PEDIATRIC MASTOCYTOSIS AND ANAESTHESIA: A THREE-CASE REPORT

Alvar, Elena; Denis, Santiago; Pérez, Raquel; Reinoso, Francisco; Gilsanz, F.
Hospital Universitario La Paz, Dept. of Anaesthesia, Madrid, Spain.

BACKGROUND: MASTOCYTOSIS

Prevalence 1:25000

Pathologic increase of mast cells both in cutaneous and extracutaneous tissues:

G-I tract, spleen, liver, bone marrow, lymphoid tissues.

Different clinical manifestations depending on:

- age of onset (pediatric vs adult)
- disease variant (systemic vs cutaneous)
- severity
- associated hematologic disorders

Typical symptoms: gastroesophageal reflux, flushing, urticaria and hypotension.

CONSEQUENCE

All patients are at risk for anaphylaxis during anesthesia, as opioids, muscle relaxants, analgesics and volatile anesthetics may directly or indirectly degranulate mast cells.

There is little information about the anesthetic management of pediatric mastocytosis.

MANAGEMENT

SPANISH MASTOCYTOSIS NETWORK PROTOCOL:

AVOID:

Stress, anxiety
NSAIDs
Opioids
Muscle relaxants
Coloids
Some Hypnotics

IMPOSSIBLE DURING ANAESTHESIA!

TOLERATED AGENTS:

Premedication: corticoids, dexchlorpheniramine, ranitidine, benzodiazepines.

Hypnotics: etomidate, propofol, ketamine, sevoflurane, desflurane.

Muscle relaxants: vecuronium (ideal), rocuronium.

Opioids: fentanyl and remifentanyl ONLY if proven tolerance and premedication.

Amide local anaesthetics: regional anaesthesia.

DEEP ANAESTHESIA MAINTENANCE!

Serum tryptase: 1h before, during and 12 hours after surgery.

CASE REPORT

Three cases of children with mastocytosis we attended to in less than a month

PATIENT 1

Urgent surgery (malar fracture)
5 yo, 21 Kg, no allergies.
Cutaneous mastocytosis (notified at the door of the OR) and asthma.
IV antibiotic already administered.
Premedication (iv):
•ranitidine 30 mg (1.5 mg/Kg)
•dexchlorpheniramine 1mg (0.05 mg/Kg)
•methylprednisolone 28 mg (1.4 mg/kg)
•midazolam 1 mg (0.05 mg/Kg).
Inhalatory induction: sevoflurane
Maintenance:
•sevoflurane (1.2 CAM)
•iv ketamine 20 mg (1mg/kg)
•fentanyl (total dose of 3 mcg/Kg)
•rocuronium 10 mg (0.5 mg/Kg)
OTI: easy, cuffed standard 5.5 tube
Mechanical ventilation (PC).

NO COMPLICATIONS

PATIENT 2

Elective plastic surgery (otoplasty).
14 yo, 48 Kg. No allergies. Febrile seizures, controlled cutaneous mastocytosis with normal serum tryptase, tolerated NSAIDs.
Premedication (iv, 1 hour before surgery):
•ranitidine 50 mg (1 mg/Kg)
•dexchlorpheniramine 5 mg (0.1 mg/Kg)
•methylprednisolone 50 mg (1mg/kg)
•Midazolam 10 mg orally (30 min before surgery)
Tryptase levels: 1h before, during and 12 h after surgery.
Inhalatory induction: sevoflurane
Auricular block: 4 ml mepiv. 2% + bupiv. 0.25%
Maintenance:
•sevoflurane (1.5 CAM).
•fentanyl (total dose 100 mcg, 2 mcg/Kg)
•remifentanyl perfusion at 0.1 mcg/Kg/min.
LMA n.3. Mechanical ventilation (IPPV).
IV antibiotic prophylaxis (amoxicillin-clav. 2g).
PO analgesia: paracetamol 500 mg (10 mg/Kg).

NO COMPLICATIONS

PATIENT 3

Urgent finger reimplantation surgery (time: 4 h)
9 yo, 30 Kg. Solitary mastocytoma on the elbow (notified at the door of the OR), no histamine release reactions, had never taken analgesics. Ischemia through gravity. Total ischemia: 1h45'
Premedication (iv):
•midazolam 1 mg (0.03 mg/Kg)
•ranitidine 45 mg (1.5 mg/Kg)
•dexchlorpheniramine 3 mg (0.1 mg/Kg)
•methylprednisolone 28 mg (1 mg/Kg).
IV antibiotic prophylaxis (amoxicillin-clav 1.5 g)
IV induction:
•propofol 180 mg (6 mg/Kg)
•fentanyl 90 mcg (3 mcg/Kg)
•rocuronium 20 mg (0.6 mg/Kg)
OTI: easy, cuffed standard 5.5 tube.
Mechanical ventilation (PC).
Maintenance:
•desflurane (0.8 CAM)
•remifentanyl perfusion at 0.1 mcg/Kg/min
•fentanyl (total dose 155 mcg, 5 mcg/Kg).
Intraoperative heparin 900 IU (instead of aspirin).

NO COMPLICATIONS

DISCUSSION

This protocol is designed for elective surgery, as it suggests to administer corticoids and antihistamines an hour before surgery, and benzodiazepines half an hour before surgery. This is not always possible in case of urgent surgery, especially when the disease is notified at the door of the operating room. We had two urgent cases where the premedication had to be administered just before anesthetic induction, but fortunately we did not have any complication. %

LEARNING POINTS

Although mastocytosis is a rare disease with a low incidence, we have attended three cases of its pediatric forms in less than a month. It is important for the anesthesiologist to know the management of this disease. Premedication with corticoids, antihistamines and benzodiazepines is key to prevent a histaminic release that can lead to an anaphylactic reaction in the operating room.

REFERENCES

Pediatric Mastocytosis: Routine Anesthetic Management for a Complex Disease. Anesth Analg 2008;107:422–7. %
Protocolos de tratamiento en las mastocitosis. REMA. <http://www.mastocitosis.org/protocolos-de-tratamiento.html>