Minimally Invasive Management of Orbital Lymphangioma with Sclerotherapy: A Case Report

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AUTHORS' CONTRIBUTIONS

Author A contributed to the conception and design of the study, supervised the project, and approved it for submission. Author B provided radiological interpretation and contributed to the description of imaging findings. Author C performed the literature review, assisted in manuscript preparation. Author D revised the content critically for important intellectual input and reviewed the final version of the manuscript. Author E collected patient clinical data, and drafted the initial manuscript. Author F contributed to the clinical management of the patient, including diagnostic evaluation and treatment planning.

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DISCLOSURES

The authors declare no conflict of interest.

CONSENT

Did the author obtain written informed consent from the patient for submission of this manuscript for publication? Yes, written informed consent was obtained from the patient's legal guardian for the publication of this case report and any accompanying images.

ETHICAL STATEMENT

Informed consent was obtained from the patient's legal guardian for publication of this case report and all accompanying clinical and imaging materials. This report does not involve any experiments on humans or animals performed by the authors. All clinical procedures and reporting were conducted in accordance with institutional ethical standards and the principles of the Declaration of Helsinki.

ABSTRACT

Orbital lymphangioma (OL) is a benign vascular disorder affecting the orbit and surrounding structures of the eye, typically appearing and progressing in childhood. Sclerotherapy has been reported to be the primary approach for OL, but with minimal evidenced reported in Indonesia. Therefore, this case report aims to present the case of a 3-year-old boy diagnosed with orbital lymphangioma who was treated with sclerotherapy. A 3-year-old male patient presented with progressive right eye enlargement over two months. The patient had a history of a congenital lump above the right ear, previously excised. Nutritional assessment showed age-appropriate growth with adequate dietary intake and no signs of malnutrition. The child attends pre-school with no concerns for peer bullying, and family/social support was good, with only intermittent school absence due to clinic visits. Examination showed right eye proptosis without visual disturbances or systemic symptoms. Imaging revealed a retro-orbital lymphangioma compressing orbital structures and signs of cerebral venous sinus thrombosis. Histopathology confirmed hemangio-lymphangioma. Sclerotherapy under general anesthesia was performed, followed by Trans Arterial Chemo Infusion (TACI) and Transcatheter Arterial Chemoembolization (TACE) evaluation. Post-treatment MRI showed reduced mass size and improvement in proptosis. Vision was preserved, recovery was uncomplicated, and functional/aesthetic outcomes improved, enabling return to normal daily activities and school participation. The patient continues under multidisciplinary care with favorable outcomes. There were functional and aesthetic improvements in a 3-year-old with orbital lymphangioma after sclerotherapy. Therefore, further research needs to focus on treatment protocols, molecular mechanisms, and long-term outcomes of sclerotherapy in lymphangioma orbitalis.

CASE REPORT

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BACKGROUND

Orbital lymphangioma (OL), also known as lymphatic malformation (LM), is a benign vascular disorder affecting the orbit and surrounding structures of the eye. Orbital lymphangioma is a malformation of the lymphatic system that is not enclosed and consists of numerous cysts. This mass is highly infiltrative, carries a risk of spontaneous bleeding and long-term complications, including amblyopia, neuropathy, secondary glaucoma, and acute hemorrhage leading to blindness [1]. Globally, orbital lymphangioma is a rare condition, reported to occur in approximately 1-3% of all orbital tumors or masses [2,3]. The incidence of ocular or ophthalmic lymphoma has also been reported to occur frequently in the orbit, with a prevalence ranging from 47% to 54% with an increase in cases in the last 2 decades globally [4]. Orbital lymphangioma typically appears and progresses in childhood, with >50% of cases diagnosed before <16 years old [5].

The clinical manifestations of OL vary widely, depending on the location, size, and vascular component of the lesion. The most common symptoms include proptosis, ptosis, periorbital swelling, impaired eye movement, and pain or discomfort [6,7]. The primary goal of orbital lymphangioma management is to prevent complications and thus preserve vision, especially since most patients are diagnosed before visual maturity. In cases with asymptomatic lesions, management may consist of close observation and monitoring. However, intervention is still necessary if symptoms appear or worsen, including lymphorrhea, bleeding, recurrent swelling, pain, and cosmetic complaints. Interventions for OL include intralesional sclerotherapy, systemic medication, and surgical excision or tumor debulking [8].

