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Case Presentation

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INTRODUCTION

- 41 years old, female, caucasian
- **Presentation:** Loss of sense in hands and skin hyperpigmentation for 6 months
- **History:** Preeclampsia (13 years), Hypertension(13 years), No history of surgery
- **Habits:** 5 packs year smoker (non for 8 months), non-alcoholic
- **Medication:** Amlodipin 5 mg/day
- **No Family History**

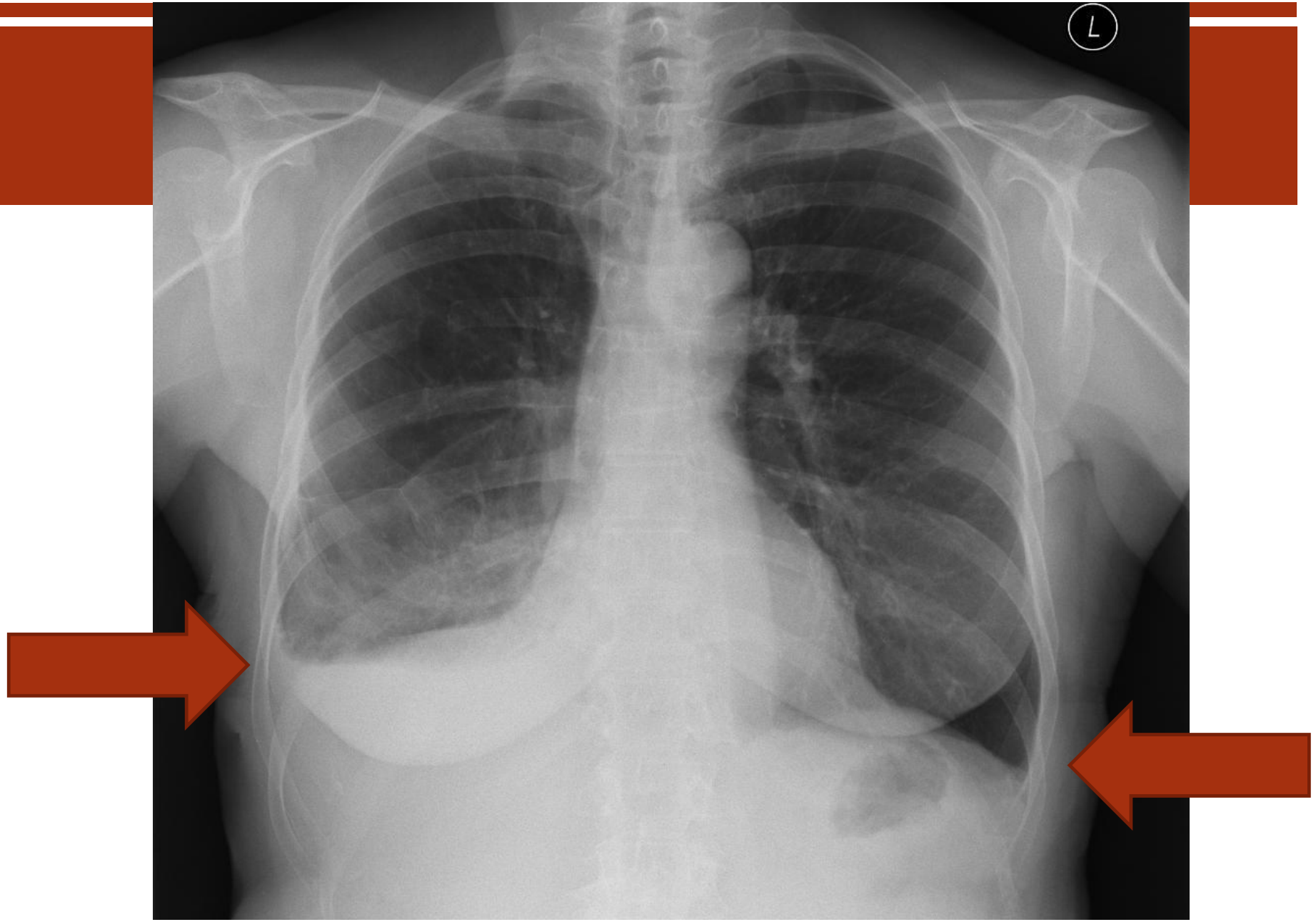
PHYSICAL EXAMINATION

- Vital Signs: Normal
- Hyperpigmentation
- Decreased lung sounds
- Decreased heart sounds
- Hepatosplenomegaly (+)
- Multiple Axillary-inguinal lymphadenopathies
- Pretibial edema + / +
- Quadriparesia, Muscle strength 3/5 upper extremities, 2/5 lower extremitities
- Loss of the Deep Tendon Reflexes for all extremities

