International Journal of Health Sciences and Research
Volume 14; Issue: 5; May 2024
Website: www.ijhsr.org
Case Report *ISSN: 2249-9571*

A Case Report of Kasabach-Merritt Syndrome with Splenomegaly and Splenic Hemangioma

Beny Margrat[1], A. Priya[2], K. Arun Chander[3]

[1,2]Department of Clinical Pharmacology, Apollo Children's Hospital, Greams Road, Thousand Lights, Chennai
[3]Consultant and Head, Department of clinical pharmacology Apollo Children's Hospital, Greams Road, Thousand Lights, Chennai

Correspondence: Dr. K. Arun Chander

DOI: https://doi.org/10.52403/ijhsr.20240527

ABSTRACT

Kasabach-Merritt syndrome is characterized by a combination of thrombocytopenia, microangiopathic hemolytic anemia, hemangioma, and hypofibrinogenemia. KMP (Kasabach-Merritt Phenomena) usually presents with an enlarging purpuric lesion involving the trunk extremities, face, and retroperitoneum. Hemangiomas are benign neoplasms characterized by the abnormal proliferation of blood vessels. It can occur anywhere in the body. Hemangioma can involve numerous solid organs, including the liver, spleen, gastrointestinal tract, brain, and lungs. The term hemangiomatosis may be applied in the setting of very large or numerous hemangiomas. Intralesional fibrinolysis is the main reason for abnormal laboratory values, including elevation of the international normalized ratio and D-dimer level. As a consequence, Kasabach-Merritt syndrome can result in severe disturbances of blood coagulation, such as disseminated intravascular coagulation. Disseminated intravascular coagulation is characterized by the widespread activation of coagulation, which results in the intravascular formation of fibrin and ultimately the thrombotic occlusion of small and midsize vessels. The aim of this report is to describe the clinical course of the patients before and after the treatment, which will hopefully guide future treatment in Kasabach-Merritt syndrome with splenomegaly and splenic hemangioma. There is no universally applicable treatment or procedure for Kasabach-Merritt syndrome, but based on several studies, some treatments have been found to be effective, which include embolization, radiation therapy, laser therapy, sclerotherapy, the use of corticosteroids, interferon, and chemotherapy.

Keywords: Kasabach-Merritt syndrome, Video-assisted thoracoscopic surgery, Disseminated intravascular coagulation, Kaposiform hemangioendothelioma, Tufted Angioma, Fresh Frozen Plasma.

INTRODUCTION

Kasabach-Merritt syndrome is characterized by a combination of thrombocytopenia, microangiopathic hemolytic anemia, hemangioma, and hypofibrinogenemia. It is a rapidly growing vascular tumor with consumptive coagulopathy. It was first described in 1940 by Kasabach-Merritt, who treated an infant with thrombocytopenic purpura and a giant capillary hemangioma. It

is a rare disease that affects babies from birth and also appears later in childhood when the vascular malformation grows. Hemangiomas are benign neoplasms characterized by the abnormal proliferation of blood vessels. It can occur anywhere in the body. Hemangioma can involve numerous solid organs, including the liver, spleen, gastrointestinal tract, brain, and lungs. The term hemangiomatosis may be applied in the setting of very large or numerous hemangiomas. Intralesional fibrinolysis is the main reason for abnormal laboratory values, including elevation of the international normalized ratio and D-dimer level. As a consequence, Kasabach-Merritt syndrome can result in severe disturbances of blood coagulation, such as disseminated intravascular coagulation. Disseminated intravascular coagulation is characterized by the widespread activation of coagulation, which results in the intravascular formation of fibrin and ultimately the thrombotic occlusion of small and midsize vessels.

The pathophysiology of KMP is platelet trapping, activation, and consumption within the abnormal vascular structure, leading to the consumption, degradation and activation of fibrinogen and the consumption of other coagulation factors. Preoperative diagnosis of the same can be confirmed by imaging studies such as CT, MRI or ultrasound. The diagnosis is based on three main findings: thrombocytopenia, enlarging hemangiomas, and consumption coagulopathy.

