Case Report

Anaesthetic management of a neonate with Kasabach-Merritt syndrome

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ABSTRACT

Kasabach-Merritt syndrome is characterised by giant haemangioma, thrombocytopenia and coagulopathy. Triggering of disseminated intravascular coagulation along with the need for massive blood transfusion is the major intraoperative complication. A 1-month-old boy was scheduled for excision and split skin grafting of a giant haemangioma over the left thigh. Investigations revealed severe anaemia with thrombocytopenia that was uncorrected despite multiple blood transfusions. Other treatment modalities were also unsuccessful and the neonate was taken up for excision of the haemangioma in order to correct the consumptive coagulopathy. Standard anaesthesia was administered and all appropriate measures to reduce blood loss were instituted. Massive blood transfusion was required but the intraoperative and post-operative period was uneventful and followed by a significant improvement in the haemoglobin and platelet counts in the post-operative period.

Key words: Anaesthesia, Kasabach-Merritt syndrome, massive blood transfusion

INTRODUCTION

Kasabach-Merritt syndrome (KMS)[1] is a rare disorder characterised by giant haemangioma, thrombocytopenia and coagulopathy. Various treatment modalities have been tried with limited success. Resection of the tumour, whenever feasible is usually curative with a complete reversal of coagulopathy.[2] These cases usually present in infancy; excessive bleeding with triggering of disseminated intra-vascular coagulation is the major intra-operative complication. The following case report illustrates the various concerns implicated in the anaesthetic management of a neonate with KMS.

CASE REPORT

A 2.5 kg 1-month-old boy, delivered at term, afflicted with KMS presented for excision and split skin grafting of a giant haemangioma over the left thigh, extending from the inguinal region to the knee, with a circumferential girth of 32.5 cm [Figure 1]. Patient had received intravenous propranolol[3] and prednisolone[4] for 10 days and intralesional bleomycin injection once, which produced a slight, but insignificant reduction in the size of haemangioma. Apart from pallor, examination was unremarkable and there were no other co-morbidities. The patient had been transfused 150 mL of packed RBCs, 250 mL platelet concentrates and 110 mL of fresh frozen plasma but the anaemia,

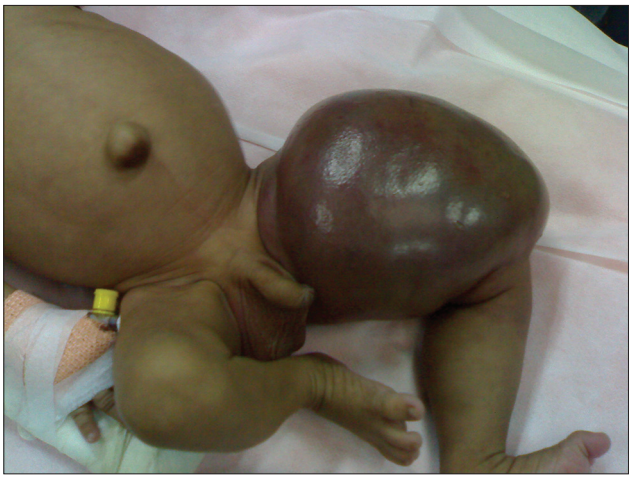

Figure 1: Giant haemangioma with circumferential girth 32.5 cm

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thrombocytopenia and coagulopathy persisted. Pre-operative haemoglobin was 7.6 g/dL, platelets 19,000/cumm, bleeding time 9 min and international normalized ratio (INR) was 1.6. Other biochemical investigations including arterial blood gas analysis were all within normal limits. Appropriate radiological investigations were carried out to rule out haemangioma in other visceral organs.

Intravenous access was secured with two 24 G intravenous cannulae. The neonate was pre-medicated with injection hydrocortisone 10 mg IV and injection fentanyl 2.5 μg IV. After standard induction using sevoflurane and atracurium, an uncuffed endotracheal tube, size 3.0 mm ID was inserted. Paracetamol 70 mg suppository was administered for post-operative analgesia. Normothermia was maintained with the use of Mediprema® Infant Operating Table and HOTLINE® Blood and Fluid Warmer. Since, the application of a conventional tourniquet was technically impossible in an attempt to minimize blood loss, we requested the surgeon to tie a Foley's catheter around the proximal most part of thigh (tourniquet time 35 min). The haemangioma was excised and raw area was covered with a skin graft taken from the excised haemangioma itself, in an attempt to further decrease the blood loss. Intraoperative blood loss was 300 mL and was appropriately replaced with 150 mL packed cells, 50 mL fresh frozen plasma and 50 mL platelet concentrates. Patient was haemodynamically stable throughout the procedure and had adequate urine output. Arterial blood gas analysis and serum electrolytes performed prior to extubation were within normal limits. Recovery from anaesthesia was smooth and uneventful. In the post-operative period, patient received additional 200 ml of packed cells and 50 mL of platelet concentrates. On the second post-operative day, the haemoglobin was 9.9 g/dL, platelets had increased to 64,000 cells/cumm and INR had improved to 1.3. Further post-operative course was uneventful.