Sclerotherapy has been reported to be the primary approach in the management of orbital lymphatic malformations due to its minimally invasive nature and effectiveness as an alternative or complement to surgery. The mechanism of sclerosing involves injecting a sclerosing agent into the cystic cavity, which induces endothelial damage, inflammation, and thrombosis, leading to fibrosis, thereby shrinking the lesion [8]. The effectiveness and response to sclerotherapy are reported to depend on the type of sclerogenic agent (sclerosant) used, lesion characteristics (type, size, and depth), and involvement of optic nerve structures [9,10].

However, sclerotherapy carries a number of risks and complications. Common complications include infection, bleeding, injury to orbital structures, and post-injection inflammation. Some of the agents used, including bleomycin, can also cause transient vomiting, skin hyperpigmentation, and pulmonary fibrosis when given in high cumulative doses [11,12]. Furthermore, data regarding the outcomes of patients with orbital lymphangioma after sclerotherapy in Indonesia are minimal, particularly in patients aged <5 years. Therefore, this case report aims to present the case of a 3-year-old boy diagnosed with orbital lymphangioma who was treated with sclerotherapy.

CASE REPORT

Anamnesis

A 3-year-old male with normal nutritional status patient was brought by his parents to the Emergency Department of R. Syamsudin Regional General Hospital with a chief complaint of an enlarged right eye. This complaint had been felt for the past 2 months. The patient denied complaints of eye pain, facial pain, blurred vision, double vision, weight loss, or similar complaints. The patient had a history of a lump above his right ear since birth. The patient denied any lump the size of a quail egg, pain, tenderness, mobility, discharge, or bleeding. The patient is the second of two children, born spontaneously, at full term, with clear amniotic fluid, and a complete immunization history. The patient had no family history of tuberculosis, heart disease, malignancy, chemotherapy, or radiotherapy. The patient had previously undergone surgery to remove a tumor above his right ear at R. Syamsudin Regional General Hospital.

Therefore, based on anamnesis, the leading consideration was an orbital lymphatic malformation (lymphangioma) given the painless, subacute proptosis in a 3-year-old with a congenital adjacent mass. Close differentials include venous malformation/ low-flow vascular malformation, dermoid/epidermoid cyst, and infantile capillary hemangioma. Further evaluation was performed to establish the diagnosis.

Physical examination

The patient's present status examination showed a general condition of moderate illness, fully alert, heart rate of 120 beats/ minute, respiratory rate of 32 breaths/minute, temperature of 36.8°C, SpO2 99% on room air, body weight of 15 kg, and height of 97 cm. The patient's general status examination showed an enlarged right eye with slight anemic conjunctiva on left eye, while the right eye was difficult to evaluate (Figure 1). Thoracic examination showed symmetrical chest wall shape and movement with normal vesicular breath sounds on both sides. No additional breath sounds were found. On abdominal examination, the abdomen appeared flat and soft with normal bowel sounds. Tenderness was noted in the right upper quadrant (RUQ) without rebound tenderness or muscular guarding. The Murphy's sign was negative, suggesting no acute cholecystitis; Murphy's sign is a clinical test used to assess for gallbladder inflammation, typically indicating cholecystitis. The Courvoisier sign was also negative, indicating no biliary obstruction; Courvoisier's sign is a clinical sign where a palpable, non-tender gallbladder suggests biliary obstruction, often due to malignancy. Rectal examination revealed strong anal sphincter tone, smooth mucosa, and a non-collapsed rectal ampulla, with no tenderness or palpable mass. Rectal examination is performed to assess for any abnormalities in the rectum, including masses or signs of rectal pathology. The stool was negative for blood or mucus upon examination. The extremities were warm, with a capillary refill time (CRT) of less than two seconds, which is considered normal, indicating adequate peripheral circulation; CRT is used to assess peripheral circulation, and a CRT of less than two seconds is considered normal.

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Laboratory examination

Laboratory examination results showed a low hemoglobin level of 11.0 g/dL, hematocrit of 34.9%, and erythrocyte count of 4.42 million/µL, with MCV 79.0 fL, MCH 24.9 pg, and MCHC 31.5%. Leukocytes showed 12,760/µL and platelet count increased to 550,000/µL. Coagulation parameters, liver function, and electrolye was within normal limits with SGOT 34 U/L and SGPT 16 U/L. Renal function was decreased with urea 12.5 mg/dL, creatinine 0.45 mg/dL and estimated glomerular filtration rate (eGFR) of 91 mL/min/1.73 m².

