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Hirsutism will be not Considered as Sign of Progressive Disease after Autologous Transplant in POEMS Syndrome

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Abstract

POEMS (polyneuropathy, organomegaly, endocrinopathy, M-protein and skin changes) syndrome is a rare multisystem disease characterized by plasma cell dyscrasia and overproduction of vascular endothelial growth factor (VEGF). Skin changes are usually multiple cutaneous angiomas, leukonychia, necrotizing vasculitis, cutaneous thickening of sclerodermiform type and hypertrichosis. VEGF is assumed to be useful in monitoring disease activity, because VEGF levels usually decrease after treatment. We described the case of a young woman with POEMS syndrome characterized by hirsutism and hypertrichosis who underwent autologous bone marrow transplantation. Evolution was hallmarked by an improvment of radiological signs and biological markers specially VEGF, while hirsutism didn't regress.

Keywords: POEMS; Autologous transplant; Hyrsutism; VEGF

Introduction

POEMS syndrome is a rare (prevalence 0.3 per 100 000) multisystemic disorder frequently mistaken for other diseases as "Chronic Inflammatory Demyelinating Neuropathy (CIDP)". The diagnosis of POEMS was made considering the concomitance of sensorimotor demyelinating polyneuropathy splenomegaly, hypothyroidism, monoclonal gammopathy, skin changes and an increase of VEGF blood level [1-3].

Ninety percent of patients had a recognized cutaneous manifestation. Hyperpigmentation and hemangioma were most common (47%), followed by hypertrichosis (38%); these manifestations seems to be not evaluable to the response of therapy [3-5].

Hirsutism is a frequent manifestation (26-74%) [1] as also the skin changes that has been reported in 46-93% of the patients. The most important parameters to evaluate the patient at the diagnosis and after the therapy are: M-protein, fluorodeoxyglucose (FDG) avidity on a PET scan and VEGF plasma level [6-8].

In POEMS syndrome improvement in peripheral neuropathy, extravascular fluid overload (ascites/effusions/edema), or pulmonary function tests, are predictive of response. The most common progression events have been Forse laboratory followed by rising VEGF [9].

In May 2014 a thirty-eight years old woman was admitted in our hematology department with the diagnosis of POEMS. Her clinical history began in july 2013 when she was admitted in neurological ward with progressive weakness, tingling and numbness in lower limbs for six months.

After the electrophysiological examination suggestive for axonal demyelinating polyneuropathy the diagnosis of CIDP was made and patient received steroids therapy obtaining a little improvement of the disease and then in november 2013 for progressive strength loss and paresthesia involved the legs, causing inabilityv to walk, she received a new cycle of steroid associated to immunoglobulin therapy(2 g/Kg during 5 days). On that time physical examination showed bilateral pedal edema, skin thickeness and an evident hypertricosis.

Cognitive functions were normal. Examination of the fundi revealed bilateral papilledema, but otherwise cranial nerve examination was unremarkable; test showed a mild trombocytosis (663x10^9/ microl) with a normal iron load and hemoglobin levels. Bcr-abl

and Jak 2 rearrangement were done and did not show any alteration. The total serum protein count and protein electrophoresis was normal (gamma 12.5%) without monoclonal M spike. Immunoelectrophoresis using the immunofixation method revealed a monoclonal IgA lambda band. Thyroid function tests revealed mild hypothyroidism. The prolactin level was very high (56.3ng/ml normal range 3.5-26.5). Immunological and mycrobyological tests were normal a part of the positivity of HbsAg and anti HbcAb without HBV Dna reduplications. Abdominal ultrasonography showed hepatomegaly (16 cm) and moderate ascites. The bone marrow biopsy from right iliac crest showed increase of plasma cells (35-40%) with light hypoplasia of normal haematopoiesis. Total bone X ray and cerebral-spine MRI did not show bone lesion. The VEGF level was high 4164 pg/ml. The diagnosis of POEMS syndrome was done.

She was included in our planned protocol and she received after lamivudine prophilaxis two cycle of intermediate dose of cyclophosphamide (4800mg) monthly in June and July 2014, and autologous peripheral stem cell collection.

In August the neurological visit described a inability of the patient to move and to maintain erect position autonomally and paralysis of lower limbs and of the distal and semi-distal muscles of the upper limbs and of the neck. The areflexia was generalized sign.

In August 2014 she received autologous peripheral stem cell infusion after a conditioning chemotherapy with melphalan 200 mg / m^2. The engraftment was obtained at day 20 for neutrophils (> 500/mmc)land at day 32 for platelets count (20x109/mmc).

Then patient started a long reabilitative therapy. Bone marrow biopsy was any VEGF assay returned in normal range (758pg/ml) Five months after transplantation despite a gradually neurological improvement the patient showed a dramatical worse of hypertricosis suspect for relapsing disease (Figure 1 and 2).

