CASE REPORT

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A case report of Kaposi Hemangioendothelioma a cause of thrombocytopenia in a neonate- Propranolol a miracle drug

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Abstract

We describe a case of kasabach – Merritt syndrome secondary to hemangioendothelioma. He was born to a G2P1L1 mother by LSCS, had a hemangioma involving the right lower leg and foot with thrombocytopenia and anemia. He was started on propranolol. Within 3 weeks of therapy, the tumor size was reduced and platelet count and the hemoglobin level were normalized. There were no side effects to propranolol encountered.

Keywords: Kaposi haemangioendothelioma, Thrombocytopenia, Anemia, Propranolol

INTRODUCTION

Kasabach- Merritt syndrome (KMS) is a rare and known etiology of thrombocytopenia in a neonate. It is mainly a clinical diagnosis. The index case was managed successfully with medications.

CASE REPORT

A 1 -day old neonate with a history of the swollen and discolored right lower limb at birth (Image 1) was admitted into the neonatal intensive care unit. He was delivered by normal vaginal delivery to G 6 P 1 L1, APLA positive mother. His head circumference, weight, and length were between 15th and 50th centile.

On examination he was pink, active, breathing well in the air with the normal oxygen saturation. The right limb was swollen and also discolored (Image 1). All the peripheral pulses were well felt. Septic markers were within the normal limit. The complete blood count showed thrombocytopenia and anemia with normal coagulation profile. The clinical diagnosis of KMS was made. The MRI of the right leg revealed Kaposi hemangioendothelioma (image 2).

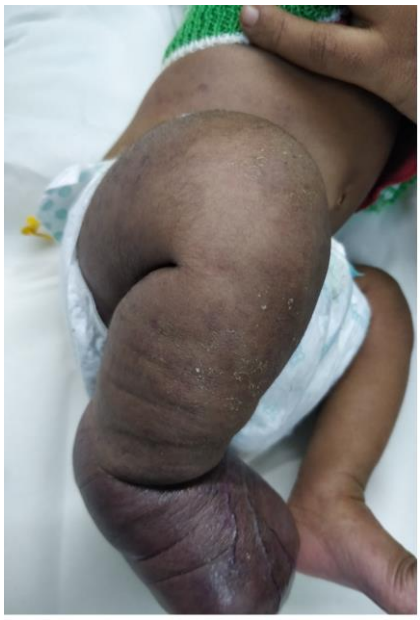


Image 1: The right leg is swollen with prominent vasculature

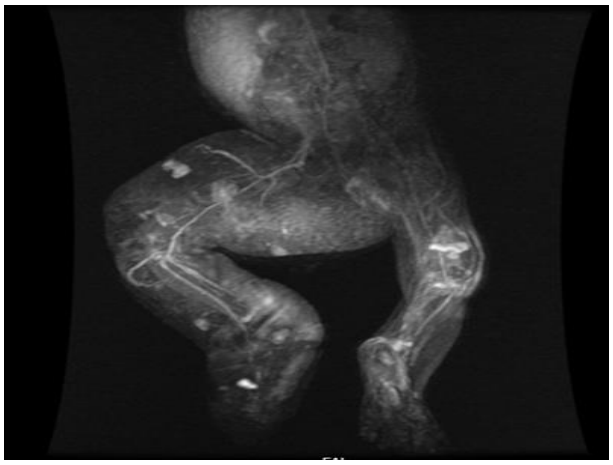


Image 2: MRI with angiogram and venogram of right lower limb showed diffuse thickening of subcutaneous tissue of right lower limb with intermuscular plane extension and a few arterial enhancing foci within, with possible involvement of the talus and calcaneum suggestive of a diffuse vascular tumor of the right lower limb, likely Kaposiform hemangioendothelioma

