

Orbital Lymphangioma: Case Series and Revue of Literature

Ouail Ilhami^{1,2}, Kenza El Yacoubi^{2*}, Houria El Yazidi¹, Abdelhakim Oukerroum^{1,2} and Faiçal Slimani^{1,2}

¹Faculty of Medicine and Pharmacy, Hassan II University of Casablanca, B. P 5696, Casablanca, Morocco

²Department of Stomatology and Maxillofacial Surgery, Hospital 20 Août 1953, University Hospital Ibn Rochd, Casablanca, Morocco

Citation: Ilhami O, El Yacoubi K, El Yazidi H, Oukerroum A, Slimani F. Orbital Lymphangioma: Case Series and Revue of Literature. *Medi Clin Case Rep J* 2025;3(2):1003-1007. DOI: doi.org/10.51219/MCCRJ/Kenza-El-Yacoubi/263

Received: 14 June, 2025; **Accepted:** 17 June, 2025; **Published:** 19 June, 2025

***Corresponding author:** Kenza El Yacoubi, Department of Stomatology and Maxillofacial Surgery, Hospital 20 Août 1953, University Hospital Ibn Rochd, Casablanca, Morocco

Copyright: © 2025 El Yacoubi K, et al., This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Introduction

Orbital lymphangioma (OL) is a rare malformation of the lymphatic and vascular systems which accounts for 0.3%- 4% of all orbital tumors¹. Although benign, it is considered to be a severe anomaly because of its infiltrative and hemorrhagic nature associated with high morbidity rate. Proptosis, diplopia and restricted eye movement, loss of visual acuity and ocular pain are the main clinical manifestations of OL². Early diagnosis and treatment are crucial in order to prevent amblyopia and compression of the optical nerve. Diagnosis confirmation of OL relies essentially on radiological imaging which allows an accurate evaluation of its global extension. Due to its friable unencapsulated nature and connection with vital orbital structures, surgical removal is often uncomplete and carries a high risk of recurrence, therefore sclerotherapy is an effective and successful alternative treatment.

We, herein report four cases of orbital lymphangiomas and the outcome of surgical and medical treatment by Bleomycin injection.

Cases Presentation

Case-1

An 8-year-old female child reported with progressive right proptosis over the past 8 months. Clinical examination revealed a mild right palpebral swelling, a right axial proptosis with no conjunctival hyperemia nor inflammatory signs (**Figure1**). The

ophthalmological examination showed preserved visual acuity with normal eye motility and no diplopia. The orbito-cerebral MRI showed a hyperintense heterogeneous right retro and latero-ocular mass in T2 that englobes the optic nerve and oculomotor nerves related to a lymphangioma (**Figure 2**). An incisional biopsy was taken and histopathological examination was in favor of a cystic lymphangioma. The patient underwent surgical excision of the tumor under general anesthesia. Pathological examination confirmed the diagnosis of orbital lymphangioma. The outcome was favorable after 6 months.



Figure 1: (A): Clinical photograph of a 8-years-old girl with Orbital Lymphangioma (OL) suffering from proptosis of the right eye, (B), (C) magnetic Resonance Imaging (MRI) axial and coronal images showing a heterogeneous right retro and latero-ocular mass.

Case-2

A 18 years old boy reported to our department with left

ptosis and progressive vision loss. Clinical examination revealed a left palpebral swelling with advanced ptosis, a painful nasal conjunctival budding mass, reddish and bleeding on contact and conjunctival hyperemia (**Figure 1**). The ophthalmological examination showed a positive light perception on the left, a limitation of the left globe elevation and a complete cataract disturbing the funduscopy. An orbito-cerebral MRI was realized showing heterogeneous Intra- and extra- conical left orbito-palpebral tissue process with T1 hypo signal and areas of hypersignal related to hemorrhagic changes, encompassing the oculomotor muscles and optic nerve (**Figure 2**). An incisional biopsy was taken and histopathological examination was in favor of a conjunctival lymphangioma.