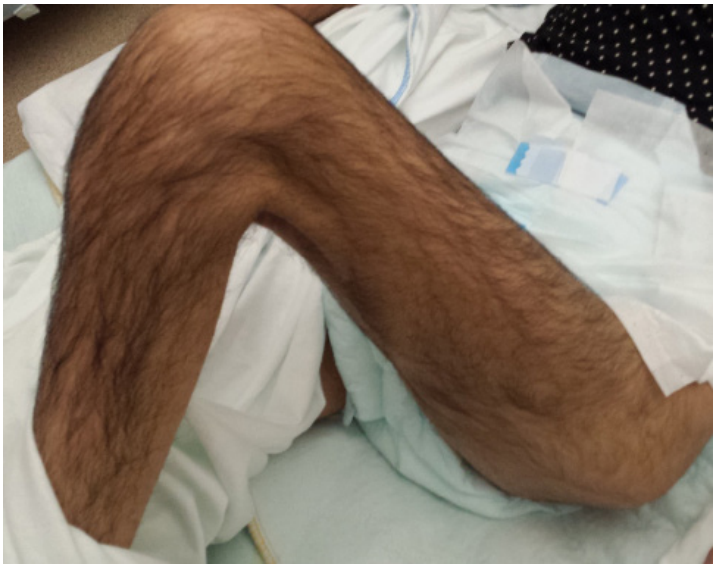

Figure 1: Patient legs after transplantation

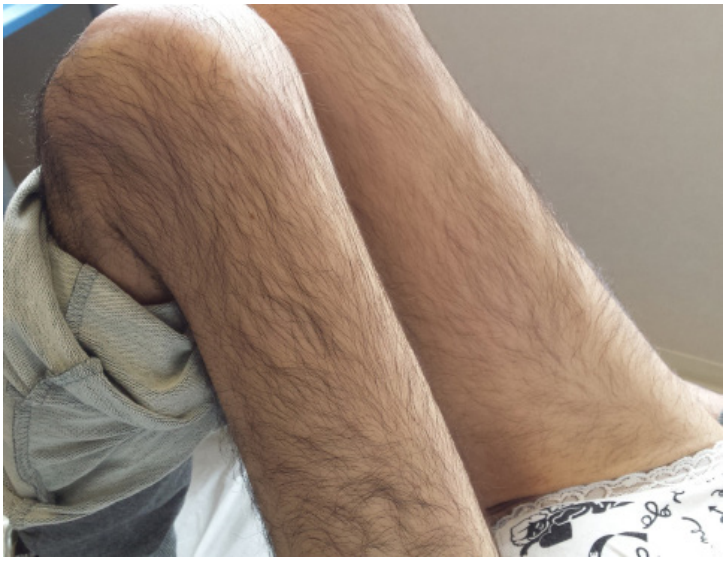

Figure 2: Patient legs after transplantation

The haematological value and bone marrow biopsy was absolutely normal persisting only a little monoclonal gammopathy IgA lambda. Endocrinologist consultant made diagnosis of hyrsutism not related to an increased androgen levels because of the normal level of all the sex hormone (Serum hCG, FSH, LH, estradiol, prolactin, testosteron and DHEAS).

Only after eight months post transplantation the excessive body hair decreased and patient is now in complete remission 24 months from transplantation with her normal body hair (Table 1).

	Diagnosis	After 24 months
Monoclonal Component	IgA lambda	none
IgA level	1164	114
VEGF level	4164	768
Bone marrow plasma cells %	20	2
Platelets count *109/l	663	239

Table 1: Diagnosis after 24 months

Discussion

Here we report a case of progressive hyrsutisms appeared after 5 months from transplant who did not correlate with progressive disease. Hypertrichosis and hyrsutisms seems to be correlate with particular light chain variable region (IGLV) genes in patients with POEMS syndrome as IGLV 1-40 that had more severe neuropathy, hypertrichosis, and papilledema [10].

Patients with POEMS syndrome present with a complex conglomerate of symptoms and signs, and early diagnosis and a prompt multidisciplinary approach increase the likelihood of reduced long-term irreversible morbidity [12]. The best choice of therapy, is alkylators, either in the form of low dose conventional therapy or high dose with stem cell transplantation. Lenalidomide shows promise with manageable toxicity. Thalidomide and bortezomib also have activity, even if their risk of exacerbating the peripheral neuropathy will be conidered. The benefit of anti-VEGF antibodies is conflicting [2].

During the follow-up, the reduction or normalization of VEGF, serum M-spike, PET scan and bone marrow or peripheral blood values, will be periodically tested for its correlations with disease activity.

On the opposite, neurological problems, and skin changes need of more time to show an improvements and sometimes probably due to the therapies could show a transitory worsened. For this reason the neurological and dermatological problems do not be consider as parameter to test the response to therapy during the first follow-up; at the same time their impairment could not consider as a sign of progression.

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