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Sirolimus as a therapeutic option for a large cervical hemolymphangioma: Report of two cases

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Abstract

Sirolimus has shown promise as a treatment for extensive lymphatic malformations, especially when surgical excision is not feasible due to size, infiltration, or location. Although the literature remains limited and heterogeneous, most studies report partial clinical responses and pain relief, with few cases of complete remission. Adverse effects are generally manageable but can necessitate dose adjustments or discontinuation. The absence of standardized dosing guidelines and variability in treatment duration limit the comparability of published data.

Despite these limitations, sirolimus represents a potentially effective and well-tolerated option, particularly for patients with complex, refractory, or inoperable malformations. Individualized treatment plans remain essential, taking into account lesion characteristics, patient age, and potential risks. Further high-quality studies are needed to establish optimal dosing regimens, safety profiles, and long-term efficacy.

Keywords: Hemolymphangioma; Tumor; Lymphatic malformation; Sirolimus; pediatric; Case report

1. Introduction

Hemolymphangioma is a rare tumor characterized by developmental anomalies of the lymphatic system, associated with vascular malformations.

The obstruction of the venolymphatic communication between dysembrioplastic vascular tissue and the systemic circulation may contribute to the formation of hemolymphangioma (1). Lymphatic vessel injury as a result of trauma or surgery results in inadequate lymph fluid drainage, which is regarded as another cause of hemolymphangioma.

The baseline management of cystic lymphatic malformations depends on their severity, location, and whether they are macro or microcystic, isolated or syndromic. Sirolimus is becoming the primary treatment for painful inflammatory manifestations, and mixed and/or complex lymphatic malformations.

Due to the rarity of hemolymphangioma, a limited number of cases of the disease have been reported in the literature thus far. This case report presents two patients with cervical hemolymphangioma who were successfully treated with sirolimus in our department.

2. Case report

A 10-year-old female patient presented to our institution with a slowly enlarging swelling on the right side of the neck, first noticed 10 months prior. The swelling was painless and not associated with any other symptoms. The patient had

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a history of submandibular adenectomy performed over a year ago, the histopathological examination confirms a reactive lymph node. According to the family, the swelling began to increase in size approximately five months after the surgery. A solitary, well-circumscribed 5 × 4 cm swelling in the right submandibular region caused noticeable facial asymmetry (Fig1).

The second case concerns a 3-year-old girl with no significant medical history, who presented with a six-month history of a painless, progressively enlarging cervical swelling in the right peri-auricular region (Fig2). On clinical examination, the mass was firm in consistency and non-tender.

Both patients underwent cervical doppler ultrasonography and magnetic resonance imaging, which confirmed the diagnosis of cystic hemolymphangioma. A pre-therapeutic workup was performed for each patient prior to initiating treatment with sirolimus. Given the challenging anatomical location and the surgical risks involved, medical management was preferred over surgery. Sirolimus was administered orally at an initial dosage of 0.8 mg/m^2 twice daily. A significant clinical improvement was observed after two months of treatment. Therapy was continued for six months with good tolerance and a sustained, satisfactory clinical response throughout the treatment period.

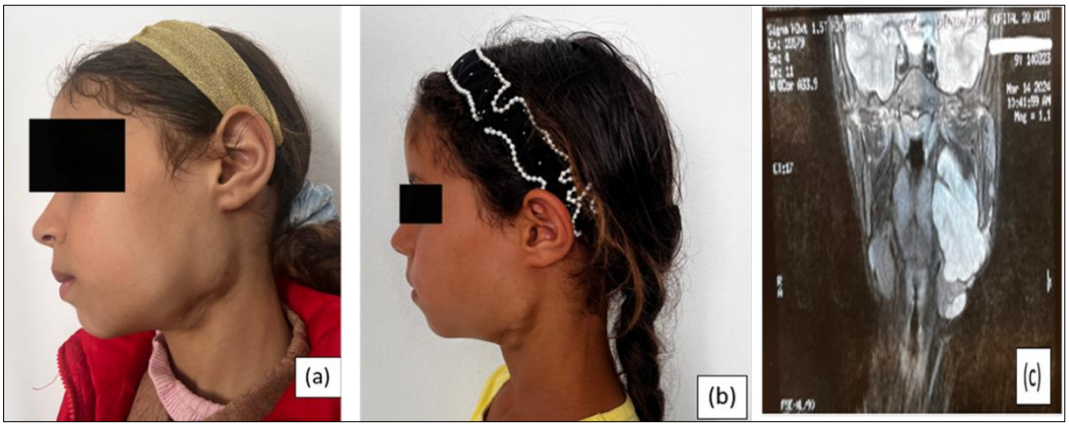

Figure 1 A 10-year-old female patient with a history of submandibular adenectomy presented with progressive right-sided cervical swelling. (a): Baseline presentation; (b) Two months after initiating treatment with sirolimus. (c): magnetic resonance imaging of the case

