Anesthesia Management of Children With Head and Neck Hemangiomas Associated With the Kasabach–Merritt Phenomenon

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Research article

Keywords: Kasabach–Merritt phenomenon, general anesthesia, anesthetic management

DOI: https://doi.org/10.21203/rs.3.rs-59556/v1

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Abstract

**Background:** We reported the anesthesia management of the patients with head and neck hemangiomas associated with the Kasabach–Merritt phenomenon (KMP).

**Methods:** All 12 young patients with KMP involving the head and neck region who admitted in Henan Provincial Hospital for surgery between June 2012 and December 2016 were included in the study. The data during preoperative preparation including platelet infusion and glucocorticoid treatment, anesthetic management and postoperative recovery were harvested and analysed.

**Results:** Of the 12 patients, the platelet counts were less than $40 \times 10^9 / L$ on admission but no gender difference and three of them responded to glucocorticoid treatment extremely well. The patients who did not respond glucocorticoid treatment well and whose platelets <40× $10^9 / L$ were transfused platelets 12 hours prior to surgery to correct the platelet count to be equal or more than $100 \times 10^9 / L$. The patients who had pneumonia before surgery had a prolonged hospital stay. All 12 patients had surgical excision successfully. After surgery, the platelet counts were increased rapidly in all patients.

**Conclusion** Careful pre-anesthesia assessment and preparation, and thoughtful anesthesia management are needed for KMP patients to receive surgical excision.

Background

In 1940, Kasabach and Merritt described the first case of an infant with a rapidly enlarging congenital vascular mass, which was considered to be capillary hemangioma associated with thrombocytopenic purpura, hemolytic anemia and coagulopathy which was named as Kasabach–Merritt phenomenon (KMP) [1] but the pathogenesis of this disease remains unclear. KMP is a rare but life-threatening vascular condition of infancy characterized with giant hemangiomas, severe thrombocytopenia, consumptive coagulopathy, microangiopathic hemolytic anemia and disseminated intravascular coagulation. Its incidence is much higher in the infants than in the adults, with most of the disease stage typically occurring in infants under six months of age.

KMP may progress rapidly and be accompanied by a high mortality rate due to hemorrhage, infection, vital structure invasion and multiple organ failure [4, 5]. Complete hemangioma removal can control the reduction of platelets and improve coagulation function by complete hemangioma removal but other optional treatments including steroids, cytotoxic agents, interferon, vincristine, radiation and embolization are also applied clinically [6–9] although surgery appears to be the most effective.

During surgery, anesthetic management is challenging and has not been often reported [2]. Herein, we reported our anesthetic management experience of 12 cases of these patients, who had KMP with the head and neck region involvement and received surgery.

Methods
After written informed consent from their parents, the clinical data of 12 children (6 males/6 females) with head and neck involvement of KMP who underwent surgical excision between June 2012 and December 2016 were analysed. One of those patients is showed in Fig. 1. Their demographic data are presented in Table 1. 5 patients had pneumonia before surgery.

**Perioperative management**

If his or her platelet count was lower than $40 \times 10^9/L$, the patient was transfused platelets to reach a platelet count of $100 \times 10^9/L$ 12 hrs prior to surgery. After established peripheral intravenous line, they received 5% glucose solution in the ward accompanied by their parents. On arrival in the operating room, they received heating blankets to prevent hypothermia. They were then administered crystalloid fluid infusion at a rate of 20 ml/kg.

Before anesthesia induction, patients received methylprednisolone sodium succinate 1.6 mg/kg. If the preoperative assessment revealed no airway concerns, after an adequate pre-oxygenation of 5 min, general anesthesia was induced with midazolam 0.05 mg/kg, sufentanil 0.4ug/kg, and cisatracurium 0.15 mg/kg. If the preoperative evaluation found airway difficulties, after an adequate pre-oxygenation of 5 min, general anesthesia was induced with high concentrations of sevoflurane, and the patient retained spontaneous breathing. Once the patient was sufficiently anesthetized, the endotracheal tube was inserted.

The patients were placed radial arterial lines for continuous arterial blood pressure monitoring and intraoperative arterial blood gas and serum electrolyte measurements, and double lumen catheters inserted into the internal jugular or femoral veins under ultrasound guidance to monitor central venous pressure (CVP). The patients were given a mixture of oxygen–air (60%) to ventilate the lungs and 1–1.5 vol% of sevoflurane combined 50 mg/h of propofol to maintain anesthesia. The end expiratory CO$_2$ and inspiratory O$_2$ concentrations were monitored. They were infused blood cells, plasma or cryoprecipitate as necessary during surgery to keep their hemoglobin more than 100 g/L.

Postoperatively, patients were transferred to the pediatric intensive care unit with mechanical ventilatory support. When their condition was stable and they were fully conscious, the endotracheal tube was removed.

