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Anesthetic Management of a Case of Pediatric Rhabdomyosarcoma

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Abstract: ***Rhabdomyosarcoma, a tumor of skeletal muscle origin is the second most common soft tissue sarcoma seen in childhood after osteosarcoma. Common sites of occurrence are the genitourinary tract, retro peritoneum, head and neck and to a lesser extend, the extremities. RMS is a high-grade malignant tumor with extensive local invasion as well as early hemorrhagic and lymphatic dissemination. In spite of aggressive approaches including surgery, dose intensive combination chemotherapy and radiation therapy, patient with metastatic disease will usually have a poor outcome. Here we describe the management of a 13 year old child with RMS of fallopian tube requiring extensive staging laparotomy. Due to the extensive surgical resection intraoperative massive blood transfusion was required. Hemorrhagic shock in pediatric surgery is challenging to the anesthesiologist as well as surgeons. Current guidelines in managing major blood loss in adults involve initial resuscitation with crystalloid fluids followed by infusion of blood components as necessary. In contrast to adult population, the pediatric massive transfusion guidelines are vague and mostly it is managed almost in a similar fashion like an adult patient. Here, we attempt to identify challenges posed due to massive transfusion in pediatric age group.***

Keywords: Rhabdomyosarcoma, pediatric age group, massive blood transfusion, anesthetic management

1. Introduction

Rhabdomyosarcoma (RMS), a tumor of skeletal muscle origin, is the second most common soft tissue sarcoma seen in childhood.

Being a high-grade malignant tumor with extensive local invasion and lymphatic dissemination, aggressive treatment approaches like chemoradiation along with surgery are needed. Surgery requires extensive resection and is thus associated with massive intraoperative blood loss, hypothermia and hemodynamic disturbances.

Here we report the successful management of 13-year-old girl with Rhabdomyosarcoma (embryonal-type) of fallopian tube requiring massive transfusion intraoperatively during the cytoreductive surgery.

2. Case Report

A 13yearold girl child who had come to pediatric oncology OP (outpatient) with complaints of abdominal pain, distension, increased urine output fever spikes and early satiety was diagnosed with an abdomino-pelvic mass 13*11*19 cms on CT scan. The histopathology report (HPR) showed Embryonal Rhabdomyosarcoma. On initial assessment it was found to be surgically unresectable due to extensive local spread of tumor (peritoneal thickening, nodularity and enhancement). As she had developed bilateral hydroureteronephrosis (R>L) along with worsening renal function, B/L ureteric stenting was done. She was started on chemotherapy (VAC regimen- vincristine, Adriamycin and cyclophosphamide). Due to poor response to chemotherapy and an increase in tumor size surgical management was planned after explaining the risks.

Cytoreductive surgery was planned.

Pre anesthetic evaluation was unremarkable except for the grossly distended abdomen leading to a decreased air entry bilaterally and difficulty in walking. She was poorly nourished. Her vitals were stable. She weighed 40 kg (Height 163 cm BMI 15.3) She had a PICC line (peripherally inserted central catheter) through which she was started on parenteral nutrition to improve her general condition (as her intake was poor). Airway assessment was normal. CT thorax and abdomen showed tiny indeterminate nodules along fissure of left lung, left ureteral obstruction, moderately hydronephrotic Left kidney. Cardiac evaluation revealed good LV function and Ejection Fraction of 65%. Her blood investigations were within normal limits (Hb 9.2g% and Serum Albumin was 3.5)

On pre-op day an ascitic tapping was done (1Litre) which was replaced with IV Human Albumin 5% 500ml. She was on IV antibiotics Meropenem and Amikacin.

She didn't have any fever spikes 48hrs prior to surgery. She was kept NPO 6hrs for solids and 2hrsfor clear liquids prior to surgery as it is our hospital protocol.

She was given premedication T.Pantoprazole 40mg HS and at 7am on day of surgery and T.Alprazolam0.25mg HS and at 7am on day of surgery. Adequate blood and blood products were arranged after crossmatching. Maintenance IV fluid Ringer Lactate at 80ml/kg was started from 7 am on the morning of surgery.

In the operating room standard ASA monitors were attached. The child was then premedicated with Inj. Glycopyrrolate 0.2mg (10mcg/kg) and Inj.

Midazolam1mg (0.05mg/kg) given intravenously. After preoxygenation for 3 minutes, child was induced with Inj. Fentanyl 80mcg (1-2mcg/kg), Inj.

Lignocaine 60mg(1.5mg/kg) and Inj. Propofol 100mg(2-3mg/kg).

After ensuring bag and mask ventilation she was given Inj. Succinyl Choline 100mg(1-2mg/kg). After 1 minute of BMV, under direct laryngoscopy, she was intubated with cuffed ETT of size 6.5mm. IPPV was commenced with Sevoflurane 1-2% (1 MAC), air and oxygen (50:50). Inj.

