Reference Haematologica 2009; 94[suppl.2]:681 abs. 1793 Title ?SUCCESSFUL USE OF VINCRISTINE IN THE MANAGEMENT OF KASABACH-MERRITT PHENOMENON
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Abstract ?Kasabach-Merritt Phenomenon (KMP) is a life threatening clinical picture characterized by thrombocytopenia, consumption coagulopathy with a low fibrinogen level and microangiopathic hemolytic anemia.in the presence of a rapidly enlarging vascular lesion. Affected infants may manifest high output cardiac failure as a result of increased blood flow through the vascular lesions. It can be lethal; the estimated overall mortality rate ranges from 10-37%. Kaposiform hemangioendothelioma is the responsible lesion most of the time, Retroperitoneal involvement is significant determinant of the mortality. Early treatment is important to prevent fatal bleeding.MRI and ultrasonography are both effective tools for determining the extension of the lesion hence the prognosis and and for assessing their response to therapy.We would like to present a 2 month old boy with KMS, successfully treated with vincristine therapy. Case report. 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. Once a week induction treatment for a month was followed by once a month maintenance for 2 months. The remission was maintained and no adverse effects were observed during the treatment. The patient did not relapse 2 months after vincristine was stopped. Conclusion. This case and review of the literature point to vincristine as a first line treatment and suggest that multidrug regimens with additional drugs should be hold for bleeding emergencies.