



Multiple Myeloma Relapse Revealed by a Solitary Skin Plasmocytoma

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Abstract

We describe an unusual case of early multiple myeloma relapse as a solitary skin plasmocytoma, in a 65-year-old woman in very good partial response (VGPR)* after induction regimen and autologous stem cell transplantation.

*International Myeloma Working Group (IMWG) 2010 Uniform Response Criteria for Multiple Myeloma.

Clinical Image

A 65-year-old woman diagnosed with an IgG Lambda multiple myeloma (MM) eight months earlier attended a follow-up hematologic consultation. She underwent a high-dose melphalan chemotherapy and autologous stem cell transplantation, after induction regimen with four cycles of bortezomib, lenalidomide and dexamethasone, five months earlier. She had recently begun a lenalidomide maintenance therapy. A bone marrow biopsy realized one month prior to consultation did not show remnant clonal plasma cells. A serum immunosubtraction revealed a slight IgG Lambda unquantifiable band, together with a normal free light chain ratio. A cytogenetic analysis revealed the persistence of a complex and hyperploid karyotype including a chromosome 1p gain. These abnormalities were already present at diagnosis.

At this follow-up consultation, a round 4 cm large mass surrounded by a dense reticular small vessel net WAS found at clinical exam (Figure 1). A biopsy of the lesion revealed highly proliferative malignant plasmocytes (MIB-1 staining >95%) with vascular plasma cell thrombi. Immunostaining for MYC was intensely strong but negative for BRAF. The later was confirmed by RT-PCR. A FISH analysis did not reveal a MYC rearrangement. PET-CT restaging revealed the described lesion (SUVmax= 4.6 g/ml) but also a centimetric subcutaneous lesion of the right thigh (SUVmax=1.9 g/ml) and a lymph node of the left external iliac region (SUVmax=4.4), together with disseminated highly metabolic bone lesions (SUVmax between 5 g/ml and 6.6 g/ml). The cutaneous lesion was treated with radiotherapy (10 Gy x 3 Gy) with a good response (Figure 2). Systemic treatment was then pursued with carfilzomib, lenalidomide and dexamethasone, until a diffuse soft-tissue, cutaneous and lymph node spread, together with a biological relapse, surged 6 months later.

Extrasosseous myeloma (EM) may surge during the disease course in almost one fifth of patients [1,2]. Its incidence is increasing, due to more extensive radiological staging, extended survival and suspected selection of more aggressive myeloma cell clones after allogenic stem cell transplant [3,4]. A shift from intact immunoglobulin towards light chain only secretion is possible [5].

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Figure 1: Plasmocytoma of the skin (left thigh). Note the dense reticular vascular bed surrounding the lesion.



Figure 2: Plasmacytoma of the skin (left thigh), 8 weeks after radiotherapy (10 Gy × 3 Gy) and pursue of systemic treatment with carfilzomib, lenalidomide and dexamethasone.

Although several breakthroughs have been made in MM treatment, this subgroup of patients still carries a poor prognosis [6]. Liver is the more frequent extraosseous site of disease spread at relapse [7]. Skin involvement is rare in the relapse setting and more prevalent at diagnosis [7]. Underlying its rarity, a recent literature review gathered 44 published cases of EM involving extremities soft tissues [8].

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