Case Report

Kasabach–Merritt phenomenon

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ABSTRACT

An association between a vascular lesion and a life threatening coagulopathy is called Kasabach-Merritt phenomenon (KMP). It includes thrombocytopenia, microangiopathic hemolytic anemia, and disseminated intravascular coagulopathy. We cannot overstate the need for excellent and careful screening. Treatment modalities of KMP have included medication, radiation, embolization and surgery. Corticosteroids have traditionally been the mainstay of treatment. We report a 4-month-old girl with an extensive vascular lesion involving the left parotid, submandibular, and parapharyngeal regions, and with KMP. We treated her with a mega dose of corticosteroids. Her coagulopathy resolved, and her vascular lesion improved.


Hemangiomas are the most common benign tumor in neonates, with an incidence of 2.5%.1,2 They are initially not present at birth; however, one-third of children will have a reddish macule, pale spot, or telangiectasia.2 Hemangiomas consist of mesodermal endothelial cells that are precursors of venous channels. The lesion undergoes rapid postnatal growth (proliferative phase) during the first 8-12 months, most often in the first 4 weeks.3 The increase in size is due to proliferation of endothelial cells and pericytes, followed by canalization of the solid mass of cells, and neovascularization. This proliferation is thought to be stimulated by elevated levels of angiogenic peptide basic fibroblast growth factor.3 The lesion then slowly regresses over the next 5-8 years (involutive phase). Of the approximately 2.5% of neonates that have a hemangioma, one in 300 develops a life threatening coagulopathy disorder that Kasabach and Merritt4 first described in 1940. They reported the presence of a rapidly growing hemangioma that was associated with thrombocytopenic purpura in a newborn baby. Our current understanding of the components of Kasabach-Merritt phenomenon (KMP) includes thrombocytopenia, microangiopathic hemolytic anemia, and disseminated intravascular coagulopathy. In this paper, we present an infant with a vascular lesion, and KMP. The aim is to draw the attention of physicians, especially in the pediatric, and otolaryngology specialties to this rare phenomenon, and to review the relevant literature.

Case Report. A 4-month-old female infant was admitted to the hospital due to left-sided facial mass, and feeding difficulty. The patient was the term product of a spontaneous vaginal delivery. A flat discolored vascular lesion on the face was observed at the time of birth. This lesion increased in size by 10 days of age. Her mother reported that the child had become increasingly irritated while feeding, and would latch off the bottle as though she were not getting enough air. The mother denied any history of cyanosis or difficulty breathing but noted recent history of bruising, and petechiae. Physical
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Figure 1 - Photograph of the infant showing left parotid swelling and bruises around the eyes.

Although the pathogenesis of KMP is not yet established, the pathophysiology is generally presumed to be that of platelet trapping by abnormally proliferating endothelium within the hemangioma. This results in the activation of platelets with a secondary consumption of clotting factors. The thrombocytopenia is usually profound, with counts often less than 20 x 10^9/L, and the platelet half-life is drastically shortened to between 1-24 hours.6

Symptoms of KMP can present immediately after birth or months later as the hemangioma rapidly proliferates. In the acute phase, infants typically present with a rapidly enlarging hemangioma, which can appear taut, shiny, or discolored and feel warm to touch. It could therefore be mistaken for cellulitis. There is usually no audible bruit. Petechiae, ecchymosis, and pallor are evident. Prolonged bleeding from puncture sites, oozing from the umbilical cord or circumcision, hematuria, and epistaxis are further evidence of the presence of a bleeding diathesis. Furthermore, laboratory evaluation is necessary to diagnose KMP. In addition to the severe thrombocytopenia, hypofibrinogenemia is prominent, and fibrin degradation products are raised. Some degree of clotting factors derangement, and microangiopathic hemolysis is usually present.1,6

The need for excellent, and meticulous screening cannot be overstated, especially in defining the extent of the lesion, and whether it is amenable to surgery. Ultrasound is a rapid and simple method to recognize and monitor most vascular lesions. Hemangioma is seen as steadily, and intensely bright homogeneous masses on contrast-enhanced CT scan. An MRI of the hemangioma displays well-circumscribed, densely lobulated masses with an intermediate signal intensity on T1-weighted images, and a moderately hyperintense signal on T2-weighted images. The MRI findings in KHE show diffuse enhancement with ill-defined margins.
(cutaneous thickening with strands of subcutaneous fat in cutaneous lesions), hemosiderin deposits, and small feeding, and draining vessels. Angiography is invasive but helpful for establishing the size, patency, and number of feeder and collateral vessels before embolization.6

Treatment modalities of KMP have included medication, radiation, embolization, and surgery. Corticosteroids have traditionally been the mainstay of treatment; however, they are frequently ineffective as a single therapeutic modality, and often require concomitant therapy. Interferon-α (IFN-α) working as an antiproliferative/antiangiogenic agent has shown promise in the treatment of KMP, especially in steroid non-responders. More than half of the patients treated with IFN-α will have some response. However, recent reports of spastic diplegia in children treated with IFN-α (2-20%) confirm that this drug should be reserved for life-threatening situations and used for shorter periods (less than 6 months), with close monitoring of neurological condition. Vincristine, a chemotherapeutic agent, has been used for life-threatening hemangiomas resistant to steroids, and recent studies have shown it to be a safe drug.6,7 Anticoagulant, antiplatelet, and antifibrinolytic agents are other pharmaceutical options.

There has been renewed interest in early surgery of lesions of KMP, possibly due to development of new technologies, functional indications, and cosmetic issues. In well-circumscribed, superficial vascular lesions associated with KMP, surgical excision should be considered as a therapeutic option. Reconstruction using skin expansion or local flaps may be an option in some cases. No single modality is universally helpful and combined therapies are often necessary in the treatment of this potentially life-threatening condition.7

References