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Case Presentation

Cutaneous involvement in multiple myeloma: a case report with an unusual location

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Abstract

Multiple myeloma (MM) is a rare cancer. Cutaneous involvement is uncommon with fewer than 100 cases described in the literature. We report herein a patient with MM, subtype IgA kappa, with unusual clinical presentation including the lower lip.

Keywords: Multiple myeloma, skin manifestations, mouth mucosa.

Introduction

Multiple myeloma (MM) is a rare cancer characterized by the presence of a paraprotein combined with varying degrees of organ impairment most commonly anemia, renal failure hypercalcemia, bone pain and recurrent infections [1]. Cutaneous involvement is uncommon with fewer than 100 cases described in literature. We present herein a new case with some particularities.

Case synopsis

An 86-year-old woman, with a past medical history of hypertension, was hospitalized on May 2010 in the nephrology department for chronic renal failure, progressing for 18 months, associated with anorexia, asthenia, and weight loss. Multiple explorations revealed an erythrocyte sedimentation rate > 100 mm/hour, normochromic normocytic anemia, and renal failure (urea: 17 mmol/l (normal range 2- 6.7 mmol/l) and creatinemia: 248 umol/l (normal range 20-120 umol/l), normal calcemia and phosphatemia. Serum protein electrophoresis showed a monoclonal peak of immunoglobulin isotype IgA kappa. Bone marrow aspirate contained 79% dystrophic plasma cells. Urine tests were negative. A skull X-ray showed multiple “punched-out” holes. Based on these clinical, biological, and radiologic findings the diagnosis of multiple myeloma (MM) subtype IgA kappa was made. Consequently, she received 3 cycles of chemotherapy with melphalan (0.1 mg/kg/day) and prednisone (1 mg/kg/day), but the disease continued to progress. On July 2010 (3 months after diagnosis) multiple firm, fixed violaceous nodules appeared on the back, abdomen, and arms, associated with multiple small sub-cutaneous skin-colored nodules (Figure 1). There was also, a skin- colored mass of 4 to 5 cm in front of the right clavicle (Figure 2) and a 0.5 cm violaceous plaque on the inferior lip (Figure 3). Skeletal survey was normal.
Multiple biopsies at different sites, including the lip, were done. The histological study showed a diffuse interstitial pattern with a trabecular disposition of neoplastic cells all over the deeper dermis and hypodermis (Figure 4 A), showing plasmacytic differentiation. These plasmablastic cells showed mild nuclear atypia with numerous mitoses (Figure 4 B). The immunohistochemical study showed strong immunoreactivity for CD138 (Figure 4 C) and epithelial membrane antigen (EMA). However, immunoreactivity for CD79a was weak (Figure 4 D). Based on all these findings, the diagnosis of an MM with cutaneous involvement was established. The patient died 1 month after appearance of the skin lesions.

Figure 1. Multiple skin-colored and erythematous cutaneous and subcutaneous nodules on the back

Figure 2. A giant tumor of 4-5 cm in front of the right clavicle with no adjacent bony lesions
Figure 3. An angiomatoid plaque on the inferior lip.

Figure 4. A diffuse interstitial infiltrate involving the entire dermis with a trabecular disposition of neoplastic cells (H and E, x 40). B. Plasmablastic cells showing mild nuclear atypia but numerous mitoses (H and E, x200). C. Strong expression of CD138 establishes the
plasmacyte lineage of the cells (CD138 stain, x 200). D. weak expression of CD79a (CD79a stain, x 200) (These histological features are of the lip lesion).

Discussion

Cutaneous involvement in patients with MM is very uncommon with fewer than 100 cases described in the literature [2-8]. Metastatic cutaneous lesions generally appear late in the course of the disease. Nevertheless, they may occur as the first manifestation of the disease [5] or at an earlier stage. In our reported case the skin lesions appeared 3 months after diagnosis of the MM. Cutaneous involvement in MM occurs most frequently in middle age to older males with the IgG and IgA subtype[4]. It is associated in 56% of cases with IgG subtype MM [6]. Skin lesions appear commonly as multiple papules, plaques, and/or cutaneous and subcutaneous nodules, with firm consistency, smooth surface, and skin-colored, red or violaceous color [7]. The skin lesions may appear at any site of the skin, but have been reported most commonly on the back, abdomen, and less frequently, on the scalp, face, and extremities. Only one case reported in the literature was localized to the oral mucosa, the tongue [3]. Our case purports to be the first to illustrate MM of the inferior lip. The cutaneous lesions from MM, occur most commonly as direct extension from underlying bone lesions. The occurrence of metastatic skin lesions is a rare phenomenon [3, 4, 5, 6]. As it was in the case of our patient, the skeletal survey did not show any bony lesions underlying the cutaneous lesions.

Histopathologically, the skin lesions of MM show 2 distinct patterns: interstitial and nodular. This latter is the most prevalent [3]. The histological aspect in our case showed a diffuse interstitial pattern with a trabecular disposition of neoplastic plasma cells.

Recent cytogenetic studies [3, 4, 8], whether in bone marrow plasma cells or in skin lesions, have demonstrated that the deletion of the rb-1 retinoblastoma gene is a great marker of aggressive clinical and biologic course and poor response to chemotherapy. Unfortunately, this cytogenetic study was not done in our case. The clinical course was characterized by a rapid progression of the illness and the death of the patient 1 month after the onset of the skin lesions. This supports the belief that cutaneous involvement in MM is a poor prognosis factor.

Conclusion

Cutaneous involvement in MM is rare, characterized by broad spectrum of clinical aspects that can affect any area of the skin. We report herein another case with multiple metastatic MM skin nodules with unusual site on the lip, with no underlying bone lesions.

References