

## CASE STUDY

# Amyloidosis of the Nasopharynx<sup>☆</sup>

## Amiloidosis localizada en nasofaringe

Elena Sánchez Legaza,<sup>a,\*</sup> Concepción Cervera Oliver,<sup>b</sup>  
Rosario Guerrero Cauqui<sup>c</sup>

<sup>a</sup> Servicio de ORL, Hospital de Algeciras, Cádiz, Spain

<sup>b</sup> Servicio de ORL, Hospital San Rafael, Cádiz, Spain

<sup>c</sup> Servicio de Anatomía Patológica, Hospital de Algeciras, Cádiz, Spain

Received 12 November 2011; accepted 13 February 2012

## Introduction

The term “amyloidosis” encompasses various clinical entities of unknown aetiology, characterised by extracellular deposit of amyloid (acellular protein material that presents yellowish-green birefringence under polarised light after Congo red staining).

Clinical signs and symptoms depend on the anatomic distribution and intensity of the amyloid deposit. There are 2 main forms: systemic and localised. Amyloidosis can present as a diffuse process or as a nodular mass, simulating a neoplasia, called amyloid tumour, amyloidosis tumoral or amyloidoma.<sup>1</sup>

To diagnose it, the presence of amyloid must be demonstrated. Findings on CAT scans are unspecific.

It is important to determine whether the amyloidosis is systemic or localise, given the different prognosis for each. Systemic amyloidosis has poor life expectancy (especially if

it is associated with myeloma). In contrast, if the localised form is diagnosed early and given appropriate treatment, the quality of life can improve dramatically, prolonging survival.

## Clinical Case

We present the case of a 43-year-old male patient without relevant history and lacking toxic habits, who presented continual moderate nasal impairment, sensation of a foreign body upon swallowing, and mucopurulent rhinorrhoea for the last year and a half. There was also epistaxis upon coughing strongly. The patient did not report nasal puritus, sneezing attacks, fever, asthenia or weight loss.

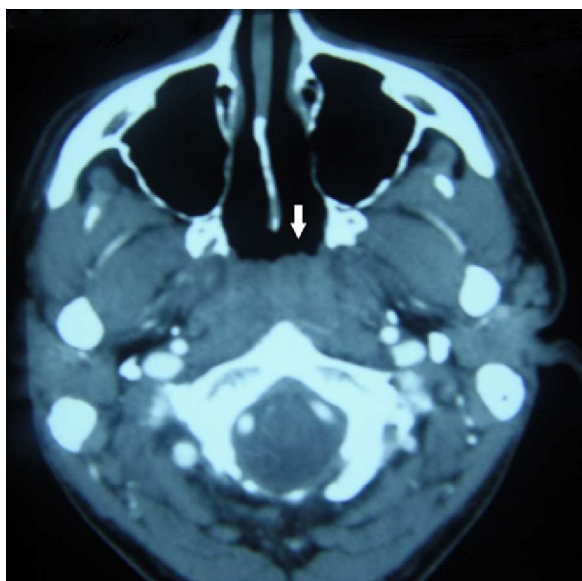
Otoscopy was normal, while the endoscopic examination revealed a rounded mass with a polypoid appearance, except a wine-red ulcerated area beginning under the fossa of Rosenmüller from left nasal fossa to uvula. A slightly polypoid mass in cavity was observed on CAT scan (Fig. 1).

This mass was removed through nasal endoscope surgery under local anaesthesia, being reported as type AL localised amyloidosis (Fig. 2), after ruling out secondary amyloidosis. No recurrence has occurred in the 2 years since the operation.

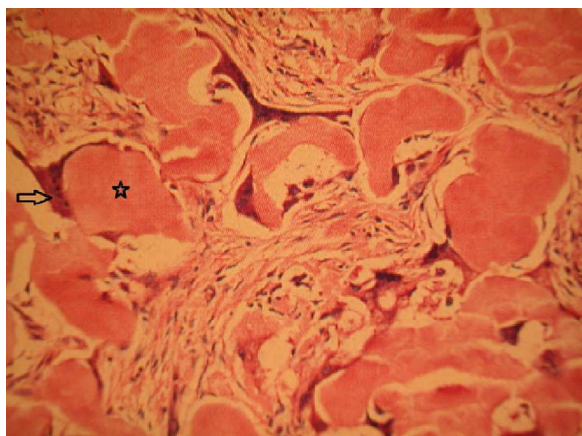
<sup>☆</sup> Please cite this article as: Sánchez Legaza E, Cervera Oliver C, Guerrero Cauqui R. Amiloidosis localizada en nasofaringe. Acta Otorrinolaringol Esp. 2013;64:377–378.

