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Kasabach-Merrit syndrome with congenital hemangioma.

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Abstract

Haemangioma are vascular lesions resulting from abnormal proliferation of blood vessels they are the most common paediatric neoplasm. Haemangioma are congenital lesions that are common in new-borns infants and children. They are generally benign and often resolve spontaneously. However, in 3 to 5% of cases, they can cause complications inherent to their size, to involvement of vital organs and to the concomitant coagulopathy like Kasabach-Merritt syndrome. Kasabach Meritt Syndrome (KMS) is a potentially life threatening coagulopathy characterized by enlarging haemangioma with severe thrombocytopenia and consumption coagulopathy. KMS is associated with kaposiform hemangioendothelioma (KHE), tufted angiomas and rarely with congenital haemangiomas (CHs). Almost 200 cases have been reported in the literature since Kasabach and Merritt described the first case in 1940. More than 80% of cases occur within the first year of life.

Keywords: Kasabach Merrit Syndrome, Haemangioma.

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Introduction

Kasabach merrit syndrome was first noted by kasabach and merrit in 1940 [1]. Kasabach merrit syndrome is a rare, locally aggressive, vascular tumor. It is characterized by a rapidly enlarging vascular anomaly, consumptive coagulopathy, thrombocytopenia, prolonged PT& APTT, hypofibrinogenemia, the presence of D- dimer and fibrin split products with or without microangiopathic haemolytic anaemia. In most patients, the site of the haemangioma is obvious, but retroperitoneal and intraabdominal haemangioma may require body imaging for detection [2].

Case Report

We are reporting a rare case of an 8 day old girl baby who presented to us with a huge swelling on left shoulder extending from shoulder to mid forearm and back to the entire scapular region from birth & yellowish discolouration of body. On evaluation baby was found to have serum total bilirubin to be 36.5, direct 2.5, platelet-4000, PT, INR prolonged with bleeding from puncture site, along with increase in size of haemangioma. This baby was clinically diagnosed to have Kasabacah Merrit Syndrome and was given platelets transfusion, FFP transfusion, intensive photo therapy was started, with this bilirubin level decreased. Then baby was started on methylprenisolone and propranalol. As the baby came with recurrences we took paediatric surgery opinion, where in paediatric surgeon advised to add vincristine and once the tumour size gets reduced plan to excise the lesion.

Discussion

Kasabach Meritt Syndrome (KMS) is a potentially life threatening coagulopathy characterized by enlarging haemangioma with severe thrombocytopenia and consumption coagulopathy. KMS is associated with kaposiform hemangioendothelioma (KHE), tufted angiomas and rarely with congenital hemangiomas (CHs) [3]. Only about 200 cases have been reported in the literature since Kasabach and Merritt described the first case in 1940.

Though hemangioma was commonly diagnosed in new-born period, presentation with KMP in early neonatal period is rare.

Pathogenesis

Although massive and deep-seated hemangiomata are frequently in reports of KMS, the majority are still cutaneous of varying sizes [4]. The factor that determines the catastrophic haematological disturbances is the angiogenic factor basic fibroblast growth factor (bFGF), known to be elevated in patients with active angiogenesis. bFGF was raised in the urine of the infants with proliferating endotheliomas regardless of whether they had KMS or not [5] and fell drastically in cases with good clinical response to therapy [6]. Most haemangiomata that proliferate rapidly subsequently undergo a period of slow spontaneous involution. Angiogenesis appears to be shut off, either by a decrease in angiogenic factors or by an increase in the endogeneous inhibitors of angigeonesis [7].

The pathophysiology of Kasabach-Merritt syndrome is generally presumed to be that of platelet trapping by abnormally proliferating endothelium within the haemangioma. This results in the activation of platelets with a secondary consumption of clotting factor. Various findings support the ‘platelet trapping hypothesis`, including early isotope studies using 51cr-labelled platelets, immune histochemical staining with anti-CD61 antibodies and indium-111 platelet scintigraphy used to identify occult lesions. Continued consumption of both platelets and clotting factors along with the initiation of fibrinolysin, eventually results in intraleisional bleeding which manifest as rapid enlargement of the haemangioma.

Intraleisional thrombosis occurring as part of the DIC-like picture is not often clinically apparent, but would explain the occasional spontaneous resolution of some lesions [7].

Treatment aims to involute the tumors to prevent significant morbidity or mortality, or in response to a life threatening event surgical excision is curative but most lesions are not amendable to this options . Historically the first line of treatment has been high dose systemic corticosteroids. However, up to two-third of lesions will not respond to corticosteroids, or will quickly relapse once treatment is discontinued [8].

Which is similar to the study conducted by Ping Wang et al. on 17 neonates of KMS, where in intravenous steroid therapy was initially effective in 6 patients and ineffective in 11. The 11 patients with a poor response to steroids and the 3 who relapsed underwent arterial embolization therapy, which was effective in 9 patients and ineffective in 4. Subsequently, four patients in whom arterial embolization therapy was ineffective and one with relapse was treated with vincristine. Steroid therapy was effective in 35% of patients, but relapse rate was 50%. Arterial embolization was effective in 64.3% of patients and vincristine was effective in 80% [9].

A number alternative therapies have been tried with variable results including interferon alfa-2a and 2b [10], radiation therapy and chemotherapeutic agents such as vincristine and actinomycin. The most promising recent option available for treatment of infantile haemangioma is propranolol [11].

Conclusion

In conclusion, neonatal KMP is a rare phenomenon. Early diagnosis and institution of treatment is associated with favorable outcome. Steroids are considered as the most effective 1st line treatment. Vincristine, interferon alpha, antifibrinolytics, radiation, embolization, and surgery are subsequent treatment options in steroid non-responders based on affordability, availability, and feasibility of a particular modality.

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