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Cutaneous Involvement in Multiple Myeloma: A Case Report

Multipl Myelomda Deri Tutulumu: Bir Olgu Sunumu

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Özet

Multipl myelom (MM) kemik iliğinden köken alan ve monoklonal immunglobulin üreten plazma hücreleri ile karakterize hematolojik bir malinignitedir. MM'li hastalarda deri tutulumu çok nadir görülmektedir. Yetmiş üç yaşında, IgG lambda MM tanısı olan hasta deri lezyonları nedeniyle Dahiliye bölümü tarafından bölümümüze konsülte edildi. Yapılan dermatolojik muayenede üst ekstremitelerde, gövde ön ve arka yüzde dağınık yerleşimli, çapları 2-5 cm arasında değişen, eritemli viyolase renkte toplam 10 adet tümöral lezyon izlendi. Lezyonlardan alınan biyopsinin histopatolojik incelemesinde lambda monoklonaliteli diffüz atipik plazma hücre infiltrasyonu saptanması nedeniyle hastanın lezyonları MM deri tutulumu olarak değerlendirildi.

Anahtar Kelimeler: Deri tutulumu, metastaz, multipl myelom.

Abstract

Multiple myeloma (MM) is a hematological malignity that originates from the bone marrow and is characterized by plasma cells that produce monoclonal immunoglobulin. Cutaneous involvement is very rare in patients with MM. A 73-year-old, male patient, who has a diagnosis of IgG lambda MM, was referred to our clinic from the Department of Internal Medicine due to his skin lesions. The dermatological examination revealed a total of 10 erythematous violaceous-tumoral lesions with a diameter varying between 2-5 cm, on upper extremities, the front and back of the body. The lesions of the patient were considered as cutaneous involvement of MM because histopathological examination of skin biopsy is showed diffuse atypical plasma cell infiltration with lambda monoclonality.

Keywords: Skin involvement, metastasis, multiple myeloma.

Introduction

Multiple myeloma (MM) is a hematological malignity that originates from the bone marrow and is characterized by plasma cells that produce monoclonal immunoglobulin. Cutaneous involvement is very rare in patients with MM (1). It usually occurs at the late stages of MM as a reflection of increased tumor cells and indicates a poor prognosis. Here, we present a case of MM with skin involvement.

Case Report

A 73-year-old, male patient was referred to our clinic due to his skin lesions. There was bone involvement in the patient who had been under follow-up by the Department of Internal Medicine with the diagnosis of MM since October 2010. The patient was unresponsive to melphalan and prednisolone treatment. The skin lesions appeared one year after the establishment of the diagnosis. The dermatological examination revealed a total of 10 erythematous violaceous-tumoral lesions with a diameter varying between 2 and 5 cm, on upper extremities and the front and back of the body (Fig. 1a).

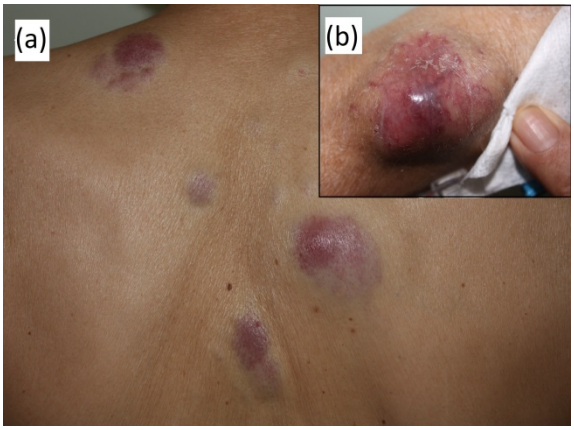

Figure 1. (a) Violaceous-erythematous tumors 2-5 cm in diameter on the back; **(b)** Vascular dilatations were noted on some of the lesions.

Vascular dilatations were noted on some lesions (Fig. 1b). Histopathological examination of skin biopsy is showed tumor formation in subepithelial tissue (Fig. 2a).

