

Primary Localized Amyloidosis Of The Neck - A Unique Differential Of Neck Masses.

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Citation

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Abstract

Amyloidosis is a group of disorders characterized by an extracellular deposition of an abnormal amount of proteins in a variety of organs. It typically presents as disseminated deposits. However rarely, localized deposits of amyloidosis are seen. Tumor like localized presentation of amyloidosis, referred to as amyloidoma, is uncommon, especially in the head and neck. We report a case of a 40year old man who presented with a large mass on the left side of the neck.

INTRODUCTION

Amyloidosis in its diverse types most often presents as a systemic form with multiorgan insufficiency. Single organ amyloidosis without evidence of generalized involvement is known as localized amyloidosis.^[1] It is an uncommon benign condition, regarded to be the result of local synthesis rather than the deposition of light chains produced elsewhere in the human body.^[2] Localised amyloidosis affects many organs, however affliction of the soft tissues of the neck is a rarity.^[3]

The differential diagnosis of enlarging neck masses is extensive ranging from granulomatous lesions to metastatic deposits. Amyloidosis, because of its infrequent occurrence is rarely considered. However, rapid growth, a striking feature of the amyloid, warrants it in the differential diagnosis of a neck masses.^[4] A case of localized amyloidosis with a clinicopathological review is presented with particular attention to its head and neck manifestation.

CASE HISTORY

A 40year old man presented to the otorhinolaryngology clinic with the history of an enlarging neck mass for the past six months. The lesion initially was a small nodule which the patient ignored and failed to seek medical attention. The mass enlarged to its present size, the clinical impression of a neck metastasis from an unknown primary was considered and an incisional biopsy of the mass was performed elsewhere, a histopathological diagnosis of inflammatory pseudotumor was offered. The patient was referred to us. On physical examination, a nontender, soft to slightly firm mass on the left lateral aspect of the neck was seen. An excisional

biopsy was undertaken and sent for evaluation.

Grossly, the surgically resected mass measured 7x5x3cm in size. Its cut surface was homogenous, firm and grey white in appearance. Microscopic examination revealed a diffuse deposition of amorphous, acellular, eosinophilic material on haematoxylin-eosin stained slides. Mixed chronic inflammatory infiltrate, composed of scattered as well as collections of plasma cells and lymphocytes was present. This histological appearance suggested amyloid deposition, hence, Congo red stained sections were evaluated, which exhibited apple-green birefringence under polarized light reiterating the diagnosis of amyloid. Ultra structural examination showed that the deposits were composed of collection of unbranched, straight and slightly bent, rigid looking filamentous structures. [Figure1] The filaments were arranged in an interlacing meshwork along with areas of parallel alignment also [Figure2]. Immunohistochemical staining for lambda and kappa demonstrated positive expression in both.

Figure 1

Figure 1. Ultrastructural image showing cell with adjacent amyloid fibrils.(12000X)

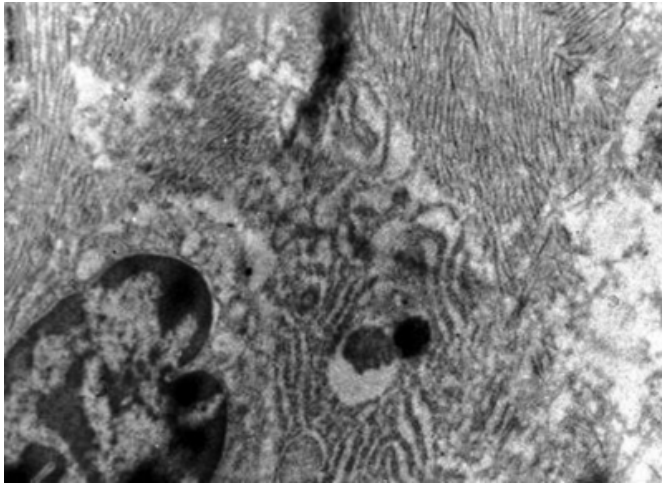
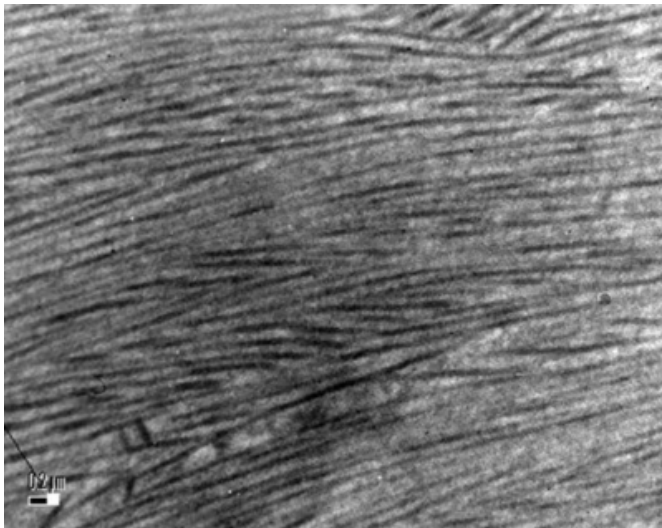


