CASE REPORT

A case of Kasabach-Merritt Syndrome in a nine-month-old: A rare complication of haemangioma in the young

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Abstract

Kasabach-Merritt Phenomenon (KMP) is a life-threatening consumptive coagulopathy that commonly occurs in infants and young children. It is a combination of an enlarging vascular lesion, thrombocytopenia, microangiopathic haemolytic anaemia, and hypofibrinogemia. The case of a nine-month-old child who presented with a tuft haemangioma leading to the clinical features of KMP is presented. The investigation which was performed and pharmacological treatment initiated at different stages of presentation and the characteristic response to treatment are also discussed.

Keywords: Haemangioma, Vincristine, Consumptive coagulopathy, KMP, Thrombocytopenia, Haemangio-endothelioma.

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Introduction

Kasabach-Merritt Phenomenon is a rare life-threatening syndrome manifesting itself usually by a multitude of disorders involving thrombocytopenia, hypofibrinogenaemia, haemolytic anaemia, and consumptive coagulopathy, and is usually associated with two types of haemangiomas: Tuft and Kaposiform Haemangio-endothelioma (KHE). Affecting 80% of the cases in their first year of life, it is safe to say that it is primarily a disease of infancy.[1,2] It was first described by Kasabach and Merritt in 1940 and around 200 cases have since been reported.[3] Haemangiomas most commonly occur in the head and neck area. KMP is extremely rare and is seen in only 1% of children with tuft haemangiomas. Mortality occurs secondarily to cardiac failure, infections, disseminated intravascular coagulation (DIC), thrombocytopenia leading to high propensity for haemorrhage, and the resultant shock makes the over-all mortality rate range from 10-37%.[4] Locally aggressive haemangiomas can be identified by either clinical methods or imaging techniques like MRI, as well as laboratory evidence such as coagulation profile and complete blood count picture; thrombocytopenia and anaemia, hypofibrinogenaemia and elevated D-dimer levels are of immense importance in the diagnosis of a case of KMP.[2,5] The management is both conservative and surgical. In conservative management cryoprecipitates are given for active bleeding and packed RBCs for any symptomatic anaemia. Since transfusing platelets can further augment consumptive coagulation and bleeding, their use is rather contraindicated in the management of KMP. Moreover, many pharmacological agents are also being used these days in the treatment of KMP e.g., Propranolol, Corticosteroids, Vincristine, Sirolimus, anti-platelet agents, Interferon alpha, with all the cases demonstrating an absolute diversity in their outcomes with the use of these drugs. Surgical management involves resection of the rapidly growing neoplasm but, unfortunately, it is not conceivable owing to the infiltrating aggressive nature of the tumour.[5,6] The present case report highlights the challenges in managing a benign tumour like Tuft Haemangioma (TA) when it presents with a severe complication like KMP. Unlike most reports where patients with TA-associated KMP respond well to corticosteroid therapy, this patient was unresponsive to steroids. Instead, the mainstay of treatment in this case revolved around Vincristine.

In this report, the case of a followed-up case of a nine-month-old child is presented, who was treated with pharmacological intervention involving steroids and Vincristine.

Case Report

A nine-month-old child presented to the emergency department of CMH Lahore, Pakistan, on May 27, 2022, with a history of marked swelling and bluish discolouration of his left forearm, which, the mother stated, had progressively increased since the past eight hours. He was born preterm at 34 weeks of gestation via C-section due to breech presentation and premature rupture of amniotic membranes. After birth, he had been put on oxygen temporarily. The child was fully vaccinated with no significant family history.

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Table: Timeline of Clinical Presentation and Management in Patient.

