CASE REPORT

A case of relapsed systemic multiple myeloma mimicking adenopathy and extensive skin patch overlying a plasmacytoma

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INTRODUCTION

Multiple myeloma rarely metastasizes to the skin and when it does, it usually manifests as cutaneous nodules. In this report, we describe a new clinical presentation of metastatic multiple myeloma mimicking AESOP syndrome in association with polyneuropathy, organomegaly, endocrinopathy, monoclonal protein, and skin changes (POEMS) syndrome.

CASE REPORT

A 70-year-old patient presented with a 6-month history of a slowly enlarging violaceous back plaque associated with a progressive dorsal pain. He also complained of hand and feet paresthesia. Physical examination revealed a large, ill-defined, vascularized, violaceous plaque in the middle upper portion of his back (Fig 1). A few areas of the plaque were infiltrated. There were multiple vascular-looking papules in its center and linear vessels on its borders. A voluminous supraclavicular lymphadenopathy was also noted.

The patient had a 6-year history of IgA-L multiple myeloma, for which he had an allogenic bone marrow transplant and multiple lines of chemotherapy because of relapses. At the time of his visit, he had been receiving ixazomib for >1 year.

Contrast-enhanced chest-abdomen-pelvis computed tomography revealed a voluminous infiltration of subcutaneous tissues in the upper portion of his back with an underlying progressive osseous hyperdensity extending from thoracic vertebrae 6 to 9. Numerous stable, lytic bone lesions compared with previous computed tomography scans, bilateral axillary lymphadenopathy, and pleural effusion were also noted. Thoracocentesis confirmed the presence of plasma cells in the pleural effusion.

Three cutaneous punch biopsies were performed in different areas of the plaque. The first 2 biopsies, 1 performed in a vascular-looking papule and the other in a noninfiltrated part of the plaque, revealed a massive dermal lymphoproliferation (Fig 2) with a morphology and immunophenotype (CD3+/CD0+, CD20+/CD0+, CD38+, CD138+, MUM1+, CD56+, Bc12+, EMA+, Bc11+, CD79+) compatible with a plasma-cell hemopathy. Lambda monoclonal restriction was observed (Fig 3). The third biopsy, performed in an infiltrated zone of the plaque, revealed dermal capillary dilations and mucin deposition in the papillary dermis, highlighted by Alcian blue and colloidal iron colorations (Fig 4).

Initially, adenopathy and extensive skin patch overlying a plasmacytoma (AESOP) syndrome was considered; however, because of systemic involvement of the myeloma, relapsed systemic multiple myeloma with cutaneous extension was a more accurate diagnosis. The patient also fulfilled the diagnostic criteria for POEMS syndrome.
Unfortunately, the patient died of respiratory insufficiency secondary to the progression of his multiple myeloma before initiation of a treatment.

DISCUSSION

Multiple nonspecific dermatologic entities are associated with monoclonal gammopathy. Acquired cutis laxa, cryoglobulinemia, mucinoses, neutrophilic dermatoses, necrobiotic xanthogranuloma, and normolipemic xanthoma are a few examples.1 However, cutaneous metastases of multiple myeloma are rare, with <100 cases published. They appear at an advanced stage of the disease and usually manifest as skin colored to violaceous or erythematous nodules. Immunohistochemical staining for CD79a, CD138, and epithelial membrane antigen is generally positive, whereas CD38 and CD43 exhibit more variable immunoreactivity.2,3 To our knowledge, this is the first case of systemic multiple myeloma with cutaneous metastasis mimicking AESOP syndrome to be published.

AESOP syndrome is a rare clinical entity characterized by an enlarging erythematous-to-violaceous skin patch overlying a solitary bone plasmacytoma with locoregional adenopathy.4 Most published cases were secondary to monoclonal gammopathy of unknown significance or POEMS syndrome with or without Castleman disease.5,6 The characteristic histopathologic findings of AESOP syndrome are diffuse capillary dilations that can be accompanied by dermal deposition of mucin and a mild inflammatory infiltrate of variable composition.7,8 CD31 immunohistochemical staining can also be positive.9 The exact cause of AESOP syndrome is unknown, but it is postulated that overproduction of vascular endothelial growth factor by the plasmacytoma could be responsible for the clinical manifestations of this paraneoplastic syndrome.1,6,8,9

Both multiple myeloma and AESOP syndrome can be associated with POEMS syndrome.5,8 POEMS
syndrome is defined by the presence of both peripheral neuropathy and monoclonal plasma-cell disorder plus 1 major and 1 minor criterion. The 3 major criteria for the diagnosis of POEMS syndrome are osteosclerotic bone lesions, elevated vascular endothelial growth factor levels, and Castleman disease. The 6 minor criteria are endocrine abnormalities, skin changes, organomegaly, extravascular volume overload, thrombocytosis, and papilledema. Our patient fulfilled both mandatory criteria and had an osteosclerotic bone lesion plus skin changes and extravascular volume overload.

In conclusion, this case of metastatic multiple myeloma with cutaneous involvement mimicking AESOP syndrome highlights a new presentation of multiple myeloma skin metastasis. Although both entities can present in association with POEMS syndrome, the main differences are that in AESOP syndrome the plasmacytoma is localized, and the prognosis is more favorable.

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Conflicts of interest
None disclosed.

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