Figure 2

Figure 2. Ultrastructure image showing parallel amyloid fibrils (30000X)



DISCUSSION

Amyloidoma is a rare lesion occurring as a localized mass, in absence of systemic amyloidosis. It has been observed in various locations but rarely soft tissues, especially of the neck. It may present diagnostic difficulties, as a localized mass often leads to a clinical diagnosis of neoplasm.^[5]

Tumorlike presentation of amyloidosis is a rare condition. In the region of the head and neck, amyloid deposits have been reported at various locations, namely tongue, larynx, thyroid, eyelids, cervical lymph nodes, parotid gland, oral cavity, pharynx, pinna,^[7] however rarely involvement of the nasopharynx and neck is seen. Clemons et al have reported a

case which presented with a multilobulated mass suggestive of lymphadenopathy and was finally diagnosed as amyloidoma.^[3] Zuang et al have suggested that although rare it should be considered in the differential diagnosis of these masses.^[6]

Localized amyloidosis is derived from AL [immunoglobulin light chain], AA[serum amyloid protein], AK type of amyloidosis. Although distinguishing between systemic and localized is of clinical importance, it is increasingly apparent that several amyloid proteins may be associated with both systemic and localized forms.^[7] Systemic amyloidosis is a serious and usually fatal condition, in which accumulations of amyloid fibrils in the tissues destroys normal structure and function. Conversely, localized deposition has an excellent prognosis. Hence, it is important to determine the site of deposition, systemic or localized. Evaluation for a systemic involvement is essential in cases presenting as a localized mass. This can be achieved by rectal biopsy or abdominal fat aspiration, which are positive in 75-90% of patients.^[7,8] A rectal biopsy was performed in our case which was negative for amyloid. Some authors however suggest it is not clinically efficient to make a search through biopsies for systemic amyloidosis in cases of localized form without apparent systemic symptoms such as renal function alteration.^[9] The clinical manifestations of amyloidosis are varied and depend on the biochemical nature of the fibril protein and area of involvement.

The generic diagnosis of amyloid is based on its tinctorial properties and ultrastructural appearance. Grossly, the infiltrated organs have a characteristic rubbery, firm consistency and typical waxy, gray or yellow appearance.^[7] On microscopic examination, deposits of extracellular eosinophilic amorphous material commonly shows a characteristic apple green birefringence on Congo red stain under polarized light.^[7] In the present case also the tissue specimen was firm with grayish appearance and morphologically and ultrastructurally characteristic of amyloid. Areas of calcification as seen in the present case has also been reported by Krishnan et al in 8 out of 14 cases, no clinical or pathological significance was attached to them.^[5]

An intimate association of plasma cells with amyloid deposits as was evident in the present case has been reported.^[5,10] Plasma cells have been described to be both polyclonal and monoclonal. Restriction of the light chain for both amyloid deposits and plasma cells has been observed

suggesting a localized plasma cell dysfunction or dyscrasia in amyloidoma.^[10] It may be produced by isolated, benign immunocyte clones of plasma cell rather than by a bonafide plasma cell/lymphoplasmacytic neoplasm.^[5] In the present case light chain expression for both κ and λ was seen and on further evaluation no evidence of multiple myeloma was detected. Compared to κ chain, λ chain restriction has been reported with greater frequency in literature essentially because the variable region of λ light chain is known to be most amyloidogenic.^[10]

Treatment consisting mainly of surgical excision has been advocated to amyloidosis of the head and neck as vast majority represent localized disease.^[7] However in a more extensive involvement total excision can impair organ function, thus if much morbidity is not caused and deposition is slow, conservative treatment with careful observation is suggested. The type of light chain restriction does not influence the management but is useful in the ruling out of multiple myeloma in concordance with the other World Health Organization criteria.

To summarize, amyloid deposit in the head and neck region is usually localized, however further evaluation for systemic involvement is a necessity. Localized excision is the treatment of choice as it is a slow benign process that carries an excellent prognosis. Although rare, it should be considered in the differential diagnosis of neck masses.

Indeed otolaryngologists must be aware of the various manifestation of localized amyloidosis and manage patient accordingly.

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