PAEDIATRIC DERMATOLOGY

TUFTED ANGIOMA WITH KASABACH-MERRIT SYNDROME: A REPORT OF TWO CASES SUCCESSFULLY TREATED WITH EVEROLIMUS.

M Chiguer [1] - N Farid [1] - S Amal [1] - O Hocar [1]

Chu Med Vi, Cadi Ayyad University, Dermatology, Marrakech, Morocco [1]

Background: Kasabach-Merritt syndrome (KMS) is a rare life-threatening complication of vascular tumors. Treatment of KMS remains challenging. Mammalian target of rapamycin inhibitors have shown efficacy in this condition.

Observations: Case 1: 2-month-21 day's old infant was admitted for a reddish lesion of 5 cm diameter located in the left cheek. The patient was diagnosed as KMS. Biopsy concluded to a tufted angioma (TA). Case 2: A 4-month-old male was admitted for a lesion over left shoulder, back and chest wall. The clinical and laboratory features were indicative of KMS and the histological features were diagnostic for TA. The 2 Patients were first treated by oral prednisolone (3mg /kg /day), acetylsalicylic acid (10mg/kg /day), and ticlopidine (10mg /kg /day). And in view of the non-availability of sirolimus in our hospital structure; treatment with everolimus (0.1 mg/kg /day) was started. Informed consent was obtained from the patient's parents prior to treatment. Patients showed significant improvements in hematologic parameters and a decrease in the size of lesion, with a significant reduction in intensity of color and thickness after 3 months. The first patient was being treated with everolimus for 12 months. He is healthy without evidence of relapse for 1 year after stopping everolimus. The cutaneous component of the patient's TA has now resolved as well. We did not note any metabolic disturbance on the biological assessment. Besides, respiratory symptoms were not noted during the 12 months of everolimus treatment. The second patient was being treated with everolimus for 3 months only. Unfortunately everolimus was not available.

Key message: We report the first 2 cases of TA with KMS that were successfully treated with everolimus. Long-term therapy with everolimus has shown to be effective without side effects. Our experience suggests that everolimus may be recommended as an early treatment of severe KMS.