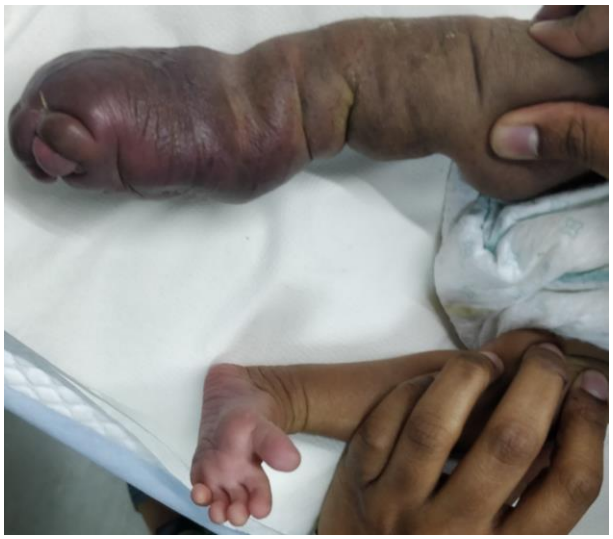
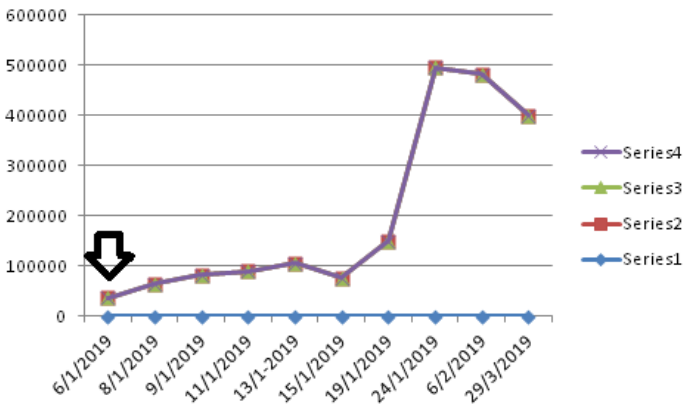
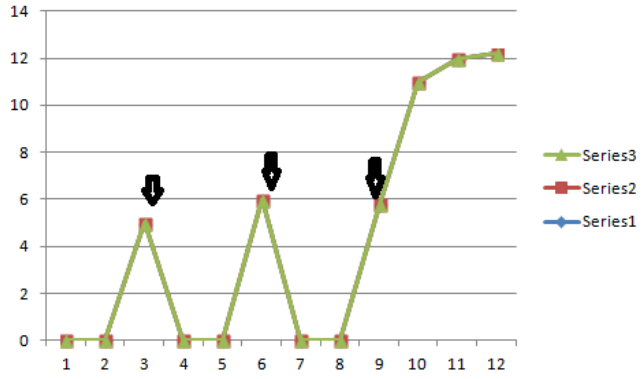


Image 3: The size and color of the right leg on D50 of Propranolol



Graph 1: The graph showing the increasing trend of platelets after the introduction of propranolol (down arrow). The X-axis depicts the day performing and the introduction of propranolol and the Y-axis depicts the platelet count



Graph 2: The graph showing the trend of hemoglobin pre and post packed RBC transfusion. The X-axis depicts the day performing the Hb level and the arrows denote the PRBC transfusion. The Y-axis denote the level of hemoglobin

He was started on propranolol at a dose of 2mg/kg/day in 3 divided doses, monitoring the side effects of the medication like heart rate, blood pressure, blood sugar, and bronchospasm. Gradually the swelling decreased to the present size (image 3) and the hemoglobin and platelet count got normalized (Graph 1 and 2). Currently, he is on propranolol and hemodynamically stable on full enteral feeds having normal milestones and anthropometry.

DISCUSSION

Kasabach- Merritt syndrome is a clinical condition associated with hemangiomas, thrombocytopenia, anemia, and hypofibrinogenemia. The syndrome was described by Kasabach and Merritt in the year 1940 [1]. The Kaposi hemangioendothelioma and tufted hemangiomas are the 2 rare vascular tumors associated with this syndrome. In 80 % of the cases, the syndrome manifests within the first year of life with the estimated mortality rate of 10-30% [2]. In 90% of cases, the syndrome is associated with kaposihaemangioendothelioma [3]. The condition can be life-threatening with severe bleeding leading to disseminated intravascular coagulation. The vascular lesion appears on the skin as a firm, indurate and purpuric, the extremities as noted in our index case, neck, thorax, abdomen and pelvis. The management includes the medications- corticosteroids, Propranolol, anti-malignant drugs, Laser, and surgery given as a stepwise manner either alone or in combination. The steroids are used in the initial step of management. In our index case, propranolol was used as an initial drug as supported by Chiu and colleagues' study on the variable response of kassabach Merritt syndrome to propralnolol [4]. The drug was used at the dose of 2mg/kg/day in three divided doses [5]. Propranolol has many advantages in the management of infantile haemangioma [6]. The reduction in the expression of vascular endothelial growth factor, down-regulation of serine-threonine protein kinase - mitogen-activated protein kinase pathway leading to decreased expression of basic fibroblast growth factor, vasoconstriction, and triggering apoptosis of capillary endothelial cells are the main mechanisms of propranolol to reduce the haemangioma [7]. Despite the multiple medications are used either alone or in combination in the management of KMS, our index case was managed well with Propranolol alone.

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