\* Corresponding author.

E-mail address: [manpro1910@hotmail.com](mailto:manpro1910@hotmail.com) (E. Sánchez Legaza).



**Figure 1** CAT: well-defined, non-vascularised mass in cavity, without infiltrating adjacent planes.



**Figure 2** Microscopic image of red-stained amorphous eosinophilic material, amyloid (\*), surrounded by multinucleated giant cells (arrow).

## Discussion

Localised amyloidosis in the head and neck and gastrointestinal tract is a rare condition, in which there is predominance of the localised AL type form (lacking evidence of previous disease or secondary to myeloma, in contrast to AA amyloidosis or reactive to inflammatory and tumour pathology). It has been described in several sites, but it predominates in the larynx, pharynx, parotid and tongue.<sup>2</sup>

Amyloidosis in nasopharynx usually surfaces in adults, in the 5th to the 7th decade of life, with no preference as to gender. Nevertheless, it has been reported in an 8-year-old child.<sup>3</sup>

Clinical signs and symptoms usually include nasal obstruction together with oral breathing, recurrent epistaxis, haematemesis and ear pathology secondary to tubaric dysfunction. At the level of the nostrils, it is implanted in the middle turbinate, although it can appear as a septal perforation.<sup>4</sup> Some patients present adenoid hypertrophy.<sup>5</sup> Many authors have described its haemorrhagic potential, attributing this to capillary fragility secondary to the infiltration by the amyloid deposit, although Kurgan et al.<sup>6</sup> described factor V and VII deficiency.

They are generally considered benign tumours, as they are slow-growing lesions, without bone destruction, intracranial extension and lymphadenopathy; however, they can be locally aggressive, producing osteolysis and extension to the base of the skull.<sup>7</sup>

The diagnosis is histological, with it being important to determine if the deposits are localised or systemic using fine needle aspiration of the fat of the anterior abdominal wall.

Immunohistochemically, it can show AL, AA and AP proteins or chain restriction, although it shows the AL type in the localised forms.<sup>8</sup> Consequently, it is recommendable to use a panel of antibodies specific to all the types of amyloid precursors, in addition to the specific search for the population of abnormal plasma cells in the deposit site, together with a study on the monoclonal production of serum immunoglobulins.<sup>9</sup>

The treatment of choice is surgical extirpation using conventional surgery or CO<sub>2</sub> laser. Relapses are common, if the lesion is not removed completely; however, given the lack of case studies and the slow development of the process, spontaneous regression can occur but it is difficult to predict the result.<sup>10</sup> Radiotherapy is contraindicated.

## References

1. Kyle RA, Bayrd ED. Amyloidosis: review of 236 cases. *Medicine*. 1975;54:271–99.
2. Wacrenier A, Fayoux P, Augusto D, Aubert S, Leroy X. Amylose localisée du cavum. *Press Med*. 2003;32:114–6.
3. Dominguez S, Wienberg P, Claros P, Vila J. Primary localized nasopharyngeal amyloidosis. A case report. *Int J Pediatr Otorhinolaryngol*. 1996;36:61–7.
4. Mufarrij AA, Busaba NY, Zaytoun GM, Gallo GR, Feiner HD. Primary localized amyloidosis of the nose and paranasal sinuses. *Am J Surg Pathol*. 1990;14:379–83.
5. Strunski V, Goin M, Guillem I. Amylose du cavum. A propos d'une observation. *Ann Oto-Laryng (Paris)*. 1991;108:49–51.
6. Raymond AK, Sneige N, Batsakis JG. Pathology consultation. Amyloidosis en the upper aerodigestive tracts. *Ann Otol Rhinol Laryngol*. 1992;101:794–6.
7. Hegarty J, Rao VM. Amyloidoma of the nasopharynx: CT and MR findings. *Am J Neurol Rs*. 1993;14:215–8.
8. Panda NK, Sharma SC, Mann SB, Radotra BD. Localized amyloidosis of the nasopharynx. *Ear Nose Throat J*. 1994;73:335–6.
9. Mozos A, Caballero M, Solé M, Candesa A. Amiloidosis del CAE. *Acta Orl Esp*. 2011;62:392–4.
10. Lim JS, Lebowitz RA, Jacobs JB. Primary amyloidosis presenting as a nasopharyngeal mass. *Am J Rhinol*. 1999;13:209–12.