Tumor-like amyloidosis of the parotid : A case report

Amylose pseudo-tumorale de la parotide : A propos d'un cas

Intissar Cherif[1], Marwa Walha[2], Soumaya Charaa[3], Mouna Guermazi[4], Sameh Marzouk[4], Souad Ferjani[3], Abdelmajid Khabir[2]

1. *Service ORL, Hôpital Habib Bourguiba de Médenine*
2. *Service Anatomo-pathologie, Hôpital Habib Bourguiba de Médenine*
3. *Service Radiologie, Hôpital Habib Bourguiba de Médenine*
4. *Service Médecine Interne, CHU Hedi Chaker Sfax*

Abstract

Introduction: Tumor-like amyloidosis or amyloïdoma is a nodular lesion related to abundant amyloid deposits that can clinically mimic a malignant tumor. Its etiologic diagnosis requires searching an underlying infectious disease, a connective tissue disorder or a lymphoma. Parotid amyloïdoma is exceptional, and only four cases have been reported in the literature from 1988 to 2021 (PubMed research).

Case report: We reported the case of a 60-year-old, diabetic and hypertensive woman, presenting an isolated swelling of the right parotid region without facial paralysis or cervical lymphadenopathy. A right superficial parotidectomy with a frozen section examination was performed. Histologically, the swelling was related to abundant amyloid deposits without tumor. On immunohistochemistry, amyloidosis was type AA. The association with the Sjögren's syndrome was confirmed.

Conclusion: The association of parotid amyloïdoma with Sjögren's syndrome is a rare condition. The histologic diagnosis may be difficult in this case. Therefore, it is necessary in the case of amyloïdoma to confirm the diagnosis and carry out an etiological investigation to search for an underlying pathology.

Keywords: Amyloïdoma, Tumor-like, Sjögren, Amyloidosis

Résumé

Introduction : L'amylose pseudo-tumorale ou amyloïdome est une lésion nodulaire en rapport avec un dépôt amyloïde abondant et pouvant mimer cliniquement une tumeur maligne. Son diagnostic étiologique nécessite la recherche d'une pathologie infectieuse, d'une maladie de système ou d'un lymphome sous-jacent. La localisation parotidienne est exceptionnelle et seulement 4 cas d'amyloïdome de la parotide ont été rapportés dans la littérature entre 1988 et 2021 (recherche faite sur PubMed).

Observation : Nous rapportons le cas d'une femme de 60 ans, diabétique et hypertendue, qui s'est présentée pour une tuméfaction isolée de la région parotidienne droite, non accompagnée de paralysie faciale ni d'adénopathie cervicale. Une parotidectomie superficielle droite avec examen extemporané étaient pratiqués. Histologiquement, la tuméfaction était en rapport avec un dépôt amyloïde abondant sans lésion tumorale. A l'étude immunohistochimique, l'amylose était de type AA. L'association à un syndrome de Gougerot-Sjögren était confirmée.

Conclusion : L'association amyloïdome parotidien et syndrome de Gougerot-Sjögren est rare. Le diagnostic histologique peut s'avérer difficile et le dépôt amyloïde peut être responsable d'une lésion pseudo-tumorale. Il est donc nécessaire de confirmer le diagnostic et de faire une enquête étiologique pour rechercher une pathologie sous-jacente.

Mots clés : Amyloïdome, Pseudo-tumeur, Gougerot-Sjögren, Amylose.

Correspondance

Intissar Cherif
Service ORL, Hôpital Habib Bourguiba de Médenine
E-mail : intissar.cherif@gmail.com

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INTRODUCTION

Amyloidosis is a set of pathological entities caused by the extracellular deposition of abnormal fibrillar proteins called amyloid (1). The accumulation of amyloid may be systemic, involving many organs, or localized manifesting as infiltration of individual organs, or in the form of a focal, tumor-like lesion (2-4). Amyloidosis may develop in the setting of underlying conditions, usually chronic inflammatory diseases, in which case it is termed secondary, or it may involve no underlying disease and thus be primary or idiopathic (4). Amyloidoma constitutes a solitary, localized, tumor-like deposit of amyloid, i.e. insoluble fibrillar proteins with beta-pleated sheet arrangement, in diverse organs without evidence of systemic amyloidosis (4). Amyloidoma of the parotid is a very rare condition (4). We here reported a new case of parotid amyloidoma.