Figure 2 A 3-year-old girl with no significant medical history, who presented with a progressively enlarging swelling in the right peri-auricular region. (a) Baseline presentation; (b) Three months after initiating treatment with sirolimus. (c): magnetic resonance imaging of the case

3. Discussion

Hemolymphangiomas are rare benign tumors of lymphatic origin resulting from developmental anomalies. They account for approximately 2.5% to 5% of congenital cervical masses. These lesions typically present during childhood.

Although they can occur in various anatomical locations, they most commonly arise in the posterior cervical triangle, with mediastinal extension (2).

Biopsy is rarely required for the diagnosis of cystic lymphatic malformations (CLM), which is primarily based on clinical and radiological findings (1).

Histologically, hemolymphangioma consists of dense fibrous tissue arranged in bands between numerous vascular spaces, infiltrating subcutaneous fat and involving both blood and lymphatic vessels (2).

The diagnosis of CLM requires thorough clinical examination and imaging-based characterization. Interpretation of imaging findings must be correlated with clinical data, which may strongly guide diagnosis. For instance, the sudden appearance of a bluish swelling suggests a macrocystic lymphatic malformation complicated by intralesional hemorrhage, whereas the presence of lymphangiectasias points toward a microcystic component.

Ultrasound is essential for initial assessment, enabling the identification of features that help rule out differential diagnoses (e.g., cystic tumors or other malformations) and classify the CLM subtype (macro-, microcystic, or mixed forms). MRI is the modality of choice for evaluating anatomical extension and for better lesion characterization, particularly in cases of deep involvement and before initiating treatment (3) (4).

CLMs may become life-threatening due to upper airway compression when located cervicofacially and may cause functional impairment depending on their location.

The baseline management of cystic lymphatic malformations depends on their severity, location, and whether they are macro- or microcystic, isolated or syndromic. If observation is not an option, first-line treatment for macrocystic lymphatic malformations is sclerotherapy. Physical modalities—including CO_2 laser, radiofrequency, or electrocoagulation—may also be used; however, these require repetition and are palliative rather than curative. These are most commonly indicated for cutaneous lymphangiectasias and mucosal involvement, as they can superficially ablate lesions and control bleeding.

The management of microcystic or mixed lymphatic malformations is more complex and relies on multidisciplinary therapeutic strategies. These may include sclerotherapy, physiotherapy, surgery, sirolimus, or targeted therapy—administered successively or in combination—with intermittent periods of therapeutic abstention (5).

In most clinical experiences, sirolimus has been administered orally at an initial dosage of 0.8 mg/m^2 twice daily (6), with adjustments made to achieve a target blood concentration. Although some protocols have used alternative dosing regimens (from 0.05 to 0.1 mg/kg per dose twice daily) (7) (8).

The time to clinical response varies, with some patients improving within eight days and others requiring several weeks. The duration of therapy across studies has ranged from a few weeks to over four years, with many patients remaining on treatment at last follow-up (6) . In select cases, sirolimus has been used in conjunction with other modalities such as corticosteroids, laser surgery, or physiotherapy. Combined therapy has occasionally yielded superior results compared to monotherapy.

Sirolimus is generally well tolerated, although adverse effects have been described. These include hematologic toxicity, hyperlipidemia, gastrointestinal symptoms, and increased susceptibility to infections. Dose adjustments or discontinuation due to toxicity were relatively infrequent (8).

Overall, sirolimus appears to be an effective and generally well-tolerated therapeutic option for patients with hemolymphangioma, particularly in refractory and inoperable cases.

4. Conclusion

Hemolymphangioma is a congenital malformation of the vascular system. Due to the rarity of hemolymphangioma, a limited number of cases of the disease have been reported in the literature thus far. Ultrasound and MRI, are important for a full evaluation of the tumor in order to confirm the diagnosis.

Various therapeutic approaches have been proposed for the management of hemolymphangioma. Among them, surgical resection remains the most effective and commonly employed strategy, particularly for localized lesions. However,

alternative or adjunctive treatments have gained attention in recent years. Although current literature remains limited, sirolimus has emerged as a potentially effective and well-tolerated option, particularly for extensive, recurrent, or surgically inaccessible cases of hemolymphangioma

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare that they have no conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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