Vecuronium 4mg(0.1mg/kg) was given for muscle relaxation.

Post induction epidural catheter was put at the L1-L2 space using 18G Touhy's needle using LOR technique. Epidural catheter was fixed at 10cmafter confirming negative intravascular or intrathecal placement. The ®IJV was cannulated using 14 Fr triple lumen catheter using Seldinger's Technique. A 20 G right radial arterial line was inserted for IBP monitoring as well as for PPV monitoring. A 14 French orogastric tube was also inserted. IV fluids were administered as per Goal Directed Fluid Therapy, guided by PPV (Pulse pressure variation). IV Human Albumin 5% 500ml was also started simultaneously upon opening the abdomen in order to restrict crystalloids, so as to avoid hemodilution.

Intraoperatively muscle relaxation maintained with Inj. Vecuronium 1mg. Analgesia was maintained with 0.2% Ropivacaine at 3-5ml/hour titrated according to BP. Her maximum allowable blood loss was calculated and found to be 365ml. As the tumor resection started, there was substantial oozing.

Since the blood loss exceeded MABL, which was obtained from soaked mops and blood collected in suction bottles, massive transfusion was initiated with Packed Red Cells and products (2:1:1 volume ratio of PRBC: FFP: platelets.

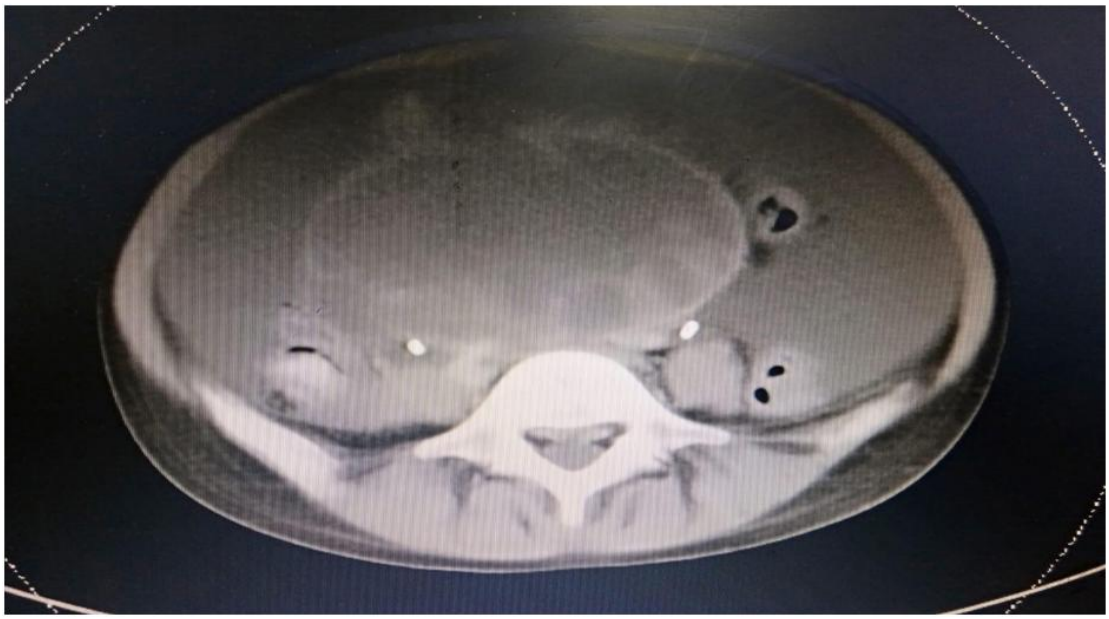
Injection Calcium Gluconate 10% 20 ml (30-100mg/kg) was given after transfusing 3 PRBCs. Additional 500mlof 5% Human Albumin was alsogiven. Because of continued ooze from raw peritoneal surface Tranexamic acid 500mg (10mg/kg) was given.

Surgery lasted for 5 hours and as the child had received quite large amounts of blood products, there was concern for development of TRALI (Transfusion related acute lung injury) as well for non-cardiogenic pulmonary edema. An ABG analysis taken after 2 hours of surgery showed a Ph: 7.4, PCO2: 52, PO2: 154, HCO3: 34, Hb: 10 and Lactate 2.1 at Fi02 of 40% followed by which ventilatory mechanics were adjusted. Urine output remained stable throughout at rate of 1ml/kg/hr. Arterial blood pressure was maintained within normal range, HR was in the range of 100-115 per minute, Etco2 in the range of 35-45 and respiratory rate at 14/min

