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Amyloidosis in ENT Presentation of a Case

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Case Report

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Abstract

Amyloidoses are a heterogeneous group of rare diseases that consist of the abnormal folding of a precursor protein that ends up being deposited in the form of fibrillar structures in various organs and systems.

It can present as a hereditary or isolated disease, localized or systemic, primary (idiopathic) or secondary to infectious processes such as tuberculosis, leprosy and osteomyelitis or chronic inflammatory processes such as rheumatoid arthritis.

Within the clinical manifestations, vague symptoms and signs appear, depending on the location and size of the deposits, since the heart, kidneys, liver, gastrointestinal tract, peripheral nervous system, lungs, soft tissues, etc. may be affected.

Therefore, it is of utmost importance that once the diagnosis is confirmed, a systemic study is performed to evaluate the extensión of the disease to other possible target organs.

We present the case of a 59-year-old man, with a lesion in the upper pole of the right tonsil, positive for amyloidosis.

Objective: Present the case of amyloidosis in an extremely rare location, for the dissemination and knowledge of the disease.

Keywords: Amyloidosis; Pathology; Palatine Tonsils

Clinical Case

A 59-year-old man, with no significant history, with a single polyp-type lesion, of three months' duration, in the upper pole of the rest of the right palatine tonsil, approximately 5 mm, pink, pedunculated, with no signs of local infection, asymptomatic. Both palatine tonsils were practically atrophic and no further lesions were found in the head and neck.

It was decided to perform exeresis under local anesthesia, with a histopathological report of: fibroepithelial polyp with amyloid deposits (positive Congo Red stain).

The patient is currently asymptomatic.

A comprehensive evaluation was requested by the Internal Medicine service to rule out involvement of target organs.

Development

There are many different proteins in our body that can misfold and cause amyloidosis. Amyloidoses are a heterogeneous group of rare diseases that consist of the abnormal folding of a precursor protein that ends up being deposited in the form of fibrillar structures in various organs and systems (heart , kidney, liver, intestine, nervous system, etc.) altering its functioning. These fibrils are identified in biopsy samples by apple-green birefringence with Red staining Congo, when viewed under polarized light [1].

The different types of amyloidosis can be classified according to the precursor protein involved and a system of abbreviations is used, where the prefix "A", which refers to amyloids, is followed by the abbreviation of the affected protein: AL chain antibody-derived amyloids light; AA serum amyloid protein A, and ATTR designates transthyretin amyloid, for example [2].

More than 36 proteins have been described that are involved in the appearance of amyloidosis variants, but the most common are AL amyloidosis (due to immunoglobulin light chains), ATTR (due to transthyretin instability due to mutations or associated with age) and AA (associated with chronic inflammatory disorders) [3].

It is essential to correctly identify the type of protein responsible for the amyloid deposit, since the treatment depends on this. For the identification of the protein, there are highly specific techniques such as mass spectrometry and immunohistochemistry with electron microscopy [4].

It can present as a hereditary or isolated disease, localized or systemic, primary (idiopathic) or secondary to infectious processes such as tuberculosis, leprosy and osteomyelitis or chronic inflammatory processes such as rheumatoid arthritis [5].

Within the clinical manifestations, vague symptoms and signs appear, depending on the location and size of the deposits, since the heart, kidneys, liver, gastrointestinal tract, peripheral nervous system, lungs, soft tissues, etc. may be affected [6].

In the head and neck, it is a very rare entity, the most frequent site of presentation is the larynx and extremely rarely, amyloid deposits can be found in the tonsils [7].

The prognosis is usually good in localized forms, unlike the systemic form which has demonstrated a rate of 5 year survival of around 16% [8].

Therefore, it is of utmost importance that once the diagnosis is confirmed, a systemic study is performed to evaluate the extensión of the disease to other possible target organs, highlighting the cardiological, digestive, renal and respiratory study. Chronic systemic diseases should also be looked for.

Conclusions

We present this case, due to its unusual presentation, to broaden diagnostic horizons and promote the dissemination of the pathology, since it is common to relate polyp-type lesions of the oral cavity only with HPV.

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