British Journal of Anaesthesia **92 (2)**: 283–6 (2004)
DOI: 10.1093/bja/aeh029

Angina bullosa haemorrhagica presenting as acute upper airway obstruction

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We report a case of acute upper airway obstruction caused by a rapidly expanding blood-filled bulla in the oropharynx (angina bullosa haemorrhagica), requiring tracheal intubation. The larynx could not be visualized by either awake fibreoptic laryngoscopy or direct laryngoscopy under anaesthesia. Surgical tracheostomy was therefore performed under general anaesthesia.

Br J Anaesth 2004; **92**: 283–6

Keywords: airway; anaesthetic techniques, fibreoptic laryngoscopy; complications, angina bullosa haemorrhagica; intubation, tracheal; surgery, tracheostomy

Accepted for publication: September 3, 2003

Angina bullosa haemorrhagica (ABH; from the Latin verb *angere*, to choke on) is a rare condition characterized by one or more blood-filled blisters or bullae in the oropharynx. To our knowledge (Pubmed search 1967–2003) it has not previously been reported to cause airway obstruction. We describe a case of ABH where a rapidly expanding bulla caused obstruction of the oropharynx. The size of the bulla and free blood in the airway hindered visualization of the larynx and intubation of the trachea by either fibreoptic endoscopy or direct laryngoscopy. The airway was secured by surgical tracheostomy. We discuss our approach to the management of the airway in this case and provide an introduction to ABH.

Case report

A 56-yr-old woman presented to the Accident and Emergency Department complaining of a weak voice, painful throat and salivary incontinence (dysphagia with drooling). She reported a similar less-severe episode 2 months previously, related to several blood-filled blisters on the buccal mucosa and hard palate, which had subsequently ruptured, leaving painful ulcers. These had been treated successfully with topical steroid (beclomethasone) and chlorhexidine mouthwash. The patient was also being treated with peritoneal dialysis for end-stage renal failure secondary to systemic lupus erythematosus (SLE), and she was taking oral prednisolone.

On examination the patient was not in any respiratory distress, and oxygen saturation on air was 99%. She was spitting blood-stained saliva. A large blood-filled bulla visible in the mouth was obstructing the left half of the oropharynx by protruding from the lateral and posterior pharyngeal wall into the lumen. In addition, there was a smaller (approximately 5 mm in diameter) haemorrhagic blister on the mucosa of the right lower lip. Assessment of the airway gave a Mallampati score of 1. The patient was able to protrude the mandible normally (Calder Class A[1]), and the thyromental distance was greater than 6.5 cm.

On flexible endoscopy shortly after admission in the Accident and Emergency Department, the nasal airway as well as the laryngeal airway beyond the oral lesion appeared normal. The vocal cords were mobile. The haemoglobin concentration was 10.3 g dl^{-1}, platelet count was 205×10^9 $litre^{-1}$ and the white cell count was 10.4×10^9 $litre^{-1}$. Coagulation tests were normal and the C-reactive protein concentration was 18 mg $litre^{-1}$ (normal value <8 mg $litre^{-1}$). The patient was admitted for observation and received oxygen via a face mask (4 litre min^{-1}). In an attempt to stop further progression of the lesions, nebulized epinephrine 1 mg and oral prednisolone 30 mg were administered.

Within 2 h of admission, the patient developed agitation and respiratory distress, with laboured breathing and inspiratory stridor. Oxygen saturation remained above 95%. On clinical examination, the oropharyngeal bulla had increased in size and now crossed the midline. Anticipating progression to possible complete airway obstruction, a decision to secure the airway with a tracheal tube was made. Three possible action plans were formulated by the anaesthetists and the ENT surgeon on call. Plan A was for awake fibreoptic intubation; plan B consisted of inhalational

induction followed by direct laryngoscopy and intubation; plan C was for surgical tracheostomy if necessary.

The patient was transferred to the operating theatre and prepared for awake fibreoptic intubation via the nasal route. We planned only one attempt because of the risk of rupturing the bulla. The increasingly agitated patient was kept in a sitting position (the only position tolerated). ECG, non-invasive arterial pressure measurement and pulse oximetry were used for monitoring. For local anaesthesia and vasoconstriction, cocaine 5%, 2 ml and lidocaine 4%, 2 ml were sprayed into both nostrils. The oropharynx was sprayed with lidocaine 10%. There was some increase in agitation, making cooperation more difficult, although the oxygen saturation remained above 95%. The patient was reassured but no sedation was used because of the risk of complete airway obstruction. The ENT surgeon and a theatre nurse stood by, both scrubbed and with a tracheostomy set opened. Endoscopy was carried out using a fibrescope with a diameter of 4.0 mm (Olympus LF-2), loaded with an armoured 6.0 mm tracheal tube. Beyond the nasopharynx, the airway or other recognizable anatomical structures could not be vizualised (one attempt only by a senior anaesthetist). The endoscopic view obtained was largely that of haemorrhagic tissue partly covered with blood.

