A 2-month old boy with Kaposiform Hemangioendothelioma of the lumbosacral region invading the retroperitoneum was admitted with Kasabach-Merritt Phenomenon. After corticosteroid and interferon treatments failed with progression of the disease to a life threatening condition, vincristine monotreatment granted a rapid rise in platelet counts, fibrinogen levels and a resolution in the enlargement of the lesion. The remission was maintained and no adverse effects were observed during the treatment. Vincristin therapy may be suggested instead of multidrugs as first line therapy in patients with KMP.