Você conhece esta síndrome?

Guida Santos[1] Sara Lestre[2] Alexandre João[2]

DOI: http://dx.doi.org/10.1590/abd1806-4841.20132266

CASE REPORT

Woman, 65-year-old, with sensory-motor polyneuropathy for 15 years, worsening in the previous 6 months. The patient had diffuse hyperpigmentation, dry skin and hemangiomas (Figures 1 and 2). She reported hypertrichosis, edema of the legs, facial lipoatrophy, leukonychia and weight loss (Figures 2 and 3). Analytically, she presented polyglobulia, thrombocytosis, hypotestosteronemia, hyperalbuminemia and elevation of IgG in the liquor. The immunochemical study emphasized IgG monoclonal gammopathy of lambda chains.

Investigations showed small adenopathies, cardiomegaly, hepatosplenomegaly, generalized edema and extensive lytic lesion of L5. A biopsy of this lesion showed a lytic proliferation of plasma cells, CD138 +, Κ chains - chains λ +. The myelogram showed hypercellularity and eosinophilia. The electromyogram showed slowing of motor and sensory nerve conduction of several nerves. An excision of the right breast angioma was performed and was compatible with capillary hemangioma. The patient has begun treatment with dexamethasone, radiotherapy, bortezomib and is waiting for a bone marrow transplant.

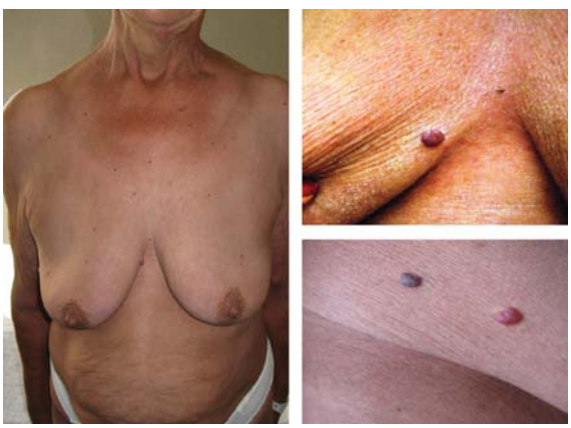

FIGURE 1: Cutaneous hyperpigmentation, cutaneous xerosis and angiomas

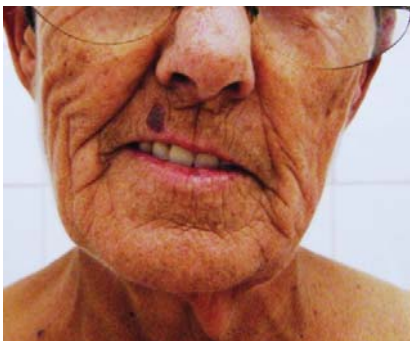

FIGURE 2: Facial lipoatrophy and angioma

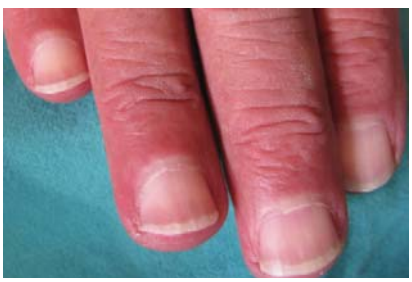

FIGURE 3: Proximal leukonychia on the hands

DISCUSSION

POEMS Syndrome or Crow-Fukase Syndrome, Takatsuki Myeloma Syndrome, Osteoclerotic Myeloma Syndrome.

In 1980, Bardwick invented the acronym for POEMS syndrome: polyneuropathy, organomegaly, endocrinopathy, M protein and skin changes.[1,2,3] No single criterion forms the basis for diagnosis of this entity. The diagnosis is made when polyneuropathy and monoclonal gammopathy (major criteria) are present, associated with other criteria (minor criteria).[4]

The syndrome is more common in males in their 50s and 60s and in Japanese people.[1,3,5,6] Although the etiopathogenesis remains unknown, it appears to be associated with Herpes Virus HHV8 and an increased production of cytokines (IL1ß, IL6, TNF-α and VEGF), leading to the production of plasmocytes.[3,4,6]

Received on 11.11.2012.
Approved by the Advisory Board and accepted for publication on 14.12.2012.
* Work performed at the Santo António dos Capuchos Hospital – Centro Hospitalar de Lisboa Central, E.P.E – Lisbon, Portugal.
Financial Support: None.
Conflict of Interests: None.

[1] MD - Intern in Dermatology and Venereology at the Dermatology and Venereology Department, Santo António dos Capuchos Hospital – Centro Hospitalar de Lisboa Central, E.P.E – Lisbon, Portugal.
[2] MD – Physician specializing in Dermatology and Venereology at the Dermatology and Venereology Department, Santo António dos Capuchos Hospital – Centro Hospitalar de Lisboa Central, E.P.E – Lisbon, Portugal.

The gammopathy is IgA, and less often IgG and light chains lambda.[1] The bone lesions have a mixed pattern: osteosclerotic and lytic.

