www.jhas-bhs.com

ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

Journal of Hematology and Allied Sciences

Case Report

Kasabach Merritt syndrome in a 28-year-old female: A strange case report

Swapan Kumar Sinha[1], Rishu Vidhatri[2], Debes Pal[1], Balaram Gupta[1]

[1]Department of Pathology, Jagannath Gupta Institute of Medical Sciences and Hospital, [2]Department of Clinical Haematology, NRS Medical College and Hospital, Kolkata, West Bengal, India.

***Corresponding author:**
Rishu Vidhatri,
Clinical Haematology, NRS Medical College and Hospital, Kolkata, West Bengal, India.

rishuvidhatrib19@gmail.com

Received: 28 August 2021
Accepted: 06 October 2021
EPub Ahead of Print: 11 Nov 2021
Published: 07 December 2021

DOI
10.25259/JHAS_22_2021

Quick Response Code:

ABSTRACT

Kasabach Merritt syndrome (KMS) is a rare disease in which a benign vascular tumor that is hemangioma grows rapidly, entraps red blood cells, platelets, and coagulation factors leading to activation of coagulation cascade resulting in life-threatening disseminated intravascular coagulation and microangiopathic hemolytic anemia. KMS affects newborns and infants. Rarely can affect older children and adults with only a few cases reported in the existing literature. Clinically patients present with large cutaneous hemangioma usually involving the extremities however visceral organs may be involved in some cases along with anemia, thrombocytopenia, coagulopathy, and bleeding. We report a case of KMS in a 28-year-old female who presented with bilateral subdural hematoma, thrombocytopenia, and consumption coagulopathy. She was given seven days course of methylprednisolone to which she responded well.

Keywords: Hemangioma, Thrombocytopenia, Kasabach Merritt syndrome

INTRODUCTION

Kasabach Merritt Syndrome (KMS) is a rare disorder associated with a high risk of mortality in absence of early intervention. It was first described in 1940 by Kasabach and Merritt in an infant who presented with a giant cutaneous hemangioma, thrombocytopenia, and bleeding manifestation.[1] It mainly affects newborns and infants.[2] Rare cases in older children and adults have been reported. There is no sex predilection.[1] Patient with KMS presents with rapidly enlarging superficial cutaneous hemangioma usually involving the extremities. Some patients may present with vascular tumors involving the visceral organ such as the spleen, liver, and brain in form of tufted angioma and Kapasiform hemangioendothelioma.[3] The syndrome is characterized by hemangioma, thrombocytopenia, and consumption coagulopathy. The vasculature of the lesion entraps the red blood cells, platelets, and coagulation factors leading to secondary activation of the coagulation cascade, fibrinogen consumption, and formation of fibrin degraded products, resulting in disseminated intravascular coagulation and features of microangiopathic hemolytic anemia such as schistocytes on peripheral blood smear which may aid in diagnosis.[1]

The mortality rate ranges from 10 to 37%.[4] Mortality is usually associated with involvement of visceral organs, hemorrhage due to invasion, severe thrombocytopenia, disseminated intravascular coagulation, and iatrogenic complications.[4]

CASE REPORT

A young female, 28-year-old presented with complaints of pallor, severe headache, and hematuria for three days. On examination, she had moderate pallor. There was no evidence of yellowish discoloration of eyes, obvious petechiae or ecchymosis, or any organomegaly. At age of 10-year, she was hospitalized for similar complaints to a local hospital where brain imaging was done which revealed a subdural hematoma. However, there was no documentation of any treatment received then and she lost to follow up as she was doing well. There was no history of related complaints in the family. Her menstrual cycle was regular with a 28-day cycle. Routine investigations were performed, complete blood count revealed normocytic anemia (hemoglobin 9.4 g/dl) and thrombocytopenia (platelet-15,000/µl). Peripheral blood examination revealed anisocytosis with hypochromasia and a few fragmented RBCs. Other laboratory parameters, including total leukocyte count, liver function test, urea, creatinine, uric acid, were within the normal biological reference interval. A urine routine examination was done which revealed 10–12 red blood cells/HPF. Prothrombin time and activated partial thromboplastin time were 32 and 36 s respectively, D-Dimer 100 mg/dl (upper limit – 40 mg/dl). Non-contrast computed tomography scan of the brain was done which revealed bilateral acute/subacute subdural hematoma [Figure 1]. Antinuclear antibody and anti-cardiolipin antibody were negative. Based on clinical presentation, examination findings, and laboratory parameters, she was diagnosed with KMS. She received platelets and fresh frozen plasma. She was given 30mg methylprednisolone for seven days to which she responded well and led to gradual improvement of symptoms. At the time of discharge, she was asymptomatic, and her complete blood count was normal (Hemoglobin-11 g/dl, Platelets-112,000/µL). The patient is on regular follow-up on an outdoor basis and is maintaining normal complete blood count parameters.

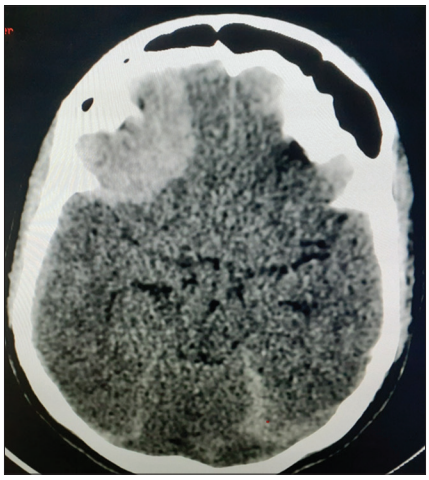

Figure 1: Non contrast computed tomography of brain showing right frontal epidural hemorrhage with left fronto-temporal thin sub dural hemorrhage.

