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Research Paper

POEMS syndrome: About a case encountered in the biochemistry laboratory at the Avicenne Military Hospital in Marrakech

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ABSTRACT:
POEMS syndrome is a paraneoplastic syndrome secondary to plasma cell dyscrasia.
It is a rare condition with a difficult diagnosis. The most common clinical presentation is progressive peripheral neuropathy associated with a monoclonal light chain, often of the lambda type. There is often an endocrinopathy, hepatosplenomegaly, condensing bone lesions, thrombocytosis or polycythemia. This clinical pleomorphism risks leading the clinician to manage each condition separately, while delaying the initiation of treatment, which would improve the functional and vital prognosis. Decisions in such cases should be made in the context of multidisciplinary consultation meetings.

KEY WORDS: *POEMS syndrome - monoclonal peak - neuropathies associated with monoclonal gammapathies*

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I. INTRODUCTION:

POEMS syndrome is an acronym for the main manifestations of the disease. It is a rare multi-visceral systemic disorder involving polyneuropathy (P), organomegaly (O), endocrinopathy (E), monoclonal gammopathy (M) and skin lesions (S). Atypical presentations may be accompanied by severe visceral involvement, particularly neurological and renal. The nature of the plasma cell dyscrasia is characterised by a monoclonal component, often in small amounts, with a lambda isotype light chain and the presence of often single condensing bone lesions.

Given the rarity of this condition, we thought it would be interesting to report the case of a patient diagnosed at the Biochemistry Laboratory of the Avicenne Hospital in Marrakech in collaboration with the haemato-oncology department, in order to recall this rare disease of multidisciplinary diagnosis.

II. CASE REPORT:

A 72 year old patient, type 2 diabetic for 2 years, recently put on insulin, admitted to the haematology department for progressive oedema and pain in all 4 limbs.

The clinical examination found a patient in poor general condition, bedridden (WHO=4), febrile with neuropathic pain in all 4 limbs, hyperpigmented skin lesions on the face and hands with snoring rales on both lung bases.

In front of this symptomatology, the initial check-up showed a hyperleukocytosis of 15000 elements/mm with PNN predominance, a CRP of 120 mg/l, the ECBU and radiothorax showed the presence of a pulmonary and urinary infection, treated by broad spectrum antibiotic therapy with gain of apyrexia and normalization of the CRP

As the symptoms did not improve and the general state of health was profoundly altered, a second check-up was requested: serum and urine protein assays, plasma protein electrophoresis (PPE), an immunological check-up (NAA and anti-CCP), an electromyogram (EMG) and a thoracic-abdominal-pelvic

scan, which revealed a left adrenal nodule that required a hormonal check-up with a pituitary MRI. Is it a POEMS syndrome? Or a malignant adrenal gland or prostate tumor with bone metastases?

The diagnosis of POEMS syndrome was therefore made on the basis of the following criteria: Hyperpigmented skin lesions on the face and hands, neuropathic pain in all 4 limbs with EMG sensory-motor axonal polyneuropathy in all 4 limbs, monoclonal gammopathy (peak in B2globulins estimated at 10g/l) of the Ig A-lambda type on serum immunofixation, with negative Bence-Jones proteinuria, normal myelogram and osteo-medullary biopsy, osteocondensing bone lesions of the spine and pelvis with hypogonadotropic hypogonadism without abnormalities on pituitary MRI.

Therapeutically, the patient was then treated according to the CyBorD protocol (Cyclophosphamide-Bortezomib-Dexamethasone), with a favourable clinical course.

III. DISCUSSION :

POEMS is more common in Japan according to Dispenzieri et al [1] and Nakanishi et al [2]. Our patient was male with an age of 72 years; in the series consulted, men are twice as affected as women, while the average age of onset is between 40 and 50 years [3].

The pathophysiology starts with plasma cell dyscrasia, leading to a true angiopathy, via an angioactive cytokine: Vascular Endothelial Growth Factor (VEGF) [4]. There are no validated criteria for POEMS syndrome. Dispenzierri proposed diagnostic criteria to exclude patients with neuropathy associated with monoclonal gammopathy of undetermined significance, multiple bone myeloma or Waldenström disease from POEMS syndrome. The presence of a monoclonal kappa component does not exclude the diagnosis of POEMS syndrome, but it makes it unlikely [5]. To retain the diagnosis, two major criteria are required: polyneuropathy, monoclonal plasma cell proliferation and at least one minor criterion among the following: osteocondensing lesion, Castleman's disease, organomegaly, oedema (peripheral, pleurisy, ascites), endocrinopathy (adrenal, pituitary, parathyroid, thyroid, diabetes), skin manifestations (hyperpigmentation, hypertrichosis, angiomas, white nails, papillary oedema) [6]

Neuropathy is the main presenting symptom, but with a difference in frequency in the 3 series studied: Dispenzierri at 95%, Soubrier at 84%, and Nakanishi at 51% [1-2-3]. In our patient, the neuropathy was peripheral, symmetrical, sensitivomotor, and progressively progressive. This is the same description found in the literature.

Plasma dyscrasia is constant; it is 100% present in Soubrier's series, 75% in Nakanishi's, and 89% in Dispenzierri's. Serum protein electrophoresis may show a narrow peak, but even in its absence, blood and urine immunoelectrophoresis or immunofixation should be requested, as the monoclonal component is often small or consists only of a light chain [7-8]. The light chain is almost always of the lambda isotype, which is similar to our case (100% of cases in the Soubrier series and in the Mayo Clinic series, 95% in the Nakanishi series). In our patient, a monoclonal IgA-lambda peak was found in the blood. The myelogram and BOM were unremarkable.

