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Multiple Primary Cutaneous Plasmacytoma: Case Report with Review of the Literature

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Abstract **Case Report**

Background*:** Primary cutaneous plasmacytoma (PCP) is an uncommon variant of cutaneous B-cell lymphoma characterized by the presence of plasma cells in the skin. It represents a localized manifestation of plasma cell neoplasia, typically presenting as a solitary lesion on the skin. The rarity of PCP poses diagnostic and management challenges, with limited clinical data available, making the establishment of standardized treatment guidelines difficult. ***Case Report*:** A 55-year-old male with no prior medical history presented with multiple purplish nodules on the back, shoulder, and face, evolving over six months. Histologic and immunohistochemical analyses confirmed cutaneous plasmacytoma. Staging investigations supported the primary nature of the disease, leading to the decision of therapeutic abstention with regular monitoring. ***Discussion: Primary cutaneous plasmacytoma involves the clonal proliferation of plasma cells in the skin. While multiple primary cutaneous plasmacytomas are rare, they have typically been treated with chemotherapy, with or without radiotherapy. In the presented case, therapeutic abstention was chosen. This decision suggests that certain indolent forms of PCP may not require immediate chemotherapy.

Keywords: Primary plasmacytoma" skin" Treatment" Abstention.

INTRODUCTION

Multiple myeloma (MM) is a malignant hematologic disorder marked by the proliferation of clonal plasma cells within the bone marrow and the production of monoclonal immunoglobulins, which can lead to anemia, renal impairment, hypercalcemia, and bone lesions. Dissemination to the skin, resulting in secondary cutaneous plasmacytomas, typically occurs in advanced stages of the disease. In contrast, primary cutaneous plasmacytoma (PCP) is a skin tumor characterized by the absence of bone marrow infiltration and the lack of systemic organ damage.

Primary cutaneous plasmacytoma (PCP) is a very rare skin tumor characterized by the proliferation of mature plasma cells in the skin and the absence of systemic involvement. It typically presents as asymptomatic, reddish-brown plaques or nodules on the skin, often affecting the trunk, face, and extremities [3].

Primary cutaneous plasmacytoma arises without multiple myeloma. Cutaneous plasmacytoma can be fatal, particularly in multiple lesions [4].

Due to its rarity and the potential for confusion with other plasma cell dyscrasias, accurate diagnosis is essential. This article aims to provide a comprehensive overview of the clinical features, pathogenesis, diagnostic criteria, and management strategies for multiple primary cutaneous plasmacytosis.

CASE REPORT

A 55-year-old man presented with purplish indurated nodules evolving over six months, starting on the back and spreading to the right shoulder, right flank, and left cheek. Lesions were painless and non-pruritic (Fig 1).

Histological examination (Fig 2) showed the presence of a dense and diffuse lymphoplasmacytic cellular infiltrate forming cellular patches with the presence of a few clusters of neutrophils and eosinophils in the periphery. Immunohistochemical analysis revealed positivity for CD138, CD38, CD56, CD79a, EMA,

MUM1, as well as negativity for CD30 and CK, with a strong expression of lambda light chains and a weak focal expression of kappa chains. The proliferative index for Ki67 was approximately 40%.

A thorough search for underlying Multiple Myeloma (MM) was conducted, a bone marrow biopsy was performed; it was negative for multiple myeloma with no significant infiltration of malignant plasma cells. There were no Bence-Jones proteins in the urine. Hemogram, biochemical blood analysis and serum protein electrophoresis were normal. Also, no lytic bone lesions are observed in Radiographic images

Once the absence of other sites of the disease was confirmed, a clinicopathologic diagnosis of multiple primary cutaneous plasmacytoma was made.

The diagnostic criteria for differentiating primary cutaneous plasmacytoma from other similar conditions include the exclusion of systemic involvement. This is supported by a negative bone marrow biopsy, absence of Bence-Jones proteins in the urine, normal serum protein electrophoresis, normal radiological findings, and Immunohistochemical evidence of monotypic restriction for either kappa or lambda light chains.

