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LOCALIZED AMYLOIDOSIS OF THE NECK: A RARE CASE AND LITERATURE REVIEW

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ABSTRACT

Amyloidosis is the term used for a group of diseases characterized by extracellular deposition of fibrillar proteinaceous substance called amyloid, in a β -pleated sheet conformation. Amyloidoma or amyloid tumor is a tumor-like localized deposit of amyloid encountered occasionally in association with multiple myeloma, various chronic inflammatory diseases and primary amyloidosis. Soft tissue amyloidoma is rare, and soft tissue amyloidoma without evidence of systemic amyloidosis is even rarer. We report a rare case of primary AL type localized amyloidoma with a clinicopathological review and particular attention to its head and neck manifestation.

Keywords: Amyloidosis, Neck mass, Histopathology.

1. INTRODUCTION

Amyloidosis is the term used for a group of diseases characterized by extracellular deposition of fibrillar proteinaceous substance called amyloid, in a β -pleated sheet conformation. It 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 etiology, treatment, and outcome of systemic amyloidosis are totally different from localized amyloidosis. The mean survival of patients with systemic amyloidosis is between 5 to 15 months, whereas patients with localized amyloidosis have excellent prognosis [4]. Here we report a case of soft tissue amyloidoma presenting as mass lesion on left lateral side of the neck.

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2. CASE REPORT

A 55 year old man presented to the otorhinolaryngology clinic with the history of a gradually enlarging painless neck mass for the past four years (Figure 1). The lesion initially was a small nodule which the patient ignored and failed to seek medical attention. There was no other complaint. Computed Tomography scan revealed well defined multiple soft tissue density masses within the left submandibular region and left posterior cervical space extending up to the cricoid cartilage. Margins of the masses are well defined with no evidence of any infiltration into any surrounding structure. Left submandibular gland was compressed and pushed anteriorly.

On examination a huge firm, nontender mass of 12×8cm was present in left submandibular, upper and mid cervical region and fixed to the underlying structures. The clinical impression of a neck metastasis from an unknown primary cancer was considered and fine needle aspiration cytology was performed. Cytology smears revealed amorphous acellular material with interspersed few inflammatory cells composed of lymphocytes and plasma cells. No atypical or malignant cell was identified.

Biopsy of mass lesion with histopathological correlation was advised. An incisional biopsy of the mass was performed. Grossly, the surgically resected mass was unencapsulated and measured 4x3x3cm in size with a homogenous, firm, grey white cut surface. Microscopic examination after treatment with haematoxylin and eosin (Figure 2) and Congo red stained sections after pretreatment with potassium permanganate (Figure 3) confirmed the diagnosis of amyloidoma. Potassium permanganate staining confirmed that the amyloid was not the AA-type amyloid, usually associated with infection, but the AL-type amyloid seen in primary amyloidosis [5].

As amyloidoma can be a manifestation of systemic disease, a thorough workup including complete blood count, liver and renal function tests, urine analysis, chest X-ray, electrocardiography, echocardiography, bone marrow biopsy, erythrocyte sedimentation rate, rheumatoid factor, anti-nuclear antibody and serum protein electrophoresis performed, were normal. A final diagnosis of primary amyloidoma (AL type) was made. The patient was managed surgically and is on close follow up for the last ten months and is doing well.

3. DISCUSSION

Primary solitary amyloidosis is characterised by localized amyloid deposition without any underlying plasma cell dyscrasia or abnormal serum proteins [6, 7]. It has been observed in various locations namely tongue, larynx, thyroid, eyelids, cervical lymph nodes, parotid gland, oral cavity, pharynx, pinna, but rarely in soft tissues like the neck [8, 9]. It may present diagnostic difficulties, as a localized mass often leads to a clinical diagnosis of neoplasm [6].

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 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 or biochemical investigations including renal function test, which are positive in 75-90% of patients [9, 10]. An abdominal fat aspiration was performed in our case which was negative for amyloid. Nine to fifteen percent of amyloidosis is of the localized type, and is usually associated with plasma cell dyscrasia [6]. However it is very rare to find primary localized amyloidosis in absence of plasma cell dyscrasia as is in our case.

In localized idiopathic AL amyloidosis, localized deposition of amyloid is regarded to be the result of local synthesis rather than the deposition of light chains produced elsewhere in the human body [2, 8]. The benign nature of localized amyloidosis suggests that a localized clone of plasma cells producing an amyloidogenic light chain may represent the pathogenetic mechanism of this disease, which appears to be a form of plasma cell dyscrasia [11].

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. Otolaryngologists and pathologist must be aware of the various manifestations of localized amyloidosis and manage the patients accordingly.

Informed consent taken: Yes

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LEGENDS TO FIGURES



Figure-1. Photograph showing a huge firm , non-tender mass of 12×8cm in left submandibular, upper and mid cervical region, fixed to the underlying structures.