LAB AND OTHER FINDINGS

- **WBC: 12040 /uL** (3040-9640/uL) Hb: 13,3 g/dl (10,8-14,9) **PLT: 676000/uL** (150000-400000/uL)
- **Urea: 65 mg/dl (0-38 mg/dl) Cr: 1.38 mg/dl (0,5-0,9 mg/dl) Uric Acid: 11.8 mg/dl (3,4-7 mg/dl)**
- Blood electrolytes: Normal LFT(Liver Function Tests): Normal
- Albumin: 3.7 g/dl (3,4-4,8 g/dl) **Globulin: 5 g/dl** (2,5-3,5g/dl) **LDH: 313 U/L** (135-225 U/L)
- CRP: 0.4 mg/dl (normal 0-0,5mg/dl) **ESR: 56mm/hour** (normal 0-20mm/hour)
- Urine analysis: Normal. No Proteinuria and hematuria
- **f T3: 1,85** pg/ml (2-4,4 pg/ml), **f T4: 0,74** ng/dl(0.93-1,7 ng/dl), **TSH: 5.19** ng/dl(0,27-4,2 ng/dl)
- ECG: Normal
- Patients Lab results were all **normal in 2012**

CHEST X-RAY



WHAT TO DO NEXT?

Abdominal US : **Hepatosplenomegaly**

Thoracoabdominal CT: **Hepatomegaly**, **in the para-aortic area and inguinal area multiple bilateral lymph nodes, diffuse free fluid in the peritoneal, pericardial areas. Bilateral pleural effusion, benign and reactive-looking multiple lymph nodes in both axillary fossas**

Peripheral smear: Erythrocytes NN, neutrophilic leukocytosis, **increased PLT, large platelets (+)**

- EMG : **Demyelination and secondary axonal sensorimotor polyneuropathy**

- ECO: **2‘ Mitral Insufficiency, 2‘ Tricuspital Insufficiency, Mild Pulmonary Hypertension, Pericardial effusion**

- LAP --- Fine Needle Aspiration→ Benign Cytology

WHAT DO YOU THINK ?

- Addisons Disease (Hyperpigmentation)? → ACTH and Basal Cortisol levels: Normal
- Hemachromatosis ?→ Iron profile and genetic analysis: Normal
- SLE(Pleural and pericardial effusion+LAP)? → ANA: Negative
- Vasculitis?(Neuropathy+LAP)→ ANCA : Negative
- Infection? → Procalcitonin: Normal, Cultures:Normal

Cr: 1.38 mg/dl, Uric Acid: 11.8 mg/dl, Globulin: 5 g/dl,
ESR: 56mm/hour, LDH: 313 U/L

MULTIPLE MYELOMA?

- IgG: 13,9 g/l(7-16 g/l) **IgA: 524 mg/dl** (45-380 mg/dl) IgM : 156 mg/dl (40-230 mg/dl)
- Free light chain kappa: **9.12 mg/dl**(0.67 - 2.24 mg/ dl)
- Free light chain lambda: **27.6 mg/dl**(0.83 - 2.7 mg/dl)
- Beta-2 microglobulin: **10271 ng/mL** (651 – 2295 ng/mL)
- Immunfixatiton Electrophoresis + Protein Electrophoresis: **IgA lambda monoclonal gammopathy**
- Bone Survey : No lytic lesions
- **But bone marrow biopsy: N**

SUMMARY

- Neuropathy
- Monoclonal gammopathy
- Endocrinopathy
- Organomegaly
- Skin changes
- Thrombocytosis
- Edema, effusions

DIAGNOSIS?