Inhalational induction using sevoflurane in oxygen 100% (flow rate 10 litre min^{-1}) was started, with the intention of performing direct laryngoscopy (plan B). Increasing concentrations of sevoflurane (0.5–6%) were used and the patient was moved to the supine position. During induction, local skin infiltration with lidocaine 1% and epinephrine 1:200 000 was performed by the ENT surgeon in preparation for a potential emergency tracheostomy (plan C). Throughout induction, the airway was maintained without the use of airway aids and the oxygen saturation remained above 95%. Direct laryngoscopy using a size-4 Macintosh blade was performed cautiously under deep anaesthesia. Again, only haemorrhagic tissue and fresh blood were seen, but no recognizable anatomical structures. No further laryngoscopy procedures were attempted.

We therefore proceeded to surgical tracheostomy (plan C). This was straightforward and without acute complications. Oxygen saturation was 95% or above throughout these procedures. After confirmation of tracheal ventilation by capnography and auscultation, anaesthesia was maintained using an infusion of propofol 150–300 mg h^{-1}, fentanyl 100 μg bolus and vecuronium 8 mg. On examination under anaesthesia, the Nikolsky sign was negative. The Nikolsky sign means that sliding pressure with a finger separates normal-looking epidermis from the dermis, resulting in a visible erosion; it is positive in pemphigus and more rarely in toxic epidermal necrolysis.[2] Biopsies were taken from the lesions and the patient was transferred to the intensive care unit.

Over the next 2 days there was a continuous ooze of blood from the oral lesions. The haemoglobin concentration decreased to 4.8 g dl^{-1} 1 day after the tracheostomy was performed. The patient received transfusions of red cells and fresh frozen plasma. Recovery was complicated by a *Pseudomonas* respiratory tract infection, which slowed weaning from artificial ventilation. Systemic steroids were continued (hydrocortisone 50 mg i.v. twice daily initially, gradually reducing to a low maintenance dose) to treat both the SLE and the mouth lesions before a histopathological diagnosis was made.

The histopathological and immunohistological appearance of the biopsies was non-specific. The pharyngeal specimen contained only clotted blood and normal tissue. The biopsy taken from the lip was normal mucosa, but there was non-specific immunofluorescence with IgG and C3. Serology was negative for bullous pemphigoid and pemphigus vulgaris. The patient was discharged to the local renal unit after 15 days on the intensive care unit.

On follow-up nasendoscopy 17 days after the acute event, there was no residual disease in the oral cavity, pharynx or larynx. The tracheostomy tube was removed and the stoma covered on the same day.

Discussion

This is a case of acute airway obstruction caused by an extensive haemorrhagic blister in the oropharynx. Biopsies showed non-specific features at both histopathology and immunofluorescence, leading to the clinical diagnosis of ABH. To our knowledge, no case of airway obstruction with respiratory compromise caused by ABH has been described in the literature (Pubmed search 1967–2003).

Badham first described ABH in 1967 as blood-filled blisters in the oral, pharyngeal and oesophageal mucosa.[3] The onset is sudden and may follow trauma caused by eating, hot drinks,[3–5] dental procedures or shouting.[3–6] The use of steroid inhalers in asthmatic patients is a possible aetiological factor.[7] In the largest published series of 30 patients, no precipitating factor was found in 47%.[5] Hosain and colleagues[8] reported a case of postoperative ABH caused by intubation and extubation, describing a patient with a single blister at the junction of the soft and hard palate that did not compromise the patient's airway. Lesions predominantly occur on the soft palate.[5] Blisters usually reach 2–3 cm in diameter and burst spontaneously, leaving ragged ulcers that heal without scarring.[5] Clinically, the lesions may recur.

The diagnosis of ABH is largely clinical, and includes the elimination of other disease processes at histology. Histopathology can reveal haemorrhagic subepithelial bullae, non-specific ulceration and a chronic inflammatory cell infiltrate in the lamina propria.[5] Biopsies taken from the lesions in our patient did not have these histopathological appearances. However, they were obtained early in the development of the blisters and may have been unrepresentative. Direct immunostaining is usually negative, but

equivocal immunostaining for IgG and C3 may be seen, as in our patient's biopsy of the lip lesion, and in four out of the 15 patients presented by Stephenson and colleagues.[5]

Although our patient suffered from SLE, which may cause oral ulceration, no vasculitis was evident on histopathology, making SLE an unlikely cause.[9] No specific immunological or histopathological features of the numerous other vesiculobullous lesions (erythema multiforme, pemphigus or pemphigoid, linear IgA disease, dermatitis herpetiformis, epidermolysis bullosa acquisita, bullous lichen planus and amyloidosis) were present. The treatment for ABH has been described as symptomatic, using a mouthwash and analgesics.[10]

