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Functional Endoscopic Sinus Surgery for Rhino-orbital mucormycosis in a pediatric patient with Acute Lymphocytic Leukemia- Anaesthetic considerations

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Abstract

Disseminated rhinocerebral mucormycosis is a lethal form of invasive fungal infection. Predisposing factors include immuno suppressed patients, diabetic ketoacidosis. Effective management consists of high index of suspicion, cytological diagnosis, optimization of comorbidities, systemic antifungal therapy with prompt surgical debridement of infected tissue. Our patient was a known case of acute lymphocytic leukemia (ALL) of pediatric age undergoing chemotherapy and presented with high grade fever and epistaxis. Following a confirmatory cytology from nasal cavity his general condition was optimized and surgical (Functional endoscopic sinus surgery (FESS) debridement done. Here we present the anaesthetic challenges in a patient of ALL affected with mucormycosis with multiple comorbidities highlighting airway management.

Keywords: Rhino cerebral mucormycosis, Acute lymphocytic leukemia, Diabetic ketoacidosis, Amphotericin B, Functional endoscopic sinus surgery, Difficult airway

Introduction

Disseminated rhino-orobital-cerebral mucormycosis is a lethal form of invasive fungal infection, which represents 8.3-13% of all fungal infections encountered during autopsies of hematological patients [1]. Predisposing factors include hematological malignancy, immunosuppressed children and diabetic ketoacidosis (DKA). Mucormycosis triad includes uncontrolled diabetes mellitus, peri-orbital infection and meningoenchephalitis [2]. Effective management consists of cytological diagnosis, optimization of predisposing conditions, systemic antifungal therapy with prompt, surgical debridement of infected tissue [3]. We highlight the anesthetic challenges in a pediatric patient of acute lymhocytic leukemia (ALL) with multiple comorbidities undergoing functional endoscopic sinus surgery for mucormycosis.

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Case Report

A male child of 8 year, and 43 kg weight, on Vincristine, Daunorubicin, Pegasparaginase and Methylprednisolone based chemotherapy for B cell ALL, presented with high grade fever, chills, breathlessness and epistaxis. Complete blood count revealed pancytopenia. Supportive therapy (including intravenous Paracetamol infusion to control fever and chills) along with broad spectrum antibiotics were started, but his general condition deteriorated with appearance of reddish blue discoloration over nasal bridge and eyelids. Black eschar appeared in the nasal cavity. Eschar cytology revealed mucor growth while blood cultures revealed pseudomonal growth. Intravenous lyophilized Amphotericin B (AmB) and anti-pseudomonal antibiotics were started. To prevent dissemination, emergency Functional endoscopic sinus surgery (FESS) guided debridement under General anaesthesia (GA) was planned. GA was induced with propofol, fentanyl and succinyl choline, and airway was secured with size 6.5 cuffed endotracheal tube. Anaesthesia was maintained with 50% oxygen in air sevoflurane, and atracurium using neuromuscular monitoring. Apart from routine monitoring, central venous and arterial pressures were monitored. Intraoperative proceedings were uneventful. Neuromuscular blockade was reversed however, consciousness level remained suboptimal. Arterial blood gas(ABG) revealed metabolic acidosis (pH- 7.067, HCO3-10.8, PaCO2 28.4, PaO2-178) and high blood sugar(456 mg/dl) with positive urine ketones indicating DKA which was aggressively optimized with Insulin, soda bicarbonate and adequate fluids. Serum electrolyte correction was done as per serial arterial blood gas and laboratory reports. Acidosis correction improved the consciousness level and child was extubated in operation theater and was shifted to post anaesthesia care unit with stable hemodynamic and respiratory parameters.

Next day, the general condition deteriorated with severe thrombocytopenia, oliguria and raised serum creatinine. Auscultation revealed bilateral rhonchi and crepitation with falling oxygen saturation (88% on

oxygen with face mask). Patient's hemodynamics were maintained on moderate dose of nor-epinephrine. Nasal eschar reappeared, so surgical debridement (FESS) was re-planned, for which random donor platelets were transfused and hyponatremia was also corrected. GA was induced with titrated doses of ketamine, etomidate, fentanyl and succinylcholine. C mac D blade was used for intubation which revealed black fungal debris at laryngeal inlet and over posterior pharyngeal wall which was quickly cleaned using tissue holding forceps. Glottic edema was noted and tracheal intubation was done with pre softened (in sterile hot water) size 6 cuffed tracheal tube with gentle maneuvering. Nasogastric tube was placed and pharyngeal pack was inserted gently. 15^0 head up tilt was given to minimize blood loss. Anaesthesia was maintained with air, oxygen and isoflurane. Tissues were friable and bled even on gentle manipulation. Nor adrenaline infusion was continued throughout intraoperative period. There was extensive black necrotic eschar extending upto adenoids with invasion of nasal and pharyngeal soft tissue and cartilage, which was debrided, which was associated with persistent ooze. Nasal packing was done after application of topical AmB ointment

Anticipating worsening of airway edema and to prevent aspiration of trickled blood and to facilitate further debridement sessions, ETT was retained postoperatively on minimal respiratory support and mild sedation. The patient showed improvement and norepinephrine was tapered. Urine output also stabilized. However, airway edema persisted over next 12 hrs Debridements were done two more times. Subsequently, intravenous steroids were administered to reduce edema. Patient was on ventilatory support all along, maintaining vital parameters with minimal inotropic support. The patient gradually improved over next 10 days.