POEMS SYNDROME

- A neoplastic syndrome and a plasma cell disorder.
- *More common F>M, Mostly between 5.-6. decades(51) , Median Survival: 165 months
- **P**oliradiculopathy
- **O**rganomegaly
- **E**ndocrinopathy
- **M**onoclonal gamopathy
- **S**kin changes

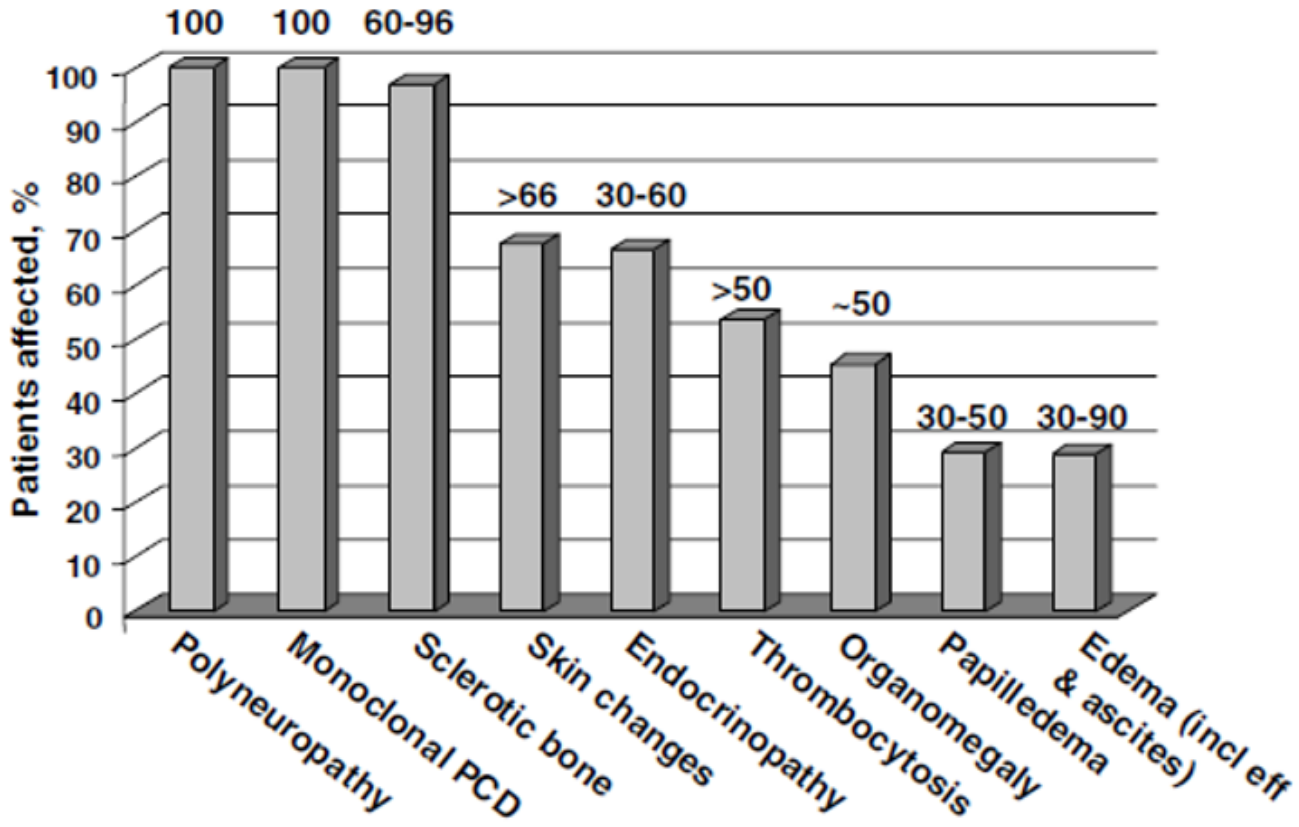
BLOOD, 1APRIL 2003 POEMS SYNDROME:ARETROSPECTIVE REVIEW 2503 VOLUME 101, NUMBER 7

