The Use of Dexmedetomidine for a Pediatric Patient with Mastocytosis

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Introduction

Mastocytosis is a rare disorder characterized by excessive mast cell proliferation, with most cases presenting during infancy or early childhood and usually resolving by puberty.

Clinical presentations include headache, cognitive disorganization, flushing, rash, nausea/vomiting, bronchospasm, cardiovascular collapse, and death. Signs and symptoms are believed to be triggered by the release of mast cell-derived mediators such as histamine, prostaglandins, heparin, neutral proteases, and acid hydrolases. Although most cases are limited to cutaneous manifestations only, life-threatening presentations have been reported during general anesthesia, citing many commonly used anesthetic agents as triggers.

We present our experience with the use of dexmedetomidine in the anesthetic management of a 3-year-old patient with confirmed mastocytosis.

Case Presentation

A 3-year-old, 16.4-kg child presented for right orchiopexy and bilateral myringotomy tube placement. Past surgical history consisted of right inguinal hernia repair at age 6 months, with development of dark brown lesions on the skin. Biopsies revealed the diagnosis of mastocytosis (urticaria pigmentosa), thought to be related to exposure to morphine. Per a history obtained from the parents, the patient had no further episodes.

Anesthesia for this case consisted of premedication with oral midazolam (10 mg), mask induction with a mixture of oxygen and nitrous oxide, IV placement, insertion of a laryngeal mask airway for airway control, and maintenance with dexmedetomidine (0.5 mcg/kg/h) and propofol (200 mcg/kg/min) infusions. A caudal block was performed with 0.2% ropivacaine (20 mg). Perioperative antibiotics were not required for this case and were not administered. The patient also received intravenous hydrocortisone (30 mg) to reduce inflammation associated with mast cell release. For additional analgesic control, the patient was given intravenous acetaminophen (220 mg) and fentanyl (20 mcg).

The case proceeded uneventfully and the patient was transferred to the postanesthesia care unit (PACU) where his caudal block receded. He was discharged home after 4 h of monitoring in the PACU.

Discussion

Management of patients with mastocytosis may prove challenging for the anesthesiologist as many routinely used anesthetic agents may be directly or indirectly associated with mast cell degranulation. Some of these described agents include lidocaine, morphine, oxymorphone, codeine, d-tubocurarine, etomidate, thiopental, succinylcholine, sevoflurane, and isoflurane. Our literature search provided no existing documentation on the use of dexmedetomidine in such cases.

We became interested in using dexmedetomidine for this specific patient because this drug is widely accepted to have a safe cardiorespiratory profile and has been demonstrated to reduce intraoperative anesthetic requirements and decrease postoperative analgesic needs. Our literature review demonstrated no documented use of dexmedetomidine, a selective alpha-2 adrenergic agonist, in patients with mastocytosis. It appears that dexmedetomidine has little to no interaction/role in mast cell degranulation. We believe dexmedetomidine can be used safely in patients with this disease process.

References