A Case of Primary Cutaneous Plasmacytoma

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A 66-year-old female patient had a firm, non-tender, dome shaped mass on the scalp. The lesion had enlarged slowly for 2 years, and measured about 4×6 cm. The histologic finding of the skin biopsy specimen demonstrated an infiltration of immature plasma cells in the dermis, which express monoclonal cytoplasmic lambda light chain by immunohistochemical stainings, and staging work-up after the biopsy revealed no evidence of disease in other foci. The mass on the scalp was treated successfully by radiation therapy, with the diagnosis of primary cutaneous plasmacytoma. (Ann Dermatol 8:(4)287~290, 1996).

Key Words : Primary cutaneous extramedullary plasmacytoma

The plasma cell neoplasms have been described as presenting in three modes ; multiple myeloma, solitary plasmacytoma (bone plasmacytoma and extramedullary plasmacytoma) and plasma cell leukemia.[1,2] The extramedullary plasmacytoma usually occurs on the upper respiratory tract, the gastrointestinal tract, the spleen and lymph nodes, but primary cutaneous lesions are very rare.[2,3,4,5,8,10,11,17] Histologically they are characterized by a dense monoclonal plasma cell infiltration in the dermis and are similar to the pattern of cutaneous B-cell lymphoma.[1,10] There are only a few cases of primary cutaneous plasmacytoma documented in the literature, since the publication of the first case by Hedinger in 1911.[2,5,9,10,20] We report herein a case of primary cutaneous plasmacytoma with immunohistochemical evidence confirming the diagnosis.

REPORT OF A CASE

A 66-year-old female patient visited our dermatologic clinic due to skin lesions on the scalp. The patient had previously been in good health and had no contributary past medical history. The main lesion had grown slowly on the scalp for the past 2 years and satellite lesions had developed in the past 5 months. The main lesion consisted of a 4× 6cm sized, firm, nontender, dome shaped mass with purplish discoloration of the overlying skin (Fig.1). The laboratory test results, including a complete blood count, urinalysis, routine chemistry, serum protein electrophoresis were within normal limits. Radiologic evaluation, abdominal ultrasonography and a bone marrow biopsy revealed no abnormal findings. A skin biopsy was done and its specimen demonstrated an infiltration of immature looking plasma cells in the upper and lower dermis with sparing on the epidermis by a narrow zone of normal collagen. Most of the tumor cells appeared poorly differentiated and they showed round, pale, centrally located nuclei with coarsely clumped chromatin and moderately abundant cytoplasm (Fig.2). To determine the nature of tumor cells, immunohistochemical stains were done, and the results suggested neoplastic plasma cells. The patient responded well with 5040cGy of radiotherapy, which almost cleared the lesion. There has been no evidence of recurrence for 8 months, since the patient first presented with her condition.

Received May 20, 1996.
Accepted for publication September 3, 1996.
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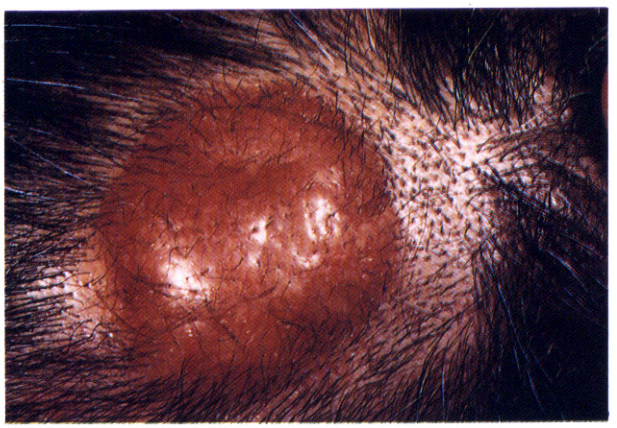

Fig. 1. 4×6cm in size, firm, nontender, dome shaped mass on scalp.

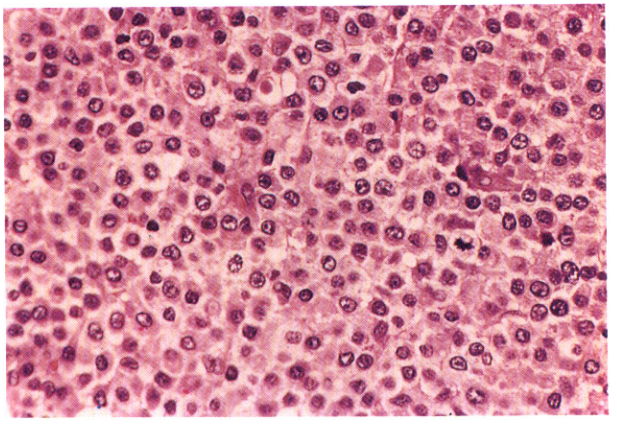

Fig. 2. The mass consists of poorly differentiated plasma cells. (H&E×400).

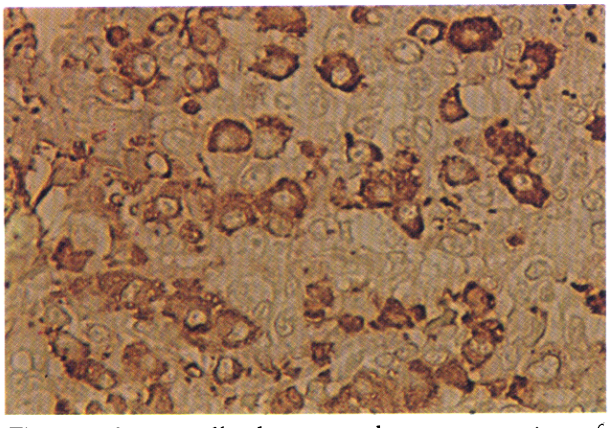

Fig. 3. Plasma cells show cytoplasmic expression of lambda light chain by immunohistochemistry .