CASE REPORT

The present case was reported according to the CARE guidelines (5). The patient gave her written consent. A 60-year-old woman, diabetic and hypertensive, was presented to the otolaryngology department with a painful swelling of the right parotid region. The physical examination had objectified a firm swelling in the right parotid measuring 4 cm in the long axis. There was no evidence of facial nerve paralysis and cervical lymphadenopathy. Magnetic resonance imaging found a heterogeneous lesion in the superficial and deep lobe of the right parotid gland, suggestive of pleomorphic adenoma (figure 1).

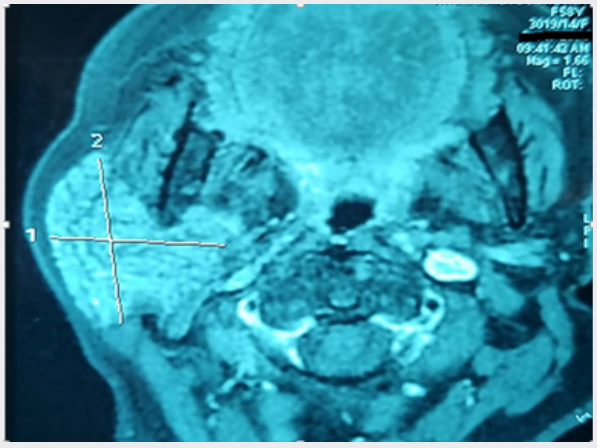

Figure 1. Axial T1-weighted Magnetic resonance imaging with fat saturation and contrast: right intra-parotid tissue process, involving the 2 lobes (superficial and deep) measuring 47 x 52 mm with moderate enhancement after injection of gadolinium.

A fine needle aspiration cytology showed rare giant cells and a few macrophage cells with no cells suspected of malignancy. A right superficial parotidectomy was done under general anesthesia. Intraoperatively, facial nerve branches were seen going into tumor tissue and were carefully dissected out. The dissection was very hemorrhagic and laborious. The deep lobe of the parotid was very adherent. The superficial lobe was sent for histopathological examination. Frozen section examination of specimen showed epithelioid and giganto-cellular granulomatous lesions in contact with an eosinophilic substance of undetermined nature accompanied by parotitis, without tumor nodule. Histologically, the substance was dense amorphous eosinophilic (Figure 2), homogeneously stained with Congo Red and exhibiting yellow-green dichroism in polarized light (Figure 3), corresponding to an amyloid deposit.

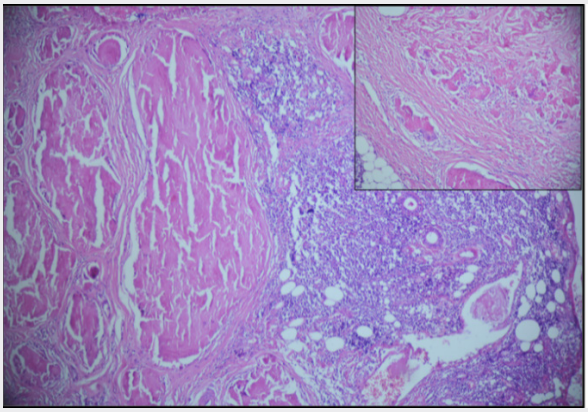

Figure 2. Abundant, eosinophilic, amorphous extracellular deposition with lymphocytic parotitis (HE x100) and foreign body type giant cell reaction (in HEx200 inset).

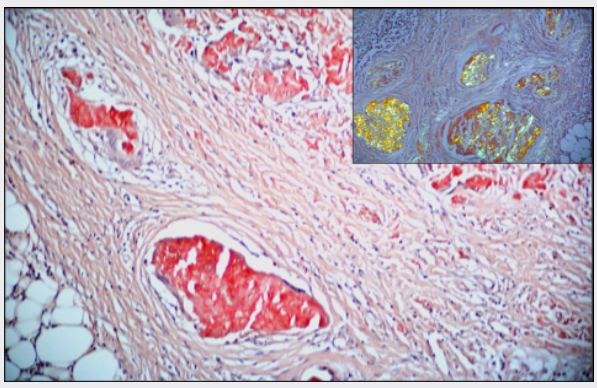

Figure 3. Brick red extracellular deposit (Congo Red x 200) with greenish-yellow birefringence in polarized light (inset x 200)

This very abundant amyloid substance was accompanied by a granulomatous inflammatory reaction and partially erased the parotid architecture. The parotid parenchyma was also the seat of a lymphoid infiltrate of variable abundance with ductal regression of the acini. There were no tumor nodules and no signs of malignancy. The diagnosis of parotid amyloïdoma was retained. On immunohistochemistry, the deposit was positive only for the AA proteins without labeling for the kappa and lambda light chains.

The patient was investigated for systemic amyloidosis. Results of laboratory studies were negative and the antinuclear antibody assay was positive. Also, infectious and neoplastic investigation were negative.

