

# Topical Sirolimus in the Management of Vulval Lymphangiectasia: A Critical Review

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## Background

- Lymphangiectasia is a rare benign condition involving the congenital (also known as microcystic lymphatic malformation) or acquired dilatation of surface lymphatic vessels.
- Lymphangiectasia is characterised by clusters of vesicles that usually affect the head/neck, lower limbs and gluteal area.<sup>1</sup> It uncommonly affects the vulva.
- Complications include significant discomfort, aesthetic impairment and infection. Standard management typically includes resection, laser therapy and sclerotherapy. Invasive management options are associated with high rates of recurrence and iatrogenic morbidity.<sup>2</sup>
- Sirolimus is a macrolide drug that reduces lymphangiogenesis by blocking the PI3K/AKT/mTOR pathway.<sup>2</sup>



**Aim:** To critically appraise the literature on topical sirolimus for vulval lymphangiectasia

## Method

A search of the literature was performed in PubMed. The search terms "microcystic lymphatic malformation", "lymphangioma circumscriptum", "lymphangioma", "lymphangiectasia", "topical sirolimus", "rapamycin", "vulval" and "vaginal" were used in "AND" and "OR" combinations. No results were retrieved.

The search was repeated, excluding anatomical location. Literature that included oral sirolimus or non-lymphatic malformation components were excluded. Six case reports were reviewed.

Figure 1: Acquired lymphangiectasia of the vulva secondary to chemoradiotherapy and surgery for alveolar rhabdomyosarcoma

## Results

\*OD- once a day; BD- twice a day

Median patient age: 15 years.

5 congenital cases; 1 acquired case.

All patients achieved "good" clinical response with topical sirolimus.

Anatomical locations involved the buttock, trunk and male genitalia.

Varying concentrations from 0.1 to 1%.

Treatment schedules ranged from OD\* to BD\* for 3 months to continuous use.

Temporary local irritation was reported in three cases.

## Conclusion

Topical sirolimus could be an effective, non-invasive, well tolerated management option for vulval lymphangiectasia. Randomised controlled trials are needed to evaluate the long-term efficacy and safety of topical sirolimus for vulval lymphangiectasia.

## References

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- 2.Teng JMC, Hammill A, Martini J, et al. Sirolimus in the Treatment of Microcystic Lymphatic Malformations: A Systematic Review. *Lymphat Res Biol*. 2022
- Leducq S, Gabeff R, Zaragoza J, et al. Topical sirolimus 1% for benign lymphangiomatous papules after radiotherapy for endometrial and breast cancers: a report of three cases. *European Journal of Dermatology*. 2021; 31: 252-4.