Imaging findings

The patient then underwent histopathological examination and was concluded as a frontoparietal hemangioma-lymphangioma. Orbital computed tomography revealed an inhomogeneous, non-hypervascular mass occupying the right retro-orbital space that obliterated the right lateral rectus muscle and the right optic nerve, abutted the posterior globe and right eyelid, and extended to the extracalvarial soft tissues of the right frontotemporoparietal region; arterial supply from the right ophthalmic artery with venous drainage to adjacent branches supported a diagnosis of lymphangioma. Computed tomography angiography (Figure 2) showed a well-defined solid mass with irregular margins in the right intraconal retrobulbar compartment extending to the right medial periorbita, displacing the globe anteriorly, compressing the extraocular muscles and orbital nerves, and producing mass-nerve conflict with the intraorbital and intracanalicular segments of the optic nerve, resulting in right-sided proptosis; compared with the prior MRI, the lesion appeared reduced in size, supporting recurrent lymphangioma. Contrast-enhanced MRI (Figure 3) releaved the same right intraconal retrobulbar mass with irregular borders extending to the medial periorbita, anterior displacement of the globe, compression of the extraocular muscles and orbital nerves, and conflict with the intraorbital and intracanalicular segments of the optic nerve, again causing proptosis; relative to the previous MRI, the lesion had decreased in size, consistent with recurrent lymphangioma. The patient was then diagnosed with hemangio lymphangioma of the right frontotemporoparietal ocular. The pediatrician planned to fulfill the patient's fluid needs according to the Holiday-Segar principle of 1,250 cc per day and has been scheduled to undergo sclerotherapy. Furthermore, the interventional radiologist planned the sclerotherapy procedure to be performed under general anesthesia.

Management

Orbital lymphangioma is a rare, benign vascular malformation of the orbit, typically presenting as a soft tissue mass that can lead to orbital proptosis, displacement of ocular structures, and visual disturbances. It arises from the abnormal development of lymphatic vessels, often resulting in cystic spaces filled with lymphatic fluid. Symptoms may include progressive proptosis, strabismus, or diplopia, with some cases associated with cranial nerve compression. Treatment options for orbital lymphangioma vary depending on the size, location, and symptom severity of the lesion. Surgical resection is typically considered, but for lesions that are difficult to access or recurrent, sclerotherapy is a preferred alternative. Sclerotherapy involves the injection of sclerosing agents into the lesion to induce fibrosis and shrinkage of the cystic spaces. In this case, sclerotherapy was performed with evaluation using Trans Arterial Chemo Infusion (TACI) or Transcatheter Arterial Chemoembolization (TACE). Clinical results post-sclerotherapy showed a reduction in the mass in the right eye, as demonstrated in (Figure 4). This was further supported by T1W1 MRI imaging (Figures 5,6), which revealed a decrease in the size of the previously identified right intraconal retrobulbar mass. The lesion's irregular borders, displacement of the globe, and compression of the extraocular muscles and orbital nerves were noted, but with significant shrinkage compared to prior imaging, suggesting a favorable response to the sclerotherapy treatment.

Follow-up

After undergoing multidisciplinary treatment, including sclerotherapy, the patient demonstrated significant clinical improvement. Proptosis in the right eye gradually improved, without any post-operative complications. Follow-up imaging revealed a significant decrease in the size of the orbital mass compared with the previous MRI. Visual function remained intact, and no additional neurological deficits were observed. Overall, the patient responded well to therapy with a satisfactory clinical outcome.

After the first sclerotherapy session, a follow-up MRI was performed after 3 to 6 weeks. The MRI results showed a reduction in the mass in the right eye, with a decrease in lesion size compared to previous scans. However, the lesion still showed irregular borders and some compression of the extraocular muscles and orbital nerves. Given these findings and the incomplete resolution of the lesion, it was decided that a second round of sclerotherapy would be necessary. The decision to proceed with a second sclerotherapy was made due to the persistence of the mass and its continued effect on ocular structures, which warranted further intervention to achieve better results. Follow-up is scheduled every 3 months during the first year and every 6 months thereafter up to 1 year, or sooner if symptoms recur. Each visit includes ophthalmic evaluation (visual acuity, refraction/amblyopia screening, proptosis/exophthalmometry, ocular motility, intraocular pressure, optic nerve assessment), surveillance for treatment-related complications, and quality-of-life review; growth and dietary intake are reassessed, and an MRI is obtained annually or if clinical progression is suspected

DISCUSSION

Etiology & Demographics

Lymphangioma is a rare benign vascular disorder found in the head and neck region, sometimes with cystic elements. Approximately 20% of cases can arise in the orbit and ocular adnexa, although they only account for 0.3–4% of all orbital

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tumors [13,14]. These lesions are estimated to account for 2–4% of all orbital occupational lesions and are primarily diagnosed in childhood. The prevalence of lymphangioma ranges from 1.1–5.3 per 10,000 live births. These lesions account for approximately 4% of all vascular tumors and approximately one-quarter of benign vascular tumors in children [5,15].

