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Case Report

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POEMS Syndrome: The Encounter Between Neurology and Hematology

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Abstract

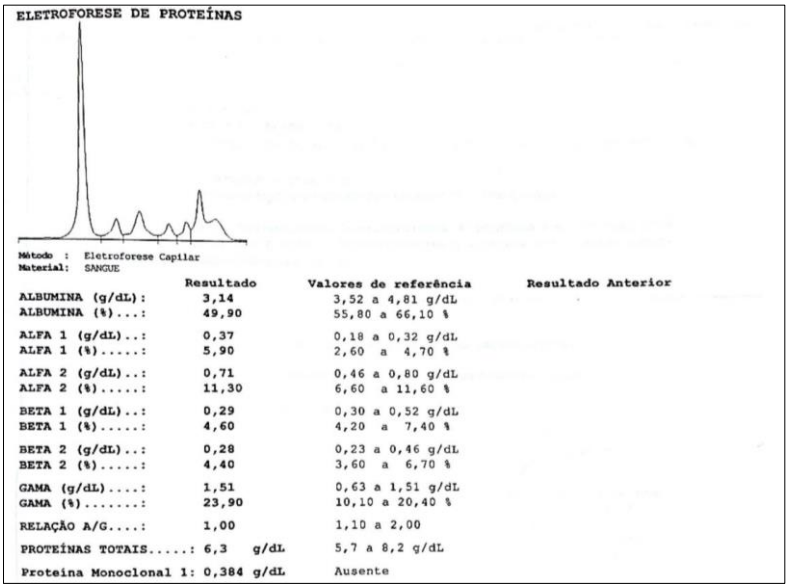
POEMS Syndrome is a paraneoplastic condition related to plasma cell dyscrasia, in which the main clinical feature is a peripheral polyneuropathy. The present report describes the case of a patient with demyelinating sensory and motor polyneuropathy, in which, after a long period of clinical investigation, from the performance of serum and urinary protein electrophoresis, a serum monoclonal peak was evidenced, followed by bone marrow (AMO) with plasma cell infiltration greater than 10%. Due to the diagnostic difficulty, it is concluded that the criteria must be broad to avoid the irreversibility of the lesions, but also meticulous enough so that the diagnosis is not established erroneously or the treatment is delayed.

Introduction

POEMS Syndrome is a paraneoplastic condition related to plasma cell dyscrasia, in which the main clinical feature is a peripheral polyneuropathy [1,2]. Due to a little-known pathogenesis, it is therefore important to assimilate the diagnostic difficulty of this condition, since, in addition to being rare, there is an unquestionable correspondence between its clinical features and that of other diseases, such as chronic inflammatory demyelinating polyradiculoneuropathy and proliferative diseases. monoclonal [2-7]. Just consider the demyelinating polyneuropathy itself, which despite denoting an important secondary axonal loss, allows demonstrating the difficulty in its recognition [1]. Nevertheless, some characteristics contribute to the clinical suspicion, among them, the fact that proximal weakness is rarely present, while the distal weakness is marked [3-7]. Furthermore, it is a painful, subacute and symmetrical sensory and motor neuropathy that does not respond to treatment with intravenous immunoglobulins [1-5,8]. Furthermore, monoclonal plasma cell disorder is also present, although its pathogenic role is not established [9]. That said, the investigation narrows down to confirmation of the presence of lambda light chain in these cells, since it is present in more than 95% of cases [3-6].