By combining clinical, histological, immunohistochemical, laboratory, and radiological findings, clinicians can distinguish between multiple myeloma and primary cutaneous plasmacytoma effectively.

After confirming the absence of systemic involvement, close monitoring was chosen in consultation with the hematology department. He was followed up with physical examination, evaluation of the monoclonal component by serum and urine protein electrophoresis and immunofixation serum-free light-chain (FLC) measurement and annual computed tomography (CT) two years of follow-up demonstrated unchanged lesions and no additional complaints

For a patient with primary cutaneous plasmacytosis, indicators that might necessitate a shift to active treatment include: Development of ulceration, bleeding, or secondary infection within the lesions. Emergence of systemic symptoms such as unexplained weight loss, fever, which could suggest systemic involvement or progression, Development of anemia, hypercalcemia, renal dysfunction, or elevated serum monoclonal protein levels that could indicate systemic progression akin to multiple myeloma, Detection of plasma cell infiltration in the bone marrow on follow-up biopsy, suggesting transformation into or coexistence with multiple myeloma, Appearance of new lytic bone lesions or other radiological evidence suggestive of systemic disease progression, Deterioration in the patient's quality of life due to the psychological or physical burden of the disease, prompting the consideration of more aggressive treatment.

Monitoring these indicators is crucial for timely intervention to prevent disease progression and manage any systemic complications effectively.

