PAEDIATRIC DERMATOLOGY

SIROLIMUS IN THE TREATMENT OF VASCULAR ANOMALIES IN MALAYSIAN CHILDREN

Hoi Ling Wong [1] - Kin Fon Leong [1] - Sabeera Begum [1]

Institut Pediatrik Hospital Kuala Lumpur, Paediatric Dermatology, Kuala Lumpur, Malaysia [1]

Introduction: Vascular anomalies are a heterogeneous group of disorders that can be categorized into vascular tumours or vascular malformations. Sirolimus, a mammalian target of rapamycin (mTOR) inhibitor has been proven to be a safe and effective therapeutic option in patients with vascular anomalies.

Objective: We aim to assess the use of oral sirolimus in patients diagnosed to have vascular anomalies in Institut Pediatrik Hospital Kuala Lumpur, Malaysia.

Method: A retrospective review of case notes of all Paediatric Dermatology patients who had been started on oral sirolimus from January 2013 to December 2017 was conducted.

Results: Forty five patients have been started on oral sirolimus during the study period. Six cases were excluded due to missing data. Sixty one percent (n=24) were female with a male to female ratio of 1:1.5.The age of starting sirolimus ranged between 0- 22. Thirty percent (n=12) of the anomalies were capillary-lymphatic-venous malformation, followed by lymphatic and lymphatic-venous malformation of 17.9% respectively (n=7). Twelve percent (n=5) had Karposiform haemangioendothelioma with 80% (n=4) having Kasabach Meritt Syndrome. Ten percent (n=4) of patients were diagnosed to have venous malformation while another 7% (n=3) had Tufted Angioma. All patients were started on sirolimus at a dose of 0.8mg/m2/dose 12hourly. Overall patients showed good clinical and radiological response. Most patients were able to achieve the desired trough level of 5-20ng/mL. Four patients developed transient hypercholesterolaemia and one patient had transient hepatitis.

Conclusion: Sirolimus is a safe and efficacious drug in the treatment for vascular anomalies. Further studies need to be conducted in order to determine the trough level for optimal sirolimus effect.