Secondary Cutaneous Plasmacytoma in a Patient with Multiple Myeloma

Joshua M. Levin MD
Lehigh Valley Health Network, Joshua_M.Levin@lvhn.org

Christian W. Oram DO
Lehigh Valley Health Network, Christian_W.Oram@lvhn.org

Published In/Presented At

Levin, J., & Oram, C. (2013). *Secondary cutaneous plasmacytoma in a patient with multiple myeloma.* Poster presentation.

Secondary Cutaneous Plasmacytoma in a Patient with Multiple Myeloma

Joshua M. Levin, M.D. and Christian W. Oram, D.O.

Lehigh Valley Health Network, Allentown, Pennsylvania and Philadelphia College of Osteopathic Medicine, Philadelphia, Pennsylvania

Case Presentation:

Patient: W.C. is a 60 year-old Hispanic male.

History of Present Illness: Patient presented with a three month history of a slowly growing, painful, violaceous nodules on the left chest proximal to a chemotherapy port.

Medical History: IgG κ-light chain restricted multiple myeloma (MM) and myeloma leukemia, depression, coronary artery disease, myocardial infarction, hypertension, hyperlipidemia, migraine headaches, chronic low back pain, hemorrhoids

Medications: Allopurinol, acyclovir, metoprolol, lorazepam, odansetron, methocarbamol, and hydromorphone

Laboratory Data: Bone marrow biopsy: >90% plasma cells (10-15% c/w MM), >15% plasma cells in the blood (<20%), +11/14 chromosomal translocation, IgG 8110 (690-1400 mg/dl), monoclonal-spike 5.3 (>3 g/dl c/w MM)

Imaging: Plain film: Ill-defined lucency at the mid-shaft of the right humerus.

Interventions: 6 cycles of bortezomib/cyclophosphamide/dexamethasone chemotherapy

Physical Examination: Several firm violaceous nodules; ranging in size from 5mm-3.0cm, located on the left chest.

Biopsy: *Lehigh Valley Health Network Laboratories* (S12-11552, 04/26/2012) Left chest: "Tumor cells are strongly reactive for CD138 and show κ-light chain restriction. Immunostain for lambda light chain is negative. These findings are consistent with a plasma cell neoplasm."

Reason for Presentation: Interest

Discussion:

Plasmacytoma is a rare tumor of plasma cells that can affect the skin and various organ systems. Plasmacytoma can be classified as 1) multiple myeloma (MM) associated plasmacytoma, 2) plasmacytoma without MM, or 3) primary cutaneous plasmacytoma. MM associated plasmacytoma can involve the skin and other organ systems, affecting approximately two percent of MM patients. Plasmacytoma without associated MM can arise within (medullary) or outside of the bone marrow (extramedullary). Medullary plasmacytoma may affect the skin by direct extension. The most common extramedullary tumor sites are the upper respiratory tract, lymph nodes, spleen, and less frequently, the skin. Primary cutaneous plasmacytoma is exceedingly rare, and diagnosis requires absence of both MM and plasmacytoma of any other organ system.

Plasmacytomas have been described as firm, erythematous, nontender nodules that can involve the neck, ears, shoulders, axillae, chest, abdomen, dorsa of the hands, and the lower extremities. Proteins associated with these skin lesions are IgA, IgD, IgG, and IgM. IgA monoclonal protein is most frequently associated with primary cutaneous plasmacytoma. Multiple skin lesions in the presence of IgA monoclonal protein portends a poor prognosis. Cutaneous plasmacytoma is more likely to develop in patients with IgD monoclonal protein and tends to follow a more aggressive course. IgG is the most common monoclonal protein implicated in secondary cutaneous plasmacytoma with MM.

Histopathologically, primary and seconday cutaneous plasmacytoma cannot be differentiated. Lesions typically demonstrate either a nodular or diffuse infiltrate of plasma cells that involve the dermis and occasionally the subcutis. A grenz zone is sometimes present. An interstitial pattern has been described in secondary cutaneous plasmacytoma, where plasma cells are located between collagen bundles with no involvement of the epidermis and papillary dermis. This pattern can be confused with a benign inflammatory infiltrate. Other reported findings include secondary cutaneous mucinosis, amyloid deposits, variable morphology and maturation of plasma cells, intracytoplasmic inclusions (Russell bodies), and intranuclear inclusions (Dutcher bodies). Immunohistochemical studies have shown the plasma cells to be CD 79A+, CD38+, and CD138+. Deletion of the rb-1 (retinoblastoma gene) using fluorescence in-situ hybridization (FISH) has been identified in neoplastic plasma cells.