Case report

A 53-year-old male patient, with progressive muscle weakness, in addition to significant paresthesia in the lower limbs, making it impossible for him to move around without the help of a wheelchair. Initially, he was diagnosed with Guillain Barré syndrome, being treated with intravenous immunoglobulin (IVIg), but without clinical improvement. In view of this, pulse therapy with corticosteroids was initiated, as prescribed by a neurologist specializing in demyelinating diseases. Following the investigation, Electroneuromyography (ENMG) was performed, showing sensitive and motor demyelinating polyneuropathy with severe axonal loss of motor predominance. Systemic secondary processes were excluded through clinical and laboratory screening. On clinical examination, he presented hepatosplenomegaly and abdominal and axillary lymph node enlargement, in addition to lytic lesions in the spine, hip and femurs. With the electrophoresis of serum and urinary proteins, a serum monoclonal peak of 0.38 grams was evidenced, with characteristic immunofixation of IgG/kappa, and dosages of free light chains with an increase in free lambda. Bone marrow aspirate (BMA) showed normocellular content for age and plasma cell infiltration greater than 10%, which is compatible with POEMS syndrome. Therefore, the CTD (Cyclophosphamide, Thalidomide and Dexamethasone) therapeutic scheme was performed, showing evolution with progressive improvement of neurological symptoms, gradually reducing asthenia and improving sensitivity. When the patient was in the fourth cycle of CTD, Velcade (Bortezomib) was introduced, since it was released by the State and, from then on, cycles of VCD (Bortezomib, Cyclophosphamide and Dexamethasone). In April 2023, the patient underwent an autologous bone marrow transplant. At her last consultation, in July 2023, she had a good clinical response to the treatment, with slight muscle weakness, but walking without the help of devices and with full recovery of sensitivity. In addition to satisfactory laboratory response, with negative monoclonal peak and normalization of free light chains and beta 2 microglobulin.

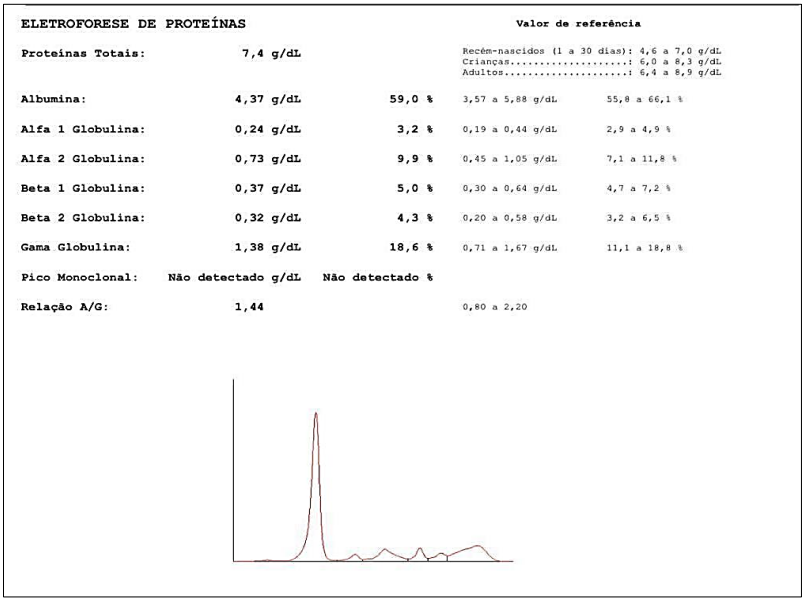


ELETROFORESE DE PROTEÍNAS

Método : Eletroforese Capilar
Material: SANGUE

	Resultado	Valores de referência	Resultado Anterior
ALBUMINA (g/dL):	3,14	3,52 a 4,81 g/dL	
ALBUMINA (%)...:	49,90	55,80 a 66,10 %	
ALFA 1 (g/dL)..:	0,37	0,18 a 0,32 g/dL	
ALFA 1 (%).....:	5,90	2,60 a 4,70 %	
ALFA 2 (g/dL)..:	0,71	0,46 a 0,80 g/dL	
ALFA 2 (%).....:	11,30	6,60 a 11,60 %	
BETA 1 (g/dL)..:	0,29	0,30 a 0,52 g/dL	
BETA 1 (%).....:	4,60	4,20 a 7,40 %	
BETA 2 (g/dL)..:	0,28	0,23 a 0,46 g/dL	
BETA 2 (%).....:	4,40	3,60 a 6,70 %	
GAMA (g/dL)....:	1,51	0,63 a 1,51 g/dL	
GAMA (%).....:	23,90	10,10 a 20,40 %	
RELAÇÃO A/G....:	1,00	1,10 a 2,00	
PROTEÍNAS TOTAIS.....:	6,3 g/dL	5,7 a 8,2 g/dL	
Proteina Monoclonal 1:	0,384 g/dL	Ausente	

Figure 1: Monoclonal peak serum protein electrophoresis (at diagnosis).



