Integrative Anesthesiology

CASE REPORT

Anesthetic Management in Stevens - Johnson syndrome: Case Report

William K Kwass and Jarva Chow*

Department of Anesthesiology and Pain Management, Loyola University Medical Center, Maywood, IL, USA

Abstract

Stephens-Johnson syndrome (SJS) is a rare, acute, disorder of the skin and mucous membranes with systemic manifestations that when severe, pose unique challenges specific to anesthetic care. There are few case reports but no current recommendations specific for the care of patients with Stephens-Johnson syndrome requiring general anesthesia. We present a case involving a 47-year-old female with severe SJS involving the entire body including head and neck presenting for an urgent ophthalmologic procedure to prevent further vision loss. The intraoperative management including monitoring, airway management, medication considerations, intraoperative ventilatory strategies, and perioperative care are discussed.

Keywords: Anesthesia, Steven's-Johnson Syndrome, Anesthetic Management, Drug reaction, Difficult airway

Introduction

Stephens-Johnson syndrome is an acute eruptive disorder of the skin and mucous membranes with systemic manifestations of variable severity. This disorder presents unique anesthetic challenges. Upon literature review there is little information to guide anesthetic care. We present a case of SJS requiring urgent general anesthesia, a brief review of the literature, and the anesthetic considerations for the patient with severe SJS. The patient has provided written consent/HIPAA authorization to publish this case report.

Case History

Our patient was a 47-year-old Hispanic female who initially presented with concern for Stevens-Johnson syndrome. Her past medical history included hypothyroidism and type 2 diabetes mellitus, for which she was taking levothyroxine and metformin and she had no known medical allergies. Four days prior to admission she had unilateral conjunctival erythema which progressed rapidly to involve her eyes bilaterally, oral mucosa, neck, back, feet, trunk, and vagina by the time she was admitted. In addition, she was experiencing fevers, sore throat, dysphagia, fatigue, and dysuria, but she denied pruritis or dyspnea. She denied any new or adjusted prescriptions/over-the-counter/herbal medications and recent prodromal illnesses or sick contacts to suggest an etiology. The Ophthalmology team recommended urgent surgery for amniotic membrane grafts to bilateral eyes in order to prevent further vision loss.

The preoperative exam was significant for poor mouth opening < 4 cm secondary to pain, mucosal sloughing and inflammation in the oropharynx with some areas of active bleeding. The thyromental distance was < 5 cm, full range of motion in the neck, and a Mallampati IV airway. Her vital signs included heart rate 110 bpm, blood pressure 141/76 mmHg, oxygen saturation 94% on room air. Day of surgery labs included hemoglobin of 12.2 g/dL and potassium 2.8 mEq/L. The anesthetic plan was to preserve spontaneous respiration and to perform an awake fiberoptic intubation followed by induction of general anesthesia after securing the airway.

Intra-Operative Events

The patient was taken back to the operating room, connected to the anesthesia monitors and she was pre-medicated with midazolam and 0.2 mg of glycopyrrolate. She was not positioned on the ophthalmology operating table due to difficulty positioning with an anticipated difficult airway. A dexmedetomidine infusion was started at 1.5 mcg/kg/hr and incremental doses of ketamine, 0.2 to 0.5 mg/kg, were given while maintaining a heart rate < 120 bpm. Additionally, topicalization was achieved with 10cc of 2% lidocaine atomized, 4% viscous lidocaine on a tongue depressor, and 2% lidocaine attached to the fiberoptic port. Additional midazolam and ketamine were given for agitation when inserting the fiberoptic scope. Although the patient began actively coughing blood prior to insertion of the fiberoptic scope, a grade 1 vocal cord view was obtained, however this quickly became obscured with blood. A second attempt was made with video laryngoscopy while maintaining spontaneous respiration and again a grade 1 vocal cord view was transiently obtained, but was obscured by profuse bleeding in the oropharynx. Blow-by oxygen was administered while the oropharynx was aggressively suctioned with a soft tipped device. The patient

Correspondence to: Dr. Jarva Chow, 5841 S. Maryland Ave, MC4028, Chicago, IL 60637, USA, Tel: 773-702-6700, E-Mail: jchow[AT]dacc.uchicago[DOT]edu

Received: Mar 11, 2019; **Accepted:** Mar 13, 2019; **Published:** Mar 15, 2019

was eventually intubated with direct laryngoscopy using a Macintosh 3 blade and an oral, cuffed 6.5 mm endotracheal tube. Once endotracheal tube placement was confirmed 80mg of propofol and 30mg of rocuronium were given to terminate spontaneous respiration. General anesthesia was maintained throughout the case with sevoflurane and a dexmedetomidine infusion. The remainder of the intraoperative course was uneventful.