Reported treatment for secondary cutaneous plasmacytoma consists of targeting the underlying multiple myeloma and excising the lesions. In the case of primary cutaneous plasmacytoma, without underlying multiple myeloma, external beam radiation continues to be the mainstay of treatment.

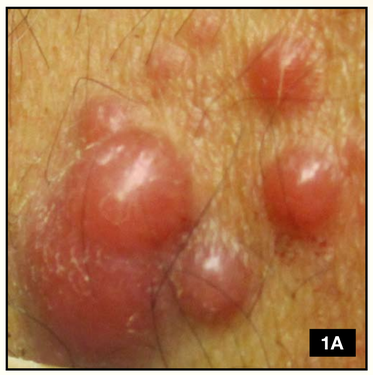

Figure 1A: Several firm violaceous nodules on the left chest.

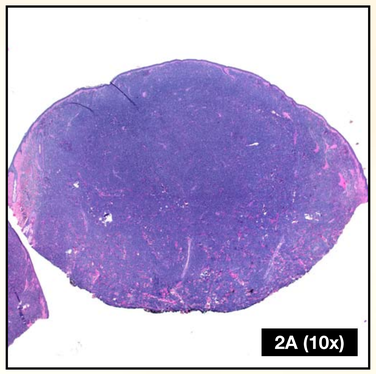

Figure 2A: Intradermal infiltrating nodule composed of basophilic cells in sheets.

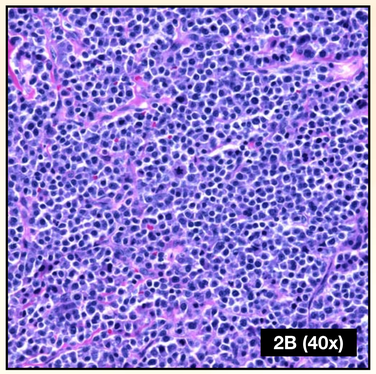

Figure 2B: H&E: Plasma cells with hyperchromatic nuclei and mitoses.

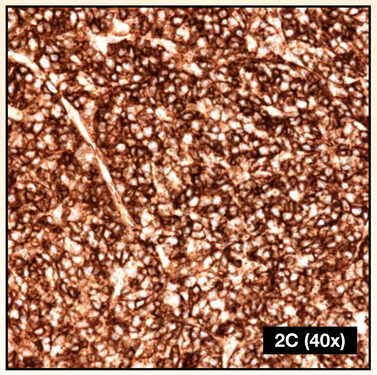

Figure 2C: Positive CD-138 stain.

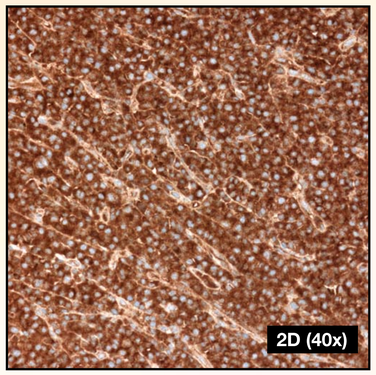

Figure 2D: Positive Kappa light chain staining.

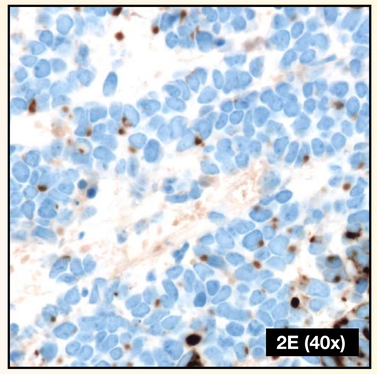

Figure 2E: Negative Lambda light chain staining.

References:

1 Daoud M, Lust J, Kyle R, Pittelkow M. Monoclonal gammopathies and associated skin disorders. *J Am Acad Derm.* 1999;40:507-534.

2 Patterson J, Parsons J, White R, Fitzpatrick J, Kohout-Dutz E. Cutaneous involvement of multiple myeloma and extramedullary plasmacytoma. *J Am Acad Derm.* 1988;19k:879-890.

3 Yoon Y, Cho W, Seo S. Case of multiple myeloma associated with extramedullary cutaneous plasmacytoma and pyoderma gangrenosum. *International Journal of Derm.* 2006;45:594-597.

4 Rongioletti F, Patterson J, Rebora A. The histological and pathogenetic spectrum of cutaneous disease in monoclonal gammopathies. *J Cutan Pathol.* 2008;35:705-721.

5 Jorizzo J, Gammon W, Briggaman R. Cutaneous Plasmacytomas. *J Am Acad Derm.* 1979;1:59-66.