The patient underwent surgical excision of the tumor under general anesthesia with a mucosal graft. Pathological examination confirmed the diagnosis of orbital lymphangioma. The one year follow up showed no sign of recurrence. However, the ptosis remained and the patient's vision was still impaired.



Figure 2: (A) and (B) : CLINICAL PHOTOGRAPHS of a 18-years-old man with Orbital Lymphangioma (OL) showing an important ptosis of the left eye and a conjunctival budding mass, (C) and (D) magnetic Resonance Imaging (MRI) axial and coronal images showing a heterogeneous intra- and extra- conical left orbito-palpebral tissue process. (E) and (F) intraoperative images showing residual cavity after mass removal and reconstruction of the defect.

Case-3

An 5-year-old girl reported with progressive upper eyelid ptosis since birth. Clinical examination showed an important

right upper palpebral swelling with a complete ptosis, a blueish, palpable, fluctuating, non-pulsating mass with no conjunctival hyperemia nor inflammatory signs (figure1). The ophthalmological examination showed preserved visual acuity with normal ocular motility. The orbito-cerebral MRI showed heterogeneous intra- and extra-conical right intra orbital solid-cystic formation with T1 and T2 hypersignal, which exerts a mass effect on the eyeball and the superior rectus muscle and extends to the subcutaneous soft tissue of the right superior eyelid and a tissue-like anteromedial component in heterogeneous T2 hypersignal. Due to a high risk of morbidity and damage of the adjacent structures, sclerotherapy sessions were scheduled rather than a surgical excision. The patient benefited from three sessions of bleomycin-based sclerotherapy. After the first injection, there was a significant regression of the palpebral tumefaction and of the ptosis, after the second injection, a reappearance of the upper palpebral tumefaction occurred concomitant with an upper respiratory infection, who has regressed after medical treatment of the pulmonary infection objectified at the following check-up. At the end of treatment, there was a clear regression of the ptosis, a small palpebral tumefaction persisted on the upper palpebral level corresponding to the mass, which had clearly diminished in size. A follow-up by orbital MRI was done to assess the impact of sclerotherapy treatment and the results indicated a substantial reduction in the size of the lesion (**Figure 3**).



Figure 3: A 5-years-old girl with Orbital Lymphangioma (OL). (A) Photograph before bleomycin injections. (B) Photograph after final bleomycin injection, (C) and (D) magnetic Resonance Imaging (MRI) axial and sagittal images showing a heterogeneous intra- and extra-conical right intra orbital solid-cystic formation.

Case-4

A 16 years old boy reported to our department with left ptosis since birth and progressive vision loss. Clinical examination revealed a left palpebral swelling with advanced ptosis, (**Figure 4**). The ophthalmological examination showed a negative light perception on the left eye and a limitation of the left globe elevation. An orbito-cerebral scan was realized showing a voluminous intra-orbital mass, intra- and extra-conical, hypodense, which infiltrates the oculomotor muscles, in particular the superior rectus and the optic nerve and is associated with another palpebral formation of well-limited rounded liquid density (**Figure 5**). Given the possibility of lesion to noble

structures, we opted for sclerotherapy rather than a surgical excision, using bleomycin. The patient underwent 3 sclerotherapy session with 1 month interval between each (**Figures 4**). At the end of treatment, the mass looked reduced in size and the ptosis was reduced significantly however, the patient's vision was still impaired due to amblyopia. it was recommended for the patient to attend regular follow-up appointments however he was lost from sight.



Figure 4: Clinical Photograph of a 16-years-old girl with Orbital Lymphangioma (OL) suffering from ptosis of the left eye.



Figure 5: Orbito-cerebral CT scan showing a voluminous intra-orbital mass, intra- and extra-conical, which infiltrates the oculomotor muscles (A) axial views (B) frontal views.