Post-Operative Course

Post-operatively the patient remained intubated for airway protection and transferred back to the Burn ICU. She was extubated on postoperative day 2 and ultimately discharged postoperative day 9. Upon further review, the patient had started taking Allopurinol several weeks prior to her initial symptoms.

Discussion

Erythema multiforme, Stevens-Johnson syndrome and Toxic Epidermal Necrolysis are blistering skin diseases considered to be a reaction to exogenous agents [1]. They are thought to be a spectrum of the same disease.

Stephens-Johnson syndrome is an uncommon acute eruptive disorder of the skin and mucous membranes with systemic manifestations of variable severity [2]. Although the true incidence of SJS is unknown, any age group and patient may be affected. This disorder concurrently with the need for general anesthesia presents several anesthetic issues. Obstacles to the provision of safe anesthetic care are many. Blood pressure cuff application may be painful and cause further epithelial damage. For this reason invasive blood pressure monitoring is often necessary [1]. Electrocardiography monitoring for arrhythmias commonly seen in SJS may be difficult to obtain due to pad adhesion and location. Body temperature is particularly important to monitor in these patients because dermal blood flow is often altered and the use of anesthesia impairs autoregulatory control often leading to hypothermia [3]. Typically, large bore peripheral IV access and invasive blood pressure monitoring are the only additional monitors necessary in patients without a significant history of cardiac dysfunction. If possible, this access is usually established over areas of normal epidermis. Central venous catheters are only placed if absolutely necessary to minimize the risk of infection through the denuded dermis.

This syndrome can be triggered by several factors. Drugs (Table 1), bacterial/viral infections, foods and vaccinations have been implicated. Currently there is no specific therapy for treatment, however, steroids and antihistamines have been advocated and are commonly used [2]. Antibiotic therapy is also frequently used given the increased susceptibility to infection [2]. Prompt withdrawal of the offending agent is often the most effective therapy along with early recognition and aggressive supportive treatment. [3] Major factors that contribute to a favorable outcome are hemodynamic stability, prevention of infection, and good nutritional status [3]. These patients are typically treated as severe second degree burn

Common Triggering Agents	
Anti-retrovirals	
	Nevirapine
	Abacavir
	Efavirenz
Anti-tuberculosis	
	Isoniazid
	Rifampin
	Pyrazinamide
Antibiotics	
	Trimethoprim-sulfamethoxazole
	Ciprofloxacin
	Levofloxacin
	Penicillins
	Cephalosporins
Anti-convulsants	
	Phenytoin
	Carbamazepine
	Valproic Acid
	Phenobarbital
	Lamotrigine
	Diazepam
Miscellaneous	
	Allopurinol
	Metronidazole
	Methotrexate
	Furosemide
	Sulfa drugs
NSAIDs	
	Oxicam
Unknown	

Table 1: Common Triggering Agents of SJS.

patients with special attention given to mucous membrane involvement for adequate fluid balance, systemic involvement leading to hemodynamic instability and dissemination of cutaneous lesions that enhance the risk of infection and sepsis.

The mucous membranes of the entire body may be involved, particularly in the respiratory tract and pleura.[4] Ulcers in the nose, oropharynx, trachea, bronchi are also observed increasing the risk of hematemesis. Ocular involvement ranges from mild conjunctivitis to pan-ophthalmitis with corneal destruction. Because there are often lesions of the oral, tracheal and laryngeal mucosa manipulation and/or trauma to these areas should be avoided if possible. Application of anesthetic mask during pre-oxygenation can abrade facial and lip epithelium. The use of soft and/or hard oral airways may also cause bleeding and push debris and blood back into the posterior airway. Endotracheal intubation is dangerous in these patients as trauma to the tracheal and laryngeal mucosa by the endotracheal tube may cause additional abrasions and bleeding that increase the risk of airway compromise after extubation.

Current Data and Recommendations for Management

There are few case reports but no current recommendations for the anesthetic care of patients with SJS [Appendix A] [3, 4]. The anesthetic care of our patient with this syndrome

who needed urgent surgery under general anesthesia has been described.

The most important factors in the anesthetic management of these patients involve aggressive fluid and electrolyte replacement, avoidance of hypothermia during procedures and adequate postoperative analgesia [3]. Regional and local anesthesia may be considered depending on the patient's skin condition overlying the point of entry and the presence of systemic infection.

General anesthesia and instrumentation of the airway presents serious risks to the patient. However, these risks must be balanced with the advantages of securing the airway electively. Intravenous agents frequently cause respiratory depression, loss of airway control, and cardiovascular depression. The literature has described successful use of ketamine, in one case as the sole anesthetic in a 14-month old infant to maintain spontaneous ventilation avoiding airway instrumentation [2]. While use of ketamine minimizes the risk of respiratory depression, limitations include sympathetic stimulation that may increase myocardial oxygen consumption and increased salivation, further complicating endotracheal intubation [4].

