were normal.

Orbital MRI showed a bilobulated orbital lesion extending into the ipsilateral mandibular region, with heterogeneous T2 signal and delayed moderate enhancement (Fig. 4B). These features were consistent with a VLM, given the heterogeneous signal, enhancement, and presence of hemorrhagic and calcified areas (areas of low signal in T2 and hyper signal in T1). Initial management with oral propranolol for four weeks produced no reduction in lesion size.

The child remained stable for 18 months until an upper respiratory infection triggered intralesional thrombosis and acute proptosis OS (Fig. 4C-E). A single session of bleomycin sclerotherapy (2 mL) resulted in significant improvement in lesion size, symptoms, and cosmetic appearance with preserved visual acuity and no evidence of optic nerve compression (Fig. 4F-H). Over time, due to the maxillary

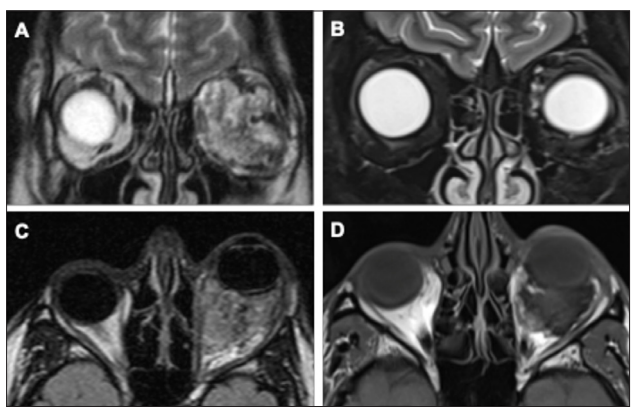


Figure 3. A) Left intraconal lesion showing hyperintense signal on T2-weighted MRI; B) Small residual area showing hyperintense signal on T2-weighted MRI compatible with sclerosis post sclerotherapy with bleomycin; C) Left intraconal lesion showing hypointense signal on T1-weighted MRI; D) Small residual area showing hypointense signal on T1-weighted MRI compatible with sclerosis post sclerotherapy with bleomycin.

component of the VLM associated with maxillomandibular hypoplasia, the child subsequently underwent five additional bleomycin sessions in the ipsilateral hemiface (total 54 mL) and has been on oral sirolimus for the past three years. Throughout 108 months (9 years) of follow-up, no adverse effects or complications were observed. The patient has remained clinically and radiologically stable, with no hemorrhagic events or recurrence.

