

UPPER LIMB LYMPHATIC MALFORMATION : FROM LIMB THREATENING TO LIMB SALVAGE WITH SIROLIMUS

¹G Felicia; ¹Wong CC; ¹A Azuhairy; ¹R Sanjeevan; ¹N Prashant

¹Department of Orthopaedic Oncology, Penang General Hospital

INTRODUCTION :

Lymphatic malformations are non-malignant masses consisting fluid-filled spaces due to abnormal development of the lymphatic system, commonly classified as microcystic and macrocystic. It imposes difficulties in treatment due to sparsity of cases seen. Sirolimus however has lately been proven as an effective way to treat macrocystic lymphatic malformation.

REPORT:

We present a case of a 7 month old girl with underlying congenital left upper limb lymphatic malformation which was diagnosed at 2 months old via MRI imaging. She was referred to us at 7 months old with an impression of limb threatening infected lymphatic malformation of the left upper limb. The upper limb circumference increased to 23cm over the wrist, 30cm over the elbow and 31 cm distal to shoulder joint. Multidisciplinary discussions were held between the surgical, dermatology and orthopaedic oncology team. Discussions include the role of sclerotherapy, initiation of sirolimus, and even to the extent of amputation if the upper limb poses to threaten life. Fortunately, circumference of the left upper limb was reduced with the help of intravenous antibiotics and was allowed discharge. She was subsequently started on sirolimus. 3 months after the initiation of sirolimus, circumference over the left upper limb was significantly reduced with the current circumference of 14 cm, 19cm and 14 cm respectively.

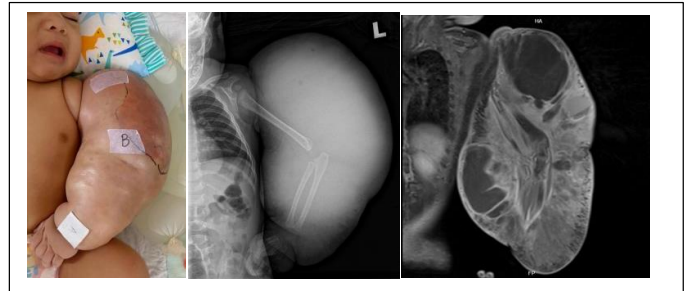


Figure 1 : initial presentation to our centre and imaging



Figure 2: three months after initiation of T sirolimus. With the latest circumference of 14 cm, 19cm and 14 cm respectively.

CONCLUSION:

Sirolimus is also known as rapamycin that is a key factor in the regulation of angiogenesis, cell growth and proliferation. Owing to its antiangiogenic and antiproliferative properties, Sirolimus could be an effective treatment in children with large complicated lymphatic malformation. Though not all patient benefits from this treatment, the initiation of sirolimus should be decided in a multidisciplinary fashion.

REFERENCES:

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC9249683/#:~:text=Sirolimus%20could%20be%20an%20effective,made%20by%20a%20multidisciplinary%20team.>

Cystic lymphangioma of the upper limb:
Clinical case and literature review