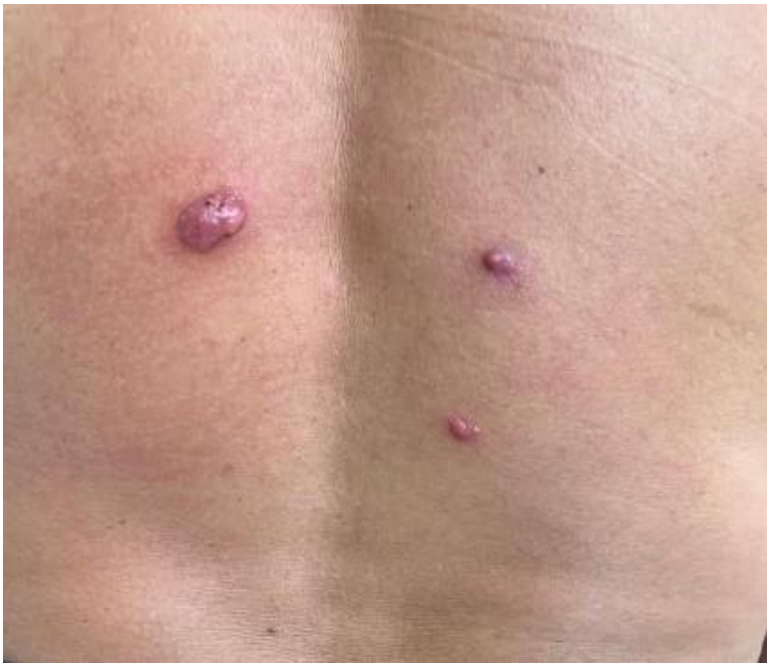

Figure 1: Erythematous nodular lesions

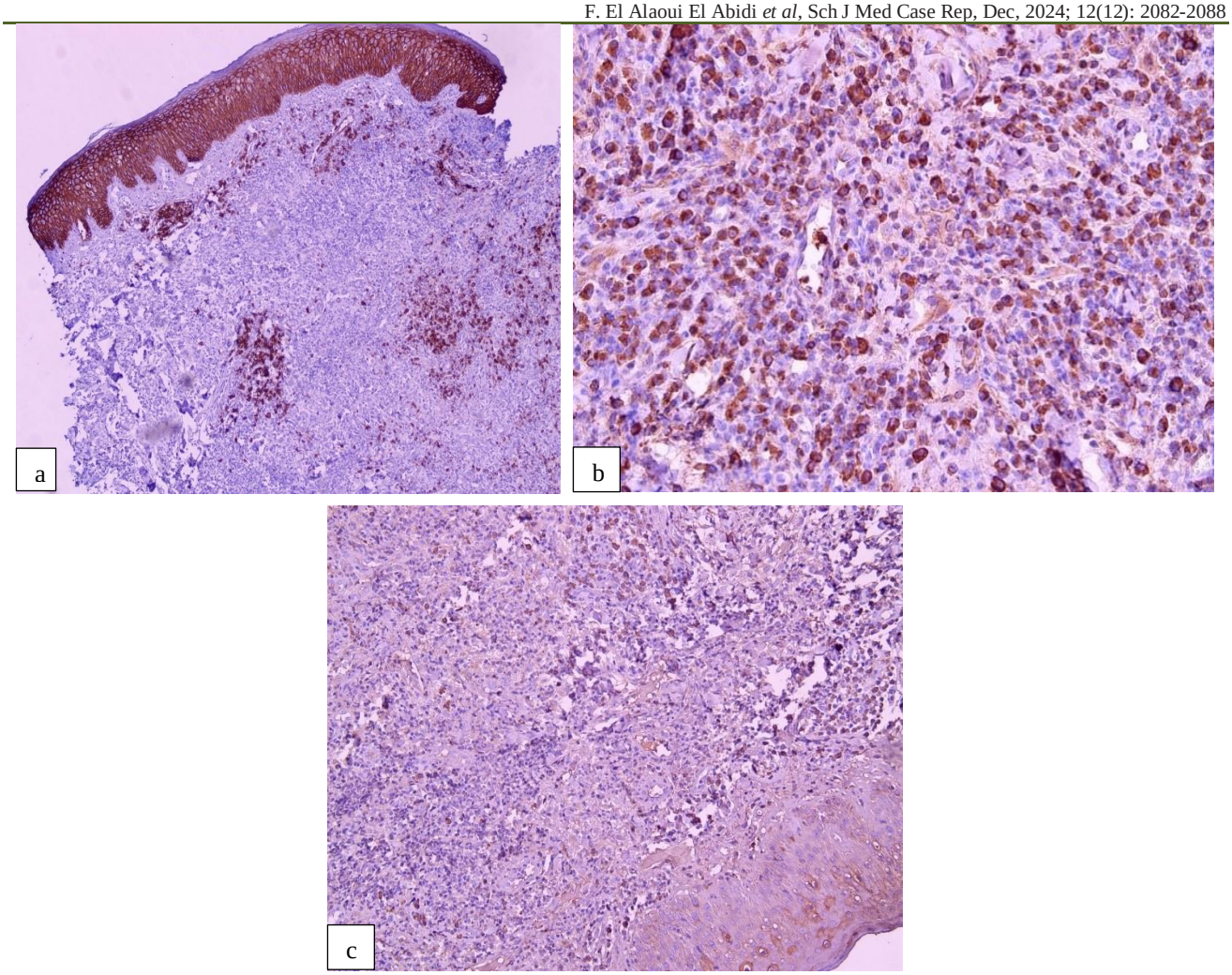

Figure 2: The plasma cells express CD138 (a), CD3 (b), and the lambda light chain (c).

Table I: Clinical data and outcome of cases published in the l

Case, Author and Year of publication	Age (years)	Sexe	Medical history/Associated disease	Clinical Presentation	Histology	IHC	Treatments
Case1 T. Green *et al*., 1992 [15]	37	M	0	infiltrated erythematous lesion	dense mixed cell infiltrate the dermis:small lymphocytes, with a few histiocytes, plasma cells, and activated lymphocytes	Immunohistochemical studies showed that the proliferation was polyclonal with a panel of pan-B and T-cell markers and antibodies recognizing immunoglobulin heavy and light chains	Chemotherapy and local radiotherapy

