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# Airway Management in a Patient with Bullous Pemphigoid

Mohammad Yasir and Fauzia Anis Khan

## ABSTRACT

Airway management in patients with pemphigoid lesions has anaesthetic implications. We report a case of a 23 years old female with bullous pemphigoid who presented with laryngeal stenosis and critical airway narrowing. The airway was initially managed with jet ventilation. Anaesthesia was maintained with propofol infusion and ventilation was performed by introducing a size 10 French gauge suction catheter through the stenotic laryngeal orifice. Thirty minutes into anaesthesia, she developed subcutaneous emphysema and decreased air entry on right side of the chest but remained hemodynamically stable. The airway was further managed by tracheostomy. This case report highlights complications that can occur during the anaesthetic management of such cases.

**Key Words:** Bullous pemphigoid. Jet ventilation. Tracheostomy. Laryngeal stenosis. Critical airway narrowing. Airway management.

## INTRODUCTION

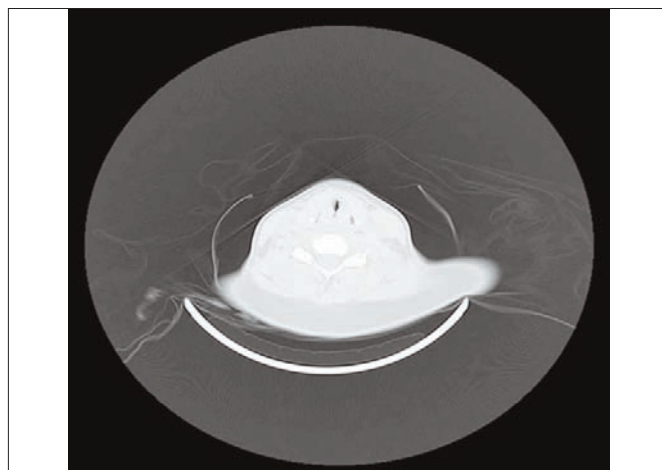
Bullous pemphigoid is a chronic autoimmune blistering disorder of skin and mucous membrane. Females are affected more than males.<sup>1</sup> It is characterized by tense bullae on the skin and mucous membrane, most commonly in the intertriginous areas but may also involve other regions.<sup>2</sup> The diagnosis is made on the basis of clinical presentation, history, and immunopathological studies. Oral mucosa can be involved in one-third of patients.<sup>3</sup> In approximately 50% of patients with oral pemphigoid the disease can progress to extra-oral sites such as the eye, larynx, pharynx or esophagus.<sup>3</sup>

We report a case of bullous pemphigoid that presented with laryngeal stenosis and critical airway narrowing that was initially managed with jet ventilation. Airway management difficulties in such a case are described and discussed.

## CASE REPORT

A 23 years old female patient weighing 53 kilograms presented with 3 years history of progressive bullous pemphigoid. She was on oral steroid 15 mg twice a day. She presented to the dermatology clinic with complaints of multiple blisters over the body and inability to open both eyes. She was referred to ophthalmology clinic where she was found to have bilateral blepharospasm and chemosis. On further enquiry and examination, she had significant cough, dyspnea and hoarseness for the past 5 months. Pulmonary function tests showed severe

obstruction without reversibility (FEV1/FVC = 32% pre-bronchodilation and 33% postbronchodilation). She had no previous history of asthma. Bronchoscopy report mentioned inability to pass the adult fiberoptic bronchoscope beyond the vocal cord and a scarred laryngeal inlet. She was referred to the ENT surgeon who performed an awake fiberoptic examination which revealed laryngeal oedema with excessive supraglottic tissue. The CT scan report revealed laryngeal stenosis at the level of glottis with lumen opening approximately 5 mm and normal vocal cords below the lesion (Figure 1). She was planned for micro-laryngoscopic excision of supraglottic lesion and blepharoplasty. The patient was then assessed by a consultant anaesthetist.



**Figure 1:** CT Scan image taken at the level of 4<sup>th</sup> cervical vertebrae showing slit like airway.

Pre-anaesthetic assessment revealed anaemia (Haemoglobin level of 10.8 mg/dl), skin blisters and restricted functional activity. Chest was normal shaped. A wheeze was audible but this appeared to be due to extra-thoracic obstruction at the level of laryngeal inlet. Chest X-ray was normal. She was not pre-medicated with benzodiazepine but was administered 100 mg intravenous hydrocortisone half hour before anaesthesia.

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Local anaesthesia to the respiratory tract was provided with 4 mls of 4% nebulized lignocaine. Standard monitors were applied with extreme caution in order to prevent skin injury and to avoid further blister formation. Her baseline respiratory rate was 20 breaths per minute with peripheral SpO<sub>2</sub> of 97% on room air. Pre-oxygenation was then started with 100% oxygen using the circle system.

She underwent inhalation induction with sevoflurane, nitrous oxide and oxygen. After checking adequacy of mask ventilation, a bolus dose of 50 mg succinylcholine was administered. Gentle laryngoscopy was performed with Macintosh laryngoscope size three blade. Vocal cords were visualized and a size 10 Nelaton catheter was introduced in the trachea.

