e-ISSN: 0975-1556, p-ISSN:2820-2643

Available online on www.ijpcr.com

International Journal of Pharmaceutical and Clinical Research 2024; 16(5); 1015-1018

Original Research Article

Kasabach - Merritt Syndrome: A Case Report

Bibhu Debbarma[1], Subham Ashok Bodhe[2], Rajesh Kishore Debbarma[3], Sukriti Bhattacharjee[4]

[1]Senior Resident, Department of General Medicine, AGMC and GBPH
[2]Post Graduate Trainee, Department of General medicine, AGMC and GBPH
[3]Professor, Department of General Medicine, AGMC and GBPH
[4]Post Graduate Trainee, Department of General Medicine, AGMC and GBPH

Received: 25-02-2024 / Revised: 23-03-2024 / Accepted: 26-04-2024
Corresponding Author: Dr. Bibhu Debbarma
Conflict of interest: Nil

Abstract:

Kasabach-Merritt syndrome is characterized by the combination of rapidly growing vascular tumors, thrombocytopenia, microangiopathic hemolytic anemia, and consumptive coagulopathy. The blood clotting disorder results from platelets and other clotting factors of the blood being used up within the tumor. Although rare, serious complications are present yielding a mortality rate of 10% to 37%. 80% of cases occur in the first year of life with 0.3% of infants affected with hemangiomas. We are reporting a case of a 19-year-old female who presented to us with a passing of black color stool.

Keywords: Kasabach-Meritt Syndrome, hemangiomas, thrombocytopenia, coagulopathy.

Introduction

It was first diagnosed in 1940 by Haig Haigouni Kasabach and Katharine Krom Merritt, who had been taking care of an infant with a giant hemangioma and thrombocytopenia purpura, Kasabach Merritt Syndrome (KMS) is a rare, but serious constellation of a hemangioma within which exist endothelial malformations that cause a consumptive coagulopathy leading to a thrombocytopenia [1,2,3].

Such lesions exist typically superficial and solitary in a male infant, which represent 80% of cases while adult KMS exists in 20% of cases where the hemangioma exists in internal organs such as the liver [4]. Patients can also have gastrointestinal hemangiomas which can lead to life-threatening upper and lower gastrointestinal bleed. Here, we are reporting a case of a 19-year-old who presented to us with complaints of passing a black color stool for several days.

Case Report

A 19-year-old female presented to the Department of Medicine, AGMC and GBP hospital with a history of passing of black color stool for 3 days. At birth, her mother noticed that she had a large skin lesion on the left side of her hand and had undergone surgery when she was 3 years of age.

As she grew up the skin lesions began to start developing in different areas of the body including the tongue. At the age of 16, she was hospitalized in a nearby hospital following the passing of a black stool, and the diagnosis was inconclusive. At the age of 19, she presented to us with a similar history and was in hypovolemic shock with a blood pressure of 80/50 mmHg and a pulse rate of 118 bpm. She is conscious, oriented, and abefrile. On examination, she was pale and had multiple hemangiomas all over the body with a distended abdomen suggestive of ascites.

Laboratory investigations and Imaging studies:

1. Complete Blood Count:- Hb-3.1gm%,Platelet count-10000/cumm,TLC-3000/cumm

2. Kidney function test:- Urea-12 mg/dl, Creatinine-0.42 mg/dl

3. Liver Function Test:- Total Bilirubin-0.9 mg/dl, ALT-14 mg/dl, AST-17 mg/dl, Albumin-2.4mg/dl

4. PT/INR and aPTT are not clotted.

5. Electrolytes:- Serum Sodium- 140mmol/l, Serum Potassium- 4.0 mmol/l

6. Random blood sugar:- 110 mg/dl

7. CT abdomen and Pelvis:

a) Hepatosplenomegaly.

b) A few subpleural ground glass nodular lesions in the apical segment of the right upper lobe and

bilateral lower lobes, could be vascular malformation.

c) Left trace pleural effusion with underlying lung collapse/consolidation.

d) CT features of pulmonary artery hypertension.

8. UGI endoscopy shows gastroduodenal hemangiomas.

9. Colonoscopy reveals colonic hemangiomas.

10. Echocardiography:- Normal study with LVEF 60%.

She was treated with three units of whole blood and iron infusion along with steroid and nor-adrenaline infusion. Her blood pressure improved to 100/60 mmHg with the latest hemoglobin of 7.2 gm%. None of her family members are suffering from a similar disease. No history of any comorbidities like diabetes, hypertension, etc. After stabilization of vitals and controlling upper GI bleeding patient has been discharged from the hospital with oral steroids and was advised to follow up in the OPD.