Clinical & Imaging Findings

Clinically, orbital lymphangioma typically presents with proptosis, limited eye movement, retroocular pain, and diplopia. Spontaneous bleeding within the lesion can cause severe headaches, eye pain, compressive optic neuropathy, and even blindness, requiring immediate decompression. Tumor growth can be exacerbated by infections of the face, nose, throat, or respiratory tract due to reactive lymphoid hyperplasia [15]. In this case, a 3-year-old male patient presented with an enlarged right eye for 2 months, without pain, visual disturbances, or weight loss. Laboratory tests revealed low hemoglobin levels. Although anemia is not a typical symptom of orbital lymphangioma, it has been reported in lymphangiomas in other organs, namely the spleen, due to hypersplenism [16]. The potential for anemia in orbital lymphangioma can arise if there is significant bleeding or if there is a systemic condition that affects blood cell production or destruction [17].

Confirmation of a lymphangioma diagnosis is most effectively performed with radiological imaging to assess the size and extent of the lesion. Typical characteristics on MRI are isointense on T1 and hyperintense on T2 with internal septa, while the absence of large vessels is present because lymphangioma is categorized as a no-flow lesion [8,18]. Histopathologically, orbital lymphangioma is a diffuse mass without a capsule, with thin-walled and irregular vascular channels, a thin endothelial lining, a discontinuous basement membrane, no pericytes, and a loose fibrous stroma. Several channels filled with proteinaceous fluid, and the absence of smooth muscle in the vessel walls with clear lymphatic fluid distinguish lymphangioma from cavernous hemangioma [3]. In this case, MRI revealed an irregular solid mass in the right intraconal retrobulbar region extending to the medial periorbital space, while CTA revealed a non-hypervascular, isohomogeneous mass compressing surrounding structures, as well as hypoplasia of the left transverse sinus and superior sagittal sinus. An orbital CT scan showed a mixed cystic mass in the right superior intraconal and extraconal areas, involving the optic nerve and causing proptosis.

Treatment & Prognosis

Although benign, the infiltrative nature of lymphangiomas makes complete surgery difficult due to the high risk of damage to vital orbital structures [3,14]. Treatment depends on the size and location of the lesion, with close observation being an option if the tumor is small and not sight-threatening. Surgery is performed when necessary due to the high recurrence rate and the difficulty of complete removal of the lesion [8].

In addition to observation and surgery, sclerotherapy is an alternative by injecting sclerosing agents to shrink the lesion [8]. Sclerosing agents used include picibanil, sodium tetradecyl sulfate, doxycycline, ethanol, pingyangmycin, and bleomycin, usually under ultrasound guidance through aspiration of cyst fluid followed by injection [19]. In this context, bleomycin is not used as a chemotherapeutic agent but as a sclerosing agent specifically targeting lymphangioma. Sclerotherapy is effective in macrocystic malformations, although less optimal in microcystic [20]. Possible risks include infection, bleeding, reactive inflammation, and increased orbital pressure that can disrupt the blood supply to the optic nerve and cause permanent blindness [8,18]. The mechanism of sclerotherapy varies depending on the agent, for example, by inducing endothelial cell death, inhibiting cell proliferation, or stimulating an immune response through cytokines [21]. Superficial lesions are easier to treat than deep or complex lesions, and cyst size plays a role in determining effectiveness, with primary recommendations for macrocysts [20, 22].

In this case, after two sclerotherapy sessions, the patient's right orbital mass showed significant shrinkage with clinical improvement, reduced proptosis, and no complications or neurological deficits. Previous reports have shown a similar pattern. In a 4-year-old female patient, three sessions of bleomycin sclerotherapy successfully reduced the mass size and proptosis, although visual impairment persisted [23]. Another case in a 15-year-old male patient showed improvement in symptoms (pain, proptosis, chemosis) and a significant reduction in lesion size after three sessions [24]. A study in a 5-year-old boy in Lahore also reported complete recovery without deformity, with minimal residual lesions on MRI, and a return to normal vision after digital subtraction angiography-based bleomycin foam sclerotherapy [25].

