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Bilateral ventricular neurofibroma of the larynx

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Abstract
Laryngeal neurofibroma (LNF) is an unusual tumour and only approximately two dozen cases have been reported previously. Isolated LNF not associated with von Recklinghausen's disease is even more unusual. According to a review of the available literature, the case presented is the first bilateral one and the first originating from the ventricles. The tumour was removed completely by microlaryngoscopy without tracheostomy. The macroscopic and light macroscopic findings are presented and a review of the literature on neurogenic tumours in the larynx is presented.

Key words: Laryngeal neoplasms; Neurofibroma

Introduction

Neurogenic tumours of the larynx are rare. Schwannomas and neurofibromas are the two major types of neoplasm. Approximately 130 cases of neurogenic neoplasms have been reported up to 1993 (Barnes and Ferlito, 1993). They can affect all age groups and are slightly more common in females (Shapshay and Rebeiz, 1993).

According to a World Health Organization (WHO) definition (Shanmugaratnam, 1991) neurofibroma is a benign tumour consisting of a mixture of neurites, Schwann cells and fibroblasts in a collagenous or mucoid matrix. Schwann cells frequently predominate. The cells have elongated dark-staining nuclei, with a wavy configuration and pointed ends (Rosai, 1989). They may occur as an isolated lesion, as in our patient, or more often as a part of von Recklinghausen's disease (Chang-Lo, 1977). Both sexes are involved in equal proportions (Barnes and Ferlito, 1993).

Macroscopically neurofibromas are firm, white-grey and well circumscribed but not encapsulated like schwannomas. Plexiform and cellular types have also been reviewed in the larynx as variants of neurofibromas (Sidman *et al.*, 1987; Stanley *et al.*, 1987). Patients with LNF are generally younger than patients with schwannoma and airway obstruction is seen more frequently (Stanley *et al.*, 1987). LNF show a tendency to occur in the aryepiglottic folds (Chang-Lo, 1977) and arytenoid cartilages (Mills and Fechner, 1985; Stanley *et al*, 1987).

In this report a neurofibroma of the larynx involving both ventricles is presented as the first case in literature. It is also, to our knowledge the first report of a LNF without any symptoms.

Case report

A 45-year-old man was admitted to the Ear, Nose and Throat-Hospital of Graz University because of chronic left-sided otitis media. During routine otorhinolaryngological examination, indirect laryngoscopy showed a polypoid mass flopping over the right vocal fold, originating from the right ventricle. The patient, who had been a heavy smoker for 25 years presented with a harsh and strained voice but himself was not aware of any voice disturbance. Laryngoscopy with the 70° magnifying endoscope and videostroboscopy confirmed that the polypoid lesion originated supraglottically from the right ventricle. A similar, but smaller lesion was suspected in the left ventricle visible in some vibratory phases. The vocal folds themselves were reddish and swollen with partially hyperplastic epithelium due to chronic laryngitis. The vocal fold vibrations showed a reduced amplitude and mucosal wave, but there was no impairment of vibrations by the supraglottic lesion. At microlaryngoscopy it was confirmed that the lesion was bilateral (Figure 1). The left-

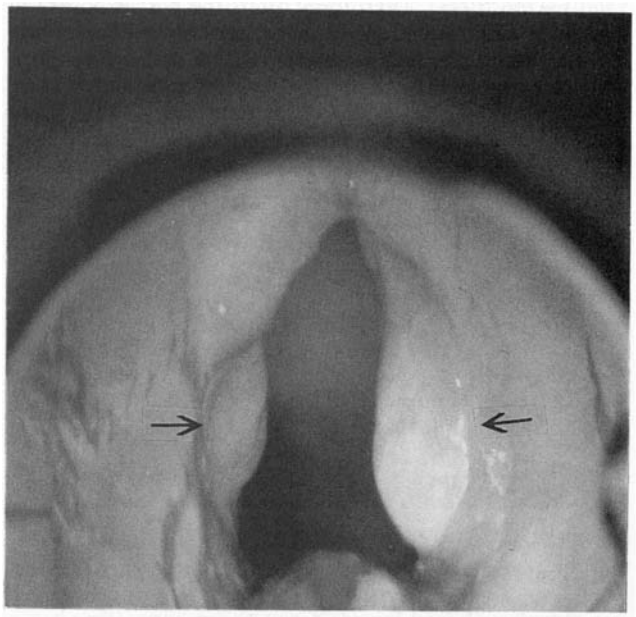

FIG. 1
Pre-operative larynx with bilateral ventricular neurofibroma (arrowed).

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sided lesion was hidden within the ventricle and was exposed when we retracted the ventricular fold. The polypoid masses were removed completely by an endolaryngeal approach and histopathological study confirmed a diagnosis of bilateral neurofibromas. Apart from the neurofibromas the vocal folds showed distinct signs of chronic laryngitis. Additional reports from the Dermatology and Ophthalmology clinics ruled out von Recklinghausen's disease. (No evidence for 'cafe au lait' macules in the skin or 'Lisch' nodules, which are pigmented hamartomas of the iris, present in over 90 per cent of patients with von Recklinghausen's disease was found). Follow-ups showed no recurrence and the patient does not have any problem at the end of first year, postoperatively. His hoarse voice was due to chronic laryngitis and post-operative stroboscopic findings remained unchanged.

Discussion

Laryngeal neurofibroma was first reported by Colledge (1930). New and Erich (1938) found only one NF out of 722 benign laryngeal lesions and Hollinger and Johnstone (1951) only one out of 1197. Kleinsasser (1987) pointed out that up to that year approximately two dozen cases had been reported.

Phang *et al.* (1987) reported the first laryngeal schwannoma originating from the ventricle. Neither a ventricular location nor bilateral ocurrence has ever been described for a neurofibroma in the available literature.

Neurofibromas are usually associated with von Recklinghausen's disease (multiple neurofibromatosis) and multiple in number, but may also occur as solitary tumours (Cummings *et al.*, 1969), as in our case. Clinical features include hoarseness, coughing or choking, noisy sleeping or stridor, weight loss (Stanley *et al.*, 1987), inspiratory dyspnoea and foreign body sensation during swallowing (Barnes and Ferlito, 1993). Only hoarseness due to chronic laryngitis was present in our patient.

Many types of surgical approach have been described (Aponte and Vicens, 1955; Schaeffer *et al.*, 1986; Barnes and Ferlito, 1993). Thyrotomy offers the best exposure in large tumours but requires a tracheostomy (Barnes and Ferlito, 1993). Sometimes extensive LNF makes laryngectomy inevitable (Mikell *et al.*, 1954; Aponte and Vicens, 1955). For selective small tumours, as in the case presented, endoscopic removal is considered appropriate. Malignant transformation does not occur in isolated neurofibromas (Enzinger and Weiss, 1995) and the outcome of surgery is excellent.

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