DIFFERENTIAL DIAGNOSIS IN PLASMA CELL DISORDERS

- Multiple Myeloma **(P, M)**
- Monoclonal gammopathy of undetermined significance MGUS **(M)**
- Solitary bone plasmacytoma **(M)**
- Waldenstrom's macroglobulinemia **(P, O, M)**
- Primary amyloidosis **(P, M, S)**
- Cryoglobulinemia **(M)**
- Chronic Inflammatory Demyelinating Polyneuropathy CIDP **(P)**

POEMS SYNDROME

- Polyneuropathy 100%
- Monoclonal gammopathy 100%
- Osteosclerotic bone lesions 60%
- Skin changes >66%
- Endocrinopathy 30-60%
- Trombocytosis (>50%)
- Organomegaly (HM, SM, LAP) 50%
- Lack of Appetite 31%
- Weight loss 37%
- Papillary edema 29%
- Pericardial, pleural effusion 30%
- Castleman Disease 15%
- Clubbing 5%



Dispenzieri A, Blood Rev, 2007;21:285

DIAGNOSIS CRITERIAS

- Mandatory Criterias : **Polyneuropathy, monoclonal gammopathy**
- Major Criterias: Sclerotic bone lesions, Castleman's disease, increased **VEGF**
- Minor Criterias: **Organomegaly, edema, endocrinopathy, skin changes** and papilledema, **thrombocytosis**

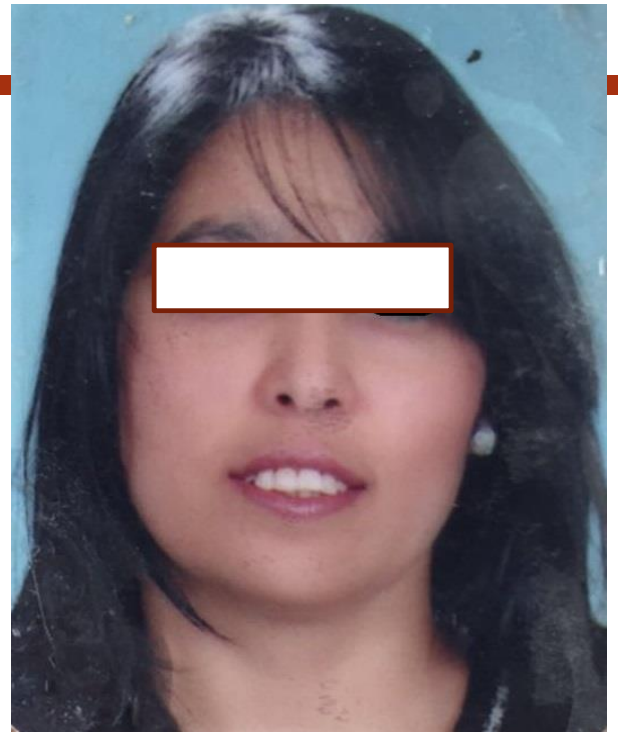
- Two mandatory, one major and at least one minor criteria for the diagnosis