The peripheral neuropathy is a bilateral sensory-motor polyneuropathy, symmetric and ascending. The disease begins with changes in the sensitivity level of the lower limbs, then progressing to motor deficit.[3] Thrombocytosis and progressive cachexia (weight loss) occur in most patients. Endocrine disorders such as hypogonadism, hyperestrogenemia, hypothyroidism, hypoparathyroidism and diabetes may also occur. Amenorrhoea is common in women, while men experience hypotestoteronemia, gynecomastia and impotence.[1,2,3] Cutaneous manifestations are numerous, heterogeneous and nonspecific,[1] with diffuse cutaneous hyperpigmentation, hypertricosis and hemangiomas being the most frequent.[1] Hyperpigmentation occurs in over 90% of the patients, predominantly in sun-exposed areas, with pseudosclerodermiform skin thickening.[2,7] Edema of the legs occurs in 90% of cases, usually early.[1,2,3,7] Hypertrichosis (trunk and face), palmoplantar hyperhidrosis, Raynaud's phenomenon, dry skin, clubbing leukonychia are also common.[1,2,3] Angiomas occur in 9-44% of cases, locating preferably in the trunk and proximal extremities. They can be capillary, lobular, cavernous, tuberous or glomeruloid hemangiomas. None of them are pathognomonic.[4,5,6,7] Glomeruloid hemangiomas are more common in Japanese patients and are violaceous or blue papules, sessile, pedunculated or cerebriform.[5,6,7]

Treatment may consist of radiation therapy, surgery, chemotherapy (melphalan and prednisone), bone marrow transplantation and/or other drugs such as interferon, lenalidomib, bevacizumab, bortezomib and thalidomide.[1,3,7] The median survival is 13.8 years.[3] ❑

Abstract: POEMS syndrome is a unique clinical entity, the diagnosis of which is made when polyneuropathy and monoclonal gammopathy occur together, associated with other changes such as organomegaly, endocrinopathy, skin changes and papilledema. Cutaneous manifestations are heterogeneous, with diffuse cutaneous hyperpigmentation, hemangiomas and hypertrichosis occurring more frequently. We report the case of a 65- year-old female patient with this syndrome, diagnosed after 15 years of disabling peripheral neuropathy.
Keywords: Hyperpigmentation; Multiple myeloma; Polyneuropathies; Syndrome

Resumo: A síndrome de POEMS é uma entidade clínica única cujo diagnóstico é colocado quando existe polineuropatia e gamapatia monoclonal associada a outras alterações tais como organomegalia, endocrinopatia, alterações cutâneas e papiledema. As manifestações cutâneas são heterogéneas, sendo as mais frequentes a hiperpigmentação cutânea difusa, os hemangiomas e a hipertricose. Relata-se o caso de uma doente de 65 anos com essa síndrome diagnosticada após 15 anos de neuropatia periférica.
Palavras-chave: Hiperpigmentação; Mieloma múltiplo; Polineuropatias; Síndrome

REFERENCES

1. Rose C, Mahieu M, Hachulla E, Facon T, Hatron PY, Bauters F, *et al*. POEMS syndrome. Rev Med Interne. 1997;18:553-62.
2. Dereure O, Guillot B, Dandurand M, Bataille R, Chertok P, Donadio D, *et al*. Cutaneous lesions in POEMS syndrome. Apropos of 3 case reports and a review of the literature. Ann Dermatol Venereol. 1990;117:283-90.
3. Pinto Neto, Jorge V. POEMS syndrome (osteosclerotic myeloma). Rev Bras Hematol Hemoter. 2007;29:98-102.
4. Phillips JA, Dixon JE, Richardson JB, Fabre VC, Callen JP. Glomeruloid hemangioma leading to a diagnosis of POEMS syndrome. J Am Acad Dermatol. 2006;55:149-52.
5. González-Guerra E, Haro MR, Fariña MC, Martín L, Manzarbeitia L, Requena L. Glomeruloid haemangioma is not always associated with POEMS syndrome. Clin Exp Dermatol. 2009;34:800-3.
6. Biswas A, Byrne JP. Violaceous papules in a patient with neuropathy. Glomeruloid haemangioma associated with POEMS syndrome. Clin Exp Dermatol. 2010;35:e81-2.
7. Barete S, Mouawad R, Choquet S, Viala K, Leblond V, Musset L, *et al*. Skin manifestations and vascular endothelial growth factor S levels in POEMS syndrome: impact of autologous hematopoietic stem cell transplantation. Arch Dermatol. 2010;146:615-23.

MAILING ADDRESS:
Guida Santos
Hospital Santo António dos Capuchos-CHLC
Alameda Santos António dos Capuchos
1069-050 Lisbon
Portugal
E-mail: guidadossantos@gmail.com

How to cite this article: Santos G, Lestre S, João AMBL. Do you know this syndrome? POEMS syndrome. An Bras Dermatol. 2013;88(6):1009-10.