Case Report

Kasabach–Merritt phenomenon

Khalid A. Al-Mazrou, *MD, FAAP,* ***Mark A. Richardson,*** *MD.*

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.

Saudi Med J 2005; Vol. 26 (8): 1286-1288

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 Merritt[4] 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

From the Department of ORL (Al-Mazrou), King Saud University and University Hospitals, Riyadh, *Kingdom of Saudi Arabia,* and the Department of Otolaryngology, Head and Neck Surgery (Richardson), Oregon Health and Science University, Portland, Oregon, *United States of America.*

Received 9th March 2005. Accepted for publication in final form 11th May 2005.

Address correspondence and reprint request to: Dr. Khalid A. Al-Mazrou, Assistant Professor of Pediatric Otolaryngology, Department of ORL, King Abdul-Aziz University Hospital, Old Airport Street, PO Box 245, Riyadh 11411, *Kingdom of Saudi Arabia.* Tel. +966 (1) 4775735. Fax. +966 (1) 4775748. E-mail:kmazrou@ksu.edu.sa

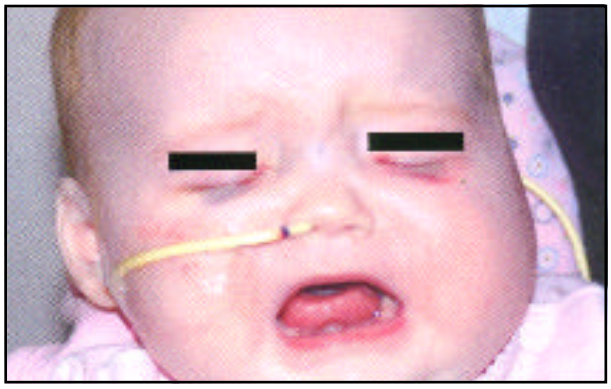

Figure 1 - Photograph of the infant showing left parotid swelling and bruises around the eyes.

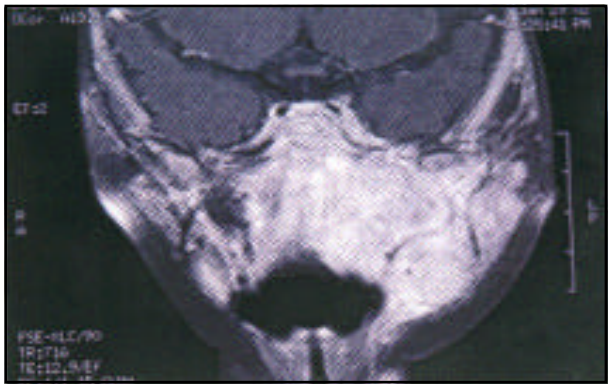

Figure 2 - Coronal T1 weighted MRI of the head demonstrates an enhanced hyperintense mass lesion located in the left parotid region and extending to the para pharynx.

examination revealed an ill-defined bluish-to-violaceous colored, firm mass located on the left parotid gland, the submandibular area, and extending anteriorly to the left eye, and posteriorly to the left ear **(Figure 1)**. The mass was non-pulsatile, and there was no bruit. Intraorally, violaceous discoloration was seen along the posterior aspect of the soft palate, and tonsillar pillars. Blood work showed a platelet count of 16,000. An ultrasound of the abdomen was normal. An axial computed tomography (CT) scan of the head with contrast showed an enhancing mass invading the left parotid gland, and submandibular area, and extending to the parapharyngeal space. A magnetic resonance imaging (MRI) scan **(Figure 2)** revealed the mass was vascular, and invading the pharynx. The child was started on prednisolone (5 mg/kg/day) with marked improvement clinically after 2 weeks. Her platelet count was normal after one month of steroids.

Discussion. The association of an enlarging hemangioma with thrombocytopenia, and consumption coagulopathy was first described by Kasabach and Merritt in 1940.[4] Since that time, more than 200 cases have been reported in the literature.[1] The KMP has it highest incidence in early infancy. Infantile vascular tumors exhibiting KMP have several features that distinguish them from "true" or "common" capillary hemangioma of infancy: 1. Equal gender ratio; 2. Predilection for retroperitoneum, deep neck, mediastinum, pelvis, upper back, and limbs; 3. Specific cutaneous findings (for example, dark purple, smooth, and shiny surface, indurated, tender, and poorly delineated); 4. Always single; 5. Typical MRI findings; 6. Morbidity of thrombocytopenia, and low fibrinogen; and 7. Pathologic characteristics of kaposiform hemangioendothelioma (KHE) and tufted angioma. Both tumors may have histologic evidence of a lymphatic component.[5]

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/I, 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]

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