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# A Case of Hypopharyngeal and Gastric Amyloidosis

KEYWORDS: Aerodigestive, amyloidosis, hypopharyngeal, dysphagia

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**ABSTRACT** *Superior aerodigestive tract amyloidosis is a rare entity mainly involving the larynx and tongue. A hypopharyngeal location is exceptional. This patient had a spectrum of many symptoms, signs of upper aerodigestive tract. Any upper Dysphagia should be fully explored, notably for malignant pathology. Laryngeal involvement tends to be isolated, while the tongue is frequently involved in a systemic AL amyloidosis with associated multiple myeloma. Phayrngeal amyloidosis is rare. Localized amyloidosis, although rare, may be implicated. Hypopharyngeal involvement is extremely rare, one type-AL hypopharyngeal amyloidosis secondary to multiple myeloma and one primitive localized amyloidosis. Management of aerodigestive tract amyloidosis requires prior identification of the cause.*

**Introduction**
Superior aerodigestive tract amyloidosis is a rare entity mainly involving the larynx and tongue. A hypopharyngeal location is exceptional. Superior aerodigestive tract amyloidosis may be associated with systemic amyloidosis and/or with myeloma or be isolated 1,2,3

We report a case of hypopharyngeal involvement, and review the epidemiological, clinical, therapeutic and evaluative features of this pathology.

**Case Report**
A 75yr old female presented in September 2009 with hoarseness of voice, loss of appetite and loss of weight since three months. Patient referred to private doctor and underwent routine blood tests, upper GI endoscopy, USG abdomen and pelvis. She was diagnosed to be having reflux esophagitis and was treated for the same. But her symptoms progressed and she developed difficulty in swallowing. She again consulted doctors in August 2010.

**On examination:**
She had pallor and cachexia, Body wt 43kg.Height 150 cm.BMI of 19.11 kg/m2. Pulse rate of 70/min, BP 130/70 mm Hg, laryngeal crepitus absent. Systolic murmur in the mitral area.

Hb-10g%, TC-7200 cells/mm3,DC-N66, L25, E04, M05Peripheral smear: Microcytic hypochromic anemia, Blood group- O positive FBS- 85 mg% PPBS-106 mg%,

**Renal function:** Blood.Urea- 36 mg%, S.Creatinine-0.8 mg%, Vit B12 assay- 183 pg/ml,

**LFT:** SGOT-21 U/L, SGPT-24 U/L, alkaline phosphatase-81 U/L, Total protein-6.5 g%, albumin- 3.5 g%, globulin-3g%,

**Lipid profile:** Total triglycerides-152 mg%, total cholesterol-178 mg%, HDL cholesterol- 42 mg%, LDL cholesterol- 106 mg%, VLDL cholesterol- 30 mg%, total cholesterol: HDL cholesterol ratio 4.2: 1.

Urine routine within normal limits.

Chest X-ray- no abnormality detected.

USG abdomen and pelvis- no abnormality detected.

ECG- sinus rhythm, right axis deviation, poor progression of R waves.

**Serum immunofixation:**
**Free light chains, Free kappa light chains:** 13.3 mg/L, Free lambda light chains: 247 mg/ L, Kappa/ Lambda ratio: 0.054

**Iron profile: Total iron:** 48 mcg/dl,Total iron binding capacity: 388 mcg/dl, Unbound iron binding capacity: 340 mcg/dl, Serum transferrin: 272 mcg/dl, Transferrin saturation: 12.37%, Ferritin: 9.09 mcg/dl. Protein electrophoresis: normal pattern

**Urine electrophoresis:** no band seen- negative for Bence Jones proteins.

**Urine for immunofixation:** negative for light chains.

**Upper GI endoscopy on 06-08-2010**
Proliferative lesion at hypopharynx. Multiple small ulcers around the pylorus. Mucosa is indurated in appearance. Biopsy taken from hypopharynx and gastric antrum. Differential diagnosis: ? ca. hypopharynx, gastric ulcer, antral carcinoma.

**HPE report suggestive of amyloidosis.**
Biopsy from hypopharynx shows stratified squamous epithelium, sub epithelium shows amorphous eosinophilic material deposited in blood vessel wall and in masses.Biopsy from gastric antrum shows gastric mucosa with lamina propria displaying amorphous eosinophilic material.Congo red stain positive with green birefringence in polarised light.

**Discussion**
Amyloidosis is a benign pathology of progressive evolution, featuring intracellular and extracellular deposits of an amyloid substance, which is the final phase in the metabolism of certain proteins. AL amyloidosis ("A" for amyloidosis, and "L" for light chains) comprises immunoglobulin kappa and lambda light chains, and is frequently secondary to lymphoplasmacytic disorders such as lymphoma or multiple myeloma [1-2]. It may involve any head and neck location, including the orbit, sinuses, salivary glands, pharynx and larynx, and most frequently the larynx or tongue. Laryngeal involvement tends to be isolated, while the tongue is frequently involved in a systemic AL amyloidosis with associated multiple myeloma[4].

Pharyngeal involvement is rare, with only 13 cases reported in the literature. Ten of these were localized and isolated, with the other three being part of a systemic amyloidosis[3,5].

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Hypopharyngeal involvement is extremely rare: extensive search of the literature retrieved only two cases, one type-AL hypopharyngeal amyloidosis secondary to multiple myeloma and one primitive localized amyloidosis [6, 7].

Clinical symptomatology and imaging are poorly specific for hypopharyngeal involvement. The lesion aspect is of a homogeneous submucosal mass with only slight if any contrast uptake. There is sometimes perilesional calcification [8,9].

Diagnosis is anatomopathological: the deposits are extracellular and pathognomic under Congo red staining [10].

Chemotherapy associating melphalan and prednisone is the usual first line treatment, but with efficacy of no more than 40 to 60% [1-5]. In secondary forms, treatment should target the underlying disorder in the relevant organs; in case of persistence of symptoms caused by the amyloid deposits, surgery may be offered.

Surgical management of an amyloidosis site is indicated only in case of complications such as hemorrhage, stenosis or persistent obstruction. Laser endoscopy has been reported in connection with a laryngeal location; for other aerodigestive tract sites, external surgery is required. Prognosis is fairer in localized than in systemic forms; 5-year survivorship in the latter is around 16% [10]. Prognosis in AL amyloidosis, especially when myeloma is associated, is poor [10]

**Conclusion**

Any upper dysphagia should be fully explored, notably for malignant pathology. Localized amyloidosis, although rare, may be implicated. Management of aerodigestive tract amyloidosis requires prior identification of the cause.

**Conflict of interest**

The authors communicated no conflict of interest.

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