Discussion

Mucormycosis was first described by Paultauf in 1885 [4]. The first case of mucormycosis from India was reported in 1963. Mortality rate may be as high as 60% in patients of rhino-cerebral mucormycosis. 1 Mucormycosis is aggressive, potentially fatal and complex disease. Mucorales are saprophytes found in soil and decaying matter. Inhaling conidia or spores may cause infection. Histo-pathologically, hyphae are broad ribbon like, angioneurally invasive, causing surrounding tissue infarction and in 90% of biopsies perineural invasion has been found explaining the propensity to cause rhino-orbital cerebral disease [5]. The fungal invasion of nasal cavity or paranasal sinuses of susceptible host causes consistent symptoms of sinusitis, periorbital cellulitis and facial numbness, followed by conjunctival congestion, blurry vision, soft tissue swelling, eschar formation and necrosis of naso-facial region, as happened in our patient (fig 1). Advancing infection spreads from ethmoid sinuses to orbit causing proptosis, chemosis and can quickly result in cavernous sinus thrombosis, carotid or jugular vein thrombosis (Lemeirre syndrome) and death [4]. Other presentations include pulmonary, gastrointestinal and cutaneous.

AmB is the cornerstone of antifungal therapy in mucormycosis. Knowledge about AmB is vital for anaesthesiologist. AmB infusion is generally given over 4-6 hrs to prevent complications, most dreaded being nephrotoxicity [6], which affected our patient. There is decreased GFR and tubular dysfunction by ischemic injury resulting in ARF (40-65%) [6]. Rapid infusion of AmB may cause severe hyperkalemia and potentially fatal arrhythmias. Continuous infusion of AmB has shown to reduce nephrotoxicity [6]. Studies have indicated that the 0•2 μm polyethersulphone (PES) filter is optimal for intravenous infusion of amphotericin B fluid to minimize the introduction of particulate matter, microbial contaminants and endotoxin into patients [7]. Sodium loading reduces incidence of nephrotoxicity, making normal saline fluid of choice in our patient [8]. Renal insult in our patient was controlled with repeated debridement, adequate fluid management and continuous infusion of AmB [3]. Serum potassium levels in our patient was monitored closely in view of anticipated renal insult, AmB administration and insulin infusion to control blood sugars. Other side effects of AmB include hypomagnesemia, fever, chills, dyspnea, hypotension, thrombocytopenia and allergic reactions. Lethal pulmonary reactions associated with AmB are known to occur [9]. Hyperbaric oxygen therapy (HBOT) has been used as an adjunct to surgical debridement. HBO exerts a fungistatic effect and aids revascularization, with subsequent healing in poorly perfused acidic and hypoxic but viable areas of tissue [9].

Kulkarni et al reported a case of an elderly diabeteic hypertensive male having mucormycosis with palatal perforation, who underwent debridement under GA [3]. Our patient developed airway edema before second debridement and selecting a smaller sized ETT for tracheal intubation proved prudent, while taking utmost care to prevent transfer of mucor colonies to lower respiratory tract by removing them before passing ETT. Here Cmac video-laryngoscope was visualization of airway, and ease of insertion of ETT. Eckman et al described a case of rhino-cerebral mucormycosis where they did not suspect epiglottitis and preoperative airway assessment was suboptimal due to trismus. There was involvement of nasal and nasopharyngeal tissues and severe epiglottic edema for which the authors had to resort to emergency tracheostomy [10]. We anticipated glottic edema before second debridement and assessed the patient for the same. We preferred intravenous induction and succinylcholine since there was no stridor or signs of airway obstruction. Otorhinolaryngologist was also informed about possibility of emergency tracheostomy. Isoflurane was preferred the second time as inhalational agent due to its fungistatic effect noted in vitro [3].

Clinical message

Opportunistic virulent airway infections commonly occur in immunocompromised patients especially with diabetic ketoacidosis. Anaesthesiologists may come across such patients as the population of immunocompromised patients grow and incidence of mucormyocosis is on the rise. We suggest the anaesthesiologist should be cautious regarding the airway as emergencies may occur and not be in hurry to extubate such patients. Also, high index of suspicion and attention and regular monitoring of basic investigation like blood sugar levels should be done diligently. Healthcare-associated outbreaks have been linked to bandages, tongue depressors and hospital linens. Anesthesia personnel are at risk of contracting the disease. We advocate proper use of masks, proper disposal or sterilization of infected instruments, linen [11].

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