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**Airway management of a patient with linear immunoglobulin A bullous dermatosis:  
A case report**

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**Abstract**

**BACKGROUND**

There is limited literature on managing the airway of patients with linear immunoglobulin A (IgA) bullous dermatosis, a rare mucocutaneous disorder that leads to the development of friable bullae. Careful clinical decision making is necessary when there is a risk of bleeding into the airway, and a multidisciplinary team approach may lead to decreased patient morbidity during these high-risk scenarios, especially when confronted with an unusual cause for bleeding.

**CASE SUMMARY**

A 45-year-old African American female presented to our ambulatory surgical center for right corneal transplantation due to corneal perforation after blunt trauma in the setting of cicatricial conjunctivitis and diffuse corneal neovascularization from linear IgA bullous dermatosis. The diagnosis of IgA dermatosis was recent, and the patient had been lost to follow-up. The severity of the disease and extent of airway involvement was unknown at the time of the surgery. Significant airway bleeding was noticed upon intubation and the otorhinolaryngology team had to be called to the operating room. The patient required transfer to the intensive care unit where a multidisciplinary team was involved in her case. The patient was extubated on postoperative day 4.

## CONCLUSION

A multidisciplinary approach to treating this disease is the best course of action before a surgical procedure. In our case, key communication between the surgery, anesthesia, and dermatology teams led to the quick and safe treatment of our patient's disease. Ambulatory surgery should not be considered for these cases unless they are in full remission and there is no mucous membrane involvement.

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## INTRODUCTION

Linear immunoglobulin A (IgA) bullous dermatosis (LABD) is an autoimmune mucocutaneous disease involving disruption of the dermoepidermal junction, resulting in the formation of blisters or bullae<sup>[1]</sup>. IgA autoantibodies attach to antigens in the skin's basement membrane and mucosa, causing these clinical findings. Lesions typically appear as clear or hemorrhagic bullae with an urticarial base on the face, trunk, buttocks, and skin overlying joint sites. In 60% to 80% of cases, mucosal membranes, including the eyes and oral cavity, are also affected<sup>[2]</sup>. Patients with mucosal involvement can experience morbidity related to corneal scarring and pharyngeal or esophageal stricture formation, requiring surgical intervention. There is limited literature on the intraoperative management of these patients, especially regarding airway instrumentation.

## CASE PRESENTATION

### *Chief complaints*

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Decreased vision in the right eye.

### *History of present illness*

A 45-year-old African American female presented to our ambulatory surgical center for right corneal transplantation due to corneal perforation in the setting of cicatricial conjunctivitis, as well as diffuse corneal neovascularization from LABD. At the time of

the surgery, the severity of the disease was unknown. The patient had been lost to follow-up with dermatology and was not taking any medications for her LABD. The severity of the patient's history of LABD diagnosis was only discovered after the operating room case and an in-depth conversation with her dermatologist. The patient's LABD was considered idiopathic per her dermatologist. She began experiencing skin rashes and blisters in 2017. In June 2019, she sought medical attention due to mucosal membrane involvement, but at that time no medical therapy was initiated. In January 2021, she developed visual problems with her right eye and, following a confirmatory skin biopsy, was officially diagnosed with LABD in February 2021. Dapsone therapy was initiated in April 2021. She was hospitalized in May 2021 for a corneal ulcer of the right eye and was treated with one dose of intravenous immunoglobulin (IVIG), rituximab infusions, and 25 mg of oral dapsone twice each day. The patient received 4 doses of rituximab and was lost to follow-up until August 2021. This case was emergent due to the risk of permanent visual loss.

### ***History of past illness***

The patient's medical history consisted of anxiety, depression, chronic benzodiazepine dependence, chronic back pain, hypertension, constipation, genital herpes, prior cocaine abuse, and LABD.

### ***Personal and family history***

The patient had a strong family history of autoimmune disease, with paternal cousins having sarcoidosis, systemic lupus erythematosus, and multiple sclerosis. The patient was married and lived a few hours away from our hospital location. She was of a lower socioeconomic background and had trouble affording medications for her treatment.

### ***Physical examination***

The patient had visible nasal ulcers and some white ulcers on her tongue (Figure 1). There was no history of bleeding in her mouth or of previous anesthetic complications. The rest of the physical exam was normal.

#### ***Laboratory examinations***

No laboratories needed for preoperative evaluation before this emergent case.

#### ***Imaging examinations***

See Figure 1 to show the preoperative oral and nasal lesions.

