ANAESTHETIC DIFFICULTIES ASSOCIATED WITH BEHÇET'S SYNDROME

Case Report

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SUMMARY

The case is described of a young woman with Behçet's disease who underwent anaesthesia for dental extraction. Due to the extensive scarring and adhesion formation in the pharynx, nasotracheal intubation was impossible and oropharyngeal intubation extremely difficult. Attention is drawn to other dermatological diseases which may also produce anaesthetic problems due to pharyngeal scarring.

Behçet's disease is a relatively rare condition which can be classified as belonging to the muco-cutaneous ocular syndromes (Walsh and Hoyt, 1969). The disease is difficult to diagnose and does not usually produce scarring of the pharynx (Price, 1969). However, this case report illustrates anaesthetic problems which can arise in this condition.

CASE REPORT

Clinical history.

A female was first admitted to Dunedin Hospital on October 2, 1960, at the age of 8 years with a 3-week history of intermittent fever, malaise and painful ulceration of her tongue and mouth. A blood test had been taken 2 weeks prior to admission and a septic spot occurred at the injection site which later broke down into an ulcer. A week later she exhibited pustular lesions around her mouth and over her arms and legs, which also became ulcerated, and complained of pain in her right foot. While in hospital she was treated with penicillin and pustules formed at the injection sites. Swabs were taken from her tongue and skin lesions, and these were sterile apart from the normal flora. Blood cultures were also negative. Later she suffered pain in her right knee and left foot. She presented a difficult diagnostic problem but it was felt at this time that she was suffering from polyarteritis nodosa. Steroid therapy by mouth was started and her condition improved dramatically so that by December 3 she was well enough to be discharged.

Over the next three years she had three episodes of ulceration of the tongue, lips and fauces and on one occasion she also had arthralgia. In 1963, at the age of 12 she had some teeth extracted under general anaesthesia and though at this time it was noted that a No. 5 Magill orotracheal tube was "a little big for her", the anaesthetic was otherwise without incident.

In December 1964 she again developed ulcers of her tongue, fauces and skin. She was in considerable pain and required hospital admission. At this stage she was diagnosed as suffering from Behçet's syndrome although she displayed no genital or ocular manifestations of the disease. During the next two years she had five episodes of ulceration of the tongue, pharynx and skin which required hospital admission on two occasions. At the end of 1966 she also developed ulcers on the labia, which like those in her mouth, were extremely painful. Over the next two years she had numerous exacerbations of mouth and throat ulcerations and these were associated with labial ulcers on three occasions and arthralgia twice.

In April 1968 she was given a course of mercaptopurine and required amitryptyline for reactive depression. It was found in 1970 that her soft palate was tethered and her nasal airway was obstructed. Throughout her illness she had received steroid therapy and during exacerbations increased steroid dosage quickly brought the disease under control.

Anaesthetic management.

On April 5, 1971, the patient was admitted to Dunedin Hospital for the extraction of four impacted wisdom teeth under general anaesthetic. On the pre-operative visit it was noted that the girl was small for her age and that she had no nasal airway. This could not be corrected with xylometazoline (Otrivine) nasal spray. Her case was discussed with her physician and it was decided not to attempt nasotracheal intubation. Unfortunately her pharynx was not examined, but since the disease was in remission no trouble was anticipated with orotracheal intubation. She was premedicated with pethidine 75 mg and atropine 0.6 mg intramuscularly 1 hour pre-operatively and was also given 100 mg hydrocortisone with the premedication.

The patient was calm on arrival in theatre and induction of anaesthesia was carried out with 2.5 per cent thiopentone 250 mg. This was followed by suxamethonium 50 mg. Fasciculations occurred and attempts were made to inflate the lungs with oxygen but without success. It was then found to be also impossible to pass an airway into the pharynx. A laryngoscope was inserted and it could be seen that the pillars of the fauces were scarred and contracted. With the help of a laryngoscope, a size 2 oropharyngeal airway was introduced and the patient (who by this time was slightly cyanosed) was ventilated with oxygen. She was then given a further 100-mg dose of thiopentone and 25 mg of suxamethonium, and the throat was again examined. Because of the shortening of the pillars it was impossible to bring the tongue forward and therefore the back of the tongue, epiglottis and vocal cords could not be visualized. The soft palate was firmly adherent to the pharyngeal wall. The airway was reinters
and the patient ventilated with nitrous oxide, oxygen and 2 per cent halothane, and then allowed to breathe spontaneously.