Age	Clinical Signs and Symptoms	Treatment
At Birth	Small reddish discolouration and swelling on his left forearm	--------
At 3 months	Swelling increased in size. Doppler Ultrasound of left upper limb was done which showed heterogeneous soft tissue lesion with increased blood flow involving poster lateral aspect of left forearm suggestive of haemangioma. The child was diagnosed as a case of tuft haemangioma at 3 months of age.	Beta blocker(1mg/kg/day) for a period of 4 weeks
At 5 months	Swelling and discolouration further increased in size. MRI of left upper limb with contrast was done which demonstrated a poorly defined, significantly enhancing lesion involving left arm, forearm, which confirmed the presence of a vascular malformation likely a tuft haemangioma.	A low dose steroid therapy(2mg/kg/day) was initiated for 4 to 6 weeks and then gradual tapering was done Packed RBCs transfusion, acetaminophen(10mg/kg/day) as needed, and IV antibiotic Linezolid(10mg/kg/dose) 8 hourly for 4 days and then it was tapered to oral syrup (100mg/5ml) and was continued for 5 days for prevention of secondary infection.
At 9 months	Deep reddish-purple swelling now extending from his forearm and involving almost the entire left upper limb, febrile and in visible distress	Vincristine was started weekly for 6 months. 1ml IV diluted in 9ml of normal saline was given in the start and then 0.3 mg once weekly was started free flow in cannula. Acetaminophen(10mg/kg/day) Co-Amoxiclav 50mg/kg/day IV divided every 8 hours.
6 months follow-up	The patient's haemangioma had completely resolved, and the patient was in good health	Patient responded remarkably to vincristine and his condition resolved over the course of 6 months

The child had a small reddish discolouration and swelling on his left forearm since birth, and it had progressively increased in size over a span of nine months and was now extending from his forearm and had involved almost the entire left upper limb. This swelling was relapsing and remitting in nature.

On physical examination, the lesion had extended from the forearm to above the elbow, it was hard in consistency with a deep reddish-purple discolouration covered with a tuft of thin hair (Figure 1). The forearm was tender, and the child was in visible distress and was febrile. There were no signs of dehydration or respiratory distress. Examination of the abdomen and cardiovascular and central nervous systems was unremarkable.

When the patient was three months of age, Doppler ultrasound of the left upper limb was done which showed heterogeneous soft tissue lesion with increased blood flow involving the poster lateral aspect of the left forearm suggestive of haemangioma. The child was diagnosed as a case of tuft haemangioma at three months of age. Treatment given at that time was supportive which included beta blocker (1mg/kg/day) for a period of four weeks, which was then gradually tapered off.

At five months of age, MRI of the left upper limb with contrast was done which demonstrated a 14.2x4.2x7.3cm, poorly defined, significantly enhancing lesion involving the left arm and forearm, which confirmed the presence of a vascular malformation, likely a tuft haemangioma.

Laboratory evaluation showed marked thrombocytopenia and decreased haemoglobin levels. Supportive treatment was initiated which included transfusion of packed RBCs, Acetaminophen (10mg/kg/day) as needed, and IV antibiotic Linezolid (10mg/kg/dose) eight hourly for four days and then it was tapered to oral syrup (100mg/5ml) and was continued for five days for prevention of secondary infection. A low dose steroid therapy (2mg/kg/day) was initiated for four to six weeks and then gradually tapered off considering the overall state of the patient and the presence of underlying haemangioma, but the patient did not show a good response to steroid treatment as the haemangioma was in the same initial state.

At nine months of age (age of presentation at CMH Lahore), the patient presented to the emergency department with the above-mentioned complaint. Doppler ultrasound was done which showed no signs of involvement of an artery or vein. His lab workup showed marked thrombocytopenia and decreased haemoglobin levels along with increased PTT and marked elevation of D dimer levels (Hb:5.5g/dl (Normal: 11-14g/dl), platelets:15 x 10^9 /L (Normal: 150-450 x10^9), D-dimers>1000<2000 (Normal: <0.5 μg/ml), after which the diagnosis of KMP was made.

The patient was admitted to the paediatric ward and underwent supportive therapy including Acetaminophen (10 mg/kg/day) Co-Amoxiclav 50 mg/kg/day IV divided every eight hours. Vincristine was included in the treatment regime. It was administered weekly for six months. Initially, 1ml IV diluted in 9ml of normal saline was given and then 0.3mg once weekly was started free flow in cannula.

The patient responded remarkably to Vincristine and his condition resolved over the course of six months (Figure 2). At the six-month follow-up post-treatment for KMP, the patient's haemangioma had completely resolved, and the patient was in good health.