Discussion

Lymphangioma (or recently called lymphatic veinous

malformation) is an unencapsulated vascular malformation of the lymphatic system³. It is not considered hamartoma since the orbit does not typically contain lymphatic vessels². They are classified as a type 1 vascular lesion (no flow) by the International Orbital Society⁴.

Lymphangiomas constitute 4% of vascular malformations and OL accounts for less than 4% of all orbital tumors with equal sex ratio^{2,3,5}. Lesions may be symptomatic at birth, but they are mostly diagnosed during the first (77%) and second (15%) decades of life, with up to 43% of cases diagnosed before the age of 6. Adult orbital lymphangiomas are rare⁶.

There are currently four theories on the development of lymphatic malformations: sequestration of lymph tissue; obstruction of lymph vessels; abnormal budding of lymph vessels; and lack of fusion with the venous system⁵. And since the orbit does not normally contain lymphatics orbital lymphatic malformations are alleged to rise from primary orbital varicose or lymphatic or venous systems of the periorbital structures^{3,7}. Lymphangioma develops as a result of either congenital or acquired lymphatic system abnormalities. The congenital form usually occurs before the age of 5 years old, it usually grows spontaneously slowly and steadily, but under certain conditions, such as infection (respiratory infection), hormonal changes or trauma, it can grow explosively^{2,3,8}. Acquired lymphangioma may develop from surgery, trauma, cancer or radiation therapy^{2,9}.

Clinically, it can present as proptosis, restricted movement of the eye, ptosis, ocular pain, diplopia, exposure keratopathy and visual impairment resulting from amblyopia and/or compression of the optic nerve⁹. In rare cases focal lesions may remain asymptomatic and be discovered by chance during neuro radiological imaging². The most prevalent symptoms of orbital lymphangiomas are proptosis (85%) and ptosis (75%) and restricted eye movement (43%)⁶, most of which occur during the first ten years of life^{1,2}. Typically, such lesions develop slowly causing mass effects on adjacent structures and compressive optic neuropathy^{2,9}. The identification of those amblyogenic factors is vital, as patients are usually seen before complete visual maturation. Complications of lymphatic malformations include mass effects from overgrowth and swelling of affected areas, infection and bleeding^{7,10}. Most lesions have both intra- and periorbital (most commonly eyelid) components and may cause bony remodeling or have associated bony anomalies⁵.

For many years, diagnosis of orbital lymphangioma has depended on histopathology. However, noninvasive diagnosis is possible based on clinical examinations and imaging results since the accuracy of the initial radiology interpretations was 77%⁶, also the confirmation of LM diagnosis can be provided by aspirating the fluid for cytology examination. On CT scan orbital lymphangioma appears typically as cyst-like masses with variable degrees of rim enhancement, it may reveal the presence of calcifications and phlebolits within the lesion and assess the condition of the orbital wall^{2,3,9}. MRI is the preferred imaging modality, LMs are usually isointense to brain on T1 and highly hyperintense on T2. It provides an excellent analysis of their extent and components, it has also been proven to detect feeder blood vessel and to differentiate between acute and chronic hemorrhages^{3,6,9,11}. Angiographic imaging show that the lesion is isolated from the arterial and venous circulations. Ultrasound can also be useful for more anterior lesions, demonstrating the cystic nature of the mass^{2,11}. Overall Lymphatic malformations

are radiographically categorized into 3 subtypes: macrocystic, microcystic and mixed type⁷.

The preservation of vision and prevention of amblyopia are top priorities when it comes to OL management, although there is still no conclusive cure. In most cases conservative observation is the recommended strategy for patients with no physical disabilities nor threatened vision^{2,9,11,12}. The main signs of intervention are: vision decrease (whether caused by amblyopia or compressive optic neuropathy), exposure keratopathy (due to excessive proptosis), glaucoma, rapid growth, cosmetic deformity or other functional problems. Treatment options also depends on location, type and size of the lymphangioma^{3,11,12}.