Attempts were made to carry out blind oropharyngeal intubation, using a size 6 Portex tube and a malleable introducer. By listening to the intensity of the breath sounds at the end of the tube, it was possible to guide it towards the vocal cords, but it became obvious that the laryngopharynx was also distorted. Anaesthesia had to be deepened by gradually increasing the inspired halothane concentration and the process repeated. Finally on the third attempt, the trachea was intubated successfully. The oropharynx was then packed with a strip of gauze and anaesthesia was maintained with nitrous oxide, oxygen and 1 per cent halothane. During the operation (which was performed without incident) efforts were made to pass a soft rubber catheter into the oropharynx via the nostrils, but these were unsuccessful.

Postoperatively the pharynx was thoroughly sucked out and an oropharyngeal airway introduced, prior to the removal of the tube. No airway problems developed and the patient awoke uneventfully.

She was seen in the ward two days after the anaesthetic. Her throat was still a little sore and it was found that the tip of her tongue could not clear her lower lip when she was requested to protrude it. On questioning, she admitted progressive dysphagia for some months prior to hospitalization. This facet of her history had not previously been obtained. On April 7, 1971, her sutures were removed, without anaesthesia, and she was discharged from hospital.

**DISCUSSION**

Behçet first described the syndrome complex which bears his name in 1940. He described aphthous changes in the mouth and genitalia associated with iritis and hypopyon, though he stated that the latter was not always present. Oral and genital lesions are usually the first to appear and may precede the ocular manifestation by many years. It is now realized that a variety of other symptoms are associated with the syndrome, and Mason and Barnes (1969) suggest the manifestations of the disease should be grouped as follows.

**Diagnostic criteria:**

**Major:** buccal ulceration; genital ulceration; eye lesions; skin lesions.

**Minor:** gastrointestinal lesions; thrombophlebitis; cardiovascular lesions; arthritis; CNS lesions; family history.

A diagnosis of Behçet’s syndrome should be made only if three major or two major and two minor criteria are present.

Often it takes several years before the diagnosis can be made because of the gradual unveiling of the clinical picture. The patient presented shows the abnormal reactivity of the skin to pinprick found in about 20 per cent of patients with this condition (Price, 1969). The aetiology of the disease is unknown but there are several theories which include viral infection (Evans, Pallis and Spillane, 1957), allergic reaction (Oshima et al., 1963) and vascular reaction (McMenemey and Lawrence, 1957). The disease is more common in the third decade but ages may range from 6 to 65 years (Oshima et al., 1963). Females are more susceptible than males.

Usually the syndrome runs a chronic course with exacerbations becoming less frequent with time. Blindness usually ensues, but life expectancy is normal except when neurological complications occur; these lead to dementia and death. Since the cause of the disease is unknown, treatment has been extremely variable and unsuccessful. Systemic and topical steroids are the mainstay of therapy and although these alter the severity of the symptoms there is little evidence that they have any effects on the course of the disease.

Behçet’s disease is a member of the mucocutaneous ocular syndromes; along with Reiter’s disease, erythema multiforme exudativum, Stevens-Johnson syndrome and Degos’ syndrome (Walsh and Hoyt, 1969). These diseases present a spectrum of signs and symptoms which tend to merge into one another and often make dogmatic diagnosis difficult. In all these conditions, scarring of the mucosa is not a prominent feature but may occur and produce anaesthetic difficulties. Other dermatological conditions are associated with scarring of the mucosa. The most important of these is benign mucous membrane pemphigus (Pemphigoid) which normally affects elderly people. Scarring is the prominent feature of all the lesions (Sneddon, 1961) and adhesions may occur between the lips, and between the soft palate and pharyngeal wall (Asboe-Hansen, 1970). This is in contrast to “true pemphigus” (pemphigus vulgaris, vegetans and foliaceus), in which there is no scarring. Epidermolysis bullosa dystrophica is a rare condition inherited as a dominant trait. Severe anaesthetic difficulties are associated with the disease since not only is there marked scarring of the mouth and pharynx but pressure results in the formation of bullae (Hamann and Cohen, 1971). Scleroderma sometimes affects the oesophagus and produces dysphagia and the pharynx too can be involved and constricted.

In all these conditions, 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. Where scarring of the pharynx exists, local anaesthesia, neuroleptanalgesia or ketamine should be employed if possible. If intubation is required, the anaesthetist must be prepared to carry this out blindly either by the nasal or oral routes.

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REFERENCES


