



Inflammatory and Autoimmune Disorders of the Larynx

Since the larynx is the crossroads between gastrointestinal and respiratory tracts it has a significant immunologic role. Due to its immune activity, the larynx participates in several inflammatory and autoimmune disorders.

Anatomy & Physiology

- Immune anatomy (compartmentalization) of the larynx: supraglottis, glottis and subglottis
- Laryngeal-associated lymphoid tissue (LALT): primarily located in the supraglottis
 - Thibeault S, et al: At the Crossroads: Mucosal Immunology of the Larynx. *Mucosal Immunol.* 2009 Mar; 2(2): 122–128. PMID: PMC2666820

Granulomatosis with polyangiitis (GPA)

- Formerly known as Wegener's granulomatosis. 10-20% of patients with GPA have subglottic involvement. Airway disease is frequently independent of the systemic disease. Therapy is systemic, yet often a localized complementary treatment is necessary.
 - Fijolek J, et al. Intratracheal Dilation-injection Technique in the Treatment of Granulomatosis with Polyangiitis Patients with Subglottic Stenosis. *J Rheumatol.* 2016 Nov;43(11):2042-2048. PMID: 27633822.

Amyloidosis

- Abnormal formation and deposition of amyloid protein fibers. Although rare, laryngeal amyloidosis is the most common amyloidosis manifestation within the head and neck. Laryngeal amyloidosis is usually a localized primary disease, yet it may be associated with an underlying disorder such as, multiple myeloma.
 - Pribitkin E, et al. Amyloidosis of the upper aerodigestive tract. *Laryngoscope* 113:2095–2101, 2003. PMID: 14660909.
 - Send T, et al. Amyloidosis of the Upper Aerodigestive Tract: Management of a Rare Disease and Review of the Literature. *Dysphagia.* 2019 Apr;34(2):179-191.
 - Rudy SF et al. Clinical characteristics of laryngeal versus nonlaryngeal amyloidosis. *Laryngoscope.* 2018 Mar;128(3):670-674.



Sarcoidosis

- A systemic granulomatous disease of undetermined etiology with a variable clinical presentation. The classical appearance of laryngeal sarcoidosis is smooth submucosal swellings of supraglottic structures.
 - Plaschke CC et al. Clinically isolated laryngeal sarcoidosis. *Eur Arch Otorhinolaryngol.* 2011 Apr;268(4):575-80. PMID: 21132317
 - Morgenthau AS et al. Sarcoidosis of the upper and lower airways. *Expert Rev Respir Med.* 2011 Dec;5(6):823-33. PMID: 22082167

Relapsing Polychondritis

- Episodic inflammation of cartilaginous structures throughout the body, involving the cartilage of the ears, nose, larynx, tracheobronchial tree, eyes, and heart's connective components. The clinical spectrum varies from intermittent inflammatory episodes leading to cartilage deformities to life-threatening airway collapse or valvular regurgitation.
 - Longo L, et al. Relapsing polychondritis: A clinical update. *Autoimmun Rev.* 2016 Jun;15(6):539-43. PMID: 26876384.
 - Vitale A, et al. Relapsing Polychondritis: an Update on Pathogenesis, Clinical Features, Diagnostic Tools, and Therapeutic Perspectives. *Curr Rheumatol Rep.* 2016 Jan;18(1):3. PMID: 26711694.

Rheumatoid nodules/Bamboo nodes

- Whitish transverse submucosal lesions within the vocal folds, commonly presents with dysphonia. Associated with various autoimmune diseases.
 - Schwemmle C, et al. Bamboo nodes associated with mixed connective tissue disease as a cause of hoarseness. *Rheumatol Int.* 2013 Mar; 33(3): 777–781. PMID: 22083614.
 - Murano E, et al. Bamboo node: primary vocal fold lesion as evidence of autoimmune disease. *J Voice.* 2001 Sep;15(3):441-50. PMID: 11575639.



Pemphigus

- Autoimmune disease of skin and mucous membranes. In the larynx, supraglottic involvement is the most common. Presents with painful erosions, typical appearance of whitish exudates or erythematous patches.
 - Ohki M et al. Nasal, oral, and pharyngolaryngeal manifestations of pemphigus vulgaris: Endoscopic ororhinolaryngologic examination. *Ear Nose Throat J.* 2017 Mar;96(3):120-127. PMID: 28346642
 - Hale EK et al. Laryngeal and nasal involvement in pemphigus vulgaris. *J Am Acad Dermatol.* 2001 Apr;44(4):609-11. PMID: 11260534

Chapters in Flint, PW, & Cummings, CW. *Cummings otolaryngology head & neck surgery* 6th Ed. (2015):

* Chapter 13: Laryngeal and Tracheal Manifestations of Systemic Disease.

* Chapter 62: Acute and Chronic Laryngitis.