### **MULTIDISCIPLINARY EXPERT CONSULTATION**

On August 10, 2021, the patient presented to the free-standing ambulatory surgical center for a corneal transplant of the right eye under general anesthesia. On the day of surgery, an anesthetic preoperative evaluation and a focused physical examination were performed. The patient had visible clear nasal ulcers and white ulcers on her tongue (Figure 1). There was no history of bleeding in her mouth or of previous anesthetic complications. General anesthesia was indicated, given the type of surgery; therefore, airway manipulation was required. The decision was made to intubate using a C-MAC for optimal airway visualization due to the unknown extent of mucosal involvement of her disease, and complete visualization of the oropharynx was desirable. A peripheral intravenous catheter was placed, and 2 mg of intravenous midazolam was administered before proceeding to the operating room. She received a standard intravenous anesthesia induction with propofol, fentanyl, lidocaine, and rocuronium.

Upon careful placement of the C-MAC Mac 3 blade, ulcerative lesions were observed throughout the oropharynx, epiglottis, and vallecula. There was spontaneous and diffuse bleeding from lesions in the vallecula, even with minimal manipulation, and the airway was quickly secured using a wire-reinforced 7.0 endotracheal tube. A wire-

reinforced tube was chosen so the endotracheal tube could be taped to the chin for surgical exposure. The ear, nose, and throat (ENT) team was called to the operating room to evaluate the bleeding, where they performed a visual examination by C-MAC, suctioned, and placed five epinephrine-soaked (0.1 mg/mL) cottonoids in the oropharynx. After a discussion between the ENT and ophthalmology teams, the decision was made to proceed with the surgery and transfer the patient intubated after surgery to our institution's main hospital surgical intensive care unit under the care of the ENT team. This hospital is located 20 min from our free-standing outpatient facility. Upon successful completion of the surgical procedure, the patient was transferred intubated to the surgical intensive care unit *via* ambulance.

The patient was transferred in stable condition to the main hospital by ambulance under the direct supervision of the attending anesthesiologist. To assess the extent of the ulcerative lesions before trial extubation, a bedside bronchoscopy was performed that revealed no ulcerative lesions in the trachea or bronchi. Ventilator weaning commenced, and extubation was planned in the operating room when clinically indicated. On postoperative day 1, the patient was taken to the operating room where a micro-suspended direct laryngoscopy was performed. Despite minimal manipulation, diffuse mucosal bleeding in the oral cavity and oropharynx was observed. The oral cavity was packed for hemostasis, and the patient remained intubated (Figure 2).

Due to the severity and extent of the disease, the dermatology team re-initiated dapsone therapy and prescribed a course of intravenous methylprednisolone and a single dose of both cyclophosphamide and IVIG. Overnight, the patient was observed to have a few isolated episodes of agitation that resulted in spontaneous bleeding; therefore, the oropharynx was repacked with epinephrine- and tranexamic acid-soaked Kerlix. On postoperative day 3, the ENT team removed the oral packing. On postoperative day 4, the patient returned to the operating room and was successfully extubated to cool, humidified air *via* a face mask. She received a dose of intravenous cyclophosphamide

and was discharged 2 d later with a course of prednisone daily and dapsone with a follow-up for a second intravenous cyclophosphamide infusion planned in 1 mo. A timeline of this case can be seen in Figure 3.

### **FINAL DIAGNOSIS**

Right corneal perforation due to cicatricial conjunctivitis and diffuse corneal neovascularization from LABD.

### **TREATMENT**

Patient was treated with dapsone and a course of prednisone. In the hospital the patient was given a course of intravenous methylprednisolone and a single dose of both cyclophosphamide and IVIG. She subsequently received monthly IV cyclophosphamide home infusions.

### **OUTCOME AND FOLLOW-UP**

The patient continues on monthly cyclophosphamide infusions and daily oral prednisone and dapsone. She is seeing her dermatologist monthly.

### **DISCUSSION**

LABD is a rare disease. In Europe, the incidence is approximately 0.1 per million. It presents in a bimodal age distribution: children aged six months to 10 years old (mean age five years old, rarely persisting past puberty) and adults over 60 years old. LABD is typically triggered by infection but can also be drug-induced by exposure to antibiotics (most notably vancomycin), antihypertensives, and nonsteroidal anti-inflammatory drugs. While some LABD presentations are idiopathic, associations with other systemic diseases have been demonstrated, such as ulcerative colitis, systemic lupus erythematosus, and lymphoproliferative disorders. Up to 5% of reported cases of LABD have been associated with lymphoid malignancies including Hodgkin's or B-cell lymphoma. Idiopathic LABD may persist for decades with episodes of

relapse/remission. In our patient, LABD was diagnosed in February 2021 and deemed idiopathic<sup>[3]</sup>, given the patient had no history of new medications or new infections.

