Primary cutaneous plasmacytoma

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Abstract

Primary cutaneous plasmacytoma without underlying multiple myeloma or bone involvement is a rare disorder. It is a rare type of B cell lymphoma which belongs to a heterogeneous group of plasma cell neoplasm. We describe a 37-year old man who presented with a large plaque lesion of the chin and small exophytic growths on bilateral alae nasi of eight months duration, showing histological features of primary cutaneous plasmacytoma. Underlying myeloma was excluded by serum protein electrophoresis, skeletal survey, and bone marrow biopsy. In view of the plasmacytoma seems completely confined to the skin, surgical excision was carried out of the lesion on the chin, while the nasal lesion too is due to be managed in a similar manner. Oncological opinion has also been sought.

Introduction

Infiltration of various organs by plasma cells is seen in multiple myeloma especially in its advanced stages. This malignant cell proliferation which is usually gener- alized can involve organs such as gastro-intestinal tract, lymph nodes, spleen, and upper respiratory tract and rarely the skin[1].

Occasionally, localized forms of plasma cell proliferation have been observed. These localized forms include solitary plasmacytoma of bone and extramedullary plasmacytoma[2]. While extramedullary plasmacytoma is predominantly localized to the sub mucous lymphoid tissue of the upper respiratory tract, those of the skin are very rare. Prognosis of Primary cutaneous plasmacytoma depends on the clinical presentation. Solitary lesions carry a favorable prognosis[3].

Case report

A 37-year old man presented with an asymptomatic lump on his chin for eight months duration. It had an insidious onset. He was free of systemic symptoms such as loss of appetite and weight, nocturnal fever, generalized pruritus or bone pain. His past medical and surgical histories were unremarkable.

Examination revealed a skin coloured, firm, well demarcated, polypoidal 4 × 4.5 cm exophytic growth on his chin and small exophytic growths on bilateral alae nasi. Rest of the clinical examination was normal (Figure 1).

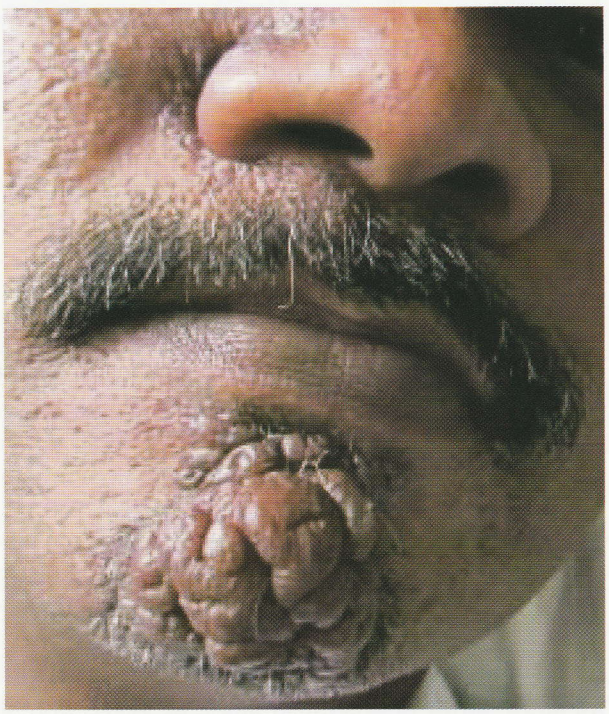

Figure 1.

Punch biopsies from both lesions revealed a dense infiltration of plasma cells from upper dermis to sub cutis. Few dysmorphic forms and scattered lymphocytes were present (Figure 2). Immunohistochemistry of the biopsy was positive for plasma cell marker CD 79a.

The results of all routine laboratory investigations were normal. Skeletal survey did not reveal any lytic lesions. Serum protein electrophoresis was negative for monoclonal band. Bone marrow aspirates and trephine biopsy revealed no abnormality.

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The patient underwent complete surgical excision of the lesion on the chin. Excision of the nasal lesions was also planned and oncological opinion has also been sought.

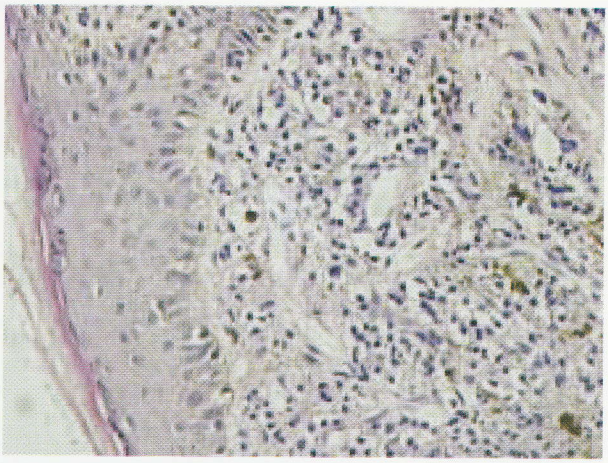

Figure 2.

Discussion

Primary cutaneous plasmacytoma is characterized by monoclonal proliferation of mature plasma cells in the skin without underlying multiple myeloma. It is a rare entity. According to World Health Organization (WHO) - European Organization for Research and Treatment of Cancer (EORTC) classification, it is classified under the Primary Cutaneous Marginal Zone B-cell lymphoma (PCMZL)[4].

A review of literature revealed a male predominance, adult onset (mean age 60.8 years) and a marginally increased prevalence of solitary lesions (58.6% vs. 41.4%)[3].

These tumours are a part of plasma cell neoplasms which represents a heterogeneous group of disorders. This group of disorders is characterized by uncont-rolled monoclonal proliferation of mature secretory B lymphocytes.

Primary cutaneous plasmacytoma should be differentiated from benign infiltration of plasma cells in the skin (primary cutaneous plasmacytosis)[5]. This differentiation relies mainly on histopathological features and demonstration of monoclonal nature of the plasma cell infiltrate[3]. Immunohistochemical studies are needed to demonstrate monoclonality which shows restriction of immunoglobulin light chain expression[1].

In our patient it was not possible to demonstrate this feature since laboratory tests to detect clonality are not available to us.

Another requirement for the diagnosis of primary cutaneous plasmacytoma is exclusion of underlying multiple myeloma by means of repeated laboratory, radiological and bone marrow investigations[1]. In our patient, all these investigations were normal and we plan to repeat them regularly during follow up.

Surgical excision, local radiotherapy, systemic corticosteroids and adjuvant chemotherapy have all been used in the management of primary cutaneous plasmacytoma. Exclusively local treatment seems to be adequate especially for solitary lesions. Surgical excision with or without local radiotherapy can be considered as valid treatment options for solitary lesions[3]. Adjuvant chemotherapy has been used in patients with multiple lesions.

Clinical presentation is an important prognostic factor. Solitary lesions show a better prognosis as they rarely progress to disseminated disease. By contrast, patients with multiple lesions have a high mortality rate. Size of the individual lesion is also important as larger lesions tend to disseminate early.

References

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