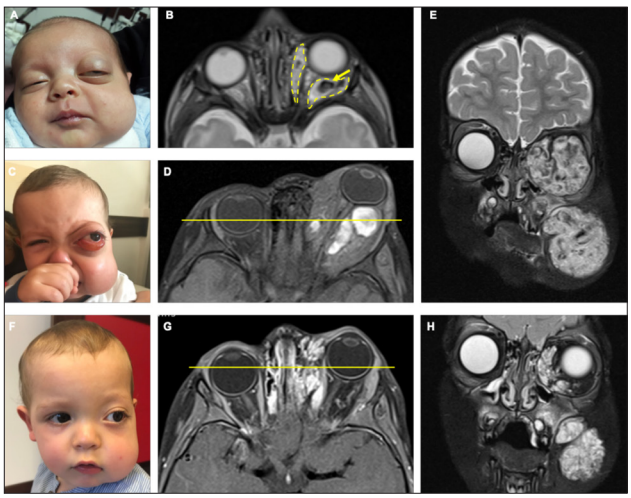


Figure 4. A) Proptosis and lagophthalmos OS at presentation; B) Bilobulated orbital lesion showing heterogeneous T2 signal: smaller one is extraconic between the orbital wall and the medial rectus and superior oblique muscles; the larger one is in the inferior and lateral quadrants, predominantly intraconal and retrobulbar, causing proptosis of the globe and orbital expansion. It displaced the inferior rectus muscle and the optic nerve medially and shows hemorrhagic foci and calcifications. It reached the orbital apex without compromising the optic canal or the superior orbital fissure; C) Acute proptosis OS following upper respiratory infection; D) and E) Marked increase in the dimensions of the left orbital lesion with associated bony remodeling and near-complete obliteration of orbital structures. The optic nerve is stretched and displaced superiorly. Inferiorly, there is extension into the masticator, parapharyngeal, and malar regions, involving the mandibular soft tissues and the ipsilateral zygomatic-temporal region, resulting in distortion of normal facial anatomy in T2; The lesion shows heterogeneous T1 signal, predominantly isointense on T1, with areas of T1 hyperintensity suggesting hemorrhagic or proteinaceous content; F) Improvement of proptosis after sclerotherapy with bleomycin; G) and H) Reduction in size of the left orbital VLM and in its internal, posterior, and inferior extension, particularly involving the orbital and periocular components. There is less proptosis of the globe and reduced stretching of the optic nerve, as well as a decrease in the mass effect on the posterolateral aspect of the globe and surrounding structures. There is also less extension into the nasosinusal and parapharyngeal regions, with a reduction of the infratemporal involvement and facial components, resulting in diminished lateral and inferior juxtamandibular prominence. A decrease in T1 and T2 signal heterogeneity is observed, with reduction of hyperintense areas, while intense and heterogeneous enhancement remains visible.

CASE 4

A 24-year-old woman was referred to the oculoplastic department with a one-week history of diplopia and ocular pain OD. Past ocular history was notable for an orbital and lower eyelid VLM diagnosed at the age of 10, for which she had undergone previous surgical interventions at another institution including surgical excisions and ethanol sclerotherapy.

On examination, there was diplopia in extreme gaze positions and proptosis (Hertel: OD 20 mm, OS 17 mm). BCVA was 20/20 OU, pupillary reflexes and fundus examination were normal. Orbital MRI revealed a multilobulated lesion within the right orbit, showing intra- and extraconal components and extension into the inferior periocular region. The lesion demonstrated marked T2 hyperintensity, isointense T1 signal, and intense, homogeneous post-contrast enhancement. It extended to the optic nerve up to the orbital apex, without evidence of optic nerve signal abnormality.

Considering the fluctuating symptoms, self-limiting recurrence episodes and imaging characteristics, the findings were consistent with a slow-flow orbital VLM, and the patient is currently being managed with clinical observation.

CASE 5

A 53-year-old man presented with a 14-year history of a VLM of the right orbit, characterized by intermittent episodes of hemorrhage and swelling that resolved spontaneously. At the time of initial diagnosis, the patient underwent surgical excision. In recent years, however, the malformation showed progressive and persistent growth, resulting in proptosis OS and ocular dryness.

On ophthalmologic examination, there was marked proptosis (Hertel: OD 33 mm, OS 25 mm), a palpable cystic mass near the inferior orbital rim, and ocular dystopia. Fundus examination revealed choroidal folds OD and BCVA was 20/32 OD and 20/25 OS. Orbital CT demonstrated a microcystic VLM in the right retrobulbar region, predominantly intraconal, with well-defined borders. The lesion caused upward displacement and stretching of the optic nerve.

The patient underwent one session of image-guided sclerotherapy with bleomycin (1.3 mL), performed by interventional radiologists. This resulted in a transient reduction in lesion size and temporary symptom relief during the first few weeks. However, proptosis persisted, prompting initiation of oral sirolimus in combination with dabigatran.[9]

After one month of sirolimus, the patient experienced a new hemorrhagic episode, accompanied by increased proptosis and orbital discomfort (Fig. 5A-D). He was

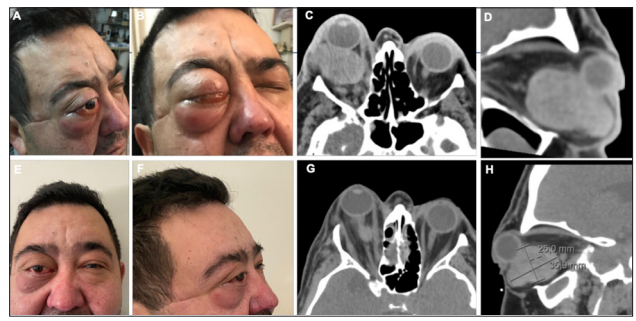


Figure 5. A) and B) Acute proptosis and lagophthalmos OS after initiation of sirolimus; C) and D) Significant increase in the size of the right inferior orbital VLM with a greater mass effect on the eyeball and increased stretching of the optic nerve. The lesion shows very faint and diffuse enhancement after contrast administration; E) and F) Improvement of proptosis and lagophthalmos OS after intravenous corticosteroids; G) and H) Intra-conal, well-defined VLM with multilobulated borders. The lesion contacts and slightly displaces the optic nerve superiorly.

admitted for five days of high-dose intravenous methylprednisolone (1 g daily), which resulted in symptomatic improvement (Fig. 5E-H). At the four-month follow-up of sirolimus treatment, there was slight improvement in proptosis (Hertel: OD 30 mm, OS 23 mm), with no other adverse events reported.