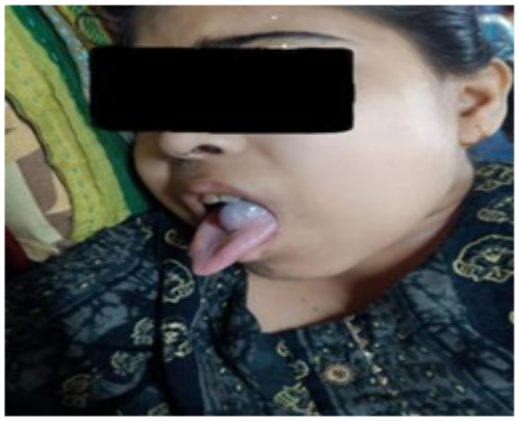


Figure 1: Hemangioma of tongue

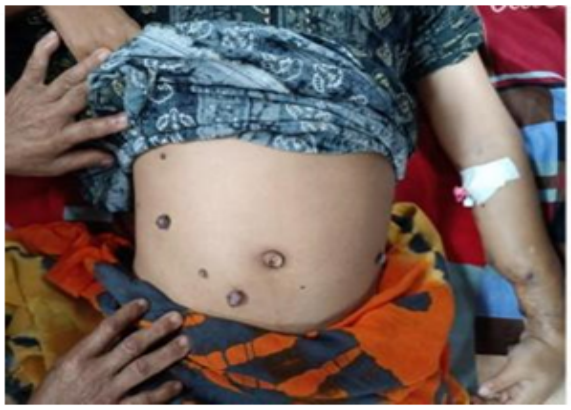


Figure 2: Hemangiomas over the abdomen

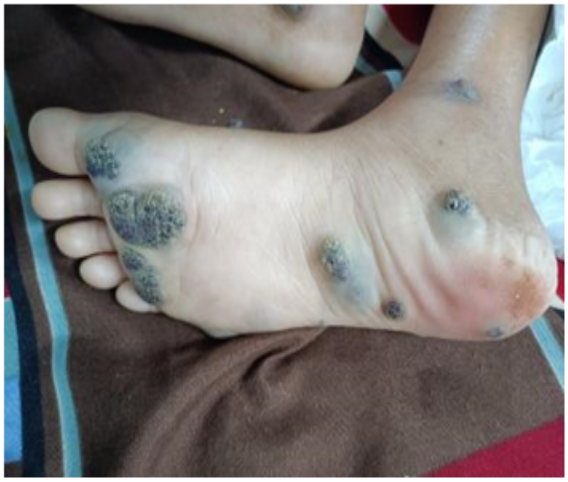


Figure 3: Multiple hemangiomas over the leg

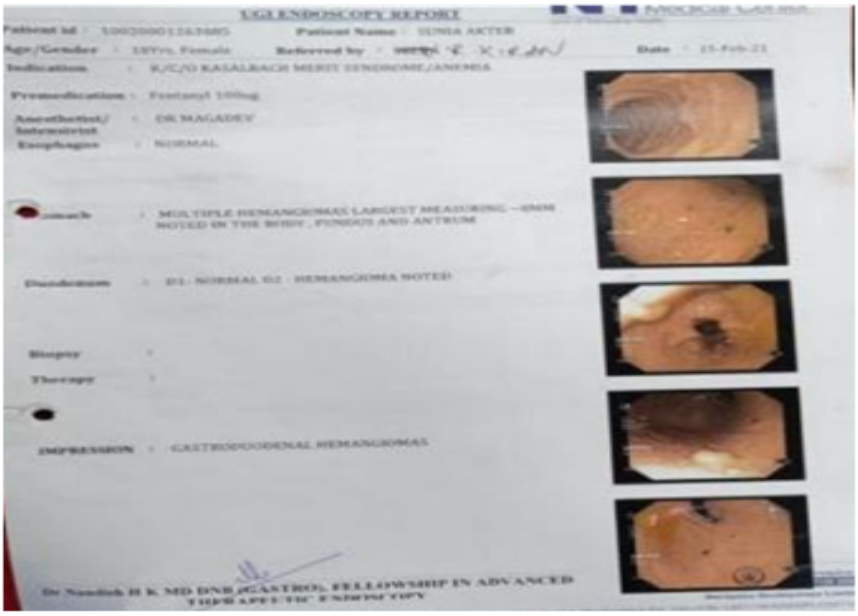
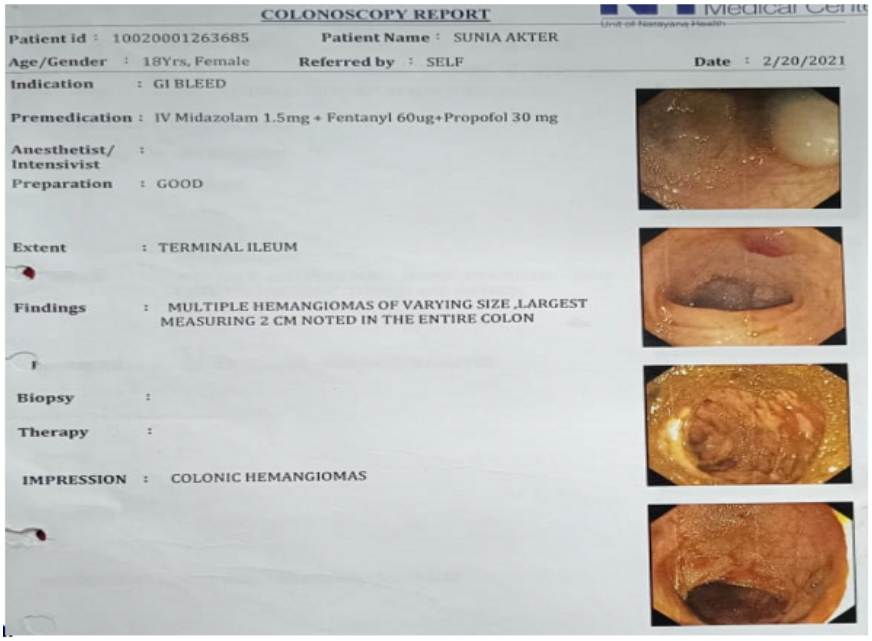


