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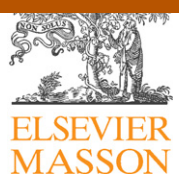

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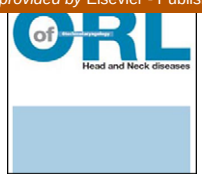

CLINICAL COMMENTARY

Hypopharyngeal amyloidosis: A case report

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KEYWORDS
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Dysphagia

Summary Hypopharyngeal amyloidosis is rare. Management depends on etiology. We report a case of hypopharyngeal amyloidosis and review the characteristics of this exceptional pathology. A 60-year-old woman with a history of diabetes and chronic cervicalgia consulted for dysphagia and deteriorated general health status, which had been evolving for 2 months. Clinical examination found two ulcerations of the lateral edge of the tongue and right pyriform sinus salivary stasis. Panendoscopy found regular swelling of the posterior wall of the hypopharynx and cervical esophagus. The pyriform sinuses and larynx were normal. Cervical CT and MRI showed thickening of the posterior wall of the hypopharynx. Biopsy found amorphous acellular eosinophil interstitial deposits, shown to be amyloid on Congo red staining, leading to a diagnosis of amyloidosis. Etiological assessment pointed to myeloma. The patient was managed by chemotherapy associating melphalan and prednisone. Evolution at 12 months' follow-up was good. Localized amyloidosis is a rare lesion of the superior aerodigestive tract, predominating in the larynx. Hypopharyngeal involvement is exceptional. Diagnosis is histological. Management depends on etiology. Local treatment is exceptional other than in case of complication. Systemic forms with associated myeloma are of poor prognosis.

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—3].

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

Observation

A 60-year-old diabetic woman, followed in Rheumatology for chronic cervicalgia and under medical treatment, consulted for upper dysphagia to solids, progressively evolving over a 2-month period and associated with generally deteriorated health status. There was neither dyspnea nor dysphonia, blood-tinged sputum or symptoms of gastro-esophageal reflux.

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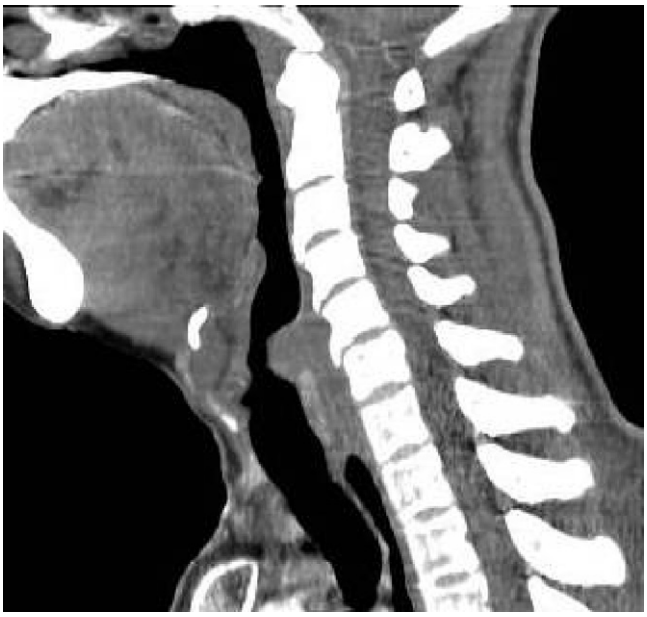

Figure 1 Cervical CT-scan, sagittal slice. Posterior hypopharyngeal wall tissue mass extending to isodense esophagus mouth.

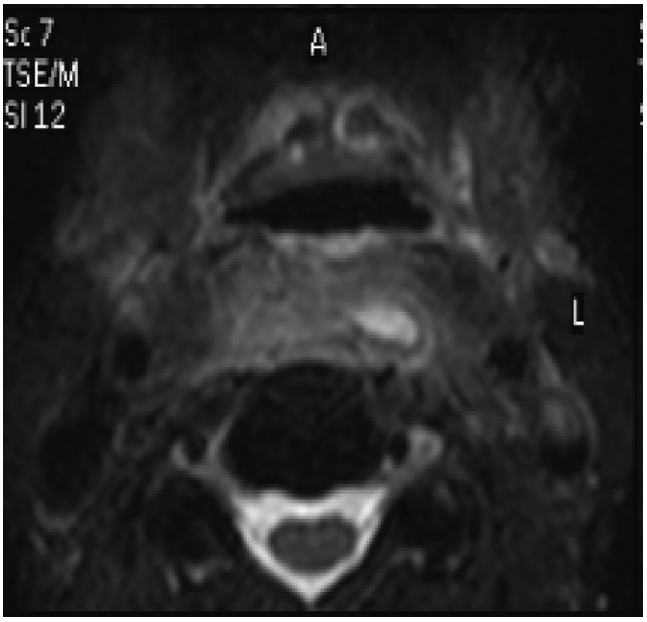

Figure 2 T1 cervical MRI with gadolinium injection, axial slice. Hypopharyngeal mass with irregular lateral contour and variable contrast uptake.

On clinical examination, the patient was thin and pale. Examination of the mouth found very poor buccodental hygiene with two ulcerations of the lateral edge of the tongue without underlying induration. The oropharynx was otherwise normal.

Nasofibroscopy found salivary stasis in the right pyriform sinus, with no suspect lesion in the larynx and conserved vocal fold mobility. Cervical examination found no palpable adenopathy; the thyroid was non-palpable. No further abnormalities appeared; there was no visceromegaly or extra-cervical adenopathy.

