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Tips & Tricks in

Tips & Tricks in Dermatologic Practice





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Foreword
PB Haribhakti

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Chapter

2 Alopecia Totalis and Systemic Amyloidosis

Aarti Shah

A 53-year-old female patient presented with alopecia totalis and loss of eyelashes since 2 years, oedema with asymptomatic papulonodular lesions on both lower limbs since 3 years and few hemorrhagic bullous lesions over both hands since 7–10 days. Facial edema and yellowish hue of face was seen and woody hard abdomen was palpated.

INVESTIGATIONS

She was anemic and had leucocytosis. The RFT showed severe proteinuria (4+).Urine examination showed presence of albumin (4+), few RBCs, pus cells. Serum A/G ratio was low. Hypothyroidism was also detected. Periorbital ecchymosis and macroglossia were absent. USG abdomen showed presence of multiple 1–2 cm sized lymphnodes in right inguinal region with subcutaneous edema in lower abdominal wall.

Serum protein electrophoresis showed albumin—45.6%, alpha 1–3.5%, alpha 2–23.1%, beta 1–13.3%, beta 2–4.7%, gamma 9.8%. No abnormal band was seen. Urine protein immune electrophoresis showed total protein—231.4, albumin 59.9, alpha 1–2.7, beta 37.4 and free kappa light chain (22.33). 24-hour urinary protein report showed reading of 4,357 (mg/day). ANA blot was negative. p-ANCA and c-ANCA were negative. Colour doppler showed no significant findings. S.HIV was negative.

Histopathological examination showed focal epidermal atrophy, hyperkeratosis and acanthosis in the epidermis. Upper dermis showed increased fibroblasts and collagen. Superficial perivascular and periappendageal lymphoplasmacytic inflammatory infiltrate and few periappendageal foreign body giant cells S/O *scleredema adultorum*.

Renal biopsy specimen showed basement membrane thickening and deposition of PAS negative material in mesangium (Congo red and Thio-T are positive). Tubular interstitial fibrosis and tubular atrophy with mild fibro-intimal

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thickening of vessels present. Immunofluorescence—C3: nonspecific mesangial staining, IgM-non specific mesangial staining, IgA and C1q, IgG—Negative. S/O renal amyloidosis.

TREATMENT

Patient was treated by oncologist with oral melphalan 10 mg daily in two divided doses and oral steroids but patient died of pleural effusion due to cardiac failure (? amyloid deposition) after 3 months of presentation at OPD.

DISCUSSION

The most common form of systemic amyloidosis is AL resulting from fibril formation by fragments of monoclonal antibody light chains resulting from plasma cell dyscrasia. The mean age of onset of primary amyloidosis is about 65 years and male preponderance is seen. Amyloidogenic immunoglobulin AL monoclonal proteins appear to be preferentially of gamma type, of lower molecular weight and lower isoelectric point. Fewer than 20% patients with AL have myeloma and 15–20% patients with myeloma have amyloidosis, the reason being production of amyloid fibrils on digestion of Bence Jones proteins. Abnormal light chain material is almost always present in serum or urine and can be demonstrated in tissue culture of bone marrow cells from affected patients. The organ tropism in AL amyloidosis may reflect germ line gene use and plasma cell burden.

CONCLUSION

We presented this case of primary systemic amyloidosis as a rare cause of alopecia totalis.

Our case was unique as it was associated with multiple myeloma and scleredema.

Editorial Comments

- Sometimes the common skin disease may reflect internal disorders proving skin to be mirror of body
- In this unique case alopecia areata turned out to be the manifestation of multiple myeloma with primary systemic amyloidosis.

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Tips & Tricks in Dermatologic Practice

This book is straight out of experiences of clinical dermatologists who share with us their own experiences in diagnosing and treating some rare to difficult to treat skin disorders.

You will be amused to find some of the out of the box treatment options, many innovations and many rare disorders. *Tips and Tricks in Dermatologic Practice* is meant to give some tips in diagnosing some rare conditions and tricks in treating them in some innovative ways. You will also find some of the finest practical tips for students and clinicians given by some of the senior most dermatologists of India.



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