CASE REPORT

Primary systemic amyloidosis of tongue with chondroid metaplasia

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ABSTRACT

Amyloid is a pathologic proteinaceous substance deposited between cells in various tissues in a variety of clinical conditions. We report a case of amyloidosis of tongue with extensive chondroid metaplasia diagnosed on incisional biopsy in a multiple myeloma patient, who underwent autologous peripheral blood stem cell transplant for the same in 2010 and now presented with disease relapse after 2 years.

Key words: Chondroid metaplasia, incisional biopsy, primary amyloidosis, tongue

INTRODUCTION

Systemic amyloidosis is described in 5-15% of patients with multiple myeloma.[1] Amyloidosis in multiple myeloma is characterized by the elaboration of excess light chains by the host. These light chains are converted to amyloid fibrils by proteolytic enzymes in macrophages and deposited in tissues. They can be deposited in connective tissue anywhere in the body and extensive deposition may cause organ dysfunction.[2] We describe an unusual case of amyloidosis of tongue with chondroid metaplasia, diagnosed on incision biopsy in a multiple myeloma patient with relapse of the disease, 2 years after autologous peripheral blood stem cell transplant. To our knowledge this is the first reported case of chondroid metaplasia in primary systemic amyloidosis. Our search of literature revealed two case reports of chondroid metaplasia in amyloidosis; of which one was in solitary amyloid tumour of tongue and the other in nodular pulmonary amyloidosis.[3,4]

CASE REPORT

A 26-year-old male, who is an iron molding laborer, was diagnosed with multiple myeloma in 2007. He was managed with thalidomide, dexamethasone, external beam radiotherapy to pelvis, and analgesics. He underwent autologous peripheral blood stem cell transplant in 2010.

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Quick Response Code:

Website: www.jomfp.in

DOI: 10.4103/0973-029X.119753

Two years later, in January 2012, he presented with dysphagia and dysarthria.

Clinical examination showed two nodules measuring 2 cm on either side of hard palate. Whole tongue was enlarged, hard with multiple nodules extending from the tip to the base of tongue and further down. Regional lymph nodes were not enlarged. Laboratory investigations revealed anemia, raised serum creatinine, and kappa light chain levels. Other parameters were within normal limits.

Incisional biopsy was done from one of the nodular lesions in the tongue with the diagnostic possibilities of carcinoma tongue/myeloma infiltration. Histopathology showed tissue lined by hyperplastic squamous epithelium with subepithelial deposits of eosinophilic amorphous material admixed with chondroid areas [Figures 1 and 2]. Congo red staining showed apple green birefringence of eosinophilic amorphous material in polarizing microscopy, thus confirming the presence of amyloid [Figure 3]. Bone marrow biopsy was done which showed sheets of immature plasma cells [Figure 4].

DISCUSSION

Amyloidosis is the deposition of complex proteins in tissues that when stained with Congo red dye shows apple green birefringence under polarized light.[1] Amyloidosis is broadly classified into systemic amyloidosis and localized amyloidosis. Systemic amyloidosis is further classified into primary amyloidosis (associated with immunocyte dyscracias), secondary amyloidosis (reactive systemic amyloidosis), hemodialysis associated amyloidosis, and hereditary amyloidosis.[1]

The incidence of systemic amyloidosis involving head and neck region varies in different studies. Kerner *et al*., reported

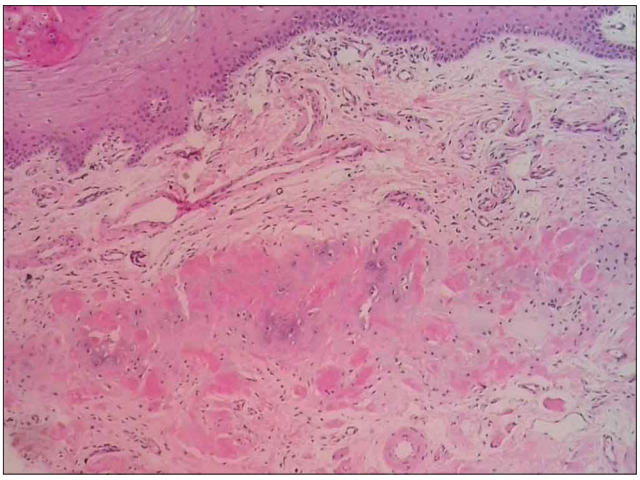

Figure 1: Subepithelial deposits of eosinophilic amorphous material admixed with chondroid areas (H&E stain, ×100)

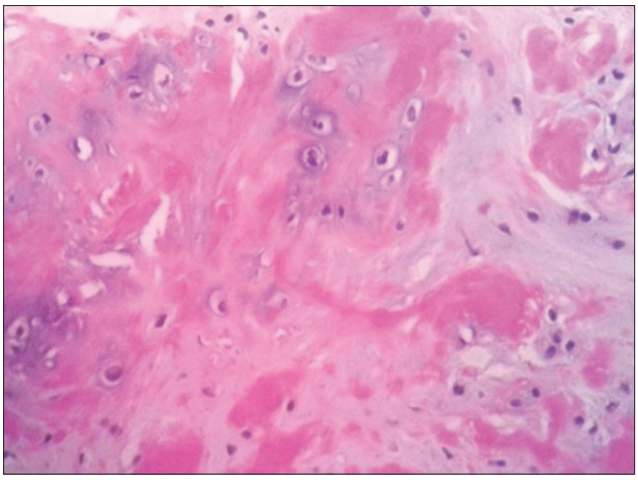

Figure 2: Eosinophilic amorphous material with chondroid areas (H&E stain, ×400)

