

## Oral Scarring in Behcet's Disease - An Airway Concern

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### Introduction

Behcet's disease is a multisystem, autoimmune disorder presenting with recurrent oral and genital ulcerations as well as ocular involvement [1]. The disease affects young males and females equally but males often have more severe form of the disease. In this disease scarring of the mucosa is not a prominent feature but may occur and produce anaesthetic difficulties [2]. Here, we want to draw attention to this rare dermatological disease that presented to us with airway difficulty secondary to mucosal scarring. Due to extensive scarring around the mouth orotracheal intubation was not possible by rigid laryngoscopy.

### Case Summary

A 14 year old young boy (weight 35 kg) suffering from Behcet's disease was admitted in Burns and Plastic department of our institute. The patient was scheduled to undergo release of contracted commissure, vermilion, excision of fibrous tissue and correction with local flap. At an age of 11 years, he developed oral ulcers, cutaneous ulcers (on side of mouth, elbows, knees and ankle) and genital ulcers and was diagnosed with Behcet's disease on the basis of diagnostic criteria by Mason and Barnes. He was put on treatment in the form of oral prednisolone. The cutaneous and genital ulcers healed but the recurrent oral ulcers resulted in microstomia due to scarring. This interfered with feeding and speech. On preoperative assessment cardiovascular, respiratory and gastrointestinal systems were normal. On airway examination there were no oral ulcers except for narrow mouth opening around one finger (1.0 x 1.25 cm). His nasal passages were normal. His investigations including Hb, BT, CT, blood sugar, blood urea, and serum electrolytes were 11 gm%, 2.08 min, 5.15 min, 74 mg%, 35 mg% 142 mEq/l sodium and 3.8 mEq/l potassium respectively. Chest x-ray and echocardiography were found to be normal. We planned to do awake nasal fibreoptic intubation. The patient was premedicated with tablet alprazolam 0.25 mg on the night before surgery. The whole procedure was explained to him. Injection glycopyrrolate 0.2 mg was given after placement of 20 G intravenous cannula. He was nebulised with 4 ml of 4% xylocaine. He was advised to do gargles with 4 ml of 2% xylocaine viscous and xylometazoline drops were put in right nostril. Simultaneously, arrangement for difficult airway cart was made. After this, the patient was transferred to the operating room.

In operating room after applying standard monitoring devices, parental sedation was given with 0.5 mg midazolam. 2% xylocaine jelly was put in right nostril. A 3.6 mm fibreoptic bronchoscope with a preloaded 6.0 mm flexometallic tube was passed through split nasopharyngeal airway in right nostril. At the level of glottic inlet 2 ml of 2% lidocaine was instilled through the working channel. After confirmation of fiberscope position in midtrachea, the well lubricated tracheal tube was advanced over the fiberscope into the trachea. Tracheal tube position was confirmed with capnograph. General anaesthesia was induced with IV propofol 80 mg, fentanyl 100 µg, atracurium 20 mg and sevoflurane. Surgery was completed without an incident. Trachea was extubated after completion of surgery. The intra-operative and recovery period were uncomplicated and patient was shifted to the ward after being observed for 2 hrs in the recovery room. Patient stay in the hospital was uneventful and was discharged home after 5 days.

### Discussion

Behcet first described the syndrome complex in 1940. He described aphthous changes in mouth and genitalia associated with iritis and hypopyon. Oral and genital lesions are usually first to appear and may precede the ocular manifestations by many years. A variety of other symptoms may also be associated with this syndrome. Diagnostic criteria as suggested by Mason and Barnes are as follows:

**Major:** Buccal ulceration, genital ulceration, eye lesions, skin lesions.

**Minor:** Gastrointestinal lesions, cardiovascular lesions, thrombophlebitis, arthritis, CNS lesions, family history.

Diagnosis of Behcet disease should be made only if three major or two major and two minor criteria are present [3]. The etiology of the disease is unknown. The main pathologic lesion is a systemic perivascularitis with early neutrophil infiltration, endothelial swelling and fibrinoid necrosis. Usually the syndrome runs a chronic course with exacerbations becoming less frequent with time [1]. There are very few case reports regarding anaesthetic management of patients with Behcet's disease. This made us to report this case.

The main preoperative anaesthetic concerns are scarring of airway leading to difficult intubation and involvement of other organ systems. Patient should be investigated according to involvement of organ systems. In our patient, there was no evidence of any major organ system involvement. Because Behcet disease is an inflammatory process, chronic use of anti-inflammatory and antineoplastic drugs is common. Blood urea nitrogen/ creatinine should be measured to identify or quantitate chronic renal disease and to reveal nephrotoxicity of treatment. With chronic corticosteroid use, supplemental corticosteroids are necessary on the day of surgery. Puncture of skin or mucous membranes is very likely to result in inflammation and nodular formation and should be kept to a minimum. This would mean that regional anaesthesia would be less ideal but not contraindicated. If spinal cord lesions are present, use of succinylcholine can result in hyperkalemia. With anaesthesia of the airway, topical application of local anaesthetics would be preferred to airway blocks because of potential compromise of the airway from inflammatory response to local injection. For the same reason and young age we used topical anaesthesia for awake fibreoptic nasal intubation. General anaesthesia can be challenging if oropharyngeal lesions are present [2]. In extreme cases, lesions can severely reduce the lumen of oropharynx and tracheostomy might be necessary for urgent surgery. For elective procedures, awake fibreoptic intubation is usually

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required. Use of LMA can aggravate lesions in the airway. Repeated attempts at laryngoscopy and intubation can cause tissue trauma and lead to oral aphthous ulcers [4]. Visual inspection of the oropharynx is indicated prior to intubation. If the patient is unable to protrude the tongue to a normal degree, an opinion from an e.n.t surgeon should be obtained prior to anaesthesia.

In conclusion, we recommend thorough and careful airway assessment in these patients before taking up for surgery as these patients can present with a difficult airway secondary to mucosal scarring. Proper visual inspection of oropharynx is indicated prior to intubating such patients. Meticulous intra-operative planning and preparation of difficult airway cart is recommended for successful management of such cases.

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