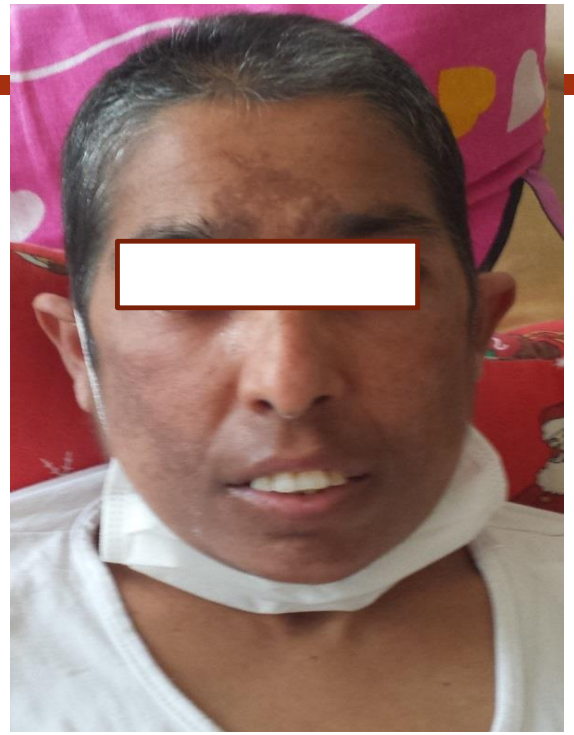
TREATMENT

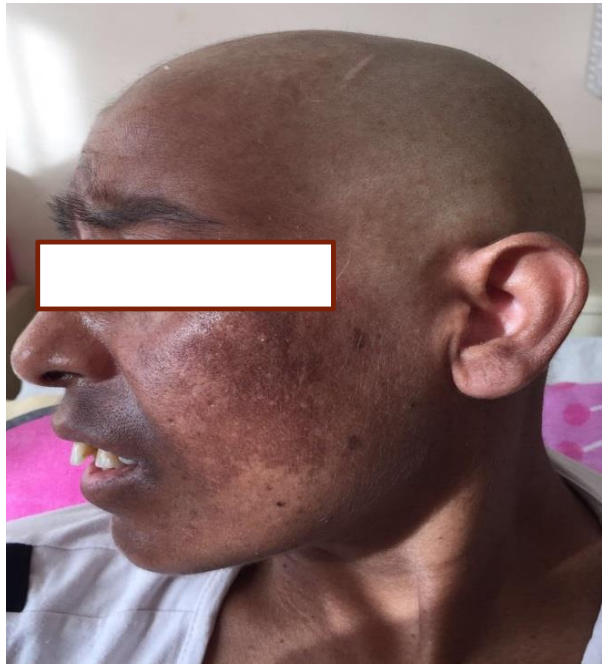
- Chemotherapy: (Melphalan + dexamethasone, melphalan + prednisone; lenalidomide / thalidomide / bortezomib-based therapies)
- Radiotheraphy for the bone lesions
- Autologous stem cell transplantation (with high-dose melphalan) ASCT
- In addition, should be given medications for volume overload , neuropathy, and endocrine abnormalities

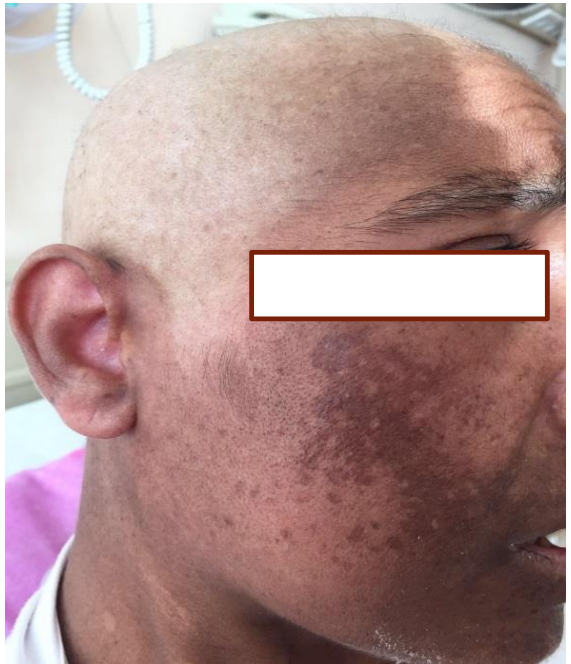
OUR PATIENT:

- 31.08.2015: First cycle VAD (Vincristine, Doxorubicin, Dexamethasone 40 mg)
- 22.09.2015: 2 cycles of VAD, after that Vincristin was stopped because of the neuropathy
- Control ECO 1’ MI, 1' TI, 1' LVH
- Hyperpigmentation is reduced after the chemotherapy.
- 06.10.2015: Autologous SCT was planned with high-dose melphalan
- 20.10.2015: High doses of cyclophosphamide with MESNA(2 days) + Mobilisation with G-CSF 25x10^6/kg of Hematopoietic stem cells were collected
- 07.01.2016: Stem Cell infusion for Autologous SCT
- Patient is still in our follow up…









TAKE HOME MESSAGE

- **Patients with peripheric neuropathy and hiperpigmentation should be evoluated for POEMS Syndrome…**

Thank you for listening…

Discover the Beauties of TURKEY…