DISCUSSION

Orbital VLMs classified as slow-flow malformations can present as venous-dominant, lymphatic-dominant, or mixed types.[4] Depending on cyst size, they are further characterized as microcystic (<2 cm), macrocystic (>2 cm), or mixed.[1,4,10] VLM are congenital lesions and although most cases present in childhood, adult-onset or late presentations have been described, particularly in long-standing and slow-growing lesions that may remain quiescent for years before symptomatic progression. In a review of 158 patients with venous anomalies, 43% of cases were diagnosed before the age of six years and 60% before sixteen years.[11]

Clinical presentation in this series of five cases was heterogeneous, reflecting the broad spectrum of orbital VLM behavior, consistent with previous reports.[1,2] The oldest patient (case 5) illustrates the chronic, relapsing course typical of adult-onset disease with intermittent hemorrhagic episodes and swelling leading to progressive proptosis and ocular discomfort. In contrast, pediatric cases, such as cases 1, 2 and 3, demonstrated acute presentations with hemorrhage or infection-related exacerbations. Indeed, acute enlargement due to hemorrhage, thrombosis or inflammation – sometimes triggered by upper respiratory infections or minor trauma – may cause functional impairments such as progressive proptosis, displacement of the globe, ptosis, restriction of ocular motility, optic nerve compression, and exposure keratopathy.[1,2] More than half of acute cases reported in the literature exhibit compressive signs such as decreased visual acuity and afferent pupillary defects.

Radiologic findings across all cases were characteristic of orbital VLMs, showing heterogeneous signal intensity on T1- and T2-weighted MRI sequences with variable enhancement after contrast administration. Lesions frequently involved both intra- and extraconal compartments, displacing orbital structures and, in some instances, compressing the optic nerve. Imaging heterogeneity reflects blood degradation products and proteinaceous fluid within cystic spaces, correlating with the mixed venous and lymphatic histology.[1,12,13]

Management of orbital VLMs remains challenging, with no universally accepted standard of care. As this series highlights, individualized and multidisciplinary approaches are essential. Treatment decisions were guided by patient age, lesion subtype, symptom severity, and risk to visual function. Conservative observation may be reasonable for minimally symptomatic or stable lesions, as in case 4, whose small, slow-flow lesion remains under clinical monitoring.[1,2,14] However, acute presentations with hemorrhage or disfiguring proptosis often warrant intervention.

Surgical excision remains an option for well-circumscribed macrocystic lesions or those accessible through orbitotomy, as demonstrated in case 1, who achieved complete recovery and remained recurrence-free for ten years. Complete resection, however, is rarely feasible for diffuse or microcystic lesions that infiltrate critical orbital structures, as exemplified by the other four cases. There is also a high risk of intraoperative hemorrhage that difficult visualization and complete excision. In such cases, sclerotherapy has become the preferred minimally invasive alternative.[1,2]

Among sclerosants, bleomycin is favored for orbital lesions because of its lower inflammatory profile and reduced risk of post-procedural swelling compared with ethanol or sodium tetradecyl sulfate.[1,2,15] Bleomycin induces endothelial apoptosis and fibrosis, leading to gradual involution of the lesion, which was used in four of the five cases, with favorable outcomes in most. Cases 2 and 3 demonstrated sustained clinical and cosmetic improvement, with no recurrences or systemic toxicity. Conversely, in case 5, two bleomycin sessions achieved only transient lesion reduction and symptom relief, consistent with literature noting limited efficacy of sclerotherapy in microcystic VLMs due to poor sclerosant penetration.[1,2,16] Reported adverse effects are usually mild and localized, and systemic toxicity such as pulmonary fibrosis occurs only at cumulative doses exceeding 400 mg. None of our cases reported any complications so far.

