

A Rare Case Report on Kaposiform Hemangioendothelioma with Kasabach Meritt Syndrome

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Abstract: *Tufted angioma and kaposiform hemangioendothelioma are rare vascular tumours that typically occur during infancy or early childhood. Kasabach-Merritt syndrome is characterised by the combination of rapidly growing vascular tumour, life threatening thrombocytopenia, microangiopathic haemolytic anemia and consumptive coagulopathy[1]. The blood clotting disorder results from platelets and other clotting factors of the blood being used up within the tumor. Here we report a newborn presenting with swelling left side of neck with bluish tinge on day 1 of life.*

Keywords: Kasabach-Merritt syndrome (KMS), Hemangioma, Thrombocytopenia, Hypofibrinogenemia

1. Introduction

The association of hemangioma, thrombocytopenia, and hypofibrinogenemia was first described in 1940 by Kasabach and Merritt [2], 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. Hemangiomas are common vascular tumors that occur in as many as 2.5% of neonates in the United States. Most are benign, and 70-80% regress by the age of 7 years. Some hemangiomas are life-threatening; 1 hemangioma in 300 is associated with coagulopathy. Tufted angiomas and KHE share several histopathological and clinical features and are thought to be of the same neoplastic spectrum [3]. 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 time and mostly in childhood, the lesions in KMS are distinctive vascular tumors that include tufted angiomas and kaposiform hemangioendotheliomas [4].

The pathophysiology is believed to be consumption of platelets and fibrinogen by intralesional thrombosis [5]. The lesions are typically superficial and solitary, but may involve internal structures such as the liver and can also lead to Cardiac failure from high-volume arteriovenous shunting. Shock, intracranial bleeding, or other internal hemorrhages

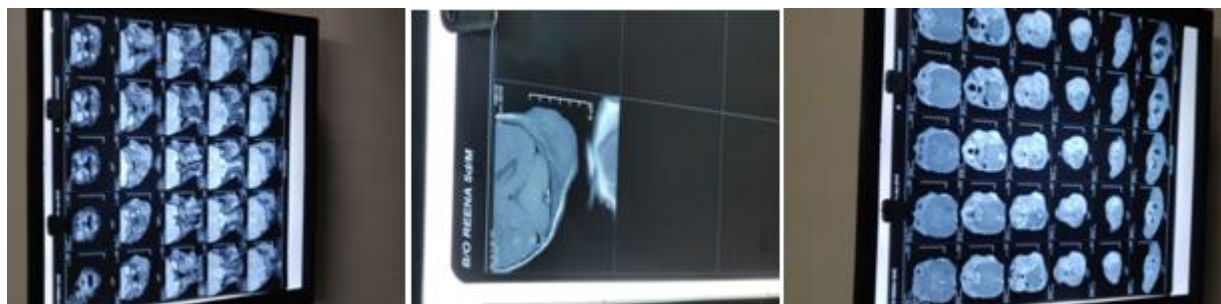
may result in mortality rates as high as 30% [6]. The management of patients with the Kasabach-Merritt phenomenon involves treatment of the tumour and hemostasis support. Surgical excision is the treatment of choice for small, localized tumours. For large, non-resectable tumors, intravenous vincristine and systemic corticosteroids or oral sirolimus with or without systemic corticosteroids [7].

2. Case Report

Here we report a newborn born to a 39 week PGR by Emergency LSCS (Breech with MSL) with birth weight 2.8kg with no birth asphyxia. At birth newborn had swelling around neck on left side with bluish tinge. Newborn developed respiratory distress soon after birth and RDS gradually settled on nasal CPAP and oxygen over the next 3 days. Rest of examination of newborn was normal. Haematological profile of newborn had thrombocytopenia 26000/dl and was given one unit of Platelet Concentrate. Usg neck of newborn revealed lobulated echogenic swelling with intralesional vascularity 4.5 *4 cm. Possibility after consultation with dermatologist of kaposiform hemangioendothelioma with Kasabach Merritt Syndrome was kept and newborn was referred to PGIMER, CHD for diagnosis confirmation and regarding further course of treatment. No family history of significance was observed.



Picture 1 and 2 .At birth, picture of swelling around neck on left side with bluish tinge.



Above are the various **MRI images1, 2 and 3 showing haemangiomas involving brain and its continuity with neck.** After consultation, newborn received weekly intravenous dose of Inj Vincristine 0.1mg intravenous and syrup omnacortil 1mg tds. Initially Child was given this treatment for initial 10 weeks. By the end of 10 weeks, neck swelling had subsided considerably and same schedule was continued for next 10 weeks .Size of the swelling reduced considerably by 1.5 *1.0cm. The infant was monitored with weekly cbc and coagulogram reports, blood pressure and rbs. During this periods infant received multiple platelet infusions and PRBC on 3 occasions. At 6 months of age, child growth parameters were wt -3.038 gm, of c 37cm and all biochemical and haematological parameter were normal. Child was developing normally for his age group. Child was kept on follow up every 2 -3 weeks in OPD and is currently doing well.

3. Discussion

Kasabach-Merritt syndrome (KMS) was first described by Haig Haigouni Kasabach and Katharine Krom Merritt in 1940 [2]. The syndrome leads to a consumptive coagulopathy [6,7] from platelet trapping and aggregation within a specific type of hemangioma, and can have a high mortality rate caused by thrombocytopenia.

The hemangioma is often within the skin but can be present anywhere, including retroperitoneal organs, the mediastinum, the pelvis, visceral organs, or the mesentery. Larger hemangiomas develop complications especially during proliferative phase, most common being ulceration and bleeding. If they are near orifices, it can cause obstruction and impairment with vital functions like vision, feeding, excretion, respiration depending on the site of hemangioma. For skin lesions, the mortality rate, with

treatment, is under 10%, but retroperitoneal tumours have a mortality rate of approximately 60% [7]. 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 [7].

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. 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 hemangioendotheliomas usually regress with time but donot 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 [8]. Also, this treatment is not without its own troubling adverse effects. Systemic steroids can be used to cause faster reduction in size and are the drug of choice especially in life threatening situation. Recommended

starting dose is 2 mg per kg body weight as single morning dose or multiple doses and gradually tapered. Steroids have direct inhibitory effect on angiogenesis and they cause enhanced expression of genes coding for markers of apoptosis. They also act by increasing the sensitivity of vessels to vasoconstrictor substances by decreasing 17 beta oestradiol. (9) A number of alternative therapies have been tried with variable results, including interferon α -2a and 2b [10], radiation therapy and chemotherapeutic agents such as vincristine and actinomycin. The most promising recent option available for treatment of infantile hemangiomas is propranolol [11]. Life threatening hemangiomas that fail to respond to systemic steroids can be treated with interferon alpha given subcutaneously. 1-3 million units/m². It acts by inhibiting angiogenesis. Flu like syndrome, spastic diplegia, neutropenia, renal failure are some of the side effects. Other immunosuppressive agents like cyclophosphamide, vincristine and intralesional bleomycin are also used for hemangiomas that are refractory to conventional treatments. Response to vincristine has been seen in about 80% of the cases.[10] When evaluating a patient with these types of malformations, we should consider syndromes associated with vascular malformations, such as Klippel-Trenaunay-Weber syndrome and Sturge- Weber syndrome.

4. Conclusion

Majority of hemangiomas are smaller lesions and these undergo spontaneous resolution. Complicated and life-threatening lesions needs immediate attention with agents that cause faster regression. Treatment option should be individualized and carefully chosen to give maximum benefit to the patients.

5. Funding

None

6. Conflict of Interest

None

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