Head and Neck Lesions

CERVICAL CYSTIC LYMPHANGIOMA IN AN INFANT TREATED WITH SIROLIMUS:

Report of a Rare Case and Literature Review

HANDIS Fatiha, BENYAHIA Samir

Department of Otorhinolaryngology (ENT), CCF, Mustapha Pacha University Hospital Center, Algiers, Algeria

Corresponding author: Prof. HANDIS Fatiha,
Department of Otorhinolaryngology (ENT), CCF, Mustapha Pacha University Hospital Center, Algiers, Algeria
Email : handis.fatiha@gmail.com
Corresponding author: HANDIS Fatiha,Department of Otorhinolaryngology–Head and Neck Surgery, CCF Unit, Mustapha University Hospital Center, Algiers, Algeria

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ABSTRACT :

Cervical cystic lymphangioma is a rare congenital lymphatic malformation in infants, often challenging to treat surgically when extensive or adjacent to vital structures. We report the case of a two-and-a-half-month-old male infant presenting with a rapidly enlarging left lateral cervical mass detected at one month of age. MRI revealed an extensive cystic lymphangioma displacing the pharyngeal wall and internal carotid artery, confirmed by pathological analysis. Due to surgical risks, a conservative treatment with oral sirolimus, combined with a short course of corticosteroids and antibiotic prophylaxis, was initiated. After ten months, the mass completely regressed without complications. Sirolimus, an mTOR inhibitor, blocks endothelial lymphatic cell proliferation and angiogenesis, explaining its effectiveness even in extensive lesions. This case demonstrates the feasibility, safety, and efficacy of medical management, as discussed in our report.

Keywords: *Cystic lymphangioma, Cervical, Infant, Sirolimus, Lymphatic malformation, Conservative treatment*

1 .INTRODUCTION :

Cystic lymphangioma (CL) is a rare congenital malformation of the lymphatic system, resulting from a failure in the canalization of primitive lymphatic vessels [1]. It accounts for less than 5% of pediatric vascular malformations and predominantly affects infants, with 50–60% of cases diagnosed before the age of two years and a slight male predominance [1,2]. The cervical region is the most common site of involvement; however, mediastinal, abdominal, and retroperitoneal localizations have also been reported [2,3]. CL may be classified as macrocystic, microcystic, or mixed, a distinction that influences both prognosis and therapeutic strategy [2,3].

Magnetic resonance imaging (MRI) is the modality of choice for assessing lesion extent and its relationship with vital structures, while histopathological examination confirms the diagnosis [3]. Conventional treatment relies on surgery or sclerotherapy; however, these options are limited in extensive lesions or when located near critical structures. Sirolimus, an mTOR inhibitor, has emerged as an effective and safe therapeutic alternative [4,5]. We report a case of cervical CL successfully treated with sirolimus in an infant and discuss this approach in light of recent evidence

2 . CASE REPORT :

A 2.5-month-old male infant was referred for evaluation of a left lateral cervical mass that had progressively increased in size. He was born prematurely at 34 weeks' gestation, with a birth weight of 3 kg. Vaccination status was up to date, and psychomotor development was normal. His medical history included prematurity and two neonatal hospitalizations for urinary tract infections.

The infant was brought by his parents for evaluation of a left lateral cervical mass that had appeared at one month of age and rapidly increased in size. On examination, he was in good general condition and well nourished. The mass was elongated, soft, non-tender, and compressible, mobile relative to both superficial and deep planes, and involved nearly the entire anterior portion of the left sternocleidomastoid muscle. The overlying skin appeared normal, and no additional cervical lymph nodes were palpable. No abnormalities were detected on ENT and general examination.

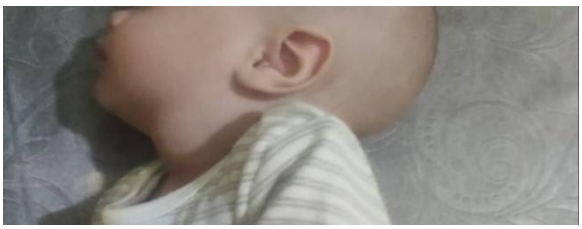

Figure 1: Lateral view showing a left cervical swelling, mobile and non-tender, involving the anterior half of the sternocleidomastoid muscle.

