Case Report

Kasabach – Merritt syndrome: A case report

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ABSTRACT

Kasabach-Merritt syndrome is characterised by the combination of rapidly growing vascular tumour, thrombocytopenia, microangiopathic haemolytic anaemia and consumptive coagulopathy. The blood clotting disorder results from platelets and other clotting factors of the blood being used up within the tumor. We report a two- and- half month old Saudi female infant who presented with epistaxis, conjunctival haemorrage and bilateral periorbital ecchymosis.

Key Words

Kasabach-Merritt syndrome; Haemangioma; Thrombocytopenia; Hypofibrinogenemia.

INTRODUCTION

The association of hemangioma, thrombocytopenia, and hypofibrinogenemia was first described in 1940 by Kasabach and Merritt [1], who took care of an infant with a giant capillary hemangioma and thrombocytopenic purpura. Kasabach-Merritt syndrome (KMS) is a rare disorder that can affect infants from the time of birth, or may appear later in infancy as the vascular malformation grows. Diagnosis of KMS is made based on the constellation of a vascular lesion, thrombocytopenia, consumptive coagulopathy, and microangiopathic hemolytic anemia. Unlike true capillary hemangiomas that regress in childhood and are a cosmetic nuisance, the lesions in KMS are distinctive vascular tumors that include tufted angiomas and kaposiform hemangioendotheliomas [2].

The pathophysiology is believed to be consumption of platelets and fibrinogen by intralesional thrombosis [3]. The lesions are typically superficial and solitary, but may involve internal structures such as the liver. Cardiac failure may result from high-volume arteriovenous shunting. Shock, intracranial bleeding, or other internal hemorrhages may result in mortality rates as high as 30% [3].

CASE REPORT

A two- and-half month old Saudi female infant was admitted to Al -Taif Children Hospital, Kingdom of Saudi Arabia, with history of nasal blockage, mild respiratory distress and noisy breathing for 20 days, associated with bilateral periorbital ecchymosis and redness of both eyes for 15 days (Figure 1A). There was no history of fever, cough, vomiting, trauma or foreign body inhalation or ingestion. She was delivered at term following normal spontaneous

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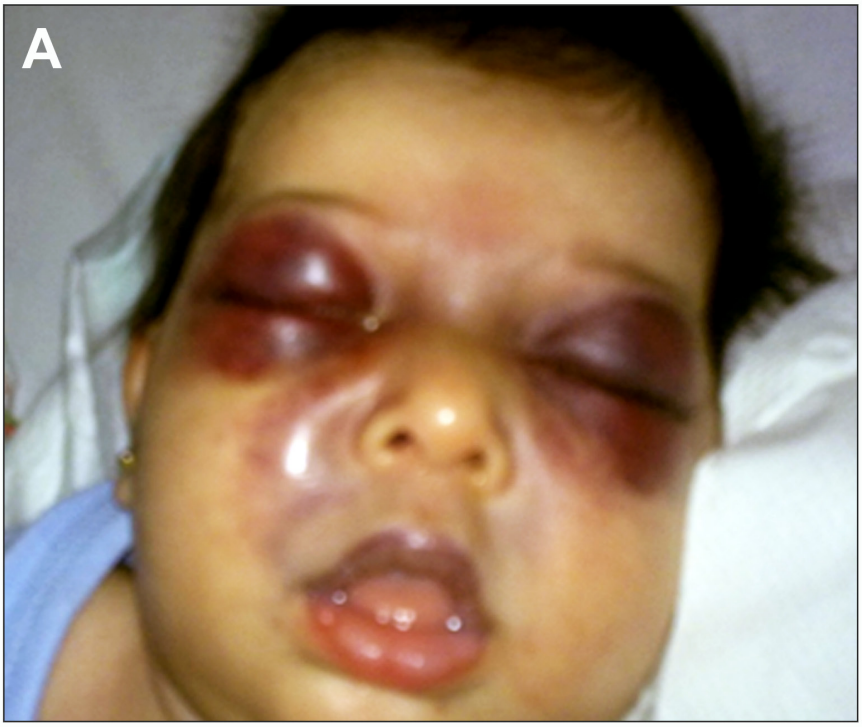

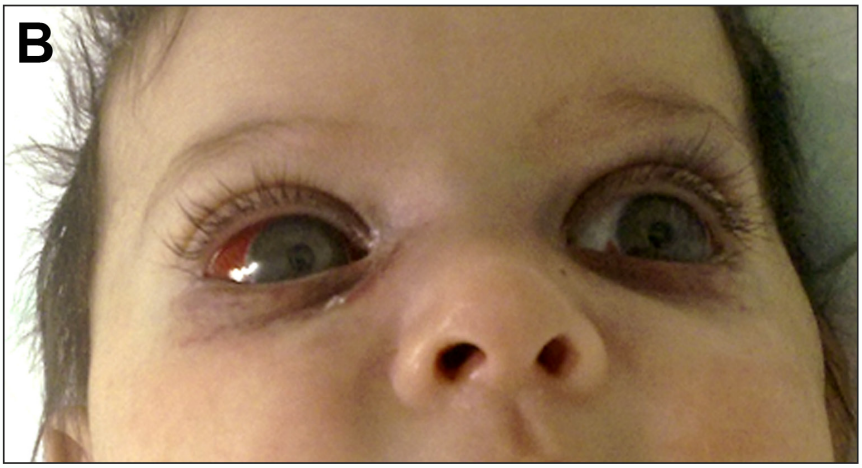

Figure 1- (A) Extensive bilateral periorbital ecchymosis

(B) Apparent bilateral subconjunctival haemorrhage

vaginal delivery to non consanguineous parents. No history of neonatal problems. No past or family history of significance.

