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Laryngeal amyloidosis with laryngocele

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Abstract
Both laryngocele and laryngeal amyloidosis are uncommon, and simultaneous occurrences of these entities are extremely rare. A case of laryngeal amyloidosis with laryngocele in which the computed tomography (CT) and magnetic resonance (MR) imaging of the larynx, clearly demonstrating both disease processes, is discussed. Diagnosis is confirmed by histopathologic specimens. Only two cases have been reported in the world literature, and this is the third case of laryngeal amyloidosis associated with laryngocele.

Key words: Larynx; Amyloidosis; Laryngocele

Introduction

Amyloid is a pathologic proteinaceous substance, deposited between cells in various tissues. Virchow (1851) was first to use the word amyloid to describe the substance (Virchow *et al.*, 1851). Borrow and Neuman (1873) first reported amyloidosis of the larynx. Since then, approximately 300 cases have been reported, the largest series of laryngeal amyloidosis consists of 235 patients (McAlpine and Fuller, 1964).

A laryngocele, as first described by Virchow in 1867, is abnormal dilatation of the laryngeal ventricle (Virchow, 1867). The association between laryngoceles and laryngeal carcinoma is well documented. The reported incidence of laryngoceles with squamous cell carcinoma varies between 4.9 per cent and 28.8 per cent (Birt, 1987; Close *et al.*, 1987; Celin *et al.*, 1991). However, the simultaneous appearance of laryngocele and laryngeal amyloidosis have been reported in two cases so far (Baker *et al.*, 1982; Schild *et al.*, 1983). We present here a case of laryngeal amyloidosis combined with laryngocele in which both disease processes were evaluated by CT and MR imaging.

Case report

A 54-year-old man presented with the complaint of increasing hoarseness and progressive dyspnoea lasting three years. Laryngoscopic examination showed a smooth, non-ulcerated, yellowish diffuse swelling of the epiglottis, aryepiglottic folds and vestibular folds (false cords), obstructing the laryngeal vestibule. CT revealed diffuse soft tissue thickening of the epiglottis, aryepiglottic folds and vestibular folds (false cords) associated with a mixed laryngocele on the left (Figure 1). MR images also revealed a mass at the same locality with a mixed laryngocele (Figure 2a, b).

The presence of amyloid was demonstrated in biopsies taken from the epiglottis and vestibular folds (false cords) (Figure 3). Complete laboratory evaluation including rectal biopsy excluded a systemic involvement. Supraglottic laryngectomy with laryngocele excision was performed. At 25 follow-up examination no recurrence was noted.

Discussion

Amyloidosis has been classified in a variety of ways, none of which is satisfactory. Current classification of the amyloidoses are based on the biochemical nature of the protein subunit (Chastonay and Hurliman, 1986; Lewis *et al.*, 1992). Type AL (immunoglobulin light chain-derived) is associated with primary systemic amyloidosis, myeloma-associated amyloid, and most localized forms of amyloid. Type AA (amyloid associated) is an immunoglobulin protein synthesized by liver. This protein is deposited in secondary amyloidosis. Several other biochemically distinct proteins have been found in amyloid deposits.

The larynx is the most common site of localized amyloidosis in the head and neck region, and constitutes less than one per cent of benign larygneal lesions.

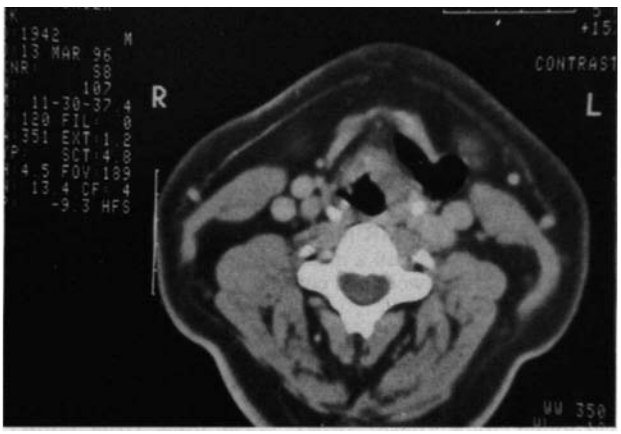

FIG. 1
Axial contrast CT scan of the larynx at the level of hyoid bone. Diffuse thickening of epiglottis and evidence of a laryngocele on the left.

