Case Report

A Newborn with Kasabach Merritt syndrome: A Case Report

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Abstract

Kasabach Merritt syndrome is an uncommon complication of large hemangioma in which there is thrombocytopenia and coagulopathy. We report a case of a newborn male baby 1 day old 2.5kg presented with large hemangioma on both the lower extremity also on back and his initial laboratory test were consistent with thrombocytopenia. He was diagnosed as a case of KMS.

Key Word: Kasabach Merritt syndrome.

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Received Date: 12/03/2015 Revised Date: 22/03/2015 Accepted Date: 25/03/2015

Access this article online	
Quick Response Code:	Website: www.medpulse.in
	DOI: 26 March 2015

INTRODUCTION

Kasabach Merritt Syndrome is characterized by a rapidly developing hemangioma[1], thrombocytopenia, micro-angiopathic haemolytic anaemia and coagulopathy as a result of platelet and RBC trapping and activation of the clotting system within the vasculature of haemangioma. This phenomenon was first described in 1940 by Kasabach and Merritt who took care of an infant with a giant capillary hemangioma and thrombocytopenic purpura. It is a rare disorder that can affect the infants from the time of birth or may appear later in infancy as the vascular malformation grows.[2] More than 80% casesmoccur within first year of life. Male and females are equally affected. Lesions are distinctive vascular tumors typically superficial and solitary but may involve internal structures that include tufted angiomas and Kaposiform haemangio-endothelioma.[3] KMS can be lethal. The estimated mortality rate ranges from 10-37%.[4] Mortality and morbidity are usually associated with visceral involvement, haemorrhage related to aggressive invasion, profoundthrombocytopenia, DIC, severe infections etc

CASE REPORT

A new born male baby, first issue of his nonconsanguineous parents presented with a large sized hemangioma on his both lower extremity and small hemangioma on the back and on parietal region of the scalp. Baby was born term at 38 weeks of gestation by normal vaginal delivary . His mother was 23 year-old-lady with good general health and was on regular antenatal checkup. There was no family history of similar illness. On examination baby was term, with birth Weight 2.5kg with large, solitary 30cmX15cm sized erythematous induration on the right lower extremity and 13x9cm on left lower extremity. Two other indurations were noted on the back (3x2cm sized) and parietal region of the scalp (2x1cm sized). These lesions were diagnosed as hemangioma. The initial laboratory data reveale normocytic normochromic anaemia(Hb% 11gm/dl), thrombocytopenia (Platelet count 10,0000/cumm of blood). Other laboratory results including WBC counts, bleeding time, clotting time, activated partial thromboplastin time (APTT), prothombin time were within the normal range. Patient was diagnosed as Kaasabach Merritt syndrome from a pre-existing hemangioma, clinical findings and laboratory values. Baby was died after 3 days.

How to site this article: Khaire P B, Bhingare R E, Vishal Mule, Patil P S, Lingayat A M. A Newborn with Kasabach Merritt syndrome:A Case Report. *MedPulse – International Medical Journal* March 2015; 2(3): 187-188. http://www.medpulse.in (accessed 28 March 2015).

DISCUSSION

Haemangioma is a commonly encountered benign vascular neoplasm.[5] It is the most common soft tissue neoplasm of infancy. KMS is coagulopathy consisting of intravascular coagulation, clotting and fibrinolysis within the haemangioma. The trigger factors for the development of KMS include surgical intervention, pregnancy, angiography and needle aspiration of haemangioma.[6] The pathogenesis of KMS is yet to be established. However, platelet trapping by abnormally proliferating endothelium within the haemangioma has been proposed as a possible mechanism. Platelet trapping can result in the activation of platelets with secondary activation of coagulation cascades, eventually leading to consumption of various clotting factors. In addition to platelet trapping, excessive blood flow and sheerstress secondary to arteriovenous shunts within the tumor may cause further platelet activation. Both thrombocytopenic status and reduction in coagulation factors eventually result in bleeding within the tumors that manifests as a rapidly growing hemangioma Management of KMS has been challenging because of its rarity and there are no well established systemic treatment strategies. Systemic corticosteroid is considered to be first line therapy in patients with KMS. If a lesion responds to steroid, the dose should be reduced slowly to prevent recurrence. The mechanism of prednisolone in controlling thrombocytopenia, coagulopathy and stabilization of hemangiomas remain unclear, although it appears to increase platelet, increase vasoconstriction, inhibit fibrinolysis and disrupt angiogenesis. In cases resistant to systemic corticosteroid, multiple treatment modalities can be used in a stepwise manner, including interferon-alpha, an antiproliferative and antiangiogenic agent and vincristine, a strong inhibitor of angiogenesis. Radiotherapy is another option which can induce embolization within the hemangioma. These options are usually recommended as second line therapies.

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Source of Support: None Declared
Conflict of Interest: None Declared