A 6 months later, the patient complained of a bilateral swelling in the parotid glands with a marked oral and ocular dryness. A biopsy of the accessory salivary glands was performed. Histological examination revealed lesions of chronic lymphocytic sialadenitis, stage 4 according to Chisholm and Mason (6). The final diagnosis was parotid amyloïdoma associated with the Sjögren syndrome. The patient was put on corticosteroid therapy at a dose of 0.5mg/kg/day for 6 weeks, then dose reduction. A 12 months later, the patient was fine without any clinical or laboratory evidence of systemic amyloidosis or multiple myeloma.

DISCUSSION

Amyloidosis is a heterogeneous group of diseases defined by deposits of abnormal extracellular fibrillary proteins that cause tissue damage (7). The most common types are AL and AA amyloidosis (8). In AL amyloidosis, deposits are composed of immunoglobulin light chains secreted in hematolymphoid disorders such as plasma cell dyscrasia or multiple myeloma (9). In AA amyloidosis, deposits are derived from serum amyloid A- (SAA-) associated protein (10). SAA protein production by the liver is increased in chronic inflammatory conditions, including autoimmune disorders such as rheumatoid arthritis, and in chronic infections (11).

Amyloidoma is the localized form of amyloidosis without evidence of systemic involvement (12). This is a rare form, described in different anatomical sites and more frequently in the lungs and upper airways (13). Localized amyloid deposits have been described in a variety of sites in the head and neck, including the orbits, nasopharynx, lips, floor of mouth, tongue, larynx, and tracheobronchial tree (14). The parotid gland represents a classic localization of systemic amyloidosis. A tumor-like amyloïdoma of the parotid gland with bilateral involvement and association with Sjögren's syndrome, without systemic involvement is exceptional and constitutes the originality of this case report (4). A parotid amyloïdoma was first described in 1988 (15). The most clinical sign is isolated parotid swelling (4,15,16). Clinical manifestations of the disease are nonspecific, increasing the need of imaging during the investigation (3).

Magnetic resonance imaging does not afford direct diagnosis since the radiological presentation is variable, ranging from the simple swelling of the gland with a modification of the signal to the characteristic of a true tumor as the case of our patient (14).

The treatment of amyloïdoma consists of complete surgical resection (4,12,13). In case of partial excision, recurrence was common (4). In the published cases of parotid amyloïdoma, diagnosis required surgery. A superficial parotidectomy with preservation of the facial nerve was performed (Table 1).

Table 1. Different described cases of parotid amyloïdoma in the literature: from 1988 to 2021 (PubMed research).

Author (Réf.)	Age	Sex	Clinical presentation	Positive diagnosis	Typing		Evolution
Stimson et al. (15) 1988	65	Male	Isolated painless swelling	On operating piece	AL	Primary	No recurrence
Vavrina et al. (17) 1995	54	Male			AL	Primary	Recurrence after 5 years (ipsilateral)
Nandapalan et al. (16)	69	Female			AL	Primary	No recurrence
Maingi et al. (4) 2020	62	Male			AL	Primary	No recurrence
The present study 2022	60	Female			AA	Secondary	Recurrence after 6 months (bilateral)

The positive histological diagnosis is based on the demonstration of amyloid deposits stained by Congo red and presenting a yellow-green dichroism in polarized light (12,13).

The typing of the deposit constitutes in a second time an essential step, it is currently based on the immunohistochemistry analysis (18). In primary and myeloma-associated amyloidosis (AL amyloidosis), the excess precursor protein is a fragment of immunoglobulin light chains. In secondary amyloidosis (AA amyloidosis), the fibrillar protein derives from a serum protein, the SAA protein, a major protein of inflammation (10). Secondary amyloidosis is observed during chronic inflammatory diseases or prolonged suppuration (4,12). In our case, it was type AA amyloidosis. Amyloidoma of the parotid gland associated with an autoimmune disease (Sjögren's syndrome) is rarely described (4). In this case, the most frequently affected sites are the skin, the lung, the tonsils and the mammary glands (17,19).

CONCLUSION

Amyloidoma parotid is a benign tumor-like lesion consisting of a nodular amyloid deposit that can clinically mimic a pleomorphic adenoma or a malignant tumor. It can be idiopathic or secondary to infection (eg; tuberculosis), systemic disease (eg; Sjögren's syndrome) or lymphoma. The clinical diagnosis can be difficult and the radiological feature is nonspecific. The diagnosis of amyloïdoma is a histological surprise requiring further investigations in order to type this amyloidosis and to verify whether it is primary or secondary amyloidosis.

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