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MYELOMA ASSOCIATED SYSTEMIC AMYLOIDOSIS

N Lakshmi Kumari, D Rama Rao, P Ramana Murthy, P Venkataramana

This is a case report of myeloma associated systemic amyloidosis in a 45 year old man. The patient presented with asymptomatic plaques and papules around the eyes, nostrils, anus, inguinal region and tongue. Purpuric lesions were noticed on and off over the lesions. Whe thorough investigations were undertaken they revealed multiple myeloma. Histopathology with amyloid stain positivity and bone marrow examination confirmed the diagnosis.

Key Words : Amyloidosis, multiple myeloma

Introduction

Systemic amyloidosis[1] is an uncommon disorder of protein metabolism showing various combinations of carpal tunnel syndrome, macroglossia, specific mucocutaneous lesions and visceral (cardiac, renal, hepatic and gastrointestinal) involvement. In primary and myeloma associated amyloidosis the fibrils are composed of protein AL (referring to light chains) and appear as a consequence of the plasma cell dyscrasis.

Histopathologically deposits of amyloid are usually superficially placed in the papillary dermis as faintly eosinophilic, amorphous often fissured masses. Amyloid deposits in the deep reticular dermis and subcutis and infitration of blood vessels will also be present.

In myeloma associated systemic amyloidosis bone marrow reveals plasma cell dyscrasia and X-ray examination reveals osteolytic lesions.

Case Report

A 45-year-old man presented with asymptomatic plaques and papules around the eyes and nostrils, groins, perianal region and tongue of 14 months duration. The lesions started as small papules around the orbits simultaneously. They were slowly increasing in number and size and few coalesced to form plaques. Later he developed similar lesions around the nostrils, followed by groins, perianal region, tongue and sides of neck gradually over a period of 14 months. He also compalained of tingling and numbness of thumb, index and middle finger of both hands, of 13 months duration.

Occasional purpuric lesions were noticed by the patient and they subsided spontaneously. He did not have any difficulty in speech and deglutition. He had no bleeding tendency, alopecia, discolouration of skin, xerostomia, dyspnoea, palpitations, giddiness, weight loss, oedema, bowel disturbances or urinary disturbances.

His general condition was good with blood pressure of 90/50 mm of Hg in both lying down and standing postures. Pulse was 80/minute and regular. Cutaneous examination revealed smooth, shiny waxy papules and plaques around the orbits extending on to the nose with a

From the Department of Dermatology, Govt. General Hospital, Kakinada - 533008, India

Address correspondence to : Dr D Rama Rao

few haemorrhagic pale papules around the angles of mouth and few papules over the right cheek were present. Verrucous/ vegitating plaques and papules were present over the under surface of the tongue and a few waxy papules on both sides of the neck, multiple waxy shiny and few skin coloured papules on both sides of groins. A verrucous lesion of 4 x 1 cm in the perianal region was present. Anaesthesia was present on both sides of the hands in median nerve distribution. Hair and nails were normal. Spleen and liver were not palpable. There was no lymphadenopathy. Lungs and heart were normal.

In urine albumin and sugar were not present. Haemogram revealed normochromic, normocytic anaemia, WBC adequate, platelets adequate, bleeding time 1.25 minutes, clotting time 2.4 minutes, 24 hours urinary protein 14.5 gm. Bence Jones proteins were present. Urinary protein electrophoresis revealed a protein band in 2 regions. Serum protein electrophoresis revealed a discrete band in the cathodal side of 2 globulin. Serum calcium was 11 mg. % serum cholesterol 90 mg.% serum creatinine 1.1 mg.%, SGOT 35 units, serum billirubin 7 mg.%. Skin biopsy showed atrophic squamous epithelium with deposition of amorphous fissured esoniphilic deposits underneath. The deposits showed greenish birefringence under polarising light after staining with cong red. Rectal biopsy showed mild nonspecific inflammatory changes in the mucosa and was negative for amyloid. Bone marrow revealed the features of multiple myeloma.

In skull osteolytic lesions were seen. Pelvis A P view showed no evidence of multiple myeloma. D L spine showed lytic lesion in the transverse process of L1. X-ray chest showed fracture of 8th rib on right side and 4th rib on left side.

Comments

Systemic amyloidosis with multiple myeloma is a rare condition. Clinically appearing like primary systemic amyloidosis it sometimes may be associated with multiple myeloma. For that reason every individual with systemic amyloidosis must be thoroughly investigated. Though systemic amyloidosis is a progressive disease and proper treatment is not available the patient can be treated for multiple myeloma.

Reference

1. Fine J, Moschella S L. Diseases of nutrition and metabolism. In: Dermatology (Moschella S L, Hurley H J, eds), 2nd edn. New Delhi: Jaypee Brothers, 1985; 1422 - 532.