Figure 4: UGI endoscopy report showing gastroduodenal hemangiomas


COLONOSCOPY REPORT

Patient id : 10020001263685 Patient Name : SUNIA AKTER

Age/Gender : 18Yrs, Female Referred by : SELF Date : 2/20/2021

Indication : GI BLEED

Premedication : IV Midazolam 1.5mg + Fentanyl 60ug+Propofol 30 mg

Anesthetist/ Intensivist :

Preparation : GOOD

Extent : TERMINAL ILEUM

Findings : MULTIPLE HEMANGIOMAS OF VARYING SIZE ,LARGEST MEASURING 2 CM NOTED IN THE ENTIRE COLON

Biopsy :

Therapy :

IMPRESSION : COLONIC HEMANGIOMAS

Figure 5: Colonoscopy report showing colonic hemangiomas

Discussion:

Kasabach-Merritt syndrome (KMS) results in a consumptive coagulopathy [1,3] from platelet trapping and aggregation within a specific type of hemangioma and can have a high mortality rate. The hemangioma is often within the skin but can be present anywhere, including retroperitoneal organs, the mediastinum, the pelvis, visceral organs, or the mesentery. For skin lesions, the mortality rate, with treatment, is under 10%, but retroperitoneal tumors have a mortality rate of approximately 60% [1.] The overall mortality rate is between 12 and 50% with death occurring from severe hemorrhage related to disseminated intravascular coagulation, local invasion of vital structures, high output cardiac failure, multi-organ failure, or sepsis [1]. Here, we have a patient who presented to us with multiple hemangiomas over the skin with thrombocytopenia and abnormal PT/INR along with gastroduodenal and colonic hemangiomas, suggestive of Kasabach-Merritt Syndrome. Historically, the first line of treatment has been high-dose systemic corticosteroids.

However, up to two-thirds of lesions will not respond to corticosteroids, or will quickly relapse once treatment is discontinued [5]. Several alternative therapies have been tried with variable results, including interferon α−2a and 2b [6], radiation therapy, and chemotherapeutic agents such as vincristine and actinomycin. The most promising recent option available for the treatment of infantile hemangiomas is propranolol [7].

Reference

1. Maguiness S, Guenther L. Kasabach–Merritt syndrome. Journal of Cutaneous Medicine and Surgery: Incorporating Medical and Surgical Dermatology. 2002 Aug; 6(4):335-9.
2. Kasabach HH, Merritt KK. Capillary hemangioma with extensive purpura: report of a case. American Journal of Diseases of Children. 1940 May 1; 59(5):1063-70.
3. Hall GW. Kasabach–Merritt syndrome: pathogenesis and management. British journal of haematology. 2001 Mar; 112(4): 851-62.
4. Reischle S, Schuller-Petrovic S. Treatment of capillary hemangiomas of early childhood with a new method of cryosurgery. Journal of the American Academy of Dermatology. 2000 May 1; 42(5):809-13.
5. Moore J, Lee M, Garzon M, Soffer S, Kim E, Saouaf R, del Toro G, Yamashiro D, Kandel J. Effective therapy of a vascular tumor of infancy with vincristine. Journal of pediatric surgery.2001 Aug 1; 36(8): 1273-6.
6. Wananukul S, Nuchprayoon I, Seksarn P. Treatment of Kasabach–Merritt syndrome: a stepwise regimen of prednisolone, dipyridamole, and interferon. International journal of dermatology. 2003 Sep; 42(9):741-8.
7. Arunachalam P, Kumar VR, Swathi D. Kasabach–Merritt syndrome with large cutaneous vascular tumors. Journal of Indian Association of Pediatric Surgeons. 2012 Jan; 17(1):33.