

Quiz

Sclerodermoid hands with waxy papules on face

A 46-year-old woman presented with asymptomatic, firm, translucent polygonal papules on the face (Figure 1), mainly around the eyes and mouth, since 2 years. She also complained of symmetrical polyarthralgia involving the wrist, elbow, knee and ankle joints along with burning pain in the hands and feet.

Both hands showed yellowish waxy papulonodular infiltrates (Figure 2) and there was limitation of hand movements. The oral cavity showed papulonodular infiltrates with petechiae (Figure 1). The tongue was moderately enlarged. Systemic examination was normal other than mild hepatomegaly. Skin biopsy is shown in Figure 3.

What is the diagnosis?

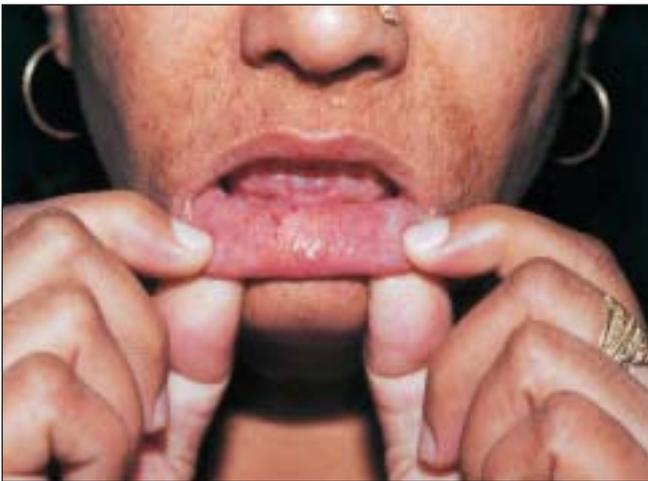


Figure 1: Waxy infiltrated papules on face and lips; note the purpura in the background



Figure 2: Yellowish waxy induration of hands

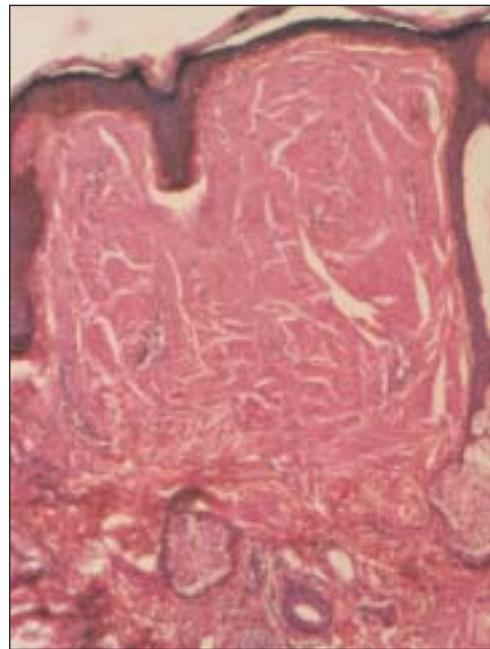


Figure 3: Skin biopsy (H&E, 100X)

How to cite this article: Singhi MK, Gupta LK, Kachhawa D, Bansal M, Gupta D. Sclerodermoid hands with waxy papules on face. *Indian J Dermatol Venereol Leprol* 2004;70:263-5.

Received: February, 2004. Accepted: May, 2004. Source of Support: Nil.



Quiz

Answer: Primary systemic amyloidosis

DISCUSSION

The skin biopsy (Figure 3) revealed fenestrated nodular deposits of eosinophilic, homogeneous material in the dermis staining metachromatically with crystal violet and revealed an apple green birefringence with alkaline Congo red staining. Bone marrow revealed plasmacytosis (10% plasma cells). Serum immunofixation electrophoresis (Figure 4) revealed an IgG monoclonal band. Myeloma proteins were absent in the serum and urine.

Amyloidosis is a metabolic disorder characterized by deposits of extracellular amorphous proteinaceous material in multiple organs impairing their functions. The disease is commonly classified as primary (occurring without antecedent or coexisting disease), secondary (associated with chronic inflammatory disease), myeloma associated, hereditary and localized.¹

Patients have a variable clinical picture depending on the predominant site of light chain deposition.