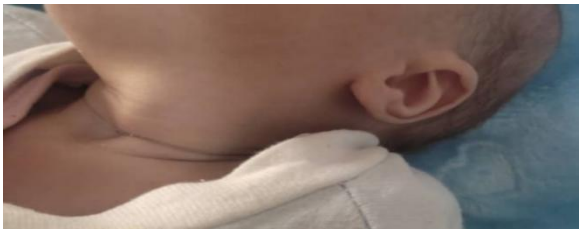

Figure 2: Anterior view showing a left cervical swelling with well-defined margins and intact overlying skin.

Cervical ultrasound demonstrated a well-defined, oval-shaped mass in the left submandibular region, which was hypoechoic and heterogeneous, containing multiple anechoic cystic spaces. The lesion measured 44 × 25 mm and showed internal vascularity on color Doppler imaging. The salivary glands, thyroid gland, and cervical lymph node regions were normal.

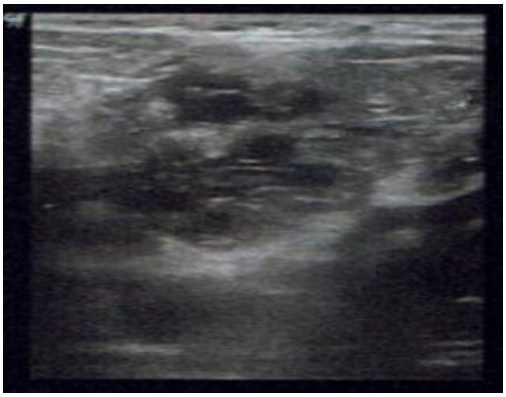

Figure 3: B-mode cervical ultrasound showing a hypoechoic, heterogeneous mass in the left submandibular region, containing multiple anechoic cystic spaces (44 × 25 mm).

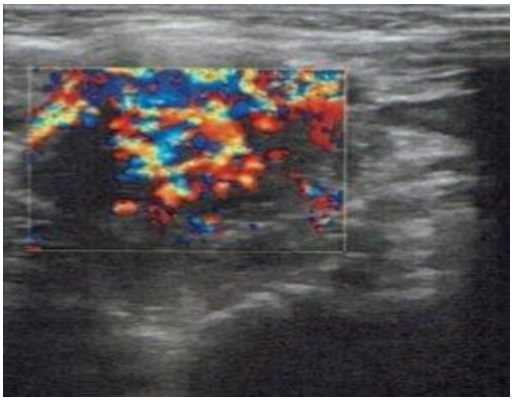

Figure 4: Color Doppler imaging demonstrating minimal internal flow within the left submandibular mass.

Cervical MRI revealed a left lateral cervical mass composed of multiple contiguous cystic spaces separated by thin septa, showing contrast enhancement. The lesion slightly displaced the pharyngeal wall medially and the internal carotid artery anteriorly, measuring 24 × 16 × 35 mm, suggestive of a cystic lymphangioma.

A fine-needle aspiration (FNA) was performed without imaging guidance, and histopathological examination confirmed the diagnosis of a cystic lymphangioma.

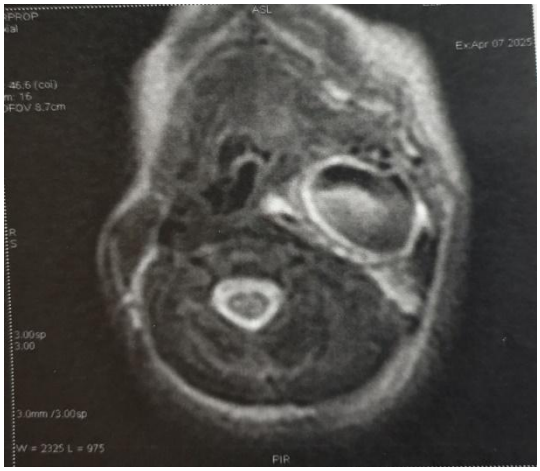

Figure 5: Axial T2-weighted image showing a multiloculated cystic lesion with hyperintense signal, delineated by thin septa, consistent with a cervical cystic lymphangioma.