In this case, a follow-up MRI was performed 3 to 6 weeks after the first sclerotherapy session. In the previous study, the imaging protocol included ultrasound examinations conducted prior to treatment, between successive treatment sessions, six weeks following the final procedure, and subsequently every six months for a two-year follow-up period [27]. Another study also reported that pediatric patients with orbital lymphangioma who received intralesional bleomycin were evaluated postoperatively at 2 weeks, with additional sessions administered every 4 weeks as needed based on clinical and radiologic response [11].

The use of sclerotherapy as a single therapy without surgery may be a primary option, given the high risk of damage to vital orbital structures if surgery is performed in this case. In this case, a good clinical response was achieved after only two sclerotherapy sessions. No side effects or post-procedure complications were observed, either clinically or radiologically. MRI imaging findings showed a reduction in lesion size in line with improvement in proptosis symptoms, with no recurrence or new visual impairment during follow-up. The findings of

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this study confirm that sclerotherapy can be considered as first-line therapy for orbital lymphangioma with macrocystic or mixed lesions, particularly in pediatric patients or those with contraindications to surgery. This method has been shown to be minimally invasive, carries fewer risks than surgery, and produces satisfactory clinical results. However, this case report has limitations, including the need for long-term follow-up to monitor for recurrence and the limited literature regarding optimal dosage and number of sessions due to the lack of standards. Nevertheless, these findings strengthen the evidence that sclerotherapy is an effective, safe, and promising treatment option. To optimize future management, large-scale prospective studies are needed to validate these findings and develop more standardized protocols.

Differential Diagnoses

The main differential diagnoses for orbital lymphangioma include venous malformation, dermoid cyst, and capillary hemangioma. Venous malformations typically demonstrate slow-flow vascular channels that enhance with contrast and may contain phleboliths, unlike lymphangiomas, which are no-flow lesions on angiography and MRI [3,8]. Dermoid cysts, on the other hand, are well-circumscribed lesions often located near the orbital rim and may show fat-fluid levels or calcifications on CT or MRI [18]. Capillary hemangiomas usually present during infancy, show rapid post-contrast enhancement, and tend to regress spontaneously with age, whereas orbital lymphangiomas persist and can cause recurrent swelling due to hemorrhage or infection[8,22]. In this case, the absence of vascular enhancement, the presence of multiloculated cystic spaces with internal septations, and the histopathological findings of irregular lymphatic channels without smooth muscle or pericytes confirmed the diagnosis of orbital lymphangioma [3,14,18].

CONCLUSIONS

This case highlights the importance of recognizing both functional and aesthetic improvements following sclerotherapy in a pediatric patient with orbital lymphangioma. These findings can serve as a basis for further research focusing on controlled clinical trials comparing various sclerosing agents and treatment protocols, as well as exploring molecular mechanisms aimed at developing standardized management guidelines. Furthermore, studies evaluating long-term outcomes are also essential to prevent recurrence and further complications, and improve patient quality of life, particularly in the pediatric population.

TEACHING POINT

Orbital lymphangioma should be suspected in children presenting with slowly progressive, non-reducible proptosis. Characteristic imaging findings include multiloculated cystic lesions with fluid–fluid levels and minimal vascular enhancement, allowing accurate diagnosis and guiding targeted treatment.

QUESTIONS

Question 1: Which of the following statements about orbital lymphangiomas is false?

1. Orbital lymphangiomas are congenital vascular malformations.
2. They are non-encapsulated and infiltrative in nature.
3. Orbital lymphangiomas are typically well-encapsulated and completely resectable. *(applies)*
4. They commonly present in childhood.
5. They may increase in size due to hemorrhage or infection.

Explanation:

1. Lymphangiomas are congenital vascular malformations that manifest during childhood.
2. They are non-encapsulated, infiltrative lesions involving multiple orbital compartments.
3. This is false: they are not well-encapsulated and complete resection is rarely feasible due to infiltration.
4. Most cases manifest in childhood, often before age 10.
5. Hemorrhage and infection can cause sudden enlargement, worsening proptosis.

Question 2: Which imaging feature is most characteristic of orbital lymphangiomas on MRI?