The endocrinopathy is polymorphic; the frequency of diabetes differs significantly in the 3 series consulted (Nakanishi: 28%, Soubrier: 41%, Dispenzierri: 3%). In general, patients have several endocrine manifestations that may be of peripheral or central origin, namely hypogonadism, hypothyroidism, adrenal insufficiency, hypo- or hyperparathyroidism, acromegaly [9]. In our case, the patient had diabetes associated with hypogonadism without other hormonal abnormalities. The skin abnormalities initially described included hyperpigmentation, skin thickening and hypertrichosis. Hyperpigmentation, found in our patient, is the most frequent skin manifestation according to the literature [10]. However, no organomegaly or PDA was noted. Abdominal ultrasound and thoracic-abdominal-pelvic CT were negative in this sense. Bone lesions are common in the literature (Dispenzierri 97%, Soubrier 68%, Nakanishi 54%), the most typical appearance being multiple condensing nodules of variable size on the pelvis, spine, ribs or skull, which is the same as in our patient's case. In addition to the tumour manifestations, other radiographic manifestations can be observed. In addition to the tumour manifestations, other radiographic manifestations can be observed. These are spiculated enthesopathic bone proliferations, irregular in the rachis and pelvis [2-3].

Treatment is based on the eradication of the abnormal plasma cell clone, Radiotherapy is the treatment of choice for plasmacytoma. Corticosteroid therapy alone improves 15% of patients and stabilises 7% in Dispenzierri's experiment. Alkylating agents improve in 40% of patients. Bone marrow autotransplantation leads to a regression of clinical manifestations and normalises VEGF levels [11-12-13].

To date, no biochemical or genetic risk factors are known to have an effect on overall survival. The prognosis of POEMS syndrome is good with a median survival of 13.8 years in the Mayo Clinic series. The number of events does not affect survival. Death is not due to progression of plasma cell dyscrasia, but mainly to cardiac, respiratory or infectious causes [1-2-3].

IV. CONCLUSION:

Although the pathophysiology is incompletely elucidated, the increase in VEGF seems characteristic of the syndrome, which may explain the multisystemic nature of the disease. The diagnosis of POEMS syndrome implies a unicist view of a set of conditions that are likely to be managed separately, delaying treatment that may improve functional and vital prognosis.

According to the case presented in this observation and in order to reduce the diagnostic and therapeutic delay and to improve the prognosis; any patient with unexplained peripheral neuropathy should benefit from electrophoresis and immunoelectrophoresis to look for a monoclonal component. If a monoclonal component is present, or if there is other evidence to suggest the diagnosis, the work-up should be completed with skeletal films, bone marrow MRI and CT scan of the pelvis to look for plasma cell lesions. An assessment of systemic involvement is then performed before considering treatment, which depends on the type of plasma cell dyscrasia.

REFERENCES:

[1]. Dispenzieri A, Kyle RA, Lacy MQ, Rajkumar SV, Therneau TM, Larson DR et al. POEMS syndrome: definitions and long-term outcome. Blood. 2003;101(7):2496-506. **PubMed** | **Google Scholar**

[2]. Nakanishi T, Sobue I, Toyokura Y, Nishitani T, Kuroiwa Y, Satoyoshi E et al. The crow-Fukase syndrome : a study of 102 cases in Japan. Neurology. 1984; 34(6):712-20. **PubMed** | **Google Scholar**

[3]. Soubrier MJ, Dubost JJ, Sauvezie BJ. POEMS syndrome: a study of 25 cases and a review of the literature. French Study Group on POEMS syndrome. Am J Med. 1994; 97(6):543-53. **PubMed** | **Google Scholar**

[4]. Dyck PJ, Engelstad J, Dispenzieri A Vascular endothelial growth factor and POEMS. Neurology. 2006; 66: 10-2.

[5]. Bardwick PA, Zvaifler NJ, Gill GN, Newman D, Greenway GD, Resnick DL. Plasma cell dyscrasia with polyneuropathy, organomegaly, endocrinopathy, M protein, and skin changes: the POEMS syndrome. Report on two cases and a review of the literature. Medicine (Baltimore). 1980; 59: 311-22.

[6]. Lagueny A, Bouillot S, Vital C, Ferrer X, Larrieu JM, Vital A Le syndrome POEMS (ou syndrome de Crow-Fukase). Rev Neurol (Paris). 2004; 160: 285-95.

[7]. T. Lenglet, La polyneuropathie du syndrome POEMS La Lettre du Neurologue Nerf & Muscle • Vol. XIII - n° 9 - octobre 2009

[8]. N. Meuleman, Pitfalls of monoclonal gammopathy Rev Med Brux 2013 ; 34 : 335-8

[9]. Dr Patrice H. Lalive Neuropathies associées aux gammapathies monoclonales Revue Médicale Suisse – www.revmed.ch – 29 avril 2009

[10]. Dispenzieri A. POEMS Syndrome: 2019 Update on diagnosis, risk-stratification, and management. Am J Hematol. 2019;94:812–827. https://doi.org/10.1002/ajh.25495

[11]. Kuwabara S, Misawa S, Kanai K, Kikkawa Y, Nishimura M, Nakaseko C et al Autologous peripheral blood stem cell transplantation for POEMS syndrome. Neurology. 2006; 66: 105-7.

[12]. Jaccard A, Royer B, Bordessoule D, Brouet JC, FerMand JP

[13]. High-dose therapy and autologous blood stem cell transplantation in POEMS syndrome. Blood. 2002; 99: 3057-9.

[14]. Dispenzieri A, Gertz MA .Treatment of POEMS syndrome. Curr Treat Options Oncol. 2004; 5: 249-57.