DISCUSSION

The diagnosis of KMS is based upon three findings: Enlarging haemangioma, thrombocytopenia and consumption coagulopathy.[1] The exposure of subendothelial elements or abnormal endothelium within the haemangioma results in platelet aggregation and activation, with secondary localised intravascular coagulation and consumption of clotting factors leading to life-threatening coagulopathy.[1,4] The haemangioma may be a cutaneous or visceral lesion and is mostly seen in neonates. Treatment is difficult and many therapeutic modalities[4] including prednisolone, propranol,[3] epsilon-amino-caproic acid, pentoxyfylline, ticlopidine, heparin, interferon, vincristine, radiotherapy, and vascular embolization have been reported, with limited success. Resection of the tumour, whenever feasible, is usually curative with a complete reversal of coagulopathy.[2]

The anaesthetic management of KMS presents various challenges. These patients usually present in early infancy and have severe anaemia (secondary to micro-angiopathic destruction of RBCs), thrombocytopenia and coagulopathy,[4] which is not amenable to correction by transfusion of blood products.[4] Platelet transfusion may not only be futile in increasing platelet counts, but may also stimulate an increase in the size of haemangioma, further worsening the consumptive coagulopathy. Therefore, in patients of KMS, platelets should be administered only in cases with active bleeding.[4]

Since excision of the haemangioma is the only definitive therapy, most patients are taken up for surgery in the presence of anaemia and coagulopathy. Therefore, it is imperative that all possible attempts be made to decrease intraoperative blood loss by various methods such as preoperative embolization, intralesional injection of sclerosants, and intraoperative use of tourniquets, wherever feasible. Recently, recombinant factor VII therapy has been used successfully to reduce the need for blood and blood products.[5] Management of disseminated intravascular coagulation (DIC), if it occurs, is of great concern, and use of anticoagulants and antiplatelet medications should be considered after careful assessment of the risks and benefits.

In spite of all above measures, patients are likely to require massive blood transfusion. Our patient was transfused nearly 2 times his blood volume during the intraoperative and post-operative period. Massive blood transfusion in a neonate may be associated with life threatening complications such as hyperkalaemia, hypocalcaemia, acidosis, citrate toxicity, circulatory overload, transfusion related acute lung injury, haemolytic reactions and hypothermia. However, none of these complications were observed in our case as evidenced by a normal arterial blood gas analysis and serum electrolytes as well as maintenance of adequate urine output and temperature. The location and size of haemangioma may itself be an anaesthetic concern. Lesions have been reported in various

anatomic locations[4] including face, neck, trunk, upper- and lower-extremities and retroperitoneum. Haemangioma on the face may preclude placement of a facemask while lesions involving the pharynx, larynx and trachea may cause bleeding or airway obstruction. Liver haemangioma may lead to jaundice. Intracranial lesions may bleed and a smooth induction and recovery are imperative to reduce the risk of intracranial haemorrhage. Large volume of blood flowing through giant haemangioma can itself precipitate high output congestive heart failure, thus, the anaesthesiologist must be vigilant for signs of heart failure.

In patients treated with steroids, perioperative steroid therapy should be given to prevent adrenal insufficiency. In view of preoperative refractory severe anaemia, intraoperative blood loss with massive blood transfusion, a period of post-operative ventilation should be considered. In our case, the patient could not be put on ventilatory support in the post-operative period due to an unforeseen unavailability of the ventilator and we had to proceed with extubation. However, the post-operative period was uneventful, highlighting the fact that removal of the haemangioma leads to a rapid recovery of the haematological profile with correction of all incrimating variables.

CONCLUSION

KMS is a rare coagulopathic disorder seen in early infancy where blood parameters are unlikely to respond to transfusion of blood or blood products and the patient will have to be operated in the presence of anaemia, thrombocytopenia and coagulopathy. Measures must be instituted to decrease intraoperative blood loss. Further, the anaesthesiologist must be watchful for DIC and complications of massive blood transfusion.

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