ELETROFORESE DE PROTEÍNAS

			Valor de referência	
Proteínas Totais:	7,4 g/dL		Recém-nascidos (1 a 30 dias): 4,6 a 7,0 g/dL Crianças.....: 6,0 a 8,3 g/dL Adultos.....: 6,4 a 8,9 g/dL	
Albumina:	4,37 g/dL	59,0 %	3,57 a 5,88 g/dL	55,8 a 66,1 %
Alfa 1 Globulina:	0,24 g/dL	3,2 %	0,19 a 0,44 g/dL	2,9 a 4,9 %
Alfa 2 Globulina:	0,73 g/dL	9,9 %	0,45 a 1,05 g/dL	7,1 a 11,8 %
Beta 1 Globulina:	0,37 g/dL	5,0 %	0,30 a 0,64 g/dL	4,7 a 7,2 %
Beta 2 Globulina:	0,32 g/dL	4,3 %	0,20 a 0,58 g/dL	3,2 a 6,5 %
Gama Globulina:	1,38 g/dL	18,6 %	0,71 a 1,67 g/dL	11,1 a 18,8 %
Pico Monoclonal:	Não detectado g/dL	Não detectado %		
Relação A/G:	1,44		0,80 a 2,20	

Figure 2: Serum protein electrophoresis after treatment and autologous bone marrow transplantation, with no evidence of a monoclonal spike.

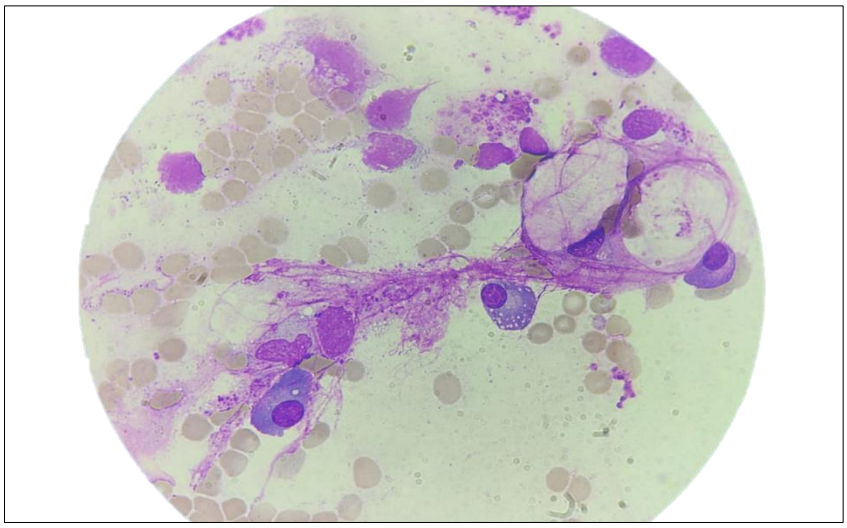


Figure 3: Photomicrograph of the patient's histological slide, containing plasmocytes and a Mott cell (plasmocyte with multiple vacuoles of immunoglobulin).

Discussion

The POEMS syndrome corresponds to a rare condition in which neurology (demyelinating polyneuropathy) and hematology (disorder of proliferation of monoclonal cells in plasma) meet, which indicates its diagnostic difficulty [7].

The incidence of this disease occurs predominantly during the 5th and 6th decades of life, with a higher prevalence in males, while in Multiple Myeloma, one of the main differential diagnoses, there is a predominance between the 7th and 8th decades [3-6].

With regard to disease activity, vascular endothelial growth factor (VEGF) becomes a target of attention, since it is a biomarker and pro-inflammatory cytokine that are elevated in this syndrome, being the main pathogenic mediator, which presupposes disease activity and response to treatment [3,4]. In addition, it is closely linked to the findings of the occurrence of microangiopathies, edema, increased pulmonary pressure, development of new blood vessels, serous effusions, polyneuropathy, in addition to leukocytosis and thrombocytosis [9]. Other findings, in addition to bone marrow infiltration, such as organomegaly, extravascular volume overload, and endocrinopathy, contribute to the diagnosis. Despite this, the latter is very unspecific [3].