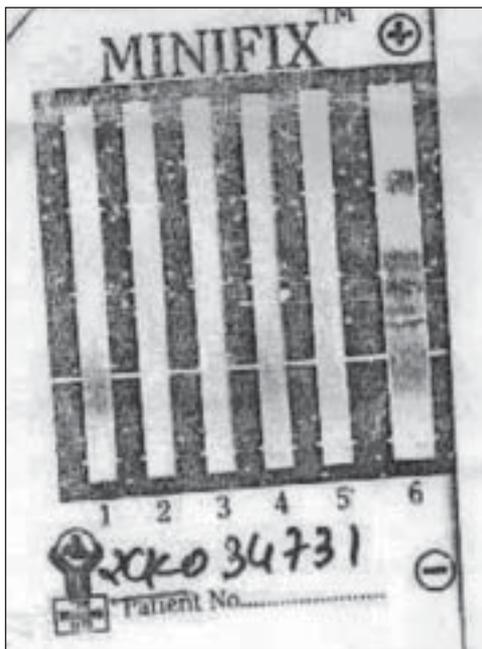


Figure 4: Serum immunoelectrophoresis showing IgG monoclonal band

Common clinical presentations include weakness, fatigue, peripheral neuropathy, periorbital purpura, dyspnea, pedal edema, syncope, light-headedness, hoarseness and dysphasia. A triad of carpal tunnel syndrome, macroglossia and cutaneous lesions is quite classical of plasma cell dyscrasia related systemic amyloidosis.²

The frequent findings of abnormal plasma cells in the bone marrow, presence of mononuclear proteins in the serum or urine in most patients, and recognition that amyloid fibrils may be derived from immunoglobulin light chain fragments suggest that the cause of disease is almost certainly an underlying plasma cell dyscrasia.² About 40% of patients have monoclonal proteins on serum electrophoresis, 68% have a monoclonal spike on serum immunoelectrophoresis and 89% have a monoclonal spike on combined serum and urine electrophoresis.³

The distinction between primary systemic amyloidosis and amyloidosis associated with multiple myeloma is difficult and may be of academic interest.⁴ About 20% of patients with primary systemic amyloidosis meet the diagnostic criteria for multiple myeloma as determined by bone marrow biopsy, skeletal survey and elevated serum calcium level. One-fourth of the patients with amyloidosis in the absence of myeloma have greater than 10% of plasma cells in the bone marrow.⁵ It is believed that the two diseases probably represent different poles of the spectrum of the same fundamental process.

The prognosis of primary and myeloma associated amyloidosis is poor, the major cause of death being cardiac and renal failure. The average survival after diagnosis in most series is 1-3 years.⁶ Melphalan, cyclophosphamide, prednisolone, colchicine and dimethylsulphoxide (DMSO) have been tried alone or in combination with variable success.⁷

M. K. Singhi, Lalit K. Gupta, Dilip Kachhawa,
Mohit Bansal, Dhruv Gupta

Department of Dermatology, Venereology and Leprosy,
Dr. S. N. Medical College, Jodhpur (Rajasthan), India.





Address for correspondence : Dr. M. K. Singhi, 3, M. D. M. Hospital
Road, Shastri Nagar, Jodhpur. E-mail: mks_2_in@yahoo.com

REFERENCES

1. Kaur I, Kumar B, Rajwanshi A, Kaur S. Primary systemic amyloidosis with bullous lesions. *Indian J Dermatol Venereol Leprol* 1983;49:29-33.
2. Black MM. Amyloid and the amyloidosis of the skin. *In: Champion RH, Burton JL, Burns DA, Breathnach SM, editors. Textbook of dermatology. 6th Ed. London: Blackwell Scientific; 1998. p. 2626-37.*
3. Demon LE. Immunohematology. *In: Adelman DC, Casale TB, Corren J, editors. Manual of allergy and immunology. 4th Ed. Philadelphia: Lippincott, Williams and Wilkins 2002; p. 362-8.*
4. Brownstein MH, Helwig EB. The cutaneous amyloidosis. *Arch Dermatol* 1970;102:20-8.
5. Gertz MA, Kyle RA, Greipp PR. Response rate and survival in primary systemic amyloidosis. *Blood* 1991;77:257-62.
6. Brandt K, Cathcart ES, Cohen AS. A clinical analysis of the course and prognosis of 42 patients with amyloidosis. *Am J Med* 1968;44:955-69.
7. Kyle RA, Gertz MA, Greipp PR, Witzig TE, Lust JA, Lacy MQ, et al. A trial of three regimens for primary systemic amyloidosis: Colchicine alone, melphalan and prednisolone, oral melphalan, prednisolone, and colchicine. *N Engl J Med* 1997;336:1202-7.

4th EDEN-IDEA International Congress on Epidemiology, Causes and Prevention of Skin Diseases “Dermato-Epidemiology, Health Care Organization and the Reshaping of Clinical Dermatology”

Venice, October, 10-12, 2004

Organized for the European Dermato Epidemiology Network and the International
Dermato Epidemiology Association

Faculty:

Dr. Luigi Naldi, Dr. Jan Nico Bouwes Bavinck, Dr. Thomas Diepgen, Dr. Robert Stern, Dr. Patrizio Sedona, Dr. M.Dennis Linder, Dr. Martin Weinstock, Dr. Hywel Williams, Dr. Berthold Rzany, Dr. Mara Maccarone

Information: Dr. Luigi Naldi, U.O. Dermatologia, Ospedali Riuniti, L.go Barozzi 1, IT-24128 Bergamo, Tel. +39-035-400625, Fax +39-035-253070. E-mail: luigi.naldi@gised.it

