Kasabach-Merritt Syndrome in a Giant Cavernous Liver Hemangioma

Kasabach-Merritt syndrome (KMS) is characterized by severe thrombocytopenia secondary to platelet trapping by an abnormally proliferated endothelium within congenital subcutaneous and/or visceral hemangiomas. This is usually accompanied by a secondary consumptive coagulopathy, which is reflected by hypofibrinogenemia, elevated levels of fibrin degradation products, and fragmentation of red blood cells. Neither the site nor the size of the hemangioma appears to reliably predict the occurrence of KMS, which is associated with a high mortality (30%-40%) as a result of uncontrollable bleeding secondary to disseminated intravascular coagulation.

This syndrome has its highest incidence in early infancy and most of the cases are secondary to single or multiple subcutaneous hemangiomas. However, those associated with a more severe phenotype are often localized in visceral organs, especially in the retroperitoneal space. The reported incidence of KMS in a giant hepatic hemangioma is very low and its occurrence in adults is exceptional. A KMS secondary to occult hemangiomas should always be considered in patients with spontaneous hematomas due to unexplained thrombocytopenia and coagulopathy. A prompt, aggressive diagnostic imaging approach is paramount to identify the presence of visceral lesions and offer proper treatment.

The gold standard treatment of KMS is the removal of the underlying hemangioma, which will correct the consumptive coagulopathy. Although many nonoperative strategies have been used to control the disease, the only curative treatment remains the surgical resection of the hemangioma. However, surgical resection of hepatic hemangiomas could be hazardous in the presence of uncontrolled coagulopathy secondary to disseminated intravascular coagulation. Perioperative substitution of consumed clotting factors with fresh frozen plasma or platelet concentrates may prevent perioperative bleeding. Hemangiectomy by means of enucleation of the tumor through dissection in a fibrous cleavage plane between the capsule of the hemangioma and surrounding normal liver parenchyma is often preferred rather than anatomical hepatic resections. This technique, when feasible, avoids resection of normal liver tissue and minimizes damage to biliary and vascular structures. Finally, a meticulous surgical hemostasis is paramount to prevent postoperative bleeding in patients with this syndrome.

In conclusion, the development of a thrombocytopenic consumptive coagulopathy in a previously healthy patient should alert physicians regarding the occurrence of a KMS. A prompt surgical removal of the underlying liver hemangioma will provide the better outcome in patients with this life-threatening syndrome.

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REFERENCES