Violaceous Plaques and Papulonodules on the Umbilicus

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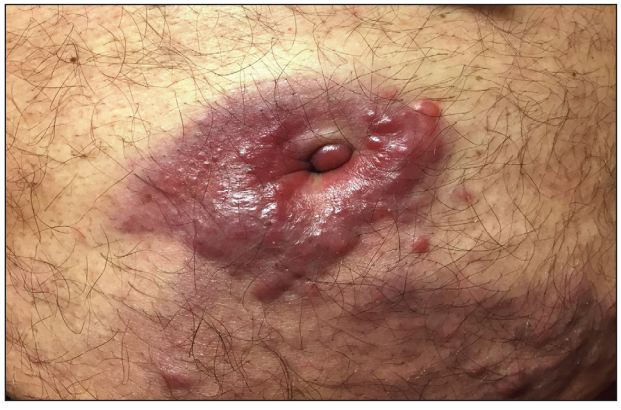

A 75-year-old man presented for evaluation of lesions on the umbilicus and lower abdomen that had developed over the past 4 weeks and were asymptomatic. His medical history was notable for plasma cell myeloma (stage III, IgA λ light chain restricted), deep vein thrombosis, and a 30-year history of smoking (20 packs per year). On physical examination, violaceous plaques and papulonodules were noted on the umbilicus. The lesions had a firm consistency and smooth surface without epidermal change. Violaceous papulonodules and subcutaneous plaques were noted on the lower abdomen. The lesions were nontender to palpation. Bilateral edema of the legs also was noted. The remainder of the skin was normal and there was no cervical, axillary, or inguinal lymphadenopathy.

WHAT'S THE **DIAGNOSIS?**

a. AIDS-associated Kaposi sarcoma
b. cutaneous deposits of myeloma
c. keloid
d. primary cutaneous marginal zone B-cell lymphoma
e. Sister Mary Joseph nodule

PLEASE TURN TO PAGE E12 FOR THE DIAGNOSIS

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The authors report no conflict of interest.
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THE **DIAGNOSIS:** Cutaneous Deposits Of Myeloma

Cutaneous deposits of myeloma are a rare skin manifestation of multiple myeloma that typically occur in less than 5% of patients.[1,2] The lesions represent monoclonal proliferations of plasma cells and arise from direct extension of a neoplastic mass or less commonly from hematogenous or lymphatic spread. This secondary cutaneous involvement by plasma cell myeloma has been referred to in the literature as metastatic or extramedullary cutaneous plasmacytoma.[1,2] This condition must be distinguished from cutaneous plasma cell infiltrates without underlying bone marrow involvement, classified by the World Health Organization as primary cutaneous marginal zone B-cell lymphoma and previously referred to as primary cutaneous plasmacytoma.[3]

Clinically, cutaneous deposits of myeloma manifest as erythematous to violaceous papules, plaques, or nodules with a smooth surface and firm consistency.[1,2] The lesions typically occur on the trunk and less commonly on the head, neck, arms, and legs. In a review of 83 cases of metastatic cutaneous plasmacytoma and primary cutaneous plasmacytoma in multiple myeloma, Kato et al[4] found that 52% (43/83) of cases occurred in IgG myelomas and 23% (19/83) in IgA myelomas.

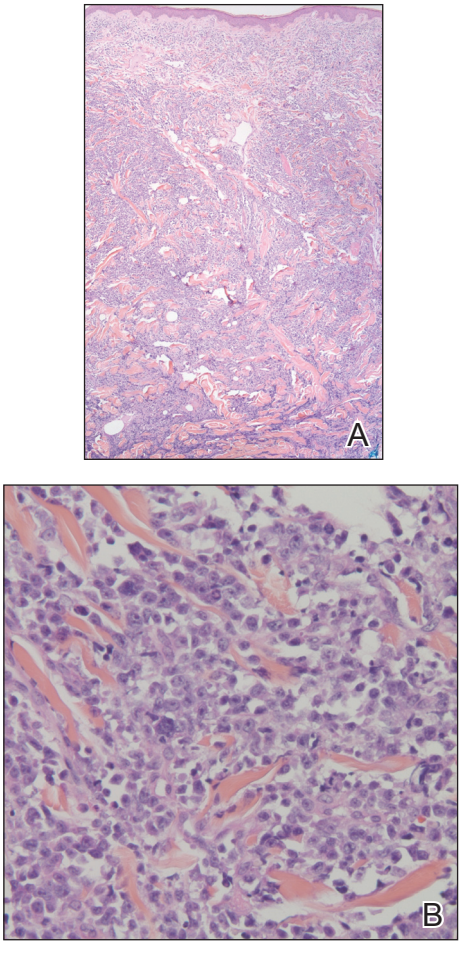

FIGURE 1. Cutaneous deposition of myeloma. A dense infiltrate of atypical plasmacytoid cells was seen through the full thickness of the dermis with nuclear pleomorphism, prominent nucleoli, and frequent mitoses (A and B)(H&E, original magnifications ×5 and ×50, respectively).