**Results**

All 12 patients with KMP from hemangiomas of the head and neck region and had surgical excision successfully. Eight patients less than 3 months, 3 patients between 3–6 months, and only 1 patient more than 6 months (Fig. 2). Of the 12 patients, the platelet counts were less than $40 \times 10^9/L$ on admission but no gender difference. (Fig. 3). Of those patients, three responded well to glucocorticoid treatment which was ineffective in the rest 9 patients. The patients who did not respond well and the platelets $< 40 \times 10^9/L$ were transfused platelets 12 hours prior to surgery to reach a platelet count of $100 \times 10^9/L$ (Fig. 4).
those who responded well to glucocorticoids, the platelet count was increased to a normal level. The glucocorticoid sensitivity was not related with gender (Fig. 5). The patients who had pneumonia before surgery had a prolonged hospital stay (Fig. 6). After surgery, the platelet counts were increased rapidly in all the patient and there were statistical significances (Fig. 7).

**Discussion**

We reported 12 cases of patients who had KMP with head and neck involvement and their anesthesia management. Our experience included careful pre-surgery assessments (airway condition, platelet count) and closely monitoring and better anaesthesia management during surgery.

Low platelets and poor coagulation function increase the perioperative anesthesia risk. We had paid close attentions to platelet count and if it is lower than $40 \times 10^9/L$, platelet transfusion and/or glucocorticoid treatment became necessary before surgery. Glucocorticoids treatment should be also considered as this treatment can help to reduce tissue edema and improve coagulation function and helped decrease the risk of hemorrhage in general.

In addition to the general health of patients, the preoperative evaluation should also include an examination for oral hemangiomas as well as assessments for anemia and thrombocytopenia, since these may require correction before surgery.

The most common complication seen in patients with KMP is severe bleeding resulting from a disturbance in blood coagulation. In addition to the severe and persistent thrombocytopenia characteristic of KMP, patients often manifest elevated D-dimer and low fibrinogen levels [10–12]. Severe anemia is also usually present. Our experience were to infusion red blood cells with amount according to the calculation $[(\text{Hb predicted value} - \text{Hb measured value}) \times \text{body weight} \times 5]$ if Hemoglobin was less than 70 g / L. When the blood loss during surgery is greater than 50% of the blood volume, plasma, 10 ~ 15 ml/kg, and then tranexamic acid 10 ~ 20 mg/kg were infused.

It is important that patients’ airway need to be carefully examined to avoid the occurrence of unexpected bleeding and difficult airway. Making sure anesthesia induction is smooth and avoiding coughing which it can cause bleeding. Mask ventilation and endotracheal intubation should be performed gently to avoid unnecessary airway injury.

Assuring temperature stability is very important for infants and young children. Heat preservation measures are very important, as hypothermia could cause increased blood viscosity, plasma concentration, reduced blood flow, impaired platelet function and inhibited coagulation factor activity, which would further increase the bleeding risk [13]. Hypothermia could also reduce immune function and increase the incidence of postoperative infection. For these reasons, our patients received warming devices to prevent hypothermia when they arrived in the operating room, and patients’ temperatures were closely monitored.
The appropriate depth of anesthesia was maintained. We paid close attention to bleeding in the surgical field and closely monitored the arterial blood pressure and blood gases. Finally, we performed volume management and maintained the internal environment stability according to the CVP, amount of intraoperative bleeding, urine volume and blood gas analysis.

**Conclusions**

KMP is a rare, life-threatening vascular condition of infancy characterized by giant hemangiomas, severe thrombocytopenia, consumptive coagulopathy, microangiopathic hemolytic anemia and disseminated intravascular coagulation. Most affected patients are newborns or infants under the age of six months. The fatality rate of the infants with KMP is high, and surgical resection is the preferred treatment. Due to blood coagulation dysfunction and other factors associated with lesion location and size, infants with KMP involving the head and neck region present great challenges for anesthesiologists and surgeons. Our experience indicated that through better preoperative preparation intraoperative anesthesia management, infants with KMP who received surgery had a good recovery.

**Abbreviations**

KMP
Kasabach–Merritt phenomenon
CVP
central venous pressure
Hb
hemoglobin

**Declarations**

**Ethics and Consent to participate**

This is a retrospective study that does not require ethical approval.

**Competing interests**

The author declare that they have no conflicts of interest.

**Funding**

Joint Research Project of Medical Science and Technology of Henan Province (2018020414); National Natural Science Foundation of China (81901110)

**Authors' contributions**

Clinical data collection: MS, NL, EC, XR,HZ,CD
Acknowledgements

We thank Prof Daqing Ma (Division of Anaesthetics, Pain Medicine and Intensive Care, Department of Surgery and Cancer, Faculty of Medicine, Imperial College London, UK) for his constructive comments during manuscript preparation.

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**Figures**

![Figure 1](image-url)
Figure 2
Figure 3

Number of platelets on admission ($\times 10^9$)

Sex and on admission

Female

Male
Figure 4

Figure 4
Figure 5
Figure 6
Figure 7