Journal Homepage: -www.journalijar.com
INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)
Article DOI:10.21474/IJAR01/5464
DOI URL: http://dx.doi.org/10.21474/IJAR01/5464

RESEARCH ARTICLE

KASABACH MERRITT SYNDROME - A PAEDIATRIC EMERGENCY IN GIANT HAEMANGIOMA.

Dr.Gomathi Vadivelu[1] MS MCh, Dr.Rohith gopinath[2] MS MCh(Resident) and Dr.J.Muthukumaran[3] MBBS DLO MS MCh.

1. Assistant Professor, Dept of Paediatric Surgery, Institute of child health ,Egmore, Chennai.
2. MCh Resident, Dept of Paediatric Surgery, Institute of child health ,Egmore, Chennai.
3. Professor, HOD, Dept of Paediatric Surgery, Institute of child health ,Egmore, Chennai.

Manuscript Info

Manuscript History

Received: 18 July 2017
Final Accepted: 20 August 2017
Published: September 2017

Key words:-
Giant haemangioma,prothrombintime,partialthromboplastintime,platelet trapping.

Abstract

KASABACH-MERRITT phenomenon has been reported only in 200 cases worldwide since 1940. It has 20% to 30% mortality due to platelet trapping , coagulopathy and haemorrhage in brain, pleura & peritoneum. We have effectively treated with steroids and alpha interferon and prevented mortality. Heparin stimulates tumour growth and worsens thrombocytopenia. Our cases were evaluated with CBC, PT, Aptt, Ultra sonogram &MRI. Skin biopsy or liver biopsy were avoided to prevent torrential bleed.

Introduction:-

Children presenting with large hemangioma in skin, liver and gastrointestinal tract must be evaluated for high output congestive cardiac failure. Anemia, thrombocytopenia and hepatomegaly are the classical triad seen in Kasabach-merritt syndrome. Common sites of hemangioma are face, chest and abdomen.

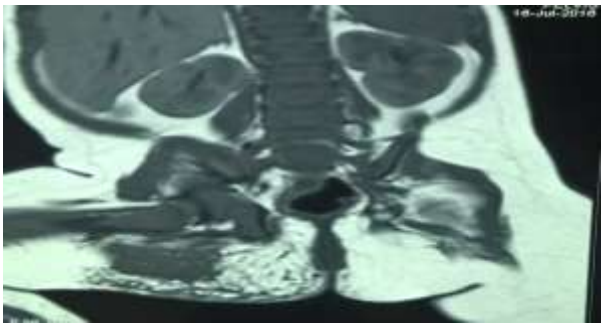
Our cases presented in early infancy with skin lesion in perineum , gluteal region and low grade fever.
On examination child activity was good, no petechiae seen. Hemoglobin was 6.4gm%, platelets-7000 lakhs/cumm Total WBC-8590, PT- NORMAL, Aptt –normal, blood enteric culture was negative. Ultra sonogram of liver and brain were normal.

DAY	TC counts/cumm	PLATELETS Lakhs/cumm			Hb mg/dl
ONE	**13000**	**5000**			7
THREE	10250	20000			9
FIVE	5800	7000			6.2
SEVEN	10,800	20000			9
NINE	9000	20000			11
ELEVEN	5770	41000			12
FIFTEEN	**8100**	**2,00,000**			**11**

Clinical picture of lesions are shown below before and after treatment respectively.

Corresponding Author:-Gomathi Vadivelu.
Address:-Dept of Paediatric Surgery, Institute of child health ,Egmore, Chennai.





MRI of the gluteal hemangioma


Before treatment After treatment

FIG-4:- Baby was started on Intravenous steroid and oral propranalol for first three days and blood transfusion on alternate days for one week

After treatment the lesion subsided completely. Children are followed up for complete regression.

Discussion:-

Endothelial defects cause platelet activation and fibrin formation leading onto platelet trapping and thrombocytopenia. 50% of Kasabach-merritt phenomenon is treated with steroids and propanalol. Alpha interferon and anti-angiogenic drugs as angiostatin and endostatin are used as second line of defence.589 nm pulsed laser can be used in future for embolisation. Fibrinolytic agents as tranexmic acid can be used to prevent platelet trapping. VINCRISTINE 1-1.5mg/m^2 weekly once can be given if lesion is intractable. AVOID heparin as it promotes angiogenesis and aggravates thrombocytopenia.

References:-

1. Kasabach-Merritt with large cutaneous vascular tumor-journal of Indian association of pediatric surgeons - 2012 Jan-Mar pg. 33-36
2. Neonatal Kasabach-Merritt syndrome-Indian journal of medical and pediatric oncology -2011, Oct-Dec pg. 238-241
3. Kasabach HD Merritt K –Capillary haemangioma with extensive purpura:report of a case AM J DID child 1940;59:1063
4. cunrieBG,ShelD,BowringAc.Gianthemangioma of the arm associated cardiac failure and Kasabach-Merritt syndrome in a neonate J paediatric surgery 1991;26:734-7
5. MalcumGP, NiclaidesE, HowardER.Giant cutaneous haemangioma with heart failure in a neonate .Successful surgical treatment .paed .surg in 1990 ;5:713
6. ShulkumBL Argentacc, Choki –Kasabach-Merritt syndrome treatment with Epsilon-Aminocapoic acid and assessment by Indium-3 platelet scintigraphy.