Case 2: Muscardin, Pulsoni, and Cerroni. 2000 [16]	55	M	0	erythematous plaques	dense, nodular infiltrates of mononuclear cells within the entire dermis and superficial subcutaneous tissues predominance of plasma cells admixed with a few lymphocytes and histiocytes. A few atypical plasma cells and rare mitotic	monoclonal expression of the immunoglobulin light chain κ	systemic chemotherapy
Case 3: V. VISEUX *et al*., 2004 [12]	77	F	monoclonal dysglobulinemia IgG lambda	Erythematous nodular lesions	Cutaneous plasmacytoma	-	Surgery Radiotherapy Local miltéfosine
Case 4 VA Fitzhugh *et al*., 2008 [17]	60		0	Multiple violaceous lesions	normal epidermis with a superficial and deep dense perivascular, peri anexal and focally interstitial infiltrate composed of pure mature plasma cells	positive for CD138 lambda clonality	Absention
Case 5: Thais Lima Saback *et al.*, 2012 [18]	87	F	Kaposi's sarcoma Alzheimer	erythematous-violaceous papules and plaques	diffuse proliferation of plasma cells in superficial and deep dermis	light chain restriction was demonstrated with a ratio of kappa/lambda 7:1 confirming the monoclonal nature of the plasma cellspositive for CD138	Abstention
Case 6: Hideki Maejima *et al*., 2014 [19]	66	M	0	Multiple subcutaneous tumors	a dense infiltrate of atypical plasmacytoid cells in the dermis.The epidermis and papillary dermis were not involved	the tumor cells were negative for T- and B-cell surface markers CD3, CD4, CD8, CD20, CD56, CD79, and HLA-DR, as well as the mature plasma cell and plasma neoplasm marker, CD138 but were positive for IgG, IgG λ light chain, bcl-2 and multiple myeloma oncogene-1 (MUM-1)	Chemotherapy
Case 7: N. Malissen *et al*., 2014 [11]	51	M	0	erythematous papulonodular lesions	monomorphic infiltrate of well-differentiated plasmacytic-like cells, with a preserved nucleocytoplasmic ratio and rare mitose	diffuse expression of the plasma cell marker CD138, no expression of CD20 and CD56 antigens. immunostaining with anti-IgA, IgG and IgM antibodies was negative.strong expression of lambda light ,a weak focal expression of kappa chains,	Abstention
Case 8: Uçmak *et al*., 2014 [20]	82	M	0	red nodular lesions	diffuse proliferation of plasma cells in the dermis	lambda immunohistochemical staining. CD20 was negative and CD 138+	Radiotherapy

Case 9: V. Gonzalez-Calle *et al.*, 2016 [21]	76	M	Solitary extramedullary plasmacytom	erythematous–violaceous papules and nodule	proliferation of plasma cells in the superficial and deep dermis ells showedan enlarged size, basophilic cytoplasm, atypical nucleus	positivity forCD79A CD138, CD56, BCL2 MUM1 as well as negativity forCD3, CD5, CD10, CD20, CD23, CD43, PAX5, BCL6, Restriction for Kappalight chains was demonstrated. Proliferative index Ki67 15%	Chemotherapy
Case 10: Giuseppe Broggi *et al.*, 2019 [22]	78	F	0	purplish cutaneous plaques	atypical oval-shaped cells with abundant cytoplasm eccentric nuclei, chromatin, infiltrating the medium and deep dermis Neoplastic cells were morphologically similar to mature plasma cells	positive to CD79a, CD138, CD56, MUM-1, and EMA, and totally negative to CD20. Immunohistochemical studies for kappa and lambda light chains revealed a monoclonal expression of immunoglobulin kappa lights chains	Chemotherapy
Our case	55	M	0	purplish indurated nodules	diffuse lymphoplasmacytic cellular infiltrate forming cellular patches with the presence of a few clusters of neutrophils and eosinophils in the periphery	positivity for CD138, CD38, CD56, CD79a, EMA, MUM1, as well as negativity for CD30, CK, there was a strong expression of lambda light chains which, associated with a weak focal expression of kappa chains, Proliferative index for Ki67 was 40%.	Abstention

DISCUSSION

Monoclonal gammopathies constitute a group of diseases characterized by the presence of a single clone proliferation of plasma cells, typically producing a monoclonal immunoglobulin.