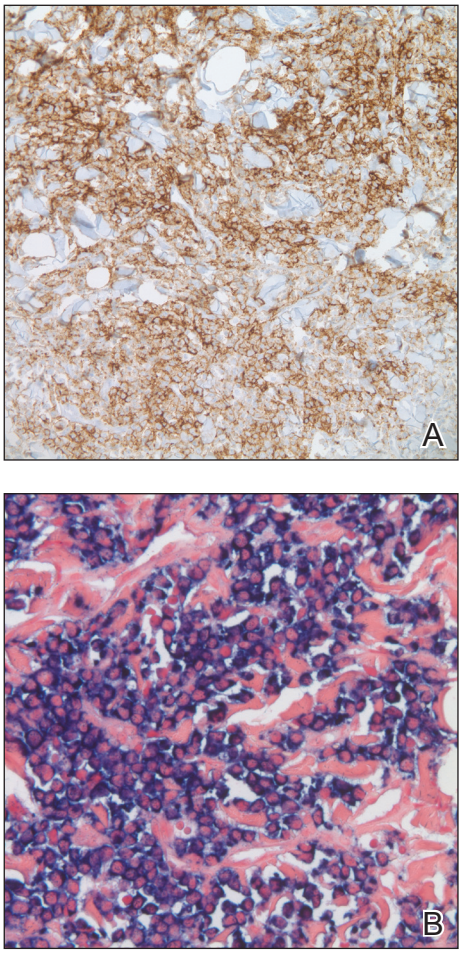

FIGURE 2. Cutaneous deposition of myeloma showing positive immunohistochemical staining for CD138 (A) and IgA λ light chain (B) (original magnification ×20 and ×50, respectively).

In our patient, a 4-mm punch biopsy of an umbilical plaque demonstrated a dense infiltrate of atypical plasmacytoid cells through the full thickness of the dermis with nuclear pleomorphism, prominent nucleoli, and frequent mitoses (Figure 1). Immunohistochemical staining was positive for IgA λ light chain (Figure 2A) and CD138 (Figure 2B) and was negative for CD20, which was consistent with the patient's known plasma cell myeloma. Positron emission tomography revealed progression of underlying disease compared to prior studies with hypermetabolic mediastinal, retroperitoneal, and pelvic side wall lymphadenopathy, as well as extensive hypermetabolic soft tissue masses with involvement of the periumbilical region.

The differential diagnosis for violaceous periumbilical plaques includes cutaneous marginal zone B-cell lymphoma (primary or secondary) or T-cell lymphoma (primary or secondary), cutaneous metastases from solid organ or hematologic malignancies (eg, Sister Mary Joseph nodule), AIDS-associated Kaposi sarcoma (plum-colored plaques that may be extensive), and cutaneous endometriosis (umbilical nodules that may develop in women after surgical excision of endometrial tissue).

The mainstay of therapy for secondary cutaneous involvement of plasma cell myeloma includes treatment with chemotherapy and local radiotherapy.[1,2,5] After the diagnosis of cutaneous deposits of myeloma was made in our patient, he was treated with bortezomib, cyclophosphamide with dexamethasone, and local radiotherapy to symptomatic bony lesions; however, he was unresponsive to therapy and the disease progressed with numerous extramedullary lesions of the mediastinum, gastrointestinal tract, and retroperitoneum 2 months later. The patient developed hydronephrosis from external renal compression necessitating nephrostomy tube and malignant pleural effusions requiring intubation. He experienced rapid clinical decline and died 3 months after the initial presentation due to multiorgan failure.

Cutaneous deposits of myeloma are a sign of underlying disease progression in plasma cell myeloma and often herald a fulminant course (eg, death within 12 months of presentation), as seen in our patient.[5] Clinicians should be aware of this rare manifestation of plasma cell myeloma and pursue aggressive therapy given the poor prognostic nature of these cutaneous findings.

REFERENCES

1. Jorizzo JL, Gammon WR, Briggaman RA. Cutaneous plasmacytomas: a review and presentation of an unusual case. *J Am Acad Dermatol.* 1979;1:59-66.
2. Bayer-Garner IB, Smoller BR. The spectrum of cutaneous disease in multiple myeloma. *J Am Acad Dermatol.* 2003;48:497-507.
3. Willemze R, Jaffe ES, Burg G, et al. WHO-EORTC classification for cutaneous lymphomas. *Blood.* 2005;105:3768-3785.
4. Kato N, Kimura K, Yasukawa K, et al. Metastatic cutaneous plasmacytoma: a case report associated with IgA lambda multiple myeloma and a review of the literature of metastatic cutaneous plasmacytomas associated with multiple myeloma and primary cutaneous plasmacytomas. *J Dermatol.* 1999;26:587-594.
5. Sanal SM, Yaylaci M, Mangold KA, et al. Extensive extramedullary disease in myeloma. an uncommon variant with features of poor prognosis and dedifferentiation. *Cancer.* 1996;77:1298-1302.