The Journal of Laryngology and Otology
December 1997, Vol. **111**, pp. 1171–1173

Asphyxial death caused by a laryngeal schwannoma: a case report

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Abstract
Neurogenic tumours of the larynx are unusual, with approximately 115 cases reported in the literature to date. Most of these lesions are benign, solitary submucosal nodules which present with hoarseness and are amenable to surgical resection. We present a case of a large pedunculated schwannoma arising in the aryepiglottic fold associated with sudden asphyxial death in an otherwise healthy young female.

Key words: Laryngeal neoplasms; Schwannoma

Introduction

Neurogenic tumours are an uncommon occurrence in the larynx and glottis, comprising less than 0.1 per cent of benign laryngeal neoplasms.

Approximately 115 cases of laryngeal neurilemmomas have been reported in the literature to date (Dekker and Haidar, 1994; Jamal, 1994; Ingels *et al.*, 1996). These tumours are usually encapsulated, submucosal and tend to arise in either the aryepiglottic folds or the true vocal folds (Ingels *et al.*, 1996). Given the common aryepiglottic location, the internal branch of the superior laryngeal nerve is the putative source for these neoplasms (Thomas and Rees, 1969; Takumida *et al.*, 1986). Most patients with a laryngeal neurogenic tumour present with either hoarseness of variable duration, dysphagia or shortness of breath. Malignant neurogenic tumours of the larynx are distinctly rare.

We describe a case of a large primary schwannoma of the larynx in a young woman associated with sudden death.

Case history

A 36-year-old white female presented to a primary care clinic with a three-day history of emesis and two days of diarrhoea accompanied by odynophagia and sinonasal congestion. Her last physician contact had been three years prior for complaints of dyspnoea and shortness of breath when in a recumbent position, with symptomatic relief when switching to a lateral position. This dysphoea was also associated with recurrent choking episodes while eating and a sense of 'tightness' when swallowing. She was otherwise healthy, and her only medication consisted of prenatal vitamins for a planned in-vitro fertilization. She was afebrile, normotensive and in no respiratory distress.

The patient was admitted to a community hospital with a diagnosis of dehydration most likely due to viral gastroenteritis. Laboratory studies at the time of admission were significant only for a pattern consistent with dehydration and an elevated leukocyte count. Six hours after admission and one hour after an unremarkable nurse visit she was found unresponsive. Resuscitative efforts were unsuccessful.

At autopsy, a large, pedunculated polyp was present arising from the right aryepiglottic fold (Figure 1). This ovoid lesion occluded nearly the entire glottis, measuring 4.4 × 2.2 × 2.0 cm in total. The stalk of the mass had a tapering configuration, about which the mass was mobile. The tumour was large enough to occlude the entire airway. The tumour had a smooth, glistening, gray-tan surface free of ulceration. The cut surface was purple-gray and slightly mottled without haemorrhage or necrosis.

The remainder of the post-mortem examination revealed no additional neoplastic lesions. No stigmata of neurofibromatosis were present. The remainder of the glottis and lower respiratory tract were unremarkable.

Post-mortem urine toxicology studies revealed no illicit or prescription substances or metabolites.

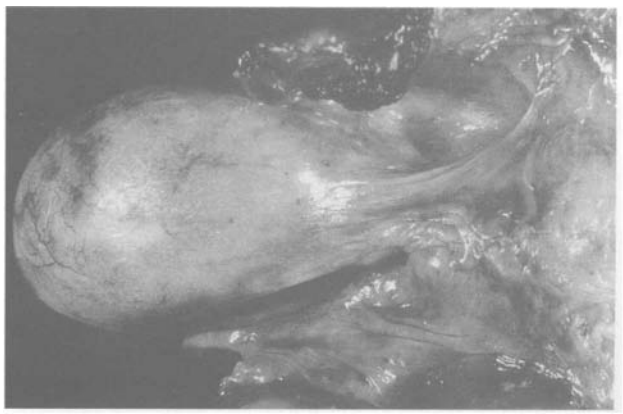

FIG. 1
Pedunculated schwannoma arising from the right aryepiglottic fold.

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Accepted for publication: 15 September 1997.

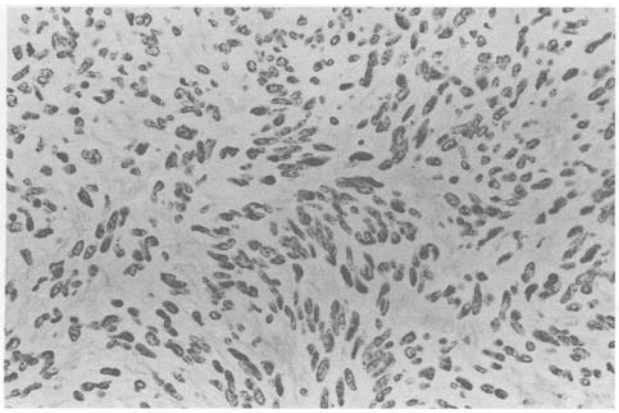

FIG. 2
A spindle cell neoplasm with highly cellular foci (H & E; × 100).