If involvement of the airway mucosa is suspected, elective endotracheal intubation should be performed in the ICU [3]. Swelling of the airway is common as aggressive fluid resuscitation is performed on admission leading to further potential airway compromise. General anesthesia with endotracheal intubation is typically the modality of choice. Care should be taken to avoid desquamation of the skin on the face during mask pre-oxygenation. Induction with etomidate or ketamine could provide better cardiovascular stability when compared to propofol or thiopental. Non-depolarizing muscle relaxants are often used if muscle relaxation is necessary to avoid succinylcholine and the associated risk of hyperkalemia that has been observed in patients with SJS. Body temperature is important to monitor in these patients because dermal blood flow is often altered and anesthesia impairs autoregulatory control leading to hypothermia. For this reason the operating room temperature should reach 28'C, warm intravenous fluids and water mattresses/forced air-warmers should be used [3]. These can sometimes only be temporizing measures and the surgical time may have to be shorted or even staged. The decision to extubate the trachea at the end of the surgery should be based on the degree ofairway involvement and intraoperative developments. The decision to proceed with surgery and anesthesia must be weighed with the substantial risks inherent in the patient with Stephens-Johnson syndrome. In our particular situation the surgical team felt that the risk of further eye injury was far too great to postpone surgery.

Appendix A: Similar Cases from Literature Review

Case 1

39-year-old female with peeling of skin around eyes, neck, anterior torso and upper and lower extremities (> 48% TBSA). She was sent to OR for skin debridement; induced with Ketamine and rocuronium and intubated orally without difficulty. General anesthesia was maintained with sevoflurane and fentanyl. She returned to the OR two more times for subsequent debridements and was anesthetized with sevoflurane and fentanyl with no issues.

Case 2

25-year-old male admitted for generalized malaise, stomatitis with sloughing of the oral mucosa, odynophagia, dysphagia and temperature of 40'C. He was admitted to the medical intensive care unit and after a few hours was sent to the OR for debridement of wounds covering 55% of the BSA. General anesthesia was induced with propofol, midazolam, and fentanyl. The patient was paralyzed with atracurium and intubated orally without difficulty. General anesthesia was maintained with sevoflurane and fentanyl. He returned to the OR 2 days later still intubated for subsequent wound debridement and was induced with fentanyl, isoflurane and paralyzed with rocuronium. Anesthesia was again maintained with sevoflurane and fentanyl. He was eventually extubated 48hrs later but required re-intubation 72hrs after surgery due to decreased Pa02, tachypnea, and fatigue due to excessive tracheobronchial secretions. His sputum cultures grew *Pseudomona aeruginosa* and he was treated with ceftazidine and tobramycin and after improvement in his clinical condition was extubated 4 days later. He was later discharged home 16 days after his initial admission.

Case 3

31-year-old male admitted for rash over the upper half of his body associated with itching and facial flushing. His physical exam was unremarkable except for skin rash, ulcers in the mouth and positive Nikolsky's sign. The next day he complained of severe oral and throat pain and had difficulties in swallowing. He developed severe ulcerations in the mouth and blisters over the chest, shoulders, and back. He went to the OR three times and was induced with propofol/fentanyl or etomidate/fentanyl and for paralysis received rocuronium or atracurium. General anesthesia was maintained with sevoflurane. After each procedure he was kept in the Burn ICU intubated, paralyzed with atracurium and sedated with morphine and midazolam. He was eventually extubated 8 days after first debridement and his condition improved gradually until he was discharged from the hospital 16 days after admission.

Conflicts of Interest/Financial Disclosures

None

Author Contribution

William Kwass: This author helped with the actual case, writing, editing, finalizing and submission

Jarva Chow: This author helped with the actual case, editing and mentorship

References

1. Wallis C, Mcclymont W (1995) Case Reports Toxic Epidermal Necrolysis with Adult Respiratory Distress Syndrome. *Anaesthesia* 50: 801-803. [View Article]

2. Cucchiara RF, Dawson B (1971) Anesthesia in Stevens–Johnson Syndrome: Report of a Case. *Anesthesiology* 35: 537-539. [View Article]

3. Rabito SF, Sultana S, Konefal TS, Candido KD (2001) Anesthetic Management of Toxic Epidermal Necrolysis: Report of Three Adult Cases. *J Clin Anesth* 13: 133-137. [View Article]

4. Smith GB, Shribman AJ (1984) Anaesthesia and Severe Skin Disease. *Anaesthesia* 39: 443-455. [View Article]

Citation: Kwass WK, Chow J (2019) Anesthetic Management in Stevens - Johnson syndrome: Case Report. Integr Anesthesiol 2: 001-004.