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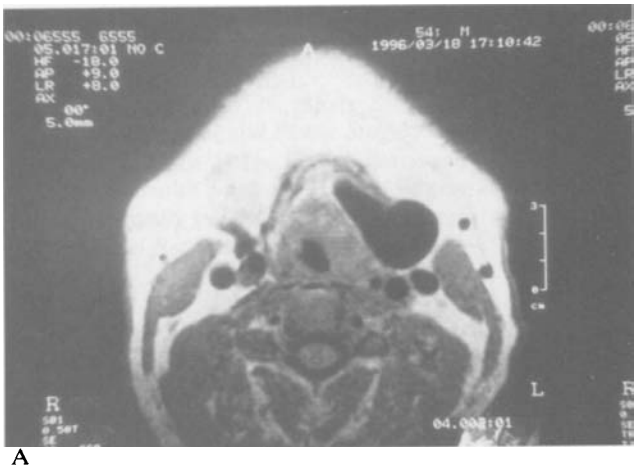

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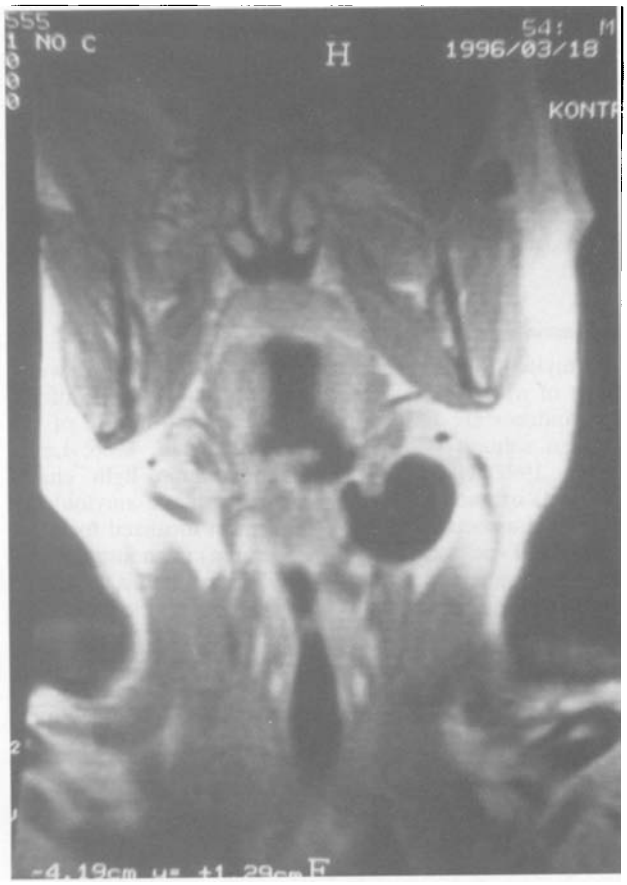

B

FIG. 2

a) T1-weighted axial MR image demonstrating thickening of the false vocal folds and a mixed of laryngocele on the left.
b) T1-weighted coronal MR image.

Localization of the lesion in the larynx is to the ventricle, vestibular folds (false cords), vocal folds (true cords), epiglottis, aryepiglottic folds, and subglottis in order of frequency (Barnes and Zafar, 1977; Mitrani and Biller, 1985). Hoarseness is the most common symptom. Progressive dyspnoea, haemoptysis and dysphagia are other presenting symptoms. The male to female ratio is 3:1 and the age range is eight to 80 years with a peak incidence in the fifth decade (McAlpine and Fuller, 1964; Barnes and Zafar, 1977).