1. Homogeneous hyperintensity on T1-weighted sequences.
2. Multiloculated cystic mass with fluid-fluid levels. *(applies)*
3. Diffuse calcification with hypointensity on all sequences.
4. Marked homogeneous post-contrast enhancement.
5. Restricted diffusion in all compartments.

Explanation:

1. T1 hyperintensity is not typical unless hemorrhage is present.
2. Correct: MRI often shows multiloculated cystic lesions with fluid-fluid levels, reflecting prior hemorrhage.
3. Calcification is not a feature of lymphangiomas.
4. Enhancement is usually heterogeneous or minimal.
5. Restricted diffusion is not a consistent finding; it is more typical of neoplasms.

Question 3: Which of the following is a recognized complication of orbital lymphangiomas?

1. Sudden proptosis due to intralesional hemorrhage. *(applies)*
2. Retinal detachment from subretinal fluid accumulation.
3. Hyperostosis of the orbital walls.
4. Bilateral cranial nerve palsy.
5. Spontaneous malignant transformation.

Explanation:

1. Correct: acute intralesional hemorrhage is a hallmark complication, often presenting with sudden painful proptosis and visual compromise.
2. Retinal detachment is not typical of orbital lymphangiomas.
3. Bony changes like hyperostosis are not characteristic.

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4. Lymphangiomas may compress the optic nerve but rarely cause widespread cranial nerve palsy.
5. Malignant transformation is not reported.

Question 4: Which of the following treatment modalities is most appropriate for orbital lymphangiomas with significant proptosis?

1. Observation only, regardless of severity.
2. High-dose systemic corticosteroids.
3. Definitive complete surgical excision.
4. Minimally invasive sclerotherapy. *(applies)*
5. Whole-orbit radiotherapy.

Explanation:

1. Observation may be appropriate for small, asymptomatic lesions, but not for severe proptosis.
2. Corticosteroids have limited efficacy in lymphangiomas.
3. Complete excision is rarely possible due to infiltrative nature.
4. Correct: sclerotherapy with agents, including bleomycin or OK-432 is an effective, minimally invasive option.
5. Radiotherapy is not a standard treatment and poses significant long-term risks in children.

Question 5: Which statement regarding the prognosis of orbital lymphangiomas is false?

1. Recurrence is common after treatment.
2. Visual compromise may occur due to optic nerve compression.
3. Long-term follow-up is required.
4. Malignant transformation into angiosarcoma is common. *(applies)*
5. Multidisciplinary management often improves outcomes.

Explanation:

1. Recurrence is common because complete excision is rarely feasible.
2. Optic nerve compression is a serious complication leading to visual loss.
3. Regular follow-up is required to monitor for recurrence and complications.
4. False: malignant transformation is not reported in lymphangiomas.
5. Multidisciplinary management (pediatrics, radiology, ophthalmology, neurosurgery) is crucial for optimal outcomes.

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FIGURES

Figure 1: 3-year-old boy with right orbital lymphangioma
Findings: clinical photographs before sclerotherapy showing right-sided axial proptosis with globe displacement and eyelid swelling (front, right, and left lateral views). **Technique:** standard digital photography, frontal and lateral orientations, under uniform ambient lighting and without flash reflection.

Figure 2: 3-year-old boy with right orbital lymphangioma
Findings: axial contrast-enhanced computed tomography angiography (cta) in the arterial phase demonstrates a non-hypervascular, inhomogeneous mass (arrows) measuring approximately 3.5 × 2.8 × 2.5 cm occupying the right intraconal retrobulbar space, obliterating the right lateral rectus muscle and compressing the optic nerve. The lesion also adheres to the posterior aspect of the right eyeball, right eyelid, and extracalvarial soft tissue in the right frontotemporoparietal region. The mass receives blood supply from the right ophthalmic artery and drains into surrounding venous branches, supporting the diagnosis of lymphangioma. **Technique:** axial cta performed using 128-slice ct scanner, 120 kv, 250 mas, 1 mm slice thickness, arterial phase after 80 ml non-ionic iodinated contrast injection (370 mgi/ml) at 4 ml/s.