Diagnosis is confirmed by clinical, histopathological, and immunological data. Treatment varies depending on the degree of severity of disease and the potential inciting event. In suspected drug-induced LABD, removal of the offending medication may result in gradual resolution. The mainstay of treatment in LABD is dapsone (50-150 mg daily in adults), which can begin resolution of lesions within 72 h. Per a collection of case reports, median length of dapsone treatment is 26 mo<sup>[4]</sup>. Common side effects of dapsone include hemolytic anemia (G6PD deficiency), methemoglobinemia, motor neuropathy, hepatitis, cholestatic jaundice, and hypoalbuminemia. Routine complete blood count, liver function tests, and G6PD levels should be checked while on dapsone therapy. Immunosuppressants (azathioprine, methotrexate, mycophenolate mofetil, and cyclophosphamide) and corticosteroids are other modalities used for treatment. Chronic use of cyclophosphamide can lead to the onset of malignancies, such as bladder cancer<sup>[5]</sup>. Other treatment options include colchicine, tetracyclines, sulfonamides, nicotinamides, and IVIG.

Dermatology input and medication management were key to improving our patient's outcome. She continued on dapsone and was also given intravenous methylprednisolone, IVIG, and a dose of cyclophosphamide to treat her mucous membrane lesions and allow for extubation. For an anesthesiologist caring for patients with LABD, the most critical information needed is the presence of mucous membrane involvement and airway compromise. In the event of airway bleeding with this disease, ENT intervention to control bleeding with epinephrine and tranexamic acid is necessary to protect the airway as medical treatment begins to take effect. Our ambulatory surgical center and the ENT clinics are all in the same building, helping our team initiate treatment quickly.



For our case, the severity and details of the disease were unknown at the time of the surgery. The surgery was emergent and needed to proceed to try to save vision in her eye. In our health system, corneal transplants are only done at our ambulatory center; necessary supplies and equipment are not present in the hospital. The patient did not report as scheduled to the anesthesia preoperative clinic so the first time the patient was evaluated by the anesthesia team was on the day of her surgery. Given the emergent need to preserve her vision, the necessity of performing the surgery at our ambulatory center, and the absence of any history of airway bleeding, the decision was made to proceed. Going forward, such cases must be completed in the inpatient setting.

### **CONCLUSION**

To our knowledge, this is the first case report of the anesthetic implications of LABD<sup>[4]</sup> involving the mucous membranes of the nasal cavities and oropharynx. Given the lack of information on this disease and its extent in this patient, our anesthesia team was unaware of the lesions we would encounter throughout both the oropharynx and hypopharynx. Upon intubation, we understood the extent of the lesions and their friability. In retrospect, we should have consulted the ENT team preoperatively for a preoperative nasopharyngoscopy to determine the extent of the lesions. Had the patient attended her preoperative clinic appointment, our team may have had more information available.

A multidisciplinary approach to treating this disease is the best course of action before a surgical procedure. Ambulatory surgery should not be considered for these cases unless they are in full remission and there is no mucous membrane involvement. In our case, key communication between the surgical services, anesthesia, and dermatology teams occurred after the surgery and led to quick and safe treatment. We present this case to bring forward an important anesthetic considerations for this rare disease.

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This manuscript was prepared in compliance with the Health Insurance Portability and Accountability Act (HIPAA) of 1996 privacy regulations and adheres to applicable Enhancing the Quality and Transparency of Health Research guidelines (CARES [for CAse REportS] checklist). Written patient consent and written HIPAA authorization for the publication of this case report were obtained using a standardized consent form. Institutional Review Board consent was not required.

### **LEARNING POINTS**

- 1) Mucous lesions in the oropharynx and hypopharynx can be extremely friable and cause bleeding upon minimal manipulation.
- 2) Early dermatology intervention and medication administration is key to a quick recovery from an acute exacerbation.
- 3) Proper planning of operating room intervention in a hospital setting is vital to mitigate potential airway complications. These cases should not be done in an ambulatory setting.

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