Manual jet ventilation was started with clinical monitoring of chest expansion and airway pressure. No wheeze was audible on chest auscultation. Anaesthesia was maintained with propofol infusion 6 mg/kg/hour. Fentanyl (50 microgram bolus) was administered for analgesia. When the patient reversed from the effects of suxamethonium, 25 mg atracurium was administered. Approximately 30 minutes after the start of surgery, subcutaneous emphysema was observed by the anaesthetist on the right side of neck. On chest auscultation, there was decreased air entry on the right side, Nelaton catheter was pulled back but air entry still remained less. Hemodynamically, the patient remained stable. Trauma to lung was suspected and this was communicated to the surgeon. The surgeon decided to perform a tracheostomy and a size 6 tracheostomy tube was inserted. Airway pressure was kept less than 20 cmH<sub>2</sub>O. The rest of surgical procedure was completed uneventfully. At the end of procedure, both supine and lateral decubitus chest X-ray was done which showed subcutaneous emphysema but no pneumothorax. Fiberoptic bronchoscopy revealed no obvious lesion and showed normal area below tracheostomy site.

Patient was shifted to recovery room on tracheostomy mask maintaining normal oxygen saturation where she remained stable throughout. Her subcutaneous emphysema settled and she was discharged 2 days later.

## DISCUSSION

This case report discusses the management of difficult airway with jet ventilation in a patient with bullous pemphigoid. This therapy is generally used as bridging therapy in the management of laryngeal airway obstruction until a definitive therapy is available.<sup>4</sup> In this case, use of jet ventilation was associated with the complication of subcutaneous emphysema.

Pemphigoid disease includes subgroups like bullous pemphigoid, cicatricial pemphigoid, localized scarring pemphigoid and Herpes gestations.<sup>2</sup> Though similar to epidermolysis bullosa, the disease has its own unique

features. Bullous pemphigoid is a skin disorder, and predominantly a disease of fifth decade of life.<sup>2</sup> There is extensive literature on anaesthesia with epidermolysis bullosa but literature describing anaesthetic management of both bullous or cicatricial pemphigoid is rare. Cicatricial pemphigoid is a variant of bullous pemphigoid which primarily affects mucosal surfaces but 25% of patients have cutaneous lesion.<sup>5</sup>

Airway management in patients with bullous pemphigoid lesion is of particular concern. Airway management can be difficult because of pre-existing bullous lesions in the oropharynx, or ulcerations, adhesions or deformities of oropharyngeal mucosa or vocal cords as a consequence of these lesions. Patients can present with stridor or dyspnea due to upper airway obstruction when the laryngeal orifice is narrowed to three or four millimeters and some can present with acute respiratory distress in emergency.<sup>6</sup> This patient had a history of dyspnea and hoarseness but not acute stridor. There was narrowing in the supraglottic area just above the cords and there was inability to pass the fiberoptic bronchoscope beyond the narrowed laryngeal inlet. One option was a pre-operative tracheostomy under local anaesthesia, however, the surgeon was reluctant to take this route initially because of the high incidence of scarring and stenosis at the tracheostomy site.

Airway management itself may also be difficult in these cases. Airway instrumentation can cause acute bullae formation and there is risk of haemorrhage. Recommended precautions are liberal lubrication of the laryngoscope blade, and the tracheal tube as well as gentle laryngoscopy and use of smaller sized tubes.<sup>7</sup> Since this patient had severe supraglottic airway narrowing, we opted for jet ventilation through a size 10 Nelaton catheter for airway management along with total intravenous anaesthesia. Unfortunately, it resulted in subcutaneous emphysema, a known complication of jet ventilation.<sup>8</sup> The probable cause was insufficient outflow due to narrowing at the inlet which lead to excessive airway pressure and surgical emphysema. Because of the complication, the patient ended up with tracheostomy which was initially avoided. The patient was lost to follow-up post-discharge because she had come from another city.

Carbon dioxide laser technique is now used for removal of supraglottic scarring, but this is not available in our institution. One of the advantages of laser technique is the reduction in surgical time. Availability of laser could have decreased the surgical time in this patient and this may have resulted in preventing the morbidity which only occurred after 30 minutes into surgery.

In conclusion, this case highlights the problem of airway management in a patient with bullous pemphigoid and complications that can occur with jet ventilation via the catheter technique. Management of these cases

requires constant vigilance on the part of anaesthetist who should be aware of such potential complication.

### REFERENCES

1. Budimir J, Mihic LL, Situm M, Bulat V, Persic S, Tomljenovic-Veselski M. Oral lesion in patient with pemphigus vulgaris and bullous pemphigoid. *Acta Clinica Croatica* 2008; **48**:13-8.
2. Prasad KK, Chen L. Anaesthetic management of a patient with bullous pemphigoid. *Anesth Analg* 1989; **69**:537-40.
3. Iqbal M, Ahmed R, Kashef SH, Bahame PS. Laryngeal involvement in a patient with bullous pemphigoid. *Ann Saudi Med* 2006; **26**:152-4.
4. Ort SA, Goldenberg D. Transoral management of obstructing tumour of upper airway. *Operat Tech Otolaryngol* 2007; **18**:156-9.
5. Drenger B, Zidenbaum M, Reifen E, Leitersdorf E. Severe upper airway obstruction and difficult intubation in cicatricial pemphigoid. *Anaesthesia* 1986; **41**:1029-31.
6. Lazor JB, Varvares MA, Montgomery WW, Goodman ML, Makool BT. Management of airway obstruction in cicatricial pemphigoid. *Laryngoscope* 1996; **106**:1014-7.
7. Mahalingam TG, Kathirvel S, Sodhi P. Anaesthesia management of a patient with pemphigus vulgaris for emergency laparotomy. *Anaesthesia* 2000; **55**:160-2.
8. Evans E, Biro P, Bedforth N. Jet ventilation. *Cont Educ Anaesth Crit Care Pain* 2007; **7**:2-5.