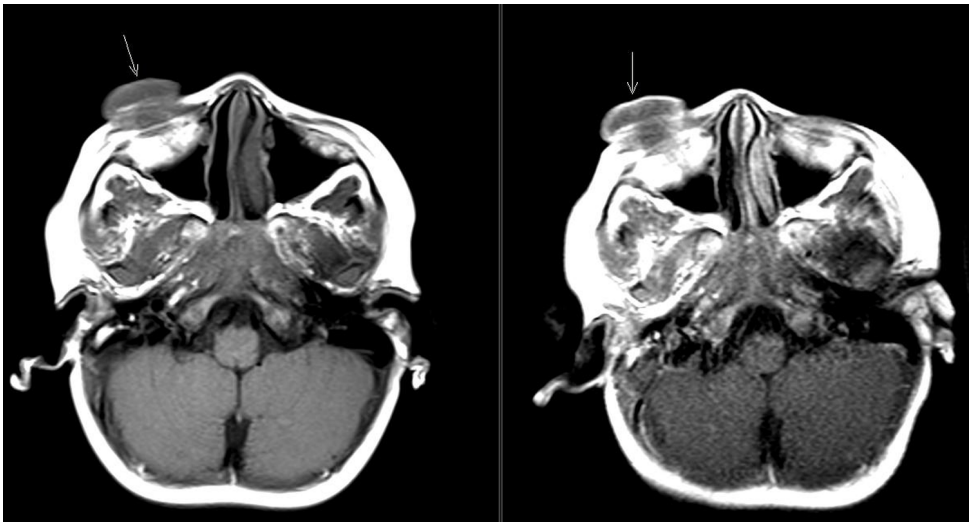

Figure 3: 3-year-old boy with right orbital lymphangioma
Findings: Pre-treatment MRI (left: pre-contrast, right: post-contrast) demonstrates a solid, multiloculated cystic mass (arrows) with well-defined borders and irregular edges in the right intraconal retrobulbar space, extending to the medial periorbita, displacing the right eyeball anteriorly, and compressing the right orbital muscles and nerves. This results in a neuromass conflict with the intracanalicular and intraorbital segments of the right optic nerve, leading to right ocular proptosis. These findings support the diagnosis of lymphangioma. **Technique:** mri obtained on 1.5 t scanner, axial and coronal t2-weighted sequence (tr = 4200 ms, te = 98 ms, slice thickness = 3 mm), pre- and post-gadolinium contrast (0.1 mmol/kg).

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Figure 4: 3-year-old boy with right orbital lymphangioma after sclerotherapy
Findings: Clinical photographs after sclerotherapy showing reduced right-eye proptosis and improved eyelid symmetry (front, right, and left lateral views). **Technique:** Standardized clinical photography as described in Figure 1.

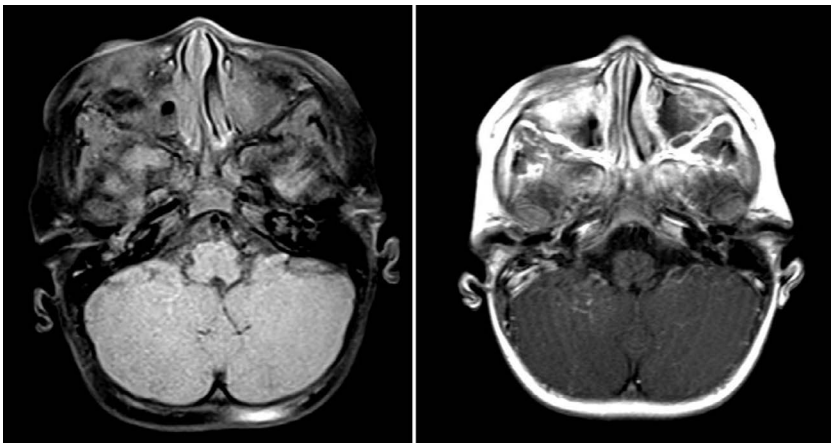

Figure 5: 3-year-old boy with right orbital lymphangioma after first sclerotherapy
Findings: Magnetic Resonance Imaging (left: pre-contrast, right: post-contrast) demonstrates a solid mass with well-defined borders and irregular edges in the right intraconal retrobulbar space, extending to the medial periorbita, displacing the right eyeball anteriorly, and compressing the right orbital muscles and nerves. This results in a neuromass conflict with the intracanalicular and intraorbital segments of the right optic nerve, leading to right ocular proptosis. These findings support the diagnosis of recurrent lymphangioma, with a decrease in size compared to the previous MRI, now measuring approximately 2.3 × 2.1 × 1.9 cm, and showing reduced mass effect on the optic nerve and extraocular muscles. **Technique:** mri performed on 1.5 t scanner, t1-weighted post-gadolinium (0.1 mmol/kg), tr = 550 ms, te = 12 ms, slice thickness = 3 mm, fat-suppressed axial plane.