Cervical CT scan showed a 32 mm × 17 mm tissular process in the posterior wall of the pharynx, extending to the mouth of the esophagus (Fig. 1), with slight contrast medium uptake.

Magnetic resonance imaging (MRI) found a medial prevertebral mass extending from the oropharynx to the mouth of the esophagus, in T2 hypersignal and T1 hyposignal, with variable contrast uptake after gadolinium injection (Fig. 2). The mass showed fuzzy, irregular lateral contours, without involving the prevertebral muscles. The larynx and cervical esophagus showed no abnormality.

Panendoscopy under general anesthesia found the pyriform sinuses to be free, with regular swelling with normal mucosa of the posterior wall of the hypopharynx extending to the mouth of the esophagus, which was blocked. Laryngoscopy found no abnormality. Deep biopsies were performed in the hypopharynx and tongue ulcerations. A gastric tube was installed at end of surgery to manage denutrition with an adapted hypercaloric diet.

Anatomopathology of the hypopharyngeal biopsy sample found amorphous acellular interstitial deposits, marked by Congo red, showing yellow-green double refraction under polarized light, as is typical in amyloidosis, with no neoplastic cells or tuberculoid granuloma. Immunohistochemistry diagnosed type-AL amyloidosis (Figs. 3 and 4). The tongue biopsies were negative.

In the amyloidosis assessment, blood count and typing found normochromic normocytic anaemia without abnormality in the platelet or red or white blood-cell lines. Biological analysis found an inflammatory syndrome (SR = 97, CRP = 28). Liver and kidney assessments were normal. Immunoelectrophoresis showed 2 monoclonal peaks of Ig G and kappa light chains. Sternal puncture found medullary invasion with 66% plasmocytes. The diagnosis was of AL-type amyloidosis with associated multiple myeloma.

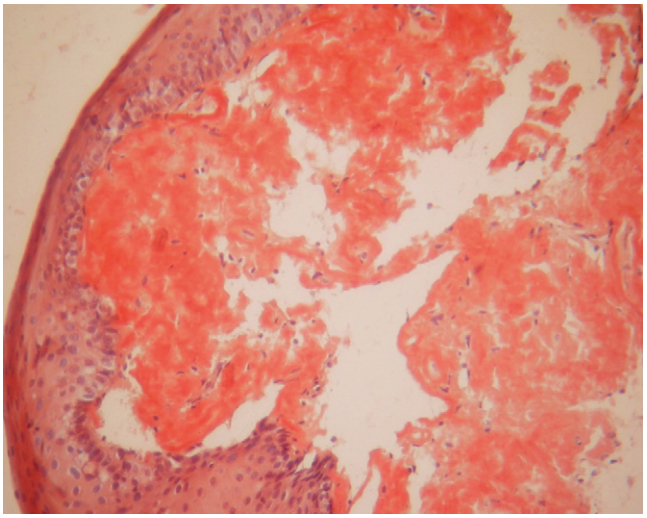

Figure 3 Histology: flaky deposit in lamina propria and vascular walls, stained by Congo red: amyloid deposits (Congo red × 400).

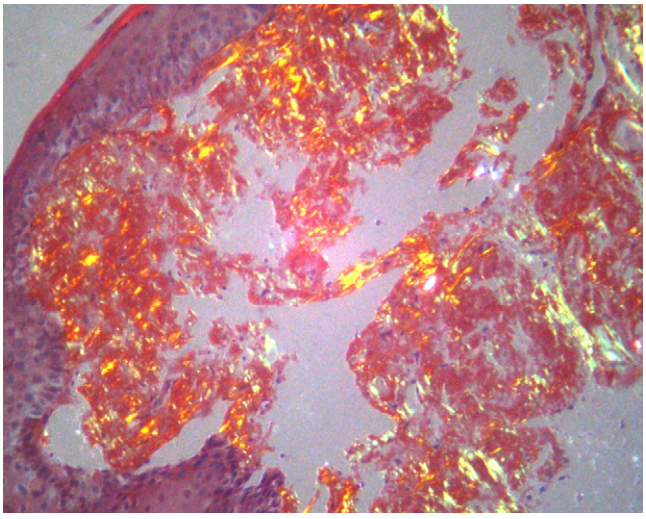

Figure 4 Histology: yellow-green double refraction, typical of amyloidosis (Congo red × 400).

For the first 4 days of each of six courses, chemotherapy associated melphalan (14.5 mg/day) and prednisone (116 mg/day).

Evolution at end of treatment showed partial improvement in dysphagia. Pharyngoesophageal surgery was not required. Resumption of nutrition and weight gain lasted for the 12 months before the patient was lost to follow-up.

Discussion

Amyloidosis is a benign pathology of progressive evolution, featuring intratissular 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 [2,4].

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]. Hypopharyngeal involvement is extremely rare: extensive search of the literature retrieved only two cases, one type-AL hypopharyngeal amyloidosis secondary to multiple myeloma [6] and one primitive localized amyloidosis [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 [1,10].

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 [1,11].

Surgical management of an amyloidosis site is indicated only in case of complications such as hemorrhage, stenosis or persistent obstruction [10]. Laser endoscopy has been reported in connection with a laryngeal location [3,4,7]; 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% [7,10].

Prognosis in AL amyloidosis, especially when myeloma is associated, is poor [12].

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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