Impending acute airway obstruction secondary to haemorrhagic blisters should be managed as for any acute upper airway obstruction, with consideration given to three main strategies: awake fibreoptic intubation, inhalational induction of general anaesthesia and intubation, or tracheostomy. Whichever strategy is selected, possible use of alternatives needs to be planned in advance. The choice of the initial strategy depends on a variety of factors. These include the degree of respiratory distress, the cooperation of the patient, the position of the disease process within the airway, the likelihood of liquid material within the airway (pus, blood or secretions), the presence of stridor and the experience of the anaesthetists and surgeons involved. In disease processes where the obstructing lesion is likely to resolve over a few days, a surgical tracheostomy is often considered as a reserve procedure, provided that it can be carried out immediately if necessary.[11]

In a moribund patient, cricothyrotomy and/or surgical tracheostomy may be needed immediately. Tracheostomy can, however, be technically difficult, particularly if a distressed patient is unable to cooperate. The patient described here developed marked agitation and was unable to lie or sit still on the operating table. Placement of a cricothyrotomy needle and performance of tracheostomy under local anaesthesia would therefore have been associated with a risk of inadvertent trauma.

Awake fibreoptic intubation in acute upper airway obstruction can be technically challenging because of abnormal anatomy, blood, pus or secretions obscuring the view, or in an agitated, non-sedated patient.[11–13] The presence of stridor, as in our patient, is thought to indicate a reduction of the airway diameter of at least 50%.[12] Fibreoptic endoscopy may completely obstruct the airway either by physically occluding the lumen further (a particular risk if stridor is present) or by causing trauma with subsequent bleeding.[11–13]

We decided to attempt an awake fibreoptic intubation in the patient described here because we had successfully performed this technique with the use of topical anaesthesia in cases of acute upper airway obstruction. With this in mind, we elected firstly not to advance the fibrescope beyond the nasopharynx blindly into the narrowed airway containing blood and secretions, and secondly to make only one attempt. If this failed, we planned to abandon the endoscopy.

Topical anaesthesia of the airway with lidocaine is a discussion point in individual cases of acute airway obstruction managed with fibreoptic intubation, as it may provoke transient laryngeal spasm and pharyngeal obstruction.[11–13] In our case, topical application of lidocaine to the back of the oral cavity and cocaine to the nasal cavity did not lead to an exacerbation of stridor or respiratory distress. Oxygen saturation (pulse oximetry) remained above 95%.

We chose cocaine 5%, 2 ml (and lidocaine 4%, 2 ml) for vasoconstriction and topical anaesthesia of the nasal cavity because of our experience in fibreoptic intubation. The dose used was well below the maximum recommended (1.5 mg kg^{-1}). Cocaine, however, may be rapidly absorbed from the respiratory mucosa into the systemic circulation and may then cause powerful cerebral stimulation, causing excitation and exacerbating any pre-existing agitation. Cocaine 2 mg kg^{-1} may cause coronary vasoconstriction.[14] Topical application of cocaine in our patient was followed by some increase in agitation before induction of general anaesthesia, although this might have been related to the patient's concerns about the planned procedure. In retrospect, alternative vasoconstrictors such as phenylephrine 1% or xylometazoline 0.1% might be considered in a similar situation. Xylometazoline has been shown to be as effective as cocaine in producing nasal vasoconstriction.[15]

Our single carefully controlled direct laryngoscopy with a Macintosh blade did not cause further bleeding, although there was clearly a risk of the blood-filled bulla bursting into the airway. Again, we made only one attempt. Close communication with the surgeon, who started disinfecting and infiltrating the skin of the anterior neck with local anaesthetic during the inhalation induction and laryngoscopy attempt, enabled quick progression to tracheostomy after direct laryngoscopy had failed.

We chose sevoflurane as the inhalational induction agent because of its fast onset and offset, and minimal arrhythmogenic potential; however, hypoventilation and apnoea may occur.[16 17] The successful use of sevoflurane in cases of airway obstruction caused by supraglottic tumours and acute epiglottitis is well described.[12 18 19]

In summary, in this case of airway obstruction caused by a haemorrhagic blister with stridor and blood in the airway, neither awake fibreoptic nor direct laryngoscopy allowed a view of the glottis, and surgery was ultimately required. Faced with a similar patient with a clear history of blood present in the mouth, and a visible lesion as a potential source, we would now consider earlier inhalational induction of anaesthesia followed directly by surgical tracheostomy or, if the patient is able to cooperate, surgical tracheostomy under local anaesthesia.

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