Table 1. Clinical findings of primary cutaneous plasmacytoma reported in the literature[4,19]

Number of patients	20
Male : Female	9:1
Age	22-88 (mean 59.5)
Extracutaneous involvement	Lymph node 3 ; mucosal site 3 ; brain 1
light chain expres sion	k 5 ; λ 2
Immunoglobulin subtype	IgA 3 ; IgG 2
Treatment	Surgery 7 ; Irradiation 5 ; Chemotherapy 4 ; Surgery + Irradiation 2 ; Chemothearpy + Irradiation 1 no treatment 1

DISCUSSION

Primary extramedullary plasmacytoma differs from plasma cell myeloma in that the primary lesion arises outside the bone marrow. Extramedullary plasmacytoma is rare, and mostly occurs in the upper respiratory tract. Only 2-4% of extramedullary plasmacytomas originate in the skin without evidence of disease in other foci.[1,12,14,15,18] The pathogenesis of primary cutaneous extramedullary plasmacytoma is not well established as in plasmacytoma of other sites. Some cases of reactive plasma cell infiltration induced by arthropod bites or injections of modified BCG, which mimic plasmacytoma have been reported.[12] So it must be differentiated whether plasma cell infiltration was due to plasma cell neoplasia or reactive plasma cell proliferation. The neoplastic nature of primary cutaneous plasmacytoma is easily confirmed by demonstration of monoclonal cytoplasmic immunoglobulin by immunohistochemical stains, together with cell morphologic findings in H&E staining, such as nuclear atypism. In addition, lack of expression of the leukocyte common antigen and B-lineage cell surface markers (EMA, L26) can help in distinguishing plasmacytoma from other types of B-cell lymphoma, such as monocytoid B-cell lymphoma, and are included in the differential diagnosis[13,18]. Two histologic patterns of infiltration have been described in the literature. The majority of the cases are reported as a well circumscribed but nonencapsulated lesion involving the deep

Table 2. Panel of antibodies used for the differential diagnosis in the case

	neoplastic plasmacytoma	reactive plasma cell infiltration	lymphoma B cell origin	lymphoma T cell origin	lymphoma monocyte origin	this case
Light chain	monoclonality (k or λ)	polyclonality				λ
L26	−/+	−	+	−	−	−
LCA*	−/+	−	+	−	−	−
EMA**	−/+	−	+	−	−	−
CD 68	−	−	−	−	+	−
Lysozyme	−	−	−	−	+	−
CD 3	−	−	−	+	−	−

* : Leukocyte common antigen
** : Epithelial membrane antigen

dermis, with sparing of the epidermis. Occasional cases may show a diffuse infiltrative pattern. Perivascular and periappendageal accentuation of the plasma cell infiltrate is sometimes noted.[4] Interestingly a high percentage of primary cutaneous plasmacytoma have been reported as nonsecretory forms in contrast with secondary cutaneous plasmacytoma associated with multiple myeloma.[7,8] In this case, the serum electrophoresis data also revealed no evidence of secretory materials from plasmacytoma. However, when we consider the small tumor burden of primary cutaneous plasmacytoma and consider the low sensitivity of the laboratory test that has existed up until now, we cannot really confide in the high percentage of the nonsecretory type.

Those reported in the literature illustrate the variable clinical course of primary cutaneous plasmacytoma. Patients with primary cutaneous plasmacytoma are mostly elderly or middle-aged (mean age 59.5 years ; median age 57.5years) and there is a male predominance (Table1).[4,9] Radiation therapy (2000-6000 rads) or surgical excision has been recommended as treatment modalities of primary cutaneous extramedullary plasmacytoma. Ninety four percent of the patients have responded to treatment and sixty two percent obtained complete responses after radiation therapy. Tumor persistence or recurrence is relatively uncommon (15%) in the literature.[3,4,17,19]

We first considered this mass on the scalp as a secondary cutaneous plasmacytoma which is more common,[14] so serum protein electrophoresis, radiological evaluation and bone marrow studies were done; but we could not find any other foci of plasmacytoma except a cutaneous lesion. Additionally, we investigated whether this primary cutaneous plasmacytoma had monoclonality or polyclonality, because if plasma cell infiltration of the skin was the result of reactive inflammatory proliferation, it should not have shown a neoplastic monoclonal pattern. Only lambda chain staining was positive in the immunoglobulin stainings (Lambda light chain, Kappa light chain, IgA, IgG, IgM)[1,6,7,8] (Fig 3). In this case the tumor cells didn' t have the typical morphology of mature plasma cells, immunohistochemical stains were done to confirm the nature of tumor cells. The tumor cells did not express EMA, L26(CD20), LCA, CD68 and lysozyme, and expressed only a cytoplasmic lambda light chain.(Table 2) [13] With these results, we could exclude the possibility of immunoblastic lymphoma, haily cell leukemia and mast cell disorders which could be included in the pathologic differential diagnosis. Therefore we concluded that this case was a primary cutaneous plasmacytoma which had monoclonality. The patient was treated with a course of radiotherapy (5040cGy) to the area. Since it was reported that the prognosis of primary cutaneous extramedullary plasmacytoma is more aggressive than noncutaneous extramedullary plasmacytoma, further follow-up was recommended.[4] Our patient has remained free of disease without evidence of recurrence or complication for 8 months of follow up.

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