RESIDENT ROUNDS: PART III

Plasma Cell Leukemia With Initial Cutaneous Presentation

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CASE REPORT

A 62-year-old Caucasian female presented with a tender, erythematous to violaceous nodular eruption for 4 weeks (Figure 1). She concomitantly reported abdominal discomfort, lower back pain, fatigue, and anorexia. Laboratory results upon initial presentation revealed acute kidney injury and she was admitted for further workup. At that time she demonstrated marked leukocytosis with blasts, eosinophilia, basophilia, and increased lactate dehydrogenase. A retroperitoneal ultrasound revealed retroperitoneal lymphadenopathy and a diffusely enlarged uterus with fluid collection.

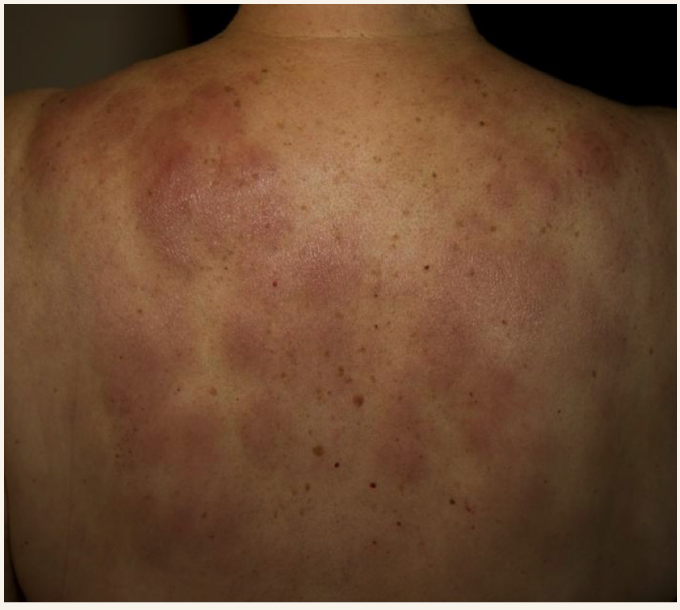
The patient underwent an endometrial biopsy, skin biopsy, and bone marrow biopsy. A bone marrow aspirate demonstrated 70% cellular marrow, 65% of which was in blast form in a diffuse interstitial pattern, hypercellularity with large blasts, and markedly decreased hematopoietic elements. Flow cytometry of the aspirate showed the atypical cells to be CD38/CD2+ with interpretation as large cell neoplasm, favoring a plasmacytic etiology. Skin and endometrial biopsies demonstrated atypical leukocytes with a plasmacytic origin, which were similar to those found in the bone marrow aspirate.

Treatment was initiated with bortezomib, doxorubicin, vincristine, lenalidomide and dexamethasone, and hemodialysis three times weekly, and although initially promising, the patient deteriorated and expired about 43 days after initial presentation. Further cytogenetic testing was not obtained prior to her demise.

Plasma cell leukemia (PCL) is a rare variant of multiple myeloma (MM), a monoclonal gammopathy, the current pathophysiology of which is not yet well understood. Our patient presented atypically with the presence of leukemia cutis, involvement of endometrium but the absence of bone pain or organomegaly. PCL may have extramedullary involvement, but cutaneous involvement as an initial presentation has only been described very sporadically in the literature.[1]

Cutaneous involvement of plasma cell dyscrasias typically occurs after chemotherapy, particularly thalidomide or stem cell transplantation, and more often in MM with later diagnosis of plasma cell leukemia rather than the initial presentation.[2] The other described cases of cutaneous involvement of PCL often describe violaceous to hemorrhagic tender, infiltrated deep-set papules and nodules. Leukemia cutis is a much more commonly observed phenomenon in lymphocytic leukemias, which is a poor prognostic factor with a one-year survival rate of 12%.[3] Subcutaneous plasmacytoma growth has been hypothesized to be a result of transformation of the adhesion molecules LFA-1 and ICAM-1 at some point during disease course, facilitating plasma cell migration from the bone marrow towards extra-

FIGURE 1. Initial presentation with tender, papular, non-pruritic, erythematous to violaceous nodules distributed along the anterior chest and back.



medullary sites.[4] Other sites that have been described as being more commonly involved include the subcutis, pleura, central nervous system and testes; the latter two possibly due to a leukemic sanctuary mechanism during treatment.[5]

Treatment options are not optimized, but therapeutic success defined as improving survival and quality of life has been described. Initial cases were treated using melphalan and prednisone; soon after, vinchristine, Adriamycin (doxorubicin), dexamethasone (VAD) was described in a number of cases as being therapeutically useful, increasing survival to 15-18 months.6 More recently, the proteasome inhibitor bortezomib, after successful studies in the treatment of MM, has shown promise, with a mean survival of 12.6 months after treatment. Stem cell transplantation has also been shown to be successful in a number of case report series and retrospective analyses with mean survival rates ranging from 34-45 months.7

CONCLUSION

PCL is an aggressive variant of MM, that rarely presents in the skin but when present portends a poor prognosis. Plasma cell dyscrasias should be considered in the differential of cases that clinically present violaceous, infiltrated deep set papules and nodules. Referral to a hematologist and potential clinical trials is warranted.

DISCLOSURES

None of the authors have declared any relevant conflicts.

REFERENCES

1. Chattopadhyay A, Nath UK, De R et al. Primary plasma cell leukemia with initial cutaneous involvement and IgA biclonal gammopathy. *Ann Hematol* 2008;87(3):249-51.
2. Alexandrescu DT, Koulova L, Wiernik PH. Unusual cutaneous involvement during plasma cell leukemia phase of multiple myeloma patient after treatment with thalidomide: a case report and review of the literature. *Clin Exp Dermatol* 2005;30:391-394.
3. Cho-Vega JH, Medeiros J, Prieto VG, Vega F. Leukemia cutis. *Anatom Path* 2008; 129:130-142.
4. Tsutani H, Sugiyama T, Shimizu S, et al. Discordant LFA-1/ICAM-1 expression in a case of secondary plasma cell leukemia associated with subcutaneous plasmocytoma. *Am J Hematol* 1993;42:299-304.
5. Bladé J, Kyle RA. Nonsecretory myeloma, immunoglobulin D myeloma, and plasma cell leukemia. *Hematol Oncol* Clin 1999;13:1259–1272.
6. Costello R, Sainty D, Bouabdallah R et al. Primary plasma cell leukaemia: a report of 18 cases. *Leuk Res* 2001;25:103–107
7. Garcia-Sanz R, Orfao A, Gonzalez M et al. Primary plasma cell leukemia: clinical, immunophenotypic, DNA ploidy, and cytogenetic characteristics. *Blood* 1999;93:1032.

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