Case 6

A 47 year – old male patient

Chief complaint Generalized,multiple,bluish-gray hyperpigmented patches

Present & Past History

In 1998,the patient developed generalized non – pitting edema on face & extremities and was diagnosed with hypertension. Before antihypertensive drug (Lasix) was started ,he noticed generalized bluish – gray hyperpigmented patches on his face & trunk .The skin lesions gradually progressed without any symptoms and specific treatment.

In 1999,he developed paresthesia of hands and feet with the problems of chronic fatigue and loss of libido. Polyglandular autoimmune failure was suspected with the evidence of hypoparathyroidism, hypoadrenalism and hypogonadism.

In 2001,he was referred from a private hospital for further management

Medications at time of consultation included

- Ca–lactale 1 tab bid
- Lasix (40) 1 tab OD
- Dexamethasone (0.5) 1 tab hs
- Hctz (50) ½ tab OD
- Allopurinol (100) 1 tab OD
- Depot testosterone 1 Amp IM

Personal history

He has been working as a goldsmith for 10 years

no other members of his family had the same condition

Physical examination

V/S afebrile, BP 140/80,PR 100

Sweating,alert,not pale,no jandice

HEENT: puffy eyelids, Mild facial swelling, Enlarged mandibular bone

Heart.Lung: WNL

Abdomen No hepato-splenomegaly, moderate ascites with umbilical hernia,

Extremities: paresthesia of both feet, no motor deficit pitting edema of both legs

Skin :Generalized ,multiple,discrete,ill-defined,bluish-gray large patches on face,trunk,extremities

no hyperpigmentation on mucous membrane and skin creases

Normal hair and nails

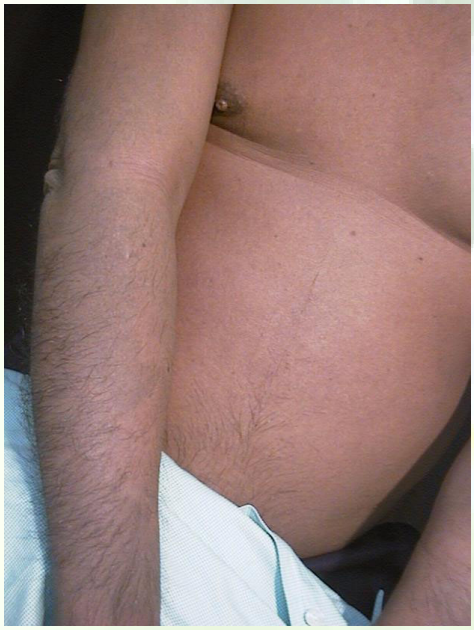

Fig. 6.1

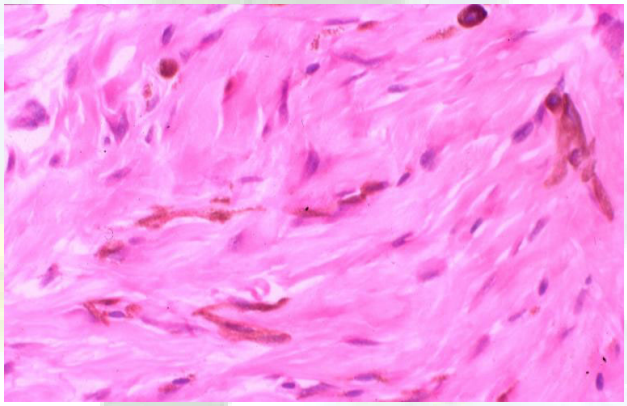

Fig. 6.2

Lab

CBC WBC 4700,HCT 41%,PLT 337000,N40,L44, M 8,Eo 5,Baso 3,MCV 26,MCHC 30

Lab chemistry

Na140 mmol/L, Uric acid 11 mg/dl, K 5.59 mmol/L, Chol 158 mg/dl, Cl 105 mmol/L, Trig 181 mg/dl

Co_2 16.3 mmol/L

LFT WNL Albumin 35 g/L (43-53) Glu 96 mg/dl BUN/Cr 11/1.2 mg/dl
UA WNL
Thyroid function test low free T4
Testosterone serum <0.2 ng/ml (2.1-17.3) FSH 9.6 miu/ml (1.3-19.3)
Serum cortisol <1.0 ug/dl(5-25) LH 7.4 miu/ml(1.2-8.5)
Chest X-ray WNL

Histopathology (S02-3027)

Scattered pigmented dendritic melanocytes among bundles of collagen, fibrotic stroma.

Diagnosis

1. POEMS syndrome
2. Acquired dermal Melanocytosis (Blue nevus)

Presenter Prachiya Jewhasuchin
Consultant Penpun Wattanakrai

Comments

POEMS Syndrome has been defined as an association of plasma cell dyscrasia with polyneuropathy, organomegaly, endocrinopathy, M protein and skin change.

Skin changes includes hyperpigmentation, hypertrichosis, hyperhidrosis, scleroderma–like skin and capillary angiomas .The patients progress to become totally bedridden and death result from the direct effect of underlying dyscrasia.

Chemotherapy or corticosteriods may improve the polyneuropathy ; plasma exchange has not been successful .

Hyperpigmentation in POEMS Syndrome is not clearly defined so the acquired dermal melanocytosis in this patient may be part of the hyperpigmentation manifestation of this syndrome or may be coincident.

References

1. Derenre O, Gnillot B, Dandurand M : cutaneous lesions in POEMS syndrome. Ann Dermatol Venereol.1990;117(4) : 283-90
2. Masson C,Krespi Y: Presse Med.1994 Apr; 9 ; 23 (14):646 - 8
3. IIO M,Kazama T,Kasahara N:Acquired multiple blue nevi scattered over the whole body. Br J Derm .2001;144 : 440 – 442
4. Gonzalez – Campora R, Galera – Davidson H : Blue nevus classical type and new related entitities. Pathol Res Pract.1994 Jun;190 (6) : 627 – 35
5. Jimenez E,Valle P,Villegas C: Unusual acquired dermal melanocytosis. J Am Acad Dermatol 1994 Feb ;30 : 277 – 8