Splenic hemangiomas, also known as splenic venous malformations, splenic cavernous malformations, or splenic slow flow venous malformations, while being rare lesions, are considered the second most common focal lesions involving the spleen after simple splenic cysts and the most common primary benign neoplasm of the spleen. They are usually found incidentally and have imaging appearances similar to hepatic hemangiomas. Splenomegaly is defined as the enlargement of the spleen measured by size or weight. The spleen plays a significantrole in hematopoiesis and immunosurveillance. The major functions of the spleen include the clearance of abnormal erythrocytes, the removal of microorganisms and antigens, and the synthesis of immunoglobulin G (IgG). The spleen also synthesizes the immune system peptides properdin and tuftsin. Approximately one-third of circulating platelets are stored in the spleen

If Kasabach-Merritt syndrome associated with splenomegaly is left untreated, it can be life-threatening with a high mortality rate. The purpose of this report is to describe the clinical courseof the patients before and after the treatment, which will hopefully guide future treatment in Kasabach-Merritt syndrome with splenomegaly and splenic hemangioma.

CASE REPORT

A 2-year-old baby boy was admitted to Apollo Children's Hospital with the complaints of long- term disseminated intravascular coagulation (DIC), low platelets, prolonged prothrombin time (PT) and partial thromboplastin time (PTT) with multiple hematomas and bruises in the backside and flanks. On physical examination, the patient's weight was noted to be 9.9 kg, the baby was afebrile and found to be pallor, the abdomen was soft, distended, flanks free, hernial orifices free, and hepatosplenomegaly was present. He had a past history of central conducting lymphatic anomaly (CCLA – presenting as chylothorax), hypothyroidism, and hepatosplenomegaly. He had a surgical history of video-assisted thoracoscopic surgery (VATS) and thoracic duct embolization. The PET CT shows splenic enlargement with multiple hypoechoic lesions (hemangiomas). The MRI shows vascular malformations in the spleen.

The initial complete blood count showed a haemoglobin level of 10.4 g/dl, a TC of 14860 million cells/cmm, packed cell volume (PCV)

of 32%, platelet count of 12000cumm, prothrombin time: control of 27/37, INR of 1.85, and PTT Time: control 11/20. The initial biochemistry showed total bilirubin 1.4mg/dl, direct bilirubin 0.4mg/dl, indirect bilirubin 1.0mg/dl, total protein 7.3g/dl, albumin 4.5g/dl, globulin 4.5g/dl, AST 43U/L, ALP 117U/L, and Gamma GTP 9U/L. The above clinical findings, imaging studies and laboratory investigations strongly suggested the diagnosis of Kasabach- merit Syndrome with splenomegaly and splenic hemangioma. So it was planned to undergo a splenectomy.

On day 1 of admission, splenectomy surgery was performed, for which the patient has been admitted. During surgery, a left subcostal incision was made, the spleen was delivered, omental adhesions to the spleen were divided, the pancreatic tail area was fused to helium and dissection was done around the helium and pancreas, the hilar area was clamped with splenic vessels and ligated separately, the short gastric artery was divided, and a splenectomy was done. Homeostasis was achieved. Inj. Cefuroxime 500mg was given as a surgical prophylactic antibiotic, and Inj Paracetamol, Inj Tramadol, Inj Pantoprazole and Inj Emest were given as treatment choices for symptomatic relief. The biochemistry report showed urea: 7mg/dl, creatinine: 0.41 mg/dl, sodium at 134 mEq/L, and potassium 3.8mEq/L.

On postoperative day 2, the child had abdominal pain, vomiting, and fever spikes. Hence, paralytic ileus with sepsis was suspected. Inj.Piptaz and Tab Pentids 100mg were started as empirical therapies. The hematological parameters of the disease were repeated after the surgery. The complete blood count showed a haemoglobin level of 12.2 gm/dl, WBC of 27260 million cells/cmm, packed cell volume (PCV) of 36%, platelet count of 95000cumm, ESR of 2mm/hr, neutrophils 87%, lymphocytes of 8%, Monocytes 5% and plasma fibrinogen 217.3mg/dl. Packed red cell transfusions, platelet transfusions, and fresh frozen plasma were given. The hematological parameters of the disease were again repeated, which showed a haemoglobin of 12.9 gm/dl, WBC 18710 million cells/cmm, packed cell volume (PCV) of 38%, platelet count 1,34000 cumm, ESR of 2mm/hr, Neutrophils of 65%, Lymphocytes 27%, monocytes of 8%, and plasma fibrinogen 375mg/dl.

