A Case of POEMS Syndrome with characteristic Skin Manifestations

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Abstract

I report the case of POEMS syndrome in a 68- year-old Caucasian woman who presented with exertional dyspnea. Progressive skin hyperpigmentation in parallel to sensorimotor neuropathy had been developed since 20 years ago.sclerodermoid changes was evident.

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Fig. no. and caption: 3 figures .Fig1. Marked hyperpigmentation and sclerodermoid changes of the face associated with dark-brown discoloration of the neck and axillary regions (a), intense pigmentation of posterior torso (b). Fig2. Intense sclerosis of phalangeal bones is seen in plain radiograph of left hand. Also a fracture line on the surface of left 4^{th} distal interphalangeal joint was evident.

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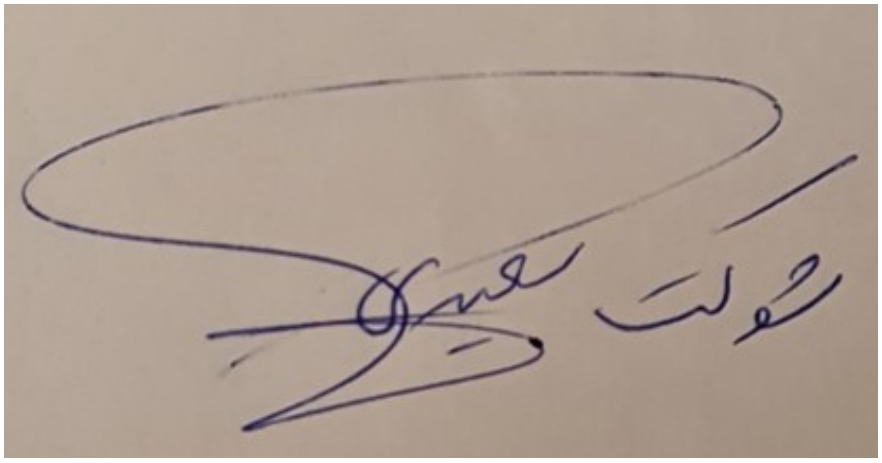

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Abstract:

Background:

POEMS syndrome is a rare paraneoplastic syndrome due to an underlying plasma cell disorder (PCD) and stands for polyneuropathy, organomegaly, endocrinopathy, monoclonal PCD and skin changes. Diagnosis are often delayed because the syndrome is rare and can be mistaken for other neurologic or dermatologic disorders[1].So awareness of its different presentations especially dermatologic ones would result to early diagnosis and better outcomes.

Case Presentation:

I report the case of POEMS syndrome in a 68- year-old Caucasian woman who presented with exertional dyspnea. Progressive skin hyperpigmentation in parallel to sensorimotor neuropathy had been developed since 20 years ago. On examination, skin hyperpigmentation, predominantly affecting face, neck and posterior torso was evident along with marked thickening of face's skin. A huge hepatomegaly also was obvious. Monoclonal band of Immunoglobin A (lambda type) detected in serum plasma electrophoresis. She had a high level of thyroid stimulating hormone (TSH) .A plain radiography of hands revealed intense sclerosis of phalangeal bones .Based on these constellations, a diagnosis of POEMS syndrome was made.

Conclusion:

This case report aims to alert clinicians about the variable presentations of POEMS syndrome especially dermatologic ones in order to make the diagnosis earlier and to search for pulmonary involvement in these subset of patients as well as preventing irreversible neurologic complications with timely treatment.

Introduction:

POEMS syndrome is a rare paraneoplastic disease. The acronym refers to several but not all of the features of the syndrome: polyneuropathy, organomegaly, endocrinopathy, monoclonal PCD and skin changes but not all of these features are required for making the diagnosis. Its prevalence is estimated to be 0.3 per 100000[1]. The pathogenesis has not clearly elucidated, but the role of vascular endothelial growth factor (VEGF) has been suggested[2].The diagnosis is based on the current Dispenzieri diagnostic criteria which requires the presence of both mandatory criteria(polyneuropathy and a PCD) and at least one major criterion(sclerotic bone lesions,castelman disease, elevated VEGF) and one minor criterion(organomegaly,edema,endocrinopathy,papilledema,thrombocytosis or polycythemia and skin changes)[2].Skin findings in POEMS syndrome are common and include hyperpigmentation, hypertrichosis, glomeruloid hemangioma,acrocyanosis,raynaud's phenomenon, sclerodermoid changes, white nails, and clubbing. A strong association between dermatologic presentations and abnormal pulmonary function tests (PFTs) has been reported which requires further diagnostic surveys in this subset[3].

Case Report:

A 68-year-old Caucasian female patient presented to cardiology clinic with a 6-month history of exertional dyspnea. Heart sounds were normal. Diminished basilar sounds of both lungs were evident. Pitting edema detected on both shins. Skin hyperpigmentation was seen predominantly affecting face, neck and posterior torso (Fig.1a) along with sclerodermoid changes of the face. (Fig1.b). Also a huge hepatomegaly detected on palpation. Skin discoloration had been emerged on her face with progression to the trunk and limbs over time without any specific diagnosis which had been worsened despite of starting corticosteroids. Leukonychia, clubbing, hypertrichosis or hemangioma were not detected .A progressive burning sensation with hyperesthesia of distal upper and lower limbs had been developed in parallel to skin discoloration since 20 years ago. Chest x-ray showed mild cardiomegaly, increased brochovascular markings consistent with heart failure. Plain radiography of hand showed intense sclerosis of phalangeal bones (Fig.2). Echocardiography revealed a preserved ejection fraction with mild diastolic dysfunction. There was a mild restrictive pattern in PFT probably due to lung congestion or the disease's complication.Electrodiagnostic studies reported a severe chronic bilateral distal sensorimotor polyneuropathy predominantly demyelinating type. Serum protein electrophoresis showed a monoclonal immunoglobin A paraproteinemia of Lambda type. A high TSH titer was reported .These findings satisfied the criteria for the diagnosis of POEMS Syndrome.

Discussion:

Skin findings are not necessary for the diagnosis of the POEMS syndrome but multiple associations between dermatologic manifestations and other characteristic findings of POEMS syndrome especially pulmonary disorders have been reported [3]. So, previous awareness about these features and seeking for them on physical examination, has important diagnostic and prognostic implications.

List of Abbreviations:

POEMS, polyneuropathy, organomegaly, endocrinopathy, monoclonal, plasma

cell dyscrasia, skin changes

PCD, plasma cell dyscrasia

TSH, thyroid stimulating hormone

VEGF vascular endothelial growth factor

PFTs pulmonary function tests

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