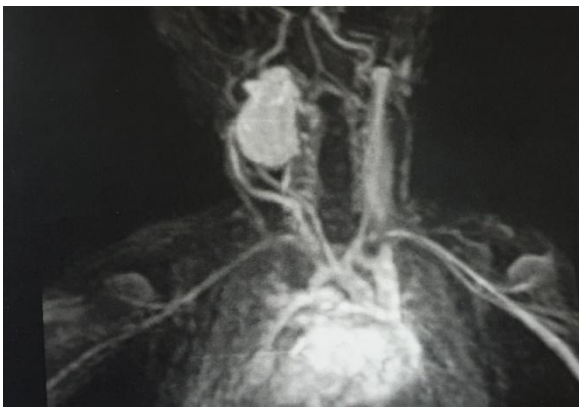

Figure 6: Cervical MRI, T1-weighted sequence with gadolinium contrast (3D-MRA), coronal view, showing a multiloculated cystic mass in the left lateral cervical region, displacing the vascular structures, consistent with a cystic lymphangioma.

A multidisciplinary team meeting involving pediatrics, dermatology, and ENT recommended conservative medical management due to the lesion's extent and its proximity to vital structures, which made surgical intervention high risk. The patient was treated with oral sirolimus solution (1 mg/mL, 0.4 mL/day administered in milk), combined with Solupred for twenty days and azithromycin for three days per month over three months. The rationale for sirolimus therapy is its mTOR-inhibitory effect, which enables regression of cystic lymphangiomas without surgical intervention.

The infant was regularly monitored clinically and biologically. At ten months of age, he showed complete regression of the mass, was in good general condition, and experienced no major complications. The successful use of medical therapy alone in an infant with an extensive cervical cystic lymphangioma is exceptional, making this case rare and of significant scientific interest.

3 . LITERATURE REVIEW AND DISCUSSION

Epidemiology

Cystic lymphangiomas are rare malformations, primarily affecting infants and young children, with a slight male predominance [1,2]. The cervical region is the most common site, accounting for approximately 75% of cases, followed by mediastinal, abdominal, and retroperitoneal locations [2,3]. Lymphangiomas are classified as macrocystic, microcystic, or mixed, based on cyst size, which influences prognosis and therapeutic strategy [2,3].

Clinical Presentation

Cervical cystic lymphangiomas typically present as soft, non-tender, mobile masses with progressive growth [2,3]. Extensive lesions may cause compressive symptoms, including dyspnea, dysphagia, dysphonia, or secondary infections. MRI is essential for evaluating the lesion's extent and its relationship with adjacent structures, while histopathological examination confirms the diagnosis and excludes other cystic or neoplastic masses [3]. In our case, the mass slightly displaced the pharyngeal wall and the internal carotid artery without significant functional symptoms, consistent with a typical presentation of a macrocystic cystic lymphangioma.

Treatment :

Surgery and sclerotherapy remain the standard treatments, but their efficacy is limited in extensive lesions or those adjacent to vital structures due to the risk of complications and recurrence [4,7,8].

Sirolimus, an mTOR inhibitor, acts on the PI3K/AKT/mTOR pathway, which is often activated in cystic lymphangiomas [5,6]. By blocking mTOR, sirolimus inhibits lymphatic endothelial cell proliferation and reduces the secretion of pro-angiogenic factors, leading to progressive cyst regression [5,6]. Its efficacy has been demonstrated in several recent series, showing rapid and significant reduction of mass volume and clinical improvement, even in infants younger than three months [5,6,7].