Further investigation showed nucleated RBC, Hematologist's opinion was obtained, and started on intravenous immunoglobulin (5 grams over 8 hours) on day 4. Corticosteroids and mTOR inhibitors were also given as treatment options for KMS. There was a continuous fever spike, andthe child was also suspected of having a COVID-19 infection. The child was continued with the above medications and treated symptomatically. Oral liquids were started and were tolerated by the child. The drain was removed on day 5. The child was found to be hemodynamically stable and hence discharged.

DISCUSSION

The most important part of KMS management is to improve our understanding on diagnosis and treatment of underlying problems, treatment is therefore started with Splenectomy. The average weight of a normal spleen is 100 to 250 grams. The size of the spleen varies from person to person and also depends on sex, age, habit, and height. The massive spleen weighs at least 500 to 1000 grams. The most common disorders associated with splenomegaly are infectious diseases, hepatic disease, Haematological disorders, congestive disorders, and inflammatory conditions. Among patients with splenomegaly, 17% had hepatic diseases, 31% had hematological disorders, and 8%had infectious diseases. In massive splenomegaly cases, the most frequently associated conditionwas found to be chronic leukaemia.[1] In India, the most common cause of splenomegaly is malaria. The other causes

include chronic myeloid Leukaemia, enteric fever, liver cirrhosis, non-cirrhotic portal fibrosis and hyper-reactive malarial splenomegaly syndrome. About 25 to 40% of the causesof splenomegaly were not identified on the usual evaluation.[2]

In 1940, Kasabach and Merritt reported a case of an infant having a huge hemangioma, thrombocytopenic-related purpuric spots, and microangiopathic hemolytic anaemia with consumption coagulopathy. So this condition was later named as Kasabach- Merritt syndrome.[3] this condition is life-threatening, and the mortality rate is in the range of 30 to 40%.[4] Diffuse cavernous Hemangiomas occur most commonly in the spleen, liver, mediastinum and bones. Kasabach-Merritt syndrome may also lead to Kaposiform Hemangioendothelioma (KHE) and Tufted Angioma (TA). So there should be a complete histological assessment of the entire lesion. Splenic hemangioma and hemangiomatosis are more common in children than in adults.

Hemangioma is a congenital benign vascular lesion that affects approximately 2-6% of the generalpopulation.[5] Hemangioma can involve numerous solid organs, including the liver, spleen, gastrointestinal tract, brain, and lungs. The term hemangiomatosis may be applied in the setting of very large or numerous hemangiomas. It can trigger high-output cardiac failure and the potentially fatal Kasabach-Merritt syndrome.[6] the pathophysiology of Kasabach Merritt syndrome is usually a localized coagulopathy leading to a giant hemangioma with thrombocytopenia. The main causeof death in Kasabach-Merritt syndrome is hemorrhage & symptomatic thrombocytopenia, which requires aggressive platelet support.[7]

There is no universally applicable treatment and procedure for Kasabach-Merritt syndrome due to its rarity.[8,6] Treatment of KMS is performed mainly to manage the coagulopathy and hemangiomas. Pharmacological therapies include glucocorticoids, vincristine, interferon alpha, sirolimus, and other supportive drugs [9] Sirolimus has been previously reported to be effective in treating KMP patients along with steroids, as it results in an increase in the platelet count and a decrease in the tumor size.[10] In this case patient has been given Cap. Sirolimus 0.25mg once daily, Tab. prednisolone 2.5mg twice daily and other supportive drugs after splenectomy.

It is necessary to use large volumes of plasma in order to correct coagulation defects associated with a prolonged APTT or PT (greater than 1.5 times the normal value) or decreased fibrinogen level (less than 1.5 g/dl). An initial dose of 15 ml/kg of FFP (fresh frozen plasma) is clinically recommended and usually administered, which has been followed in this case.[11] Beyond those treatments, the definitive treatment option for Kasabach-Merritt syndrome is surgical removal of the tumor or related organ. The embolization of the tumour may provide some temporary benefits for the condition, along with the supportive therapies.[8,6] According to several studies, the non- pharmacological treatments suggested for Kasabach-Merritt syndrome include embolization, radiation therapy, laser therapy, sclerotherapy and chemotherapy.[12] Low-molecular-weight heparin combined with a supplement of associated coagulation factor helps control consumptive coagulopathy complicated by diffuse cavernous hemangioma. Splenectomy is indicated for children with splenomegaly and splenic hemangioma.[13] In this case, splenectomy combined with other supportive treatments gave us better results for the diagnosis of Kasabach-Merritt disease. Despite his serious health problems, the baby was in good spirits after the first few days of the splenectomy in the hospital and, ironically, that lifted our spirits. Finally, we are happy that baby is recovering from his bad condition. His liver and kidney function remained normal throughout his hospital stay

and he was discharged with follow-up plan.