DISCUSSION

Hemangioma is a benign vascular tumor that commonly affects newborns and infants. It spontaneously resolves in around two-third of cases. However, it may be associated with a high risk of mortality in few cases due to associated coagulopathy and microangiopathic hemolytic anemia as seen in KMS. It may be seen uncommonly in adults with only a few cases reports in the existing literature. It clinically presents as rapidly progressing usually cutaneous lesion, disseminated intravascular coagulation, and microangiopathic hemolytic anemia. Iatrogenic factors such as attempts of surgical treatment, angiography, and fine-needle aspiration cytology may sometimes act as triggering factors. Hormonal changes during pregnancy can also exacerbate the lesion.[5-7]

The pathogenesis of KMS is not known. It is believed that proliferating vascular endothelium of tumor entraps platelets and red blood cells result in the activation of platelets with secondary activation of the coagulation cascade, leading to consumption coagulopathy. Excessive blood flow and stress within the tumor sometimes cause further platelet activation. Coagulopathy results in bleeding within the tumor which clinically presents as a rapidly enlarging haemangioma.[7]

For diagnosis complete blood count with peripheral blood smear examination, percentage of reticulocyte, lactate dehydrogenase, coagulation profile including fibrin degraded products and D-dimer level should be performed. In case of no visible or cutaneous vascular lesion a whole-body contrast-enhanced Computed Tomography scan or Magnetic Resonance Imaging should be performed which may reveal a heterogenous enhancing mass. Doppler study can be done to differentiating between solid and vascular tumors.[8]

Management of KMS is challenging because of the rarity of the disease. Systemic corticosteroid is first-line therapy.[9-12] The exact mechanism by which steroids act is unclear, but it appears to increase platelet, increase vasoconstriction, inhibit fibrinolysis, and disrupt angiogenesis. Other second-line options include interferon-alpha, chemotherapeutic drugs such as vincristine, cyclophosphamide and actinomycin D. Complete surgical excision is difficult as lesions are too large and increased risk of bleeding. However, if the tumor is small complete surgical excision might help.12Radiotherapy can be tried in severe cases.[13]

CONCLUSION

KMS is a rare syndrome associated with increased mortality without early intervention. It commonly affects infants and rarely older adults. The disease is characterized by large hemangioma either cutaneous or involving visceral organs associated with coagulopathy and microangiopathic hemolytic anemia. The diagnosis of KMS is based on clinical, laboratory, and radiological parameters. Early diagnosis and treatment can be lifesaving as in our case. The first-line agent is steroids. In cases of steroid resistance, other second-line agents like chemotherapeutic drugs, radiotherapy, and surgical intervention may be tried in selected cases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Kasabach HH, Merritt KK. Capillary hemangioma with extensive purpura: A case report. Am J Dis Child 1940;59:1063-70.
2. Martins AG. Hemangioma and thrombocytopenia. J Pediatr Surg 1970;5:641-8.
3. Beutler E, Lichtman MA, Coller BS, Williams WJ. Williams Hematology. 6th ed. New York: McGraw-Hill; 2001. p. 110-3.
4. Lewis D, Vaidya R. Kasabach merritt syndrome. In: Stat Pearls. Treasure Island, FL: Stat Pearls Publishing; 2021.
5. Esterly NB. Kasabach-merritt syndrome in infants. J Am Acad Dermatol 1983;8:505-13.
6. Karabocuglu M, Basarer N, Aydogan U, Demirkol M, Kurdogklu G, Neyzi O. Development of KMS following needle aspiration of hemangioma. Pediatr Emerg Care 1992;8:218-20.
7. Jona JZ, Kwaan HC, Bjelan M, Raffensperger JG. Disseminated intravascular coagulation after excision of giant hemangioma. Am J Surg 1974;127:588-92.
8. Gong X, Ying H, Zhang Z, Wang L, Li J, Ding A, *et al*. Ultrasonography and magnetic resonance imaging features of kaposiform hemangioendothelioma and tufted angioma. J Dermatol 2019;46:835-42.
9. Shin HY, Ryu KH, Ahn HS. Stepwise multimodal approach in the treatment of Kasabach-Merritt syndrome. Pediatr Int 2000;42:620-4.
10. Wananukul S, Nuchprayoon I, Seksarn P. Treatment of Kasabach-Merritt syndrome: A stepwise regimen of prednisolone, dipyridamole, and interferon. Int J Dermatol 2003;42:741-8.
11. Rodriguez V, Lee A, Witman PM, Anderson PA. Kasabach-Merritt phenomenon: Case series and retrospective review of the mayo clinic experience. J Pediatr Hematol Oncol 2009;31:522-6.
12. Lei HZ, Sun B, Ma YC, Li MM, Wang LF, Jiang SW, *et al*. Retrospective study on the outcomes of infantile tufted angioma complicated by Kasabach-Merritt phenomenon. Clin Chim Acta 2018;486:199-204.
13. Mitsuhashi N, Furuta M, Sakurai H, Takahashi T, Kato S, Nozaki M, *et al*. Outcome of radiation therapy for patients with Kasabach-Merritt syndrome. Int J Radiat Oncol Biol Phys 1997;39:467-73.

How to cite this article: Sinha SK, Vidhatri R, Pal D, Gupta B. Kasabach Merritt syndrome in a 28-year-old female: A strange case report. J Hematol Allied Sci 2021;1:81-3.