A corticosteroid (Solupred) was administered to reduce local inflammation and secondary reactions, while azithromycin was given prophylactically to prevent infections associated with the large mass or the aspiration procedure [5,6,7]. These adjunct therapies improved treatment tolerability and accelerated clinical regression.

The patient was monitored clinically and with laboratory tests, assessing both treatment safety (e.g., cytopenias, hyperlipidemia) and regression of cystic spaces. Although the optimal duration of therapy is not standardized, treatment is generally continued for several months until lesion stabilization or complete resolution [5,6,8]. The case demonstrates the feasibility and effectiveness of sirolimus in the management of an extensive and complex cervical cystic lymphangioma.

Ethics and Consent

Written informed consent was obtained from the patient's parents/legal guardians for the publication of this case report and any accompanying images.

The study was conducted in accordance with the ethical standards of the institutional Ethics Committee of Mustapha University Hospital, Algiers, Algeria, and with the 1964 Helsinki Declaration and its later amendments.

CONCLUSION

Cervical cystic lymphangioma in infants remains a rare malformation, which can pose therapeutic challenges due to its extent and proximity to vital structures. Sirolimus represents an effective conservative option, even in very young infants, demonstrating favorable outcomes in mass regression and good tolerability. This case highlights the value of a multidisciplinary medical approach, which can help avoid complications associated with surgery, and supports the growing role of sirolimus in the management of complex cystic lymphangiomas.

REFERENCES :

1- **Wassef, M., Blei, F., Adams, D., et al**. (2015). Vascular anomalies classification: Recommendations from the International Society for the Study of Vascular Anomalies. *Pediatric Dermatology, 32*, 221–230.

2–Saibene, A. M., Rosso, C., Felisati, G., et al. (2023). Sirolimus treatment for paediatric head and neck lymphatic malformations: A systematic review. *European Archives of Oto-Rhino-Laryngology, 280*, 3529–3540.

3–Wang, J., Meng, Y., Zhang, X., et al. (2025). A real-world study of sirolimus in the treatment of pediatric head and neck lymphatic malformations. *Journal of Vascular Surgery: Venous and Lymphatic Disorders, 13*, 102230.

4– EM-Consulte. (2021). *Le traitement chirurgical du lymphangiome kystique*. EM-Consulte. (Document no. 180256).

5– Cho, Y. J., Kwon, H., Ha, S., et al. (2024). Successful outcome with oral sirolimus treatment for complicated lymphatic malformations: A retrospective multicenter cohort study. *Annals of Surgical Treatment and Research, 106*, 125–132.

6–**Yu, S., & Guo, X.** (2025). Application of sirolimus in an infant presenting with a life-threatening lymphatic malformation of the head and neck: Case report. *Frontiers in Pediatrics, 13*, 1587330.

7–**Wu, S., Song, G., Guo, L., & Wang, Y.** (2021). Refractory head and neck lymphatic malformation in infants treated with sirolimus: A case series. *Frontiers in Pediatrics, 9*, 700.

8–Huerta, C. T., Beres, A. L., Englum, B. R., Gonzalez, K., et al. (2024). *Management and outcomes of pediatric lymphatic malformations: A systematic review from the APSA Outcomes and Evidence-Based Practice Committee*. *Journal of Pediatric Surgery, 59*(10), 161589.

9– **Railean, S., Gudumac, E., Lisița, N., Golban, R., Tsibirna, A., & Bernic, J.** (2023). *Lymphangiomas in the head and neck region in children*. Bulletin of the Academy of Sciences of Moldova. Medical Sciences, 76(2), 42–47.

10–**Assaad, M., Acharya, R., Shamsi, W., Springsted, E., Foroozesh, M., Cirino-Marcano, M. d. M.** (2025). *Mediastinal lymphangioma and intestinal lymphangiomatosis presenting with chylothorax: A systematic review of therapeutic modalities*. BMC Pulmonary Medicine, 25, 202.