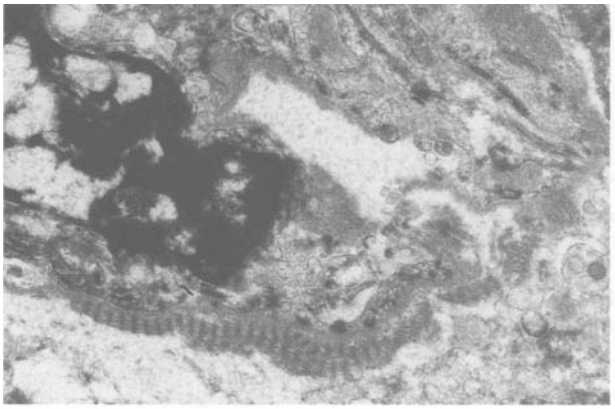

FIG. 4
Extracellular long-spacing collagen – Luse bodies (Electron microscopy × 12,000).

Histology

The encapsulated lesion was partially covered by fibrocollagenous tissue. The centre of the lesion was composed of a highly vascularized proliferation of spindle cells with hyalinized degenerative foci. The spindle cells (Figure 2) had a palisaded arrangement with Verocay bodies (Figure 3). No storiform pattern of growth, interlacing fascicles, or plexiform configuration was present. Mast cells were not present. Both cellular (Antoni A) and myxoid (Antoni B) elements were present in the mass. The individual cells strongly expressed S100 protein, and failed to express desmin, smooth muscle actin, CD68 (KP-1), synaptophysin, glial fibrillary acidic protein and HMB45.

Ultrastructural analysis revealed cells with a spindle shape and an enveloping basal lamina. Extracellular long-spacing collagen, or Luse bodies, were present in the matrix between the cells (Figure 4). Tight junctions were present at the surface of these cells, and the cells showed no surface microvilli or cytoplasmic intermediate filaments.

Discussion

Neurogenic tumours primary to the larynx include neurofibromas, schwannomas (neurilemmomas) and malignant neurosarcomas. Laryngeal examples of each of these lesions have been cited in the literature, however, the true frequency of benign tumours is difficult to ascertain as many of these reports use the term neurofibroma and neurilemmoma interchangeably in the text. The head and neck region accounts for 25–45 per cent of benign schwannomas (Jamal, 1994) with the larynx being an uncommon site.

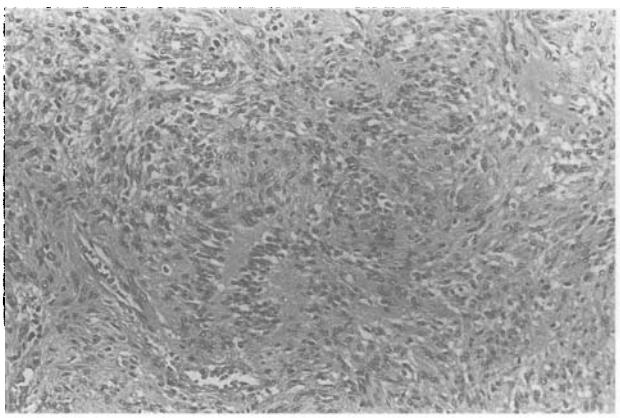

FIG. 3
Palisading Schwann cell nuclei, the so-called Verocay bodies (H & E; × 200).

Verocay first described what are now accepted as tumours derived from Schwann cells in 1908 (Whittam and Morris, 1970), terming them neurinomas. Stout recognized the Schwannian derivation shortly thereafter (Stout, 1949), and employed the term schwannoma for the same entity. The occurrence of these lesions may be associated with von Recklinghausen's disease, but most cases described are sporadically occurring lesions unrelated to neurofibromatosis.

The laryngeal presentation of neurogenic lesions is usually manifested as hoarseness, with no particular male or female predilection. These tumours occur over a wide age range. Laryngeal schwannomas are nearly always solitary, submucosal and circumscribed, and may be painful. Several reports in the literature (Thomas and Rees, 1969; Whittam and Morris, 1970; Palva *et al.*, 1975; Mehta, 1991; Newman and Youngs, 1992) cite dysphagia and/or shortness of breath as presenting symptoms of neurogenic laryngeal lesions. The clinical appearance may be similar to a laryngeal cyst or 'singer's nodule'. Laryngeal lipomas are rare entities which may also have a similar appearance at laryngoscopy due to their submucosal location. Computerized tomographic (CT) imaging may be of value in defining the size and extent of the neoplasm (Schaeffer *et al.*, 1986; Takumida *et al.*, 1986). Surgical resection is the treatment of choice (Paul-Murphy *et al.*, 1986; Takumida *et al*, 1986).

The histological pattern of an encapsulated lesion with both Antoni A and Antoni B growth patterns and S100 protein expression confirmed the diagnosis of a laryngeal schwannoma in this case. The ultrastructural Luse bodies lend support to this diagnosis. A neurofibroma would be expected to lack the alternating cellular (Antoni A) and myxoid (Antoni B) patterns, a capsule and Luse bodies.

Most patients with laryngeal schwannomas have complete resolution of symptoms and an uneventful course following surgical resection. The patient we describe had a three-year history of typical symptomatology, but no previous diagnosis, and died abruptly with a large, solitary, pedunculated lesion found in the aryepiglottic fold at autopsy. The pedunculated nature and aryepiglottic location likely resulted in a 'ball and valve' effect, with complete obstruction of the upper airway. No other visceral abnormalities were found at autopsy. Post-mortem toxicology studies revealed no illicit or prescription drugs present at the time of death to account for a decreased respiratory drive.

To our knowledge this is the first case of a benign schwannoma of the larynx associated with an asphyxial death due to upper airway obstruction. These rare tumours have a predilection for the supraglottis and may grow in such a manner as to obstruct the airway.

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