Primary cutaneous plasmacytoma characterized by the proliferation of plasma cells occurring outside the context of multiple myeloma, it's included in the category of the marginal zone B lymphomas [5].

The average age of patients with cutaneous plasmacytomas is approximately 60 years, with extremes ranging from 16 to 88 years, a clear male predominance is observed. Solitary forms are more frequent than multiple ones. The preferential localizations are the face, then the trunk and the limbs [6].

Macroscopically, PCP appears as solitary or multiple erythematous–violaceous subcutaneous papules, nodules of variable size, hemispherical, embedded in the dermis. They may be accompanied by discrete inflammatory signs or even ulcerations [9].

On the histopathological level, it is characterized by a proliferation composed of mature monomorphic plasma cells with rare cytonuclear atypia, located in the mid-dermis, occasionally extending to the subcutaneous tissue, while preserving the superficial dermis and epidermis. Immunohistochemistry reveals expression of CD138, CD79, CD38, but not CD20 or CD19. Investigation for lambda or kappa monotypia should be performed.

Plasma cell proliferations (PCP) occur in the absence of clinical and paraclinical evidence of bone or hematologic manifestations, favoring the exclusion of myeloma. A definitive diagnosis can only be proposed once myeloma is ruled out through appropriate assessment (complete blood count, serum and urine protein electrophoresis, bone marrow biopsy, and radiological exams).

In contrast to multiple myeloma, PCP is non-secretory in 75% of cases and are not accompanied by a monoclonal peak of immunoglobulin [7, 8].

The two prognostic factors for PCP are firstly the solitary or multiple character; multiple forms have a poorer prognosis than solitary forms, and secondly the size of the lesion [6, 7].

Therapy options range from surgical excision plus radiotherapy in the case of solitary plasmacytomas to systemic therapy in the case of multiple lesions [10].

Systemic therapies, including chemotherapy, immunomodulatory drugs, and targeted therapies, are typically reserved for cases with extensive skin involvement or progression toward systemic disease, their side effects profile makes them less ideal for indolent cases where localized treatments suffice [23,24].

Radiation therapy is often considered the first-

line treatment for localized cutaneous lesions; Compared to systemic therapies, radiation therapy is less likely to cause systemic side effects, making it an attractive option for localized disease [24]. Newer targeted therapies and immunomodulatory agents may offer better tolerability and specificity [23].

Multiple forms of PCP are even more rare. In 2014, Malissen *et al*., reported about 15 published observations; In 75% of cases, patients received chemotherapy (the standard treatment for myeloma), associated in 25% of the observations with radiation therapy. A quarter of the patients were treated surgically or received systemic corticosteroid therapy [11].

Treatment is not codified for multiple forms, inaccessible to surgery or radiotherapy and the combination of melphalan and prednisone would not alter the risk of transformation to multiple myeloma but would delay it. Visieux *et al.*, reported an observation of multiple primary cutaneous plasmacytomas, recurrent after surgery and radiotherapy [12]. The choice of close and active surveillance was reported by N. Malissen *et al*., in a 51-year-old patient, with a Four-year delay without any transformation [11]. Saback T *et al*., [13] reported in an 87-year-old patient no signs of systemic disease with 12 months of rigorous follow-up, the same choice also reported by VA Fitzhugh *et al*., [14] in a 60-year-old man.

The description of similar cases in the literature suggests the existence of forms of PCP that are not very progressive, indolent that should be characterized by prognostic criteria. However, the rarity of this disease and the severe prognosis of the multiple forms incite, while waiting for such markers to follow the recommendations for management of the disease.

CONCLUSION

Multiple primary cutaneous plasmacytomas are rare, and their optimal management remains challenging. The presented case, treated with therapeutic abstention, highlights the need for individualized approaches and close surveillance in certain indolent forms of PCP. Further research is warranted to identify prognostic markers and guide management recommendations for this uncommon condition.

CONSENT: The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

Conflict of Interest: The authors have no conflicts of interest to declare. All co-authors have seen and agree with the contents of the manuscript and there is no financial interest to report. We certify that the submission is original work and is not under review at any other publication.

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