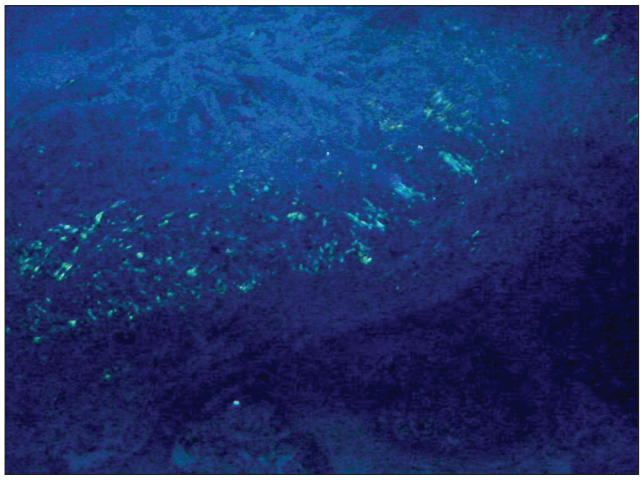

Figure 3: Amyloid deposit showing characteristic apple green birefringence under polarized light (Congo red stain, ×100)

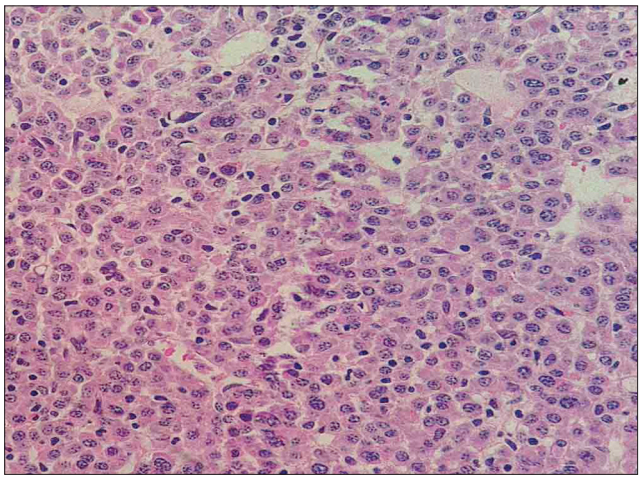

Figure 4: Bone marrow biopsy showing sheets of immature plasma cells (H&E stain, ×400)

involvement of head and neck in 19% patients in a review of 141 biopsy proven cases of amyloidosis of which most common site was tongue.[5] However, chondroid metaplasia in amyloid deposits is rare with only two case reports of which one was in the tongue and the other was in the lungs.[3,4] Although fibrosis, calcification, ossification, and chondroid metaplasia is described in secondary amyloidosis which occurs as a complication of an underlying chronic inflammatory or tissue destructive process, it is not yet specifically described in primary amyloidosis. Ours is the first case highlighting the chondroid metaplasia in amyloid deposits in a case of primary systemic amyloidosis and only the second case described in tongue with chondroid metaplasia of amyloid deposits. Of prime importance is not to overlook the eosinophilic amorphous material in chondroid areas especially in small biopsies so that diagnosis is not missed and repeat biopsies are avoided. The exact pathogenesis of chondroid metaplasia in amyloidosis is not clearly described, but most probably involves bone morphogenetic proteins which induce chondrogenic or osteogenic expression in mesenchymal stem cells.[6]

Chondroid lesions are rarely encountered in tongue. There are case reports of chondroid metaplasia in fibroepithelial polyp, chondroid lipomas, cartilaginous choristomas, cartilaginous metaplasia due to ill-fitting dentures, chondromas, and solitary amyloid tumor of tongue with chondroid metaplasia.[4,7-10] Fibroepithelial polyp, lymphangiomas, hemangiomas, and lipomas can also cause similar nodules in oral mucosa. But diagnosis of amyloidosis can easily be made by typical histopathology findings and demonstration of apple green birefringence under polarized light.

The significance of identifying amyloidosis in multiple myeloma in these patients is that it is associated with poor survival. The median survival time in these patients is assumed to be about 4 months and death usually occurs as a complication of amyloidosis affecting major organ systems. Our patient survived only for 1 month after presentation and died due to multiorgan dysfunction.

Since amyloid deposition in multiple myeloma is evaluated as a grave factor and since there are no laboratory parameters

that associate amyloidosis in these patients, a routine histopathology examination is essential for every multiple myeloma patient with suspected oral lesions.

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How to cite this article: Vasudevan JA, Somanathan T, Patil SA, Kattoor J. Primary systemic amyloidosis of tongue with chondroid metaplasia. J Oral Maxillofac Pathol 2013;17:266-8.

Source of Support: Nil. **Conflict of Interest:** None declared.