Surgical excision remains the primary approach for managing orbital lymphangiomas^{12,13}. Superficial lymphangiomas (ie eyelid and conjunctiva) are excised easily, but deep lymphangioma has venous connection, it is also diffuse and non-capsulated so surgical excision is difficult¹⁴. In addition, deep OL has a tendency to infiltrate essential orbital structures which makes surgical intervention (debulking and/or excision) often challenging and associated with a high risk of collateral damage to the said structures and a significant rate of recurrence^{2,9,14}. Due to its thin walled and easily collapsible nature, surgical management can be combined with preoperative intraluminal injection of n-butyl cyanoacrylate glue or fibrin glue to aid in dissection and improve hemostasis^{3,12,15}. The risk of hemorrhage can also be reduced by using diathermy, carbon dioxide or YAG laser^{1,3,14}.

Extreme cases of extensive deep orbital lymphatic malformation with persistent pain or a severe cosmetic disfigurement in the context of a blind eye orbital exenteration can be considered a reasonable option^{11,16}.

Nonsurgical strategies for orbital lymphatic venous malformations are sclerosing therapy, with or without ultrasound or fluoroscopic guidance, resulting in scar formation and reduced cyst and lesion size^{2,11,17}. This modality be combined with aspiration of blood¹⁰. Sclerosants include OK-432, doxycycline, ethanol, hypertonic saline, acetic acid and sodium tetradecyl sulfate, morrhuate sodium, pingyangmycin and bleomycin^{2,3,9-11,17}. Bleomycin is a cytostatic antineoplastic agent that inhibits the rapamycin (mTOR) pathway that has a sclero-embolic effect². In a study conducted by Nuruddin, et al,¹² cases of orbital lymphangioma were treated with intralesional injections of bleomycin at a dose of 0.5mg/kg bodyweight renewed every 4 weeks if required¹³.

Medical therapy with medications such as PDE-5 inhibitors (Sildenafil) or mTOR inhibitors (Sirolimus) are novel alternatives that have shown some promising results^{2,3,7,8,10,12}. Systemic cortico- steroid therapy alone was an effective management according to some authors^{10,18}.

Outcome spectrum of OL is wide, ranging between spontaneous regression to disfigurement organ dysfunction and life-threatening infection⁸. Many complications are reported after surgery, including muscle or nerve injury, hemorrhage, seroma and infection^{8,11}. Recurrence is common and related to regrowth from residual elements, it was noted in 58% of patients¹⁹. The tumor size after recurrence is usually smaller than the primary lesion^{3,5}.

Sclerotherapy could lead to a more pronounced inflammatory response causing spillover toxicity to critical structures (optic

nerve, muscles and globe) and the development of an acute post-procedure compartment syndrome¹¹.

Nuruddin and al. reported that 50% of patients had complete resolution and another 50% had resolution greater than 70% using intralesional bleomycin¹³.

Conclusion

Lymphangioma is a multicystic lymphatic and vascular malformation primarily affecting children in the head and neck regions. Orbital lymphangioma is extremely uncommon and requires early diagnosis and treatment to prevent structural and functional impairment (prevent amblyopia and preserve vision). Radiological imaging is the most effective method for confirming the diagnosis of Orbital lymphangioma by identifying and evaluating the orbital mass. OL can be challenging to manage, due to the confined bony space and proximity to noble structures and their entanglement with them. Different treatments are possible both surgical and non-surgical approach. Conservative management is recommended as the first line of treatment. Surgery should be performed for patients with threatened vision or severe cosmetic problems. Sclerotherapy is an alternative treatment for orbital lymphangiomas, that offers favorable outcomes and improve the quality of prognosis of the patients.