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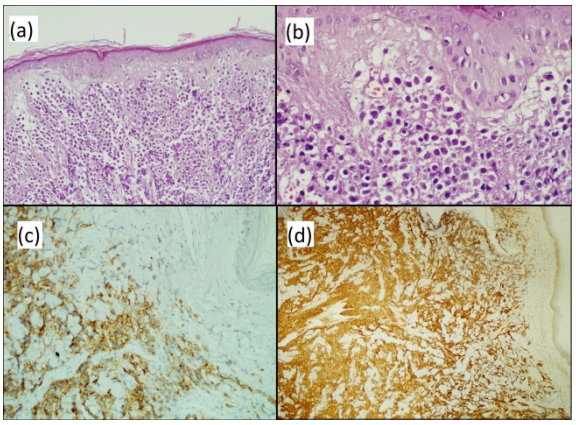

Figure 2. **(a)** Tumor infiltration in dermis (HEX10); **(b)** The tumor consisted of atypical plasmoblasts (HEX40); **(c)** Positive staining by CD38 in the immunohistochemical study (X20); **(d)** Positive staining by Lambda in the immunohistochemical study (X10).

The tumor consisted of atypical plasmoblasts with large hyperchromatic nucleus, distinct nucleolus and large eosinophilic cytoplasm (Fig. 2b). In the immunohistochemical study, the atypical cells showed a strongly diffused positive staining by CD38 and Lambda and negative staining by Kappa, CD79 alpha, CD20 and CD10 (Fig. 2c, d). Based on these results, the lesions were considered as cutaneous involvement of MM. The patient died after 6 months of the onset of skin metastases.

Discussion

The skin manifestations of MM may be observed in 3 forms as specific dermal plasma cell infiltrations, skin symptoms mediated by abnormal protein (amyloid, cryoglobulins) levels and other paraneoplastic disorders (2). Specific cutaneous involvement is very uncommon in MM and most of the cases were male and were in the age range of 36 and 81 (1). Cutaneous involvement mostly occurs as a direct extension of the underlying osteolytic bone lesions; however, it may also manifest as metastatic deposits in the form of a large number of lesions. Metastatic skin lesions occur as erythematous violaceous, hard and smooth-surfaced papules, nodules or plaques, 1 to 5 cm in diameter. In the literature, vascular tumor-like lesions have been reported (3). Cutaneous lesions occur most commonly on the back and abdomen. A large number of lesions are usually seen in patients. Lesions may be ulcerated or secondarily infected (4). In our patient there were a large number of erythematous violaceous-tumoral lesions that mostly located on the trunk; in addition, vascular dilatations were seen on some of the lesions.

The histopathological examination of skin metastasis of MM shows two infiltration patterns as nodular and diffuse involvement (4). Neoplastic dermal infiltration usually consists of immature plasmablasts that don't exhibit the typical cytological features of the plasma cells. Immunohistochemical examinations are required to establish the diagnosis. Tumor cells secrete immunoglobulin and give a positive reaction to CD79a, CD138 and epithelial membrane antigen. A correlation between serum electrophoresis and plasma cells involving the skin with respect to immunoglobulins produced and the light chain type was detected (1). In the review of 100 patients Requena et al, reported that the plasma cells secrete several paraproteins including IgG (54%), IgA (28%), IgD (12%), light chain alone (4%) and IgM (1%) and 40 of the patients were IgG subtype (20 cases kappa type, 7 cases lambda light chain type and 13 cases with unspecified light chain type) (1).
Although skin metastasis is mostly expected with the IgA subtype (4), most of the cases in the literature were of IgG subtype (1). This has been associated with the fact that IgG subtype was the most common subtype in MM. Those with light chain disease alone and cutaneous involvement of IgD subtype exhibit a more aggressive course (1). In our patient, diffused atypical plasma cell infiltration with lambda-monoclonality associated with IgG subtype was also detected by histopathological and immunohistochemical examination.

Cutaneous involvement of MM may develop at the early stages of the disease and may even be an initial finding of the disease (5); however it is usually associated with increased tumor burden, advanced disease and death occurring several months after the development of the skin lesions (1). Thus, cutaneous involvement in MM is an indicator of poor prognosis. Our patient also died shortly after skin metastasis.
In conclusion, MM cutaneous involvement may occur rarely but it may also occur as an initial sign of the disease and may indicate clinical

progression in patients with an established MM diagnosis; thus, clinicians should consider it in the differential diagnosis for appropriate cases.

Kaynaklar

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