FIG. 3

Amorphous amyloid material is seen (H&E; ×40)

The specific diagnosis of amyloidosis depends upon obtaining a tissue specimen by biopsy and demonstration of amyloid with appropriate stains. With the haematoxylin and eosin (H&E) stain and the light microscope, amyloid is seen as an homogeneous, acellular, eosinophilic, extracellular infiltrate. When a tissue specimen, stained with Congo red, is examined it reveals characteristic green birefringence. Because laryngeal amyloidosis can be a localized manifestation of systemic amyloidosis, some authors suggest an extensive work-up for systemic disease including abdominal subcutaneous fat pad aspiration or rectal biopsy, immunoelectrophoretic studies on serum or urine, chest and bone X-rays to reveal lytic lesions, serum calcium, urea nitrogen, creatinine, and uric acid levels. There should be a careful physical examination searching for tender bones and enlargement of the spleen and lymph nodes when amyloid is identified in the larynx (Barnes and Zafar, 1977; Mitrani and Biller, 1985). However, because the association of laryngeal amyloid with generalized amyloidosis is extremely rare some authors believe that an extensive work-up may not be necessary (Ryan *et al.*, 1977).

In the CT examination, amyloidosis is demonstrated as a marked thickening of the laryngeal structures with high density as measured with the Hounsfield numbers. On the other hand, the signal characteristics of amyloid on MR images resemble those of skeletal muscles. This may be an important differentiating point because muscle is an easy reference frame, and tumours do not appear in this manner on MR images (Gean-Marton *et al.*, 1991).

Squamous cell carcinoma is associated with occult laryngoceles in up to 28.8 per cent of patients (Close *et al.*, 1987). Micheau *et al.*, (1978) reported the carcinoma and laryngocele association to be 10 per cent, when the laryngectomy specimens were not selected according to site of cancer in the larynx. When only the supraglottic series were considered, the frequency of associated laryngoceles rose to 21 per cent. Because amyloidosis mainly involves the supraglottic larynx, laryngocele development might be expected more frequently. Our case was a diffuse supraglottic amyloid tumour with a mixed laryngocele. The question 'why laryngocele with amyloidosis is very rare' is still obscure. Perhaps in these cases the presence of laryngocele was not evaluated in detail.

The relationship between laryngocele and laryngeal carcinoma is speculative. Perhaps carcinoma contributes to the development of laryngoceles. For the majority of cases, laryngeal cancers are hypothesized to cause increased periods of intralaryngeal pressure. This is due to coughing and increased efforts in phonation and clearing secretions

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(Canalis, 1976; Close *et al.*, 1987). This increase in intralaryngeal pressure could then explain the development of ipsilateral, contralateral, or bilateral laryngoceles. Less frequently, a ventricular carcinoma is thought to form a valve-like mechanical obstruction leading to ipsilateral ventricular distention and laryngocele formation (Canalis, 1976; Holinger *et al.*, 1978). Although amyloid is not a tumour, its relationship with a laryngocele is much like a carcinoma.

The treatment of laryngeal amyloidosis is surgical incision. Systemic or intralesional corticosteroid application has not proven to be satisfactory neither has radiotherapy (Mitrani and Biller *et al.*, 1985). Since recurrence may occur very shortly after the operation it is essential not to leave residual tumour. Although some limited lesions can be excised endoscopically, extended cases require laryngofissure, supraglottic laryngectomy, or even total laryngectomy. Most authors suggest long-term follow-up due to the potential for recurrence (Barnes and Zafar, 1977; Mitrani and Biller, 1985; Lewis *et al.*, 1992; O'Halloran and Lusk, 1994; Tinaz *et al.*, 1994). Good results can be achieved with the use of the CO_2 laser (McIlwain and Shepperd, 1986; Talbot, 1990).

The treatment of laryngeal amyloidosis and laryngocele is surgical removal. The exact mechanism of laryngocele development in laryngeal amyloidosis is uncertain as in laryngeal carcinoma. In detection of the asymptomatic laryngoceles and predicting the extent of amyloid involvement within the larynx, CT and especially MR imaging are very sensitive.

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