References

1. Saha K, Leatherbarrow B. Orbital lymphangiomas: a review of management strategies. *Curr Opin Ophthalmol* 2012;23(5):433-438.
2. Aristiady EB, Hilman null, Nugraha HG, Caesario MD, Kartiwa A, Susanty A. Sclerotherapy in orbital lymphangioma: A case report. *Radiol Case Rep* 2023;18(12):4366-4369.
3. Jafari N, Jafari R. Orbital Lymphangioma: A Case Report and Review of Management Modalities. *J Pediatr Rev* 2021;9(4):321-328.
4. Harris GJ. Orbital vascular malformations: a consensus statement on terminology and its clinical implications. *Orbital Society. Am J Ophthalmol* 1999;127(4):453-455.
5. Nassiri N, Rootman J, Rootman DB, Goldberg RA. Orbital lymphaticovenous malformations: Current and future treatments. *Surv Ophthalmol* 2015;60(5):383-405.
6. Thavara B, Rajagopalawarrier B, Balakrishnan S, Kidangan G. A case of adult orbital intraconal lymphangioma. *Asian J Neurosurg* 2020;15(1):168-171.
7. Shoji MK, Shishido S, Freitag SK. The Use of Sirolimus for Treatment of Orbital Lymphatic Malformations: A Systematic Review. *Ophthal Plast Reconstr Surg.* juin 2020;36(3):215.
8. Liu X, Cheng C, Chen K, Wu Y, Wu Z. Recent Progress in Lymphangioma. *Front Pediatr* 2021;9.
9. Kummari S, Ranga M. Orbital Lymphangioma in an Adolescent Male: A Case Report and Review of Literature. *Cureus* 2024.
10. Woo YJ, Kim CY, Sgrignoli B, Yoon JS. Orbital Lymphangioma: Characteristics and Treatment Outcomes of 12 Cases. *Korean J Ophthalmol KJO* 2017;31(3):194-201.
11. Rootman DB, Diniz SB, Cohen LM. Clinical Assessment and Lesion-Specific Management of Orbital Vascular Malformations. *J Neurol Surg Part B Skull Base* 2021;82(1):116-128.
12. Patel SR, Rosenberg JB, Barmettler A. Interventions for orbital lymphangioma. *Cochrane Database Syst Rev* 2019;5(5):CD013000.
13. Nuruddin M, Roy SR, Singh Mudhar H. Results of Intralesional Bleomycin Sclerotherapy for Treatment of Orbital

- Lymphangiomas at a Tertiary Eye Care Centre in Bangladesh. *Ocul Oncol Pathol* 2019;5(6):412-417.
14. Iqbal SS, Islam MA, Siddique MAB, Alam AE, Rana MM, Hasanuzzaman M. Management of Vascular Malformation of Orbit in a Tertiary Eye Hospital. *Cent Med Coll J* 2021;5(1):20-24.
 15. Kahana A, Bohnsack BL, Cho RI, Maher CO. Subtotal excision with adjunctive sclerosing therapy for the treatment of severe symptomatic orbital lymphangiomas. *Arch Ophthalmol Chic Ill* 1960 2011;129(8):1073-1076.
 16. Kirkin V, Thiele W, Baumann P, et al. MAZ51, an indolinone that inhibits endothelial cell and tumor cell growth in vitro , suppresses tumor growth in vivo. *Int J Cancer* 2004;112(6):986-993.
 17. Patel KC, Kalantzis G, El-Hindy N, Chang BY. Sclerotherapy for Orbital Lymphangioma - Case Series and Literature Review. *Vivo Athens Greece* 2017;31(2):263-266.
 18. Sires BS, Goins CR anderson RL, Holds JB. Systemic Corticosteroid Use in Orbital Lymphangioma: *Ophthal Plast Reconstr Surg* 2001;17(2):85-90.
 19. Tunç M, Sadri E, Char DH. Orbital lymphangioma: an analysis of 26 patients. *Br J Ophthalmol* 1999;83(1):76-80.