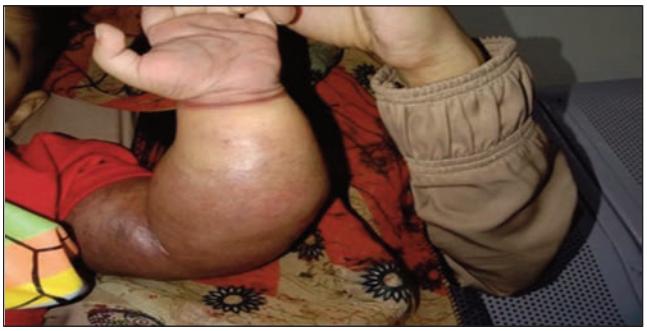

Figure-1: Pre-treatment condition.

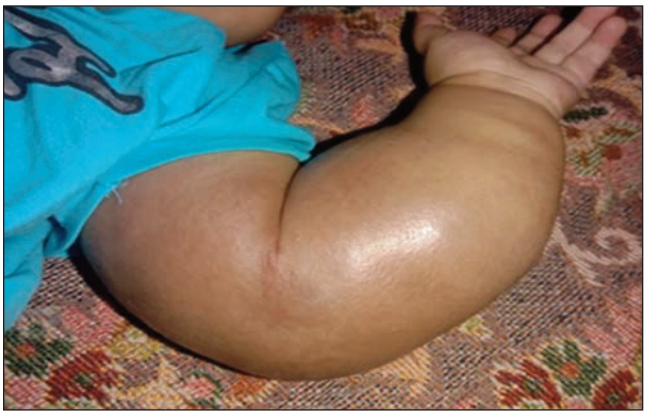

Figure-2: Post-treatment condition.

Discussion

Kasabach Merritt phenomenon (KMP) is a rare coagulative disorder. It is a consumptive coagulopathy caused by trapping of platelets and clotting factors by the abnormal proliferation of capillary endothelial cells having a mortality rate of 30%.[2] Young age, large size of the tuft, and Kaposiform haemangioendotheliomas are potential risk factors for the development of KMP. Most lesions occur below the age of 12 months.[6] The continuous entrapment of platelets and clotting factors results in intra-lesional bleeding and a consequent increase in the size of haemangioma.

The diagnosis of KMP requires lab evaluation i.e. complete blood count, coagulation profile, and PTT. Physical examination is sufficient for diagnosing the cutaneous lesion. However, if an underlying vascular tumour is suspected, Magnetic Resonance Imagining (MRI) is done for confirmatory purposes as was done in the patient under discussion. MRI will reveal enhancement of gadolinium with dermal and subcutaneous thickening.[2]

A similar case was seen in Karachi, Pakistan, in which the haemangioma appeared at the nape of the neck of a four-month-old child. This case was also first treated with Propranolol monotherapy followed by steroids. However, steroids were tapered off when the child developed Cushingoid features as a side effect of steroid use and thereafter eventually changed to Vincristine. In the present case, the initial treatment was with Propranolol and low dose steroids, however, there were no signs of recovery till Vincristine was used. Furthermore, the patient under discussion did not develop any complication as a result of the use of steroids.[7]

The aim of treatment involves two modalities which include providing a supportive treatment and curative therapy. Platelet infusion is only reserved for cases where there is active bleeding and thrombocytopenia. The first line of curative therapy is steroids. Low dose steroids were given to the patient under discussion and the response was not good. Hence, considering the critical condition of the patient, and the angiogenetic component of KMP, chemotherapy i.e. Vincristine was initiated on an urgent basis. Several patients who had not responded to steroids have shown positive response with Vincristine. A 22-week trial with Vincristine have shown 100% recovery rates.[5,8]

Conclusion

Vincristine therapy demonstrated marked improvement in the management of Kasabach Merritt Phenomenon over six months, outperforming the limited efficacy observed with beta- blockers. This case underscores Vincristine's potential as a superior treatment option, warranting further exploration in similar conditions.

Informed consent: Consent was taken from both the parents to publish this case as the child was a minor.

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Conflict of Interest: None.

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Author Contribution:

NZ, SI, SM, NBN, AUR: Design, performed experiments and data curation.

SI: Data curation, literature data, writing and editing.