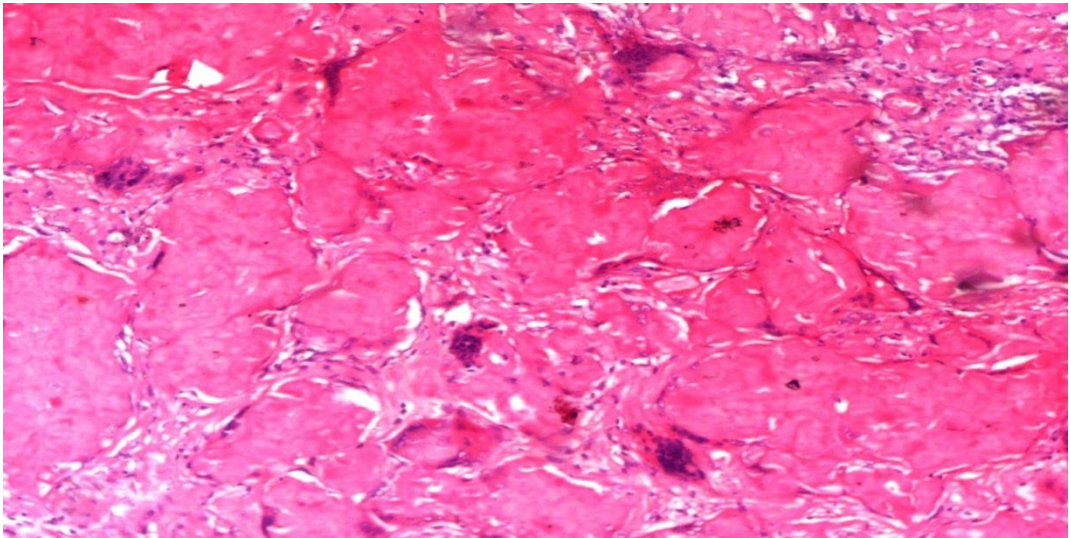


Figure-2. Microscopic examination (after pretreatment with haematoxylin and Eosin x 40) revealed a diffuse deposition of amorphous, acellular, eosinophilic material with a mixed chronic inflammatory cell infiltrate of scattered plasma cells and lymphocytes.

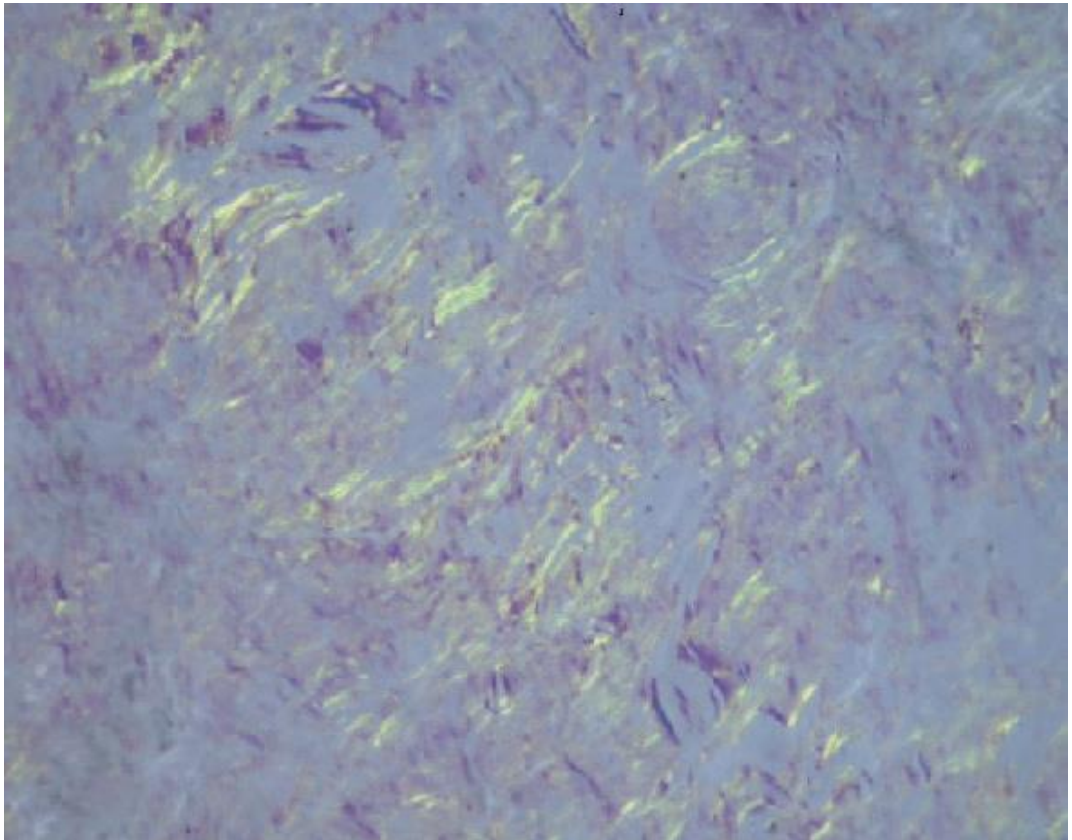


Figure-3. Congo red (x 40) stained sections after pretreatment with potassium permanganate exhibited apple-green birefringence under polarized light.

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