In recent years, systemic therapy with sirolimus, an mTOR pathway inhibitor, has gained attention as a promising option for complex vascular malformations refractory to conventional interventions.[1,2,7] Sirolimus inhibits lymphangiogenesis and endothelial proliferation by modulating the PI3K–AKT–mTOR pathway, resulting in reduced vascular permeability and lesion stabilization[1,2,7]. In this series, two patients—pediatric (case 3) with extensive facial involvement and the adult case (case 5), received sirolimus after an incomplete response to sclerotherapy. Both showed clinical improvement, with the child achieving long-term stability and the adult showing significant clinical improvement after four months. These findings align with recent studies reporting partial or complete responses in 70%–90% of patients with complex lymphatic or VLM treated with sirolimus, particularly in microcystic subtypes.[7] Importantly, both patients tolerated therapy well, with no significant adverse events.

Despite the heterogeneity in presentation and management, several patterns emerge (Table 1). First, recurrence or progression was common, even decades after initial treatment, underscoring the need for long-term follow-up. Recurrence rates in other body regions can reach 40% after incomplete excision and 17% after macroscopically complete excision[17]; however, recurrent lesions are usually smaller than the initial lesion.[18] Second, microcystic or mixed lesions respond less favorably to sclerotherapy and surgery, often necessitating systemic therapy. Third, while bleomycin remains a safe and effective first-line sclerosant, sirolimus offered a valuable second-line or combination approach for diffuse or refractory cases. Lastly, multidisciplinary intervention—involving ophthalmology, interventional radiology, maxillofacial surgery, pediatrics, and internal medicine—is critical to preserving vision and optimizing

Table 1. **Clinical features of orbital VLM, treatment, recurrence and outcome.**

No	Age	Age at presentation	Sex	Eye	Presenting symptoms	Treatment	Recurrence	Follow-up	Outcome
1	23	9	F	OD	Subconjunctival hemorrhage, inferior eyelid ecchymosis, proptosis	Surgical excision	None	108 months	Resolution
2	28	8	M	OS	Proptosis, limitation of abduction and elevation	Surgical excision and sclerotherapy (bleomycin)	2 episodes (14 y and 4y interval)	240 months	Improvement
3	9	Birth	M	OS	Proptosis	Bleomycin and sirolimus	None	108 months	Improvement
4	25	10	F	OD	Diplopia and pain	Surgical excision	Multiple self-limited episodes	18 months	Improvement
5	53	39	M	OD	Keratitis, proptosis and infra eyelid soft lesion	Surgical excision and sclerotherapy (bleomycin) and sirolimus	Multiple self-limited episodes	168 months	On going

cosmetic outcomes.

Our case series adds to the limited body of literature describing orbital VLMs managed with combined sclerotherapy and systemic mTOR inhibition. Our collective experience supports a stepwise treatment algorithm: conservative observation for mild cases, targeted sclerotherapy for localized macrocystic disease, and systemic sirolimus for diffuse, microcystic, or recurrent lesions. The generally favorable tolerance profile of sirolimus and its efficacy in long-term disease control suggest it may soon become an integral part of standard VLM management.

CONCLUSION

This case series highlights the complexity of treating orbital VLM and the necessity of a multimodal, individualized approach. Surgery and embolization remain effective for well-defined lesions, whereas bleomycin sclerotherapy provides safe, targeted control in most orbital presentations. For diffuse or microcystic forms, refractory to other treatment approaches, sirolimus represents a valuable systemic option capable of achieving durable stabilization. Collectively, these findings support a stepwise, patient-centered treatment strategy guided by lesion characteristics, symptom severity, and multidisciplinary expertise. Continued long-term follow-up remains essential to monitor for recurrence and optimize both functional and aesthetic outcomes.

CONTRIBUTORSHIP STATEMENT / DECLARAÇÃO DE CONTRIBUIÇÃO

TM: Conceptualization, formal analysis, data collection, writing original draft.

JC, FP and GC: Conceptualization, formal analysis, writing - review & editing.

All authors approved the final version of the manuscript for publication and assume responsibility for all aspects of the work, ensuring the accuracy and integrity of the data presented.

TM: Conceitualização, análise formal, recolha de dados, redação do rascunho original.

JC, FP e GC: Conceitualização, análise formal, redação — revisão e edição.

Todos os autores aprovaram a versão final do manuscrito para publicação e assumem responsabilidade por todos os aspetos do trabalho, garantindo a exatidão e a integridade dos dados apresentados.

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