

Multiple Primary Cutaneous Plasmacytoma

To the Editor;

A 66-year-old Japanese man presented with a painless skin tumor in his left axillae which had been present for several years without receiving any medical treatment. The tumor enlarged and lesions appeared on several areas on his body within a few months of April 2012 (Fig. 1). Histopathological examination of a skin biopsy specimen taken from a right inguinal tumor showed a dense infiltrate of atypical plasmacytoid cells in the dermis and subcutis (Fig. 2). The epidermis and papillary dermis were not involved. Immunohistological analysis revealed that the tumor cells were negative for T- and B-cell surface markers CD3, CD4, CD8, CD20, CD56, CD79, and HLA-DR (Fig. 3), as well as the mature plasma cell and plasma neoplasm marker, CD138 (Fig. 3), but were positive for IgG, IgG λ light chain, bcl-2 and multiple myeloma oncogene-1 (MUM-1) (Fig. 4). Laboratory investigation revealed a small amount of Bence-Jones protein in the urine and elevated serum levels of total protein (10.5 g/dL), creatinine (1.4 mg/dL), calcium (1.4 mg/dL), immunoglobulin G (5720 mg/dL), and β 2 microglobulin (8.1 mg/dL). Chest and abdominal computed tomography showed multiple subcutaneous masses; however, other organs were unaffected. Histopathological examination of a bone marrow biopsy specimen



Figure 1. Multiple subcutaneous tumors bilaterally on the shoulders, chest, and right axilla.

showed no abnormalities. The diagnosis of primary cutaneous plasmacytoma (PCP) was made based on the International Myeloma Working Group criteria (1). The patient was treated with bortezomib and combined vincristine, doxorubicin, and dexamethasone therapy. The tumors initially responded to therapy by decreasing in size and temporarily disappearing. Several months later, the subcutaneous tumors reappeared and enlarged, and the patient subsequently died from bleeding from a tumor in the abdomen 14 months after initial presentation.

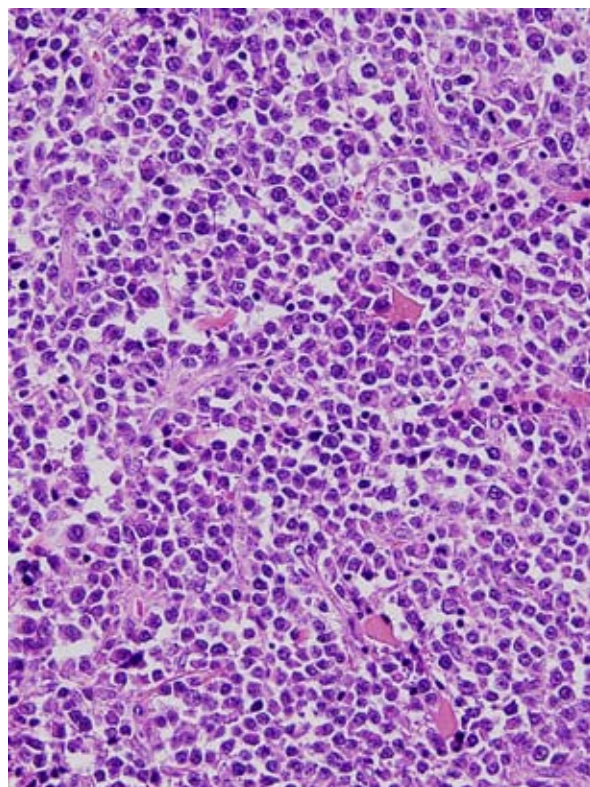


Figure 2. Dense, atypical plasma cell infiltrates were seen in the dermis and subcutaneous tissues (hematoxylin and eosin, $\times 400$).

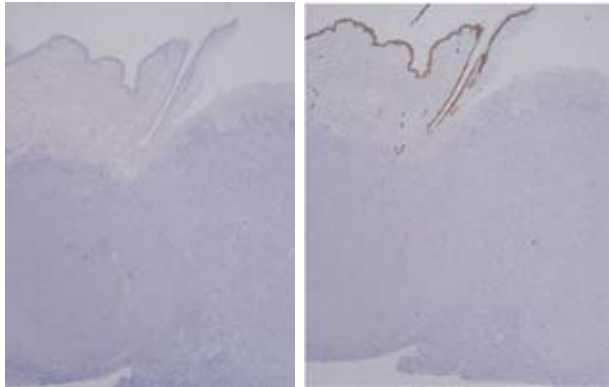


Figure 3. Negative staining of neoplastic cells for CD20 (left) and CD138 (right).

Extramedullar plasmacytoma (EMP) is a plasma cell tumor that arises outside the bone marrow and may occur in the upper respiratory tract, gastrointestinal tract, and central nervous system, but cutane-

ous involvement is rare (1). PCPs constitute approximately 2–4% of EMP (2). A prognostic difference between the solitary and multiple forms of the disease has been highlighted by some clinicians who pointed out that the latter runs a more aggressive course and has a relatively high mortality rate, as was seen in our patient (3). PCP cells usually express CD79a, CD38, and CD138, but are negative for CD20 and leukocyte common antigen. Monotypic expression of IgG light chains is usually present (3). In our case, tests were negative for CD79 and CD138, but positive for MUM-1, bcl-2, and IgG light chain. Bcl-2 is member of a family of signaling molecules with proapoptotic and anti-apoptotic activities (4). Located in the mitochondrial membrane, bcl-2 prevents cell death by inhibiting adapter molecules involved in the activation of caspases in the intrinsic pathway. Overexpression of bcl-2 is found in most human tumor types, and is associated with prolonged tumor cell survival, an