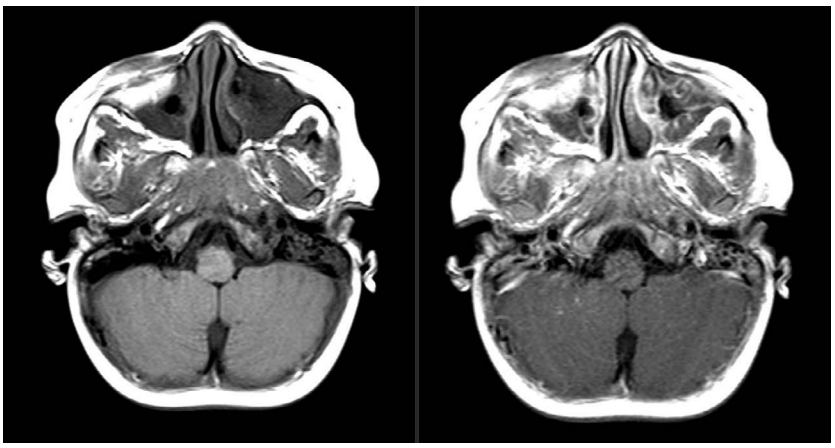

Figure 6: 3-year-old boy with right orbital lymphangioma after second sclerotherapy
Findings: Magnetic Resonance Imaging (left: pre-contrast, right: post-contrast) demonstrates a solid mass with well-defined borders and irregular edges in the right intraconal retrobulbar space, extending to the medial periorbita, displacing the right eyeball anteriorly, and compressing the right orbital muscles and nerves. This causes a neuromass conflict with the intracanalicular and intraorbital segments of the right optic nerve, resulting in right ocular proptosis. These findings support the diagnosis of recurrent lymphangioma, with a further decrease in size compared to the previous MRI, now measuring approximately 1.7 × 1.4 × 1.2 cm, and showing restoration of orbital anatomy and normalization of globe position. **Technique:** mri obtained on 1.5 t scanner, t1-weighted post-contrast (tr = 520 ms, te = 11 ms, slice thickness = 3 mm), fat-suppressed sequence in axial plane.

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Table 1: Summary of epidemiologic, clinical, therapeutic, and imaging characteristics of orbital lymphangioma based on current literature

Parameter	Description
Etiology	Congenital lymphatic malformation caused by developmental anomaly of lymphatic vessels leading to non-encapsulated, cystic proliferation within orbital tissues.
Incidence	Rare; accounts for approximately 1–3% of all orbital tumors and 2–4% of benign orbital lesions.
Gender ratio	Slight female predominance (approximately 1.2:1).
Age predilection	Primarily occurs in childhood; over 50% diagnosed before age 16.
Risk factors	None clearly established; enlargement may follow upper respiratory infections, trauma, or intralesional hemorrhage.
Clinical features	Progressive, non-pulsatile, non-reducible proptosis; periorbital swelling; diplopia; occasional acute pain or visual loss due to hemorrhage or optic nerve compression.
Treatment	Observation for small, asymptomatic lesions; sclerotherapy for macrocystic or symptomatic lesions; surgery for refractory or vision-threatening cases.
Prognosis	Generally favorable; recurrence possible due to infiltrative nature; vision preservation achievable with timely treatment.
Findings on imaging	**CT:** Non-enhancing, hypodense, multiloculated mass without calcification. **MRI T1:** Isointense or mildly hyperintense (if hemorrhage present). **MRI T2:** Hyperintense multiloculated cystic lesion with internal septations and fluid-fluid levels. **Post-contrast:** Minimal or heterogeneous enhancement. **US:** Cystic lesion with no internal vascular flow on Doppler.

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KEYWORDS

Child proptosis; clinical outcome; eye vascular malformation; percutaneous sclerotherapy; Indonesia

ABBREVIATIONS

CTA = Computed Tomography Angiography
CVST = Cerebral Venous Sinus Thrombosis
IOI = Idiopathic Orbital Inflammation
LM = Lymphatic Malformation
MRI = Magnetic Resonance Imaging
OL = Orbital Lymphangioma
TACE = Transcatheter Arterial Chemoembolization
TACI = Trans Arterial Chemo Infusion

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