Laryngeal amyloidosis: localized versus systemic disease

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OBJECTIVE

To study clinical and pathological characteristics, possibility of systemic disease, and effect of local therapy in patients with laryngeal amyloidosis.

PATIENTS

Records of all patients with localized laryngeal amyloidosis in a single tertiary referral center were examined retrospectively at diagnosis and after local therapy. Out of 188 new patients with amyloidosis between 1990 and 2003, five patients had localized laryngeal amyloidosis. One of these patients (*) already had ocular amyloidosis without systemic involvement. A sixth patient (#) known elsewhere with localized laryngeal amyloid turned out to have systemic AL amyloidosis 8 years later. This patient was added tot the study group. Patient characteristics are listed in table 1.

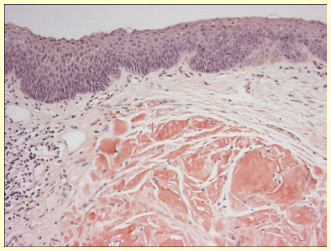

Figure 1. Congo red stained amyloid deposit

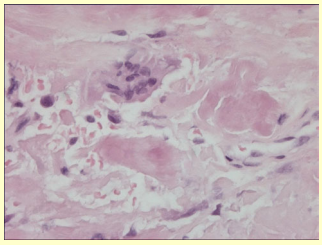

Figure 2. Giant cells at the border of amyloid deposits

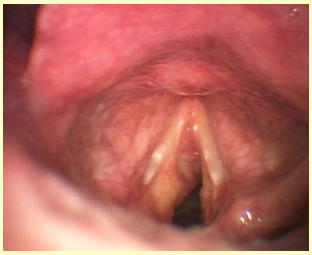

Figure 3. Glottic amyloidosis

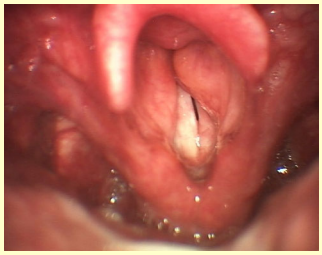

Figure 4. Supraglottic amyloidosis

Table 1.
Characteristics at initial presentation of the patients with localized laryngeal amyloidosis

	gender	anatomical characteristics		hoarseness					medical history of amyloidosis	
		location	site	G	R	B	A	S	age at first laryngeal treatment	follow-up since (years)
	male	supraglottis	unilateral right	3	3	3	0	2	24	5
*	female	supraglottis and glottis	unilateral left	2	0	2	2	1	41	1
	female	glottis	bilateral	3	3	2	0	3	42	7
	male	glottis	bilateral	3	3	3	0	3	50	5
	male	supraglottis	unilateral right	3	3	3	0	0	57	3
#	male	supraglottis	bilateral	3	2	3	0	3	46	13

G = overall grade; R = roughness; B = breathiness; A = asthenicity; S = strained quality

RESULTS

Ratio of free light chains was abnormal in two patients: the sixth patient with systemic amyloidosis (#) and the patient already known with ocular amyloidosis (*). Histological examination showed giant cells at the peripheral margins of amyloid in two patients (fig. 1 and 2). Immunohistology helped to exclude AA amyloid, confirmed the presence of SAP, but failed to detect specific reactivity for either kappa or lambda light chain. The most important indications for treatment were dramatic voice changes, objectified by phonetograms, and decreased tolerance of exertion. Amyloid interfering with laryngeal or airway function was removed during microlaryngoscopy with CO_2-laser or cold endoscopic excision. The best results were seen when glottic deposits (fig. 3) were removed by cold endoscopic excision and supraglottic deposits (fig. 4) were removed by CO_2-laser. Four patients showed recurrent disease, requiring surgery in only one patient after 7 months.

CONCLUSIONS

- **Laryngeal amyloidosis is a slowly progressive disease, which should be treated depending on the complaints of the patient.**
- **Stabilization often occurs after a number of years.**
- **A systematic work-up, including measurement of free light chains, helps to detect systemic disease.**
- **Follow-up should be yearly for at least 10 years.**