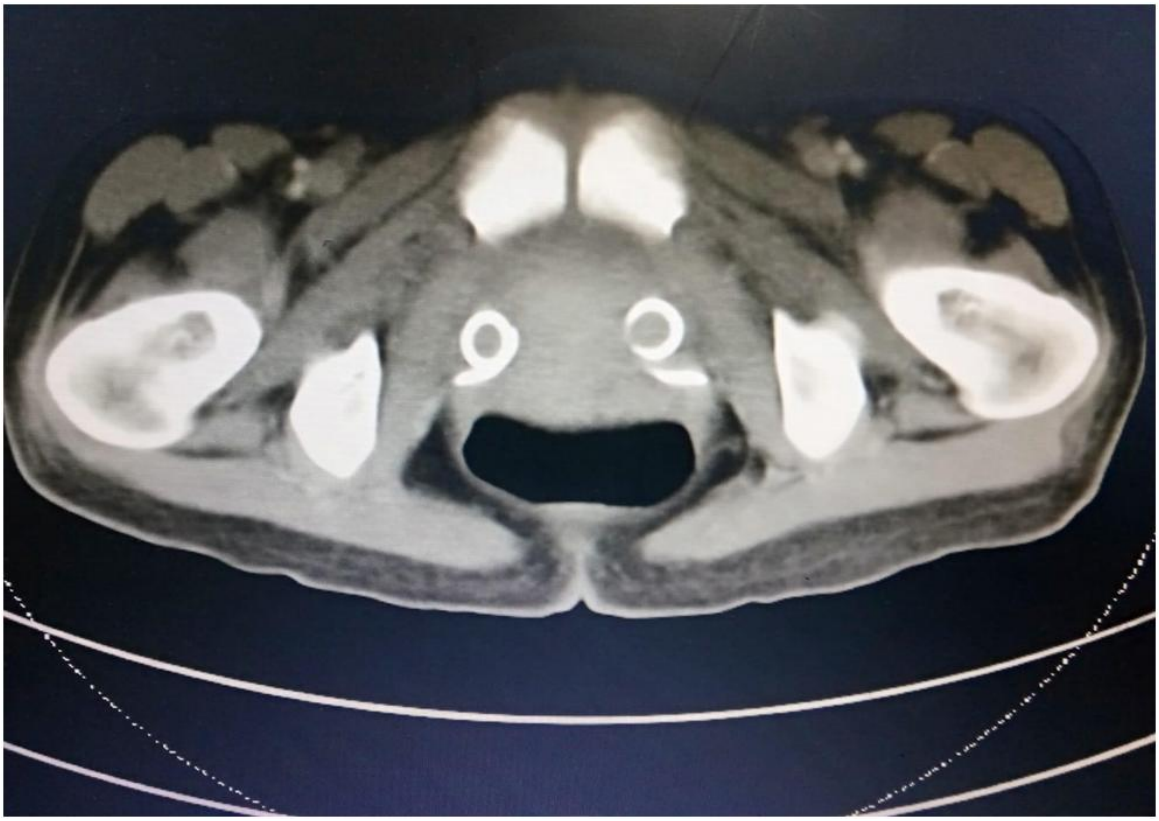
As the tumor was resected completely and hemostasis was achieved, an ABG analysis was done towards end of the surgery showing a PH: 7.34, PCO2: 45, PO2: 153, HCO3: 26, Hb: 10.2 and Lac: 2 at FiO2 40%. By this time, the patient had received a total of 8 pints of IVF Ringer Lactate, 5 units of PRBC, 3 units of FFP, 3 units of platelet concentrate and the total blood loss was around 1600ml with a total of 20 mops including 12 fully soaked mops and 8 partially soaked mops and 2 liters of ascitic fluid and blood in suction bottle. Intraoperatively normothermia was maintained with forced air warmer and fluid warmers.

In view of massive transfusion, and prolonged surgery with respect to age, decision for post-operative mechanical ventilation was taken.
On post op day 1 she was extubated after successful spontaneous breathing trial. Pain was managed with Inj. Buprenorphine 150mcg epidurally 12 hourly and continuous infusion at 3ml/hr. Her Hb on POD1 was 6.8 and 2 PRC given and Hb repeated after6hrs was 8.2, all other coagulation parameters were normal. IV fluids were given at 150ml/hr and if urine output was less than 60ml/hr IVF reduced to 100-120ml/hr, along with diuretics to avoid fluid overload. Antibiotics Inj. Meropenem and Inj. Amikacin were continued. Her body fluid balance picked up gradually. On day 3, oral fluids started and protein rich soft diet started as she tolerated gradually. On day 4 she was shifted to ward and antibiotics were stopped on day5 and epidural catheter also removed. Chest physiotherapy and incentive spirometry were also given during the post-operative period.

