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Conservative laryngectomy
(An effective treatment for severe aspiration in motor neurone disease)

by

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Summary

Four cases are presented to illustrate that conservative laryngectomy is a simple and reliable treatment for severe aspiration in patients with motor neurone disease whose symptoms are predominantly bulbar. This treatment leads to a rapid return of the patients to their home, thus reducing their dependancy on skilled nursing care.

Introduction

Aspiration pneumonia is the frequent fate of many patients with motor neurone disease. Neurologists are often faced with a case in which the symptoms are predominantly bulbar. In such cases the quality of life may be dramatically improved if the aspiration can be prevented. We describe here four cases and discuss the range of surgical remedies for aspiration.

Case Reports

Case 1

A 66-year-old retired court officer presented to the ENT Department with an eighteen-month history of progressive dysphagia, having already been in hospital for one week. On examination he was anarthric, unable to swallow solid food and all liquids were promptly aspirated. There was spasticity of the face, tongue and palate accompanied by a brisk jaw jerk. However, he had almost normal power in both flexors and extensors of all limbs.

He underwent a total laryngectomy and was discharged from hospital 30 days later having learnt to feed himself with liquidized food. This treatment gave him five months of enjoyable home life before being overcome by generalized body weakness never requiring readmission to hospital.

Case 2

This 56-year-old man presented in March 1981 with a five-month history of progressive dysarthria, drooling of saliva and dysphagia with regurgitation. On examination he was found to have ataxic nystagmus, bilateral facial weakness, poor gag reflex and a fasciculating tongue which was deviated to the right. He had normal power and range of movement in all four limbs.

In April 1981, six months after noticing his first symptoms, the excessive salivation was treated with radiotherapy to the parotid glands. A cricopharyngeal myotomy was carried out in August 1981 to relieve his increasing dysphagia and overspill. In October 1981 he was readmitted anarthric, cachetic and with pneumonia due to severe aspiration of liquids and solids.

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He had a total laryngectomy in December 1981. He returned home fourteen days post-operatively. Most of the following year was spent working in his flower shop after which he died eleven months post-operatively.

Case 3

Two years after he was diagnosed as having motor neurone disease, a retired 64-year-old tool maker presented to the ENT department. He was anarthric, cachetic and aspirated anything that he swallowed. He had normal or virtually normal power in his limb and limb girdle musculature, though fasciculation and wasting were noted of some limb muscles.

A total laryngectomy was performed the same month, from which he made a rapid recovery. He returned to a fulfilling home life where his family were able to care for him until he died ten months post-operatively.

Case 4

A 64-year-old man was referred with dysphonia, dysphagia with aspiration and pneumonia involving both lungs. He had previously been diagnosed as having motor neurone disease. He had virtually normal power in his lower limbs though there was a marked weakness of the left upper limb girdle muscles. The right arm still had excellent power such that the patient was still able to perform routine daily tasks such as feeding himself and dressing. He had lost more than 20 kg. in weight and was in a poor nutritional status.

Although there was marked incoordination of pharyngeal and laryngeal movement it was decided to perform an epiglottopexy as his voice was still understandable. This failed to heal adequately and three weeks post-operatively his lower respiratory tract was still unprotected. A total laryngectomy was performed.

He made an excellent recovery from this, being discharged from hospital 17 days post-surgery. He was readmitted to hospital 13 months later where he died of inanition within a week.

Discussion

The three basic functions of the larynx are; (1) protection of the lower airway; (2) respiration; and (3) phonation. Once the protective function is lost the patient's life is threatened by aspiration pneumonia. It is generally agreed that the ideal procedure should be simple, reliable, and reversible in patients who are expected to recover laryngeal function (Kirchner and Sasaki, 1984). In motor neurone disease, laryngeal function is gradually lost so that in the advanced stages of the disease the patient is aphonic, cachetic and aspirating the food that he tries to eat.

Cricopharyngeal myotomy is a valuable procedure in the management of motor neurone disease provided that it is carried out soon after the patient develops dysphagia but before the onset of aspiration (Loizou *et al.*, 1980). Once tongue and hypopharyngeal weakness is established it does nothing to prevent aspiration.

If there is still a useful voice but aspiration is present, epiglottopexy is then the operative procedure of choice (Vecchione *et al.*, 1975). The one case reported here failed to heal adequately and it is our experience in several other cases that continued aspiration of saliva is common, particularly when the operation is done in a patient with reduced powers of healing due to poor nutritional status.

In the aphonic patient with aspiration there is a range of surgical treatments available. The commonly used nasogastric tube with tracheostomy and cuffed tracheal tube is expensive as it requires continuous skilled nursing care or a trained coordinated patient to maintain proper inflation of the balloon cuff to prevent aspiration and yet minimize the risks of tracheal necrosis. This often means that the patient is hospitalized for frequent, long periods. Only two of the four patients presented here required readmission to hospital following laryngectomy, and only for a total of eleven days. The use of a tracheostomy with cuffed tube alone also impedes swallowing.

Montgomery (1975) recommended a simple glottic closure technique *via* a laryngofissure with a tracheostomy. Although this procedure is reversible, none of the 12

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patients he reported had undergone sufficient recovery to merit a reconstitution of the laryngeal airway. One of the patients reported also required revision surgery. This glottic closure may be reinforced by a superiorly based sternomastoid muscle pedicle (Kirchner and Sasaki, 1984).

Baron and Dedo (1980) recommended transecting the trachea below the first ring and making a permanent tracheostome. The subglottis is closed by making the first ring into two trap doors and approximating them in the midline. Pooling of saliva and food in the larynx is said to be a problem in this technique but Baron and Dedo deny this. This technique also has the advantage of being reversible.

Lindemann *et al.* (1976) also proposed a reversible technique for use in patients who may recover their laryngeal function. This involved dividing the trachea just below the larynx and anastomosing the superior stump to the oesphagus and the inferior stump to the skin as a tracheostome. Baron and Dedo (1980) claim that separation and closure of the larynx is sufficient and the tracheo-oesophageal anastomosis is unnecessary.

Once the ambulant patient with motor neurone disease has reached the stage of aphonia with severe aspiration then surgery should be offered to relieve his distress. Management by laryngectomy offers an effective treatment by a single operative procedure with a short hospital stay. Unlike a laryngectomy for malignant disease the strap muscles are preserved and are utilized to reinforce the pharyngeal repair, thus reducing the risk of a pharyngo-cutaneous fistula even in the cachetic patient. In all the four patients above healing was by primary intention and none developed a pharyngo-cutaneous fistula. The patient should then be able to return to a home-based life or even to work.

Cannon and McLean (1982) managed four patients with severe intractable aspiration by total laryngectomy and concluded that, where the prognosis for recovery of laryngeal function is poor, or where wound healing is compromised, a laryngectomy is the procedure of choice. We endorse this opinion. Conservative laryngectomy is a reliable technique that can be performed usually within a couple of hours and under local anaesthesia when necessary. It offers the best chance for these patients to be returned to their homes to live out their lives with their families and possessions around them.

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