On routine physical examination, she was ill looking, pale, not cyanosed or jaundiced. She was keeping her neck in hyperextension posture, afebrile with mild respiratory distress and well hydrated. Extensive bilateral periorbital ecchymosis was documented with bilateral subconjunctival haemorrhage with no discharge (Figure 1B). She did not have purpuric rash, bruises or ecchymosis on her skin or mucus membranes. Her vital signs and oxygen saturation were normal. Examination of the chest, cardiovascular, abdomen and central nervous systems swere normal. Soon after admission she developed epistaxis from both nostrils. Investigations revealed Hb 11.2 g/dl (N=11.5 – 15.5), WBC 6.6 X 10^3/dl (N= 5.5 – 15.5 X 10^3/dl), with normal differential count, platelets 9 x 10^3/dl (N= 150 – 400 X 10^3/dl), and a normal peripheral blood film except for thrombocytopenia.

She had normal chemistry, prothrombin time 12.9, (N= 11 – 15), INR = 1, APPT 40.7 seconds (N= 25 –40) and ESR 10 mm/hr (N= 0 – 10). C - reactive protein was negative. Blood gas result was normal and there was no bacterial growth from blood and urine cultures. Ultrasound and computed tomography (CT) of brain and abdomen were normal. X- ray neck showed enlarged adenoid glands.

While the patient was in hospital she was seen again by the ENT surgeon who assured the parents. She was also seen by the ophthalmologist who confirmed presence of bilateral perioccular and subconjunctival hemorrhage with normal anterior chamber, pupils, with clear lens and normal fundi. There were no vitreous or retinal hemorrhages, and on reevaluation after two days the presence of bilateral retinal hemorrhage was documented. The patient was stable while she was in hospital with no active bleeding from any site although her daily CBC showed decrease in Hb to 9.5 gm/dL (N= 11.5 – 15.5), and low platelet counts ranging between 7 to 15 x 10^3/dl (N=150 – 400). Bone marrow, following aspiration, was normocellular with only megakaryocytic hyperplasia which suggested Idiopathic thrombocytopenic purpura.

The patient received intravenous (IV) immunoglobulin (1 gm/kg /day for 2 days) with several platelet transfusions but without successfully increasing her platelet count. So another course of IV immunoglobulin with the same dose was given for 2 days followed by a course of prednisolone tablets (4 mg/kg for 4 days, then 2 mg/kg) but the platelets count remained very low.

She was referred to a higher specialized centre for further evaluation by the hematologist who agreed with the plan of management . However, while the patient was there, she developed severe respiratory distress with inspiratory stridor and desaturation. An urgent direct laryngoscopy examination showed a moderate to large sized haemangioma at the larynx. A tracheostomy tube was inserted immediately to maintain an adequate airway and the diagnosis of Kasabach-Merritt syndrome was made and was referred to the ENT surgeon for further investigation and treatment of the hemangioma.

DISCUSSION

Kasabach-Merritt syndrome (KMS) was first described by Haig Haigouni Kasabach and Katharine Krom Merritt in 1940 [1]. The syndrome results in a consumptive coagulopathy [4,5] 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 tumours have a mortality rate of approximately 60% [5]. The overall mortality rate is between 12 and 50% with death occurring from severe haemorrhage related to disseminated intravascular coagulation, local invasion of vital structures, high output cardiac failure, multi-organ failure, or sepsis [5].

Kaposiform hemangioendotheliomas are typically solitary tumours which appear in the soft tissues of the limbs, head and neck or retroperitoneum. They are usually seen in infants less than 2 years of age, although cases have been reported in adults. They do not spread (metastasize) but can cause serious problems because of local growth, cardiac failure or the associated Kasabach-Merritt phenomenon.

There are few reports of kaposiform hemangioendotheliomas without Kasabach-Merritt syndrome. Kaposiform haemangioendotheliomas usually regress with time but do not completely disappear. Tufted angiomas usually present before 5 years of age, although they can occur throughout life. They present as brown, red or purple areas of skin and are firm to touch. They are often painful. Spontaneous

regression is unusual. Most tufted angiomas do not cause Kasabach-Merritt syndrome and metastasis is rare.

Treatment aims to involute the tumour to prevent significant morbidity or mortality, or in response to a life-threatening event. Surgical excision is curative but most lesions are not amenable to this option. 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 [6]. Also, this treatment is not without its own troubling adverse effects. A number of alternative therapies have been tried with variable results, including interferon α-2a and 2b [7], radiation therapy and chemotherapeutic agents such as vincristine and actinomycin. The most promising recent option available for treatment of infantile hemangiomas is propranolol [8].

When evaluating a patient with these types of malformations, one must also consider syndromes associated with vascular malformations, such as Klippel-Trenaunay-Weber syndrome and Sturge-Weber syndrome.

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