She was discharged on post op day 7



CT Image: showing abdomino pelvic mass 13*11*19cms



CT image showing Bilateral DJ stent in view of hydroureteronephrosis

3. Discussion

The management of massive hemorrhage and massive transfusion has been described in adults through studies and experience in combat situations. It has been shown that tourniquet use, damage control resuscitation strategies, balanced transfusion ratios, and anti-fibrinolytic therapy have important implications in decreasing trauma related mortality. Treatment of massive hemorrhage in children is less well defined and many regimens related to its management have been extrapolated from the adult trauma literature and techniques (1)

Evidence-based strategies include awareness of patient or procedural risk factors, system and provider preparation for potential hemorrhage in high-risk situations, and intraoperative goal-directed care with a particular emphasis on prevention of the well-described lethal triad (coagulopathy, acidosis, and hypothermia) associated with massive transfusion. (1)

Pediatric massive transfusion is defined as (i) Packed red blood cell (PRBC) transfusion of 50% of total blood volume (TBV) in 3h, (ii) PRBC transfusion of 100% TBV in 3 h, (iii) PRBC transfusion of >10% of TBV/min (2).

The principles of pediatric massive blood loss are similar to adults. They rely on clinical signs, symptoms, monitoring, and investigations. As with adults, dyspnea, altered mentation, hypotension, and reduced capillary refill can be used to assess hemodynamic state. However, pediatric patients have good physiological reserve, maintaining arterial pressure even after a loss of 25– 40% of blood volume. Consequently, the clinical signs and symptoms of hypovolemia may not be good predictors of early hemorrhage. A narrow pulse pressure may be a more sensitive sign of hypovolemia than tachycardia or systolic hypotension. Lactic acidosis secondary to hypoperfusion and decreased urine output are additional indicators of hypovolemia. (2) Assessment of blood loss in theatre can be difficult (2).

Clinical signs are affected by anesthesia and blood loss calculated by direct observation, collection in suction bottles, and weighing of swabs are inaccurate and impractical, often underestimating volume lost. (2) Despite difficulty in using clinical signs and symptoms to determine pediatric blood loss, transfusion requirements using volumes of crystalloid and blood products given to maintain vital parameters (including heart rate, arterial pressure, central venous pressure) can be used to help determine the degree of blood loss (2). Blood loss being approximated as one-third of crystalloid or an equivalent volume of colloid replacement required to maintain hemodynamic stability (2).

In uncontrolled hemorrhage, damage control or hemostatic resuscitation, aims to minimize iatrogenic resuscitation injury, prevent worsening of shock and coagulopathy, and obtain definitive hemostasis, usually via surgical control. It differs from the traditional management of hemorrhagic shock by limiting crystalloid fluid resuscitation and by transfusing blood products empirically before coagulopathy is identified by testing. In the pediatric trauma setting, increased crystalloid volume replacement has been associated with increased transfusion requirements, coagulopathy (prolonged pro thrombin times), and a tendency towards increased mortality and multi-organ failure rates (2).

Permissive hypotension aiming at maintaining sub arterial blood pressure during intraoperative period is helpful to reduce blood loss, but children having a large physiological reserve compensates for blood loss with minimal clinical signs, hence it is not appropriate to control blood loss. The most common complication of transfusion, regardless of age, is metabolic disturbances. Among these, post-transfusion hypocalcemia due to chelation of calcium by the citrate preservatives in PRBCs. Important in children, but neonates are more susceptible because decreased ability to metabolize citrate.

Hypocalcemia countermeasures for this group include decreasing the rate of transfusion or providing supplemental IV calcium (3). In addition to electrolyte derangement and coagulopathy, immunologic reactions can arise resulting in ABO incompatibility, transfusion-related acute lung injury (TRALI), transfusion-associated circulatory overload (TACO), and alloimmunization which can all worsen the clinical condition (3)

Massive transfusion protocol in the ratio 2:1:1 (PRBC: FFP: Platelets) has been initiated. Of note, research into lower PRBC:FFP ratios during massive transfusion in children is yet to indicate significant improvements in morbidity and mortality. For this reason, and possibly due to difficulty in providing FFP quickly, most guidelines advocate replacement in a PRBC: FFP: platelet ratio of 2:1:1(3) Lung protective strategies like reduced tidal volume, increased respiratory rate and PEEP has been taken in this case. To avoid fluid overload judicious fluid administration based on Goal Directed Fluid Therapy on the basis of Pulse Pressure Variation, Stroke Volume Variation and cardiac output monitoring. Diuretics were administered towards the end of surgery. Post operatively, pneumatic compression devices have been given in view of increased risk of DVT, along with chest physiotherapy and incentive spirometry for early recovery and to reduce post-operative pulmonary complications.

4. Conclusion

Massive hemorrhage in pediatric patients is a stressful and hazardous situation for anesthetists as well surgeons. Management requires assessment of initial and ongoing blood loss with frequent evaluation of response to therapy. It is important to anticipate major blood loss and to ensure the availability of blood and blood products. Early resuscitation with adequate volume can prevent the complications of coagulopathy.

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