Declaration by Authors

Ethical Approval: The study was approved by the Institutional ethics committee, Apollo Children's Hospital, Chennai.

Acknowledgement: None

Source of Funding: None

Conflict of Interest: The authors declare no conflict of interest.

REFERENCE

1. Marsden P D, Hamilton P J. Splenomegaly in the tropics. Br Med J 1969; 1 :99 doi:10.1136/bmj.1.5636.99
2. O'Reilly RA. Splenomegaly in 2,505 patients in a large university medical center from 1913 to 1995. 1913 to 1962: 2,056 patients. West J Med. 1998 Aug;169(2):78-87. PMID: 9735688; PMCID: PMC1305176.
3. Pinkhas J, Djaldetti M, de Vries A, Safra D, Dollberg L. Diffuse angiomatosis with hypersplenism. Splenectomy followed by polycythemia. Am J Med. 1968 Nov;45(5):795- 801. doi: 10.1016/0002-9343(68)90213-1. PMID: 5687260.
4. Kasabach HH, Merritt KK: Capillary hemangioma with extensive purpura: report of a case. Am J Dis Child: 1063-70, 1940.
5. Maguiness S, Guenther L. Kasabach Merritt syndrome. J Cutan Med Surg 2002;6: 335-339.
6. Yildirm Karslioglu, Ali Faut Cicek, Mukerrem Safali, Ahmet Ifran, Cengiz Beyan. A splenic hemangiomatosis case manifested by Kasabach Merritt syndrome in an adult. Turkish Journal of Pathology 2008; 24 (2): 107-110.
7. Henley JD, Danielson CF, Rothenberger SS, Hsueh Y, Thelia A, McCarthy LJ. Kasabach-Merritt syndrome with profound platelet support. Am J Clin Pathol. 1993 May;99(5):628- 30. doi: 10.1093/ajcp/99.5.628. PMID: 8493954.
8. Dufau JP, le Tourneau A, Audouin J, Delmer A, Diebold J. Isolated diffuse hemangiomatosis of the spleen with Kasabach-Merritt-like syndrome. Histopathology. 1999 Oct;35(4):337-44. doi: 10.1046/j.1365-2559.1999.00726.x. PMID: 10564388.
9. Liu Y, Wu X, Ye L, Xu H. Successful treatment of a patient with Kasabach-Merritt syndrome and multiple giant hepatic hemangiomas. J Int Med Res. 2020 Jan;48(1):300060519898358. doi: 10.1177/0300060519898358. PMID: 31948308; PMCID: PMC7113715.
10. Combination therapy for pediatric patients with Kasabach–Merritt phenomenon A single-center retrospective study Li Zhang, MDa, Lang Liu, MDb, Huanmin Luoc, Wenbiao Xu, MDd, Huishan Chen, MDe, Muxia Yan, MDa, and Yiqian Wang, PhDf,
11. Wada et al. Journal of Intensive Care 2014, 2:15 Diagnosis and treatment of disseminated intravascular coagulation (DIC) according to four DIC guidelines
12. Abass K, Saad H, Kherala M, Abd-Elsayed AA. Successful treatment of kasabach-merritt syndrome with vincristine and surgery: a case report and review of literature. Cases J. 2008 May 23;1(1):9. doi: 10.1186/1757-1626-1-9. PMID: 18577262; PMCID: PMC2438320.
13. Jing Yan Tang, Jing Chen, Ci Pan, Min- Zhi Yin, Min Zhu. Diffuse cavernous hemangioma of the spleen with Kasabach- Meritt Syndrome misdiagnose as idiopathic thrombocytopenis in a child. World J Pediatr, Vol 4 No 3. August 15, 2008. 227-230.

How to cite this article: Beny Margrat, A. Priya, K. Arun Chander. A case report of Kasabach-Merritt syndrome with splenomegaly and splenic hemangioma. *Int J Health Sci Res.* 2024; 14(5):219-223. DOI: *10.52403/ijhsr.20240527*

******