With the suspicion of the syndrome, one must evaluate organomegaly, ocular fundoscopy, history and detailed neurological examination, dermatological analysis, in addition to evaluating the presence of other nonspecific gastrointestinal and endocrinological clinical manifestations [1,2]. It is recommended to start the investigation with electrophysiological studies, computed tomography of the chest, abdomen and pelvis, and bone windows, PET-CT and echocardiography [1,2,7]. It should be complemented with the hormonal dosage of testosterone, estradiol, luteinizing hormone, prolactin, serum cortisol, parathyroid hormone and TSH [7]. In addition, perform CBC, complete metabolic panel, serum immunoglobulins, electrophoresis and immunofixation, 24-hour proteinuria, VEGF, interleukin-6, biopsy and bone marrow aspirate, PCR and FISH panel for myeloma [5].

For diagnostic purposes, the presence of polyneuropathy and monoclonal peak associated with one of the major criteria is considered: osteoclerotic lesions, Castleman's disease, high levels of vascular endothelial growth factor. It is also necessary to combine at least one minor criterion, namely: organomegaly, fluid extravasation into the third space, endocrinopathy, typical skin lesions, papilledema, thrombocytosis/polycythemia [1-7].

Although the standard treatment is not yet established, it takes into account the presence of localized or disseminated bone lesions [9]. In cases where lesions occur in up to 3 sites, localized disease, the treatment consists only of radiotherapy, while, in the case of systemic involvement, this is performed through the association between chemotherapy and autologous transplantation of hematopoietic cells in eligible patients, as was done in the patient in question, who presented objective improvement of the symptomatic picture and laboratory findings. For those ineligible for transplantation, the options consist of immunomodulators associated with dexamethasone [7-9].

It should be noted that, even with well-established criteria, this is a disease whose format does not fit well with the objectivity of diagnostic parameters [10], as in the case described, in which the patient was initially treated with IVIg, according to a mistaken diagnosis of Guillain Barre.

Conclusion

The importance of understanding this rare syndrome is consolidated, since its recognition and early targeted treatment is essential for its positive prognosis, aiming at reducing the morbidity and mortality of the disease and symptomatic improvement. Incidentally, given the presence of so many traits, the criteria must be broad to avoid irreversibility of the lesions, but also meticulous enough so that the diagnosis is not erroneously established.

References

1. Angela Dispenzieri. POEMS syndrome: 2017 Update on diagnosis, risk stratification, and management: Annual clinical updates in hematological malignancies: a continuing medical education series. Am J Hematol. 2017;92:814-829. DOI: 10.1002/ajh.24802
2. Silva, R. R. P., et al. (2022). Síndrome POEMS: desafios no diagnóstico de um caso atípico. Revista De Medicina, 101(5), e-196228. https://doi.org/10.11606/issn.1679-9836.v101i5e-196228
3. Keddie, Stephen; Luna, Michael P. . síndrome de POEMS. Current Opinion in Neurology 31(5):p 551-558, outubro de 2018. | DOI: 10.1097/WCO.0000000000000610
4. Brown, R., Ginsberg, síndrome de L. POEMS: atualização clínica. J Neurol 266 , 268–277 (2019). https://doi.org/10.1007/s00415-018-9110-6
5. Scarlato, Marina; Previtali, Stefano C. . síndrome de POEMS: uma abordagem prática. Current Opinion in Neurology 24(5):p 491-496, outubro de 2011. | DOI: 10.1097/WCO.0b013e328348e107
6. Dispenzieri A. POEMS syndrome. Blood Rev. 2007 Nov;21(6):285-99. doi: 10.1016/j.blre.2007.07.004. Epub 2007 Sep 11. PMID: 17850941.
7. National Comprehensive Cancer Network.NCCN Clinical Practice Guidelines in Oncology: Multiple Myeloma. p 37- 41. agosto de 2022. www.NCCN.org
8. Hashiguchi T, Arimura K, Matsumuro K, et al. Highly concentrated vascular endothelial growth factor in platelets in Crow-Fukase syndrome. Muscle Nerve 2000;23:1.051-1.056.
9. Pinto Neto, J. V.. (2007). Síndrome de POEMS (mieloma osteoesclerótico). Revista Brasileira De Hematologia E Hemoterapia, 29(1), 98–102. https://doi.org/10.1590/S1516-84842007000100018
10. Stanley S. Levinson. POEMS syndrome: Importance of the clinical laboratory practitioner's role. Clinica Chimica Acta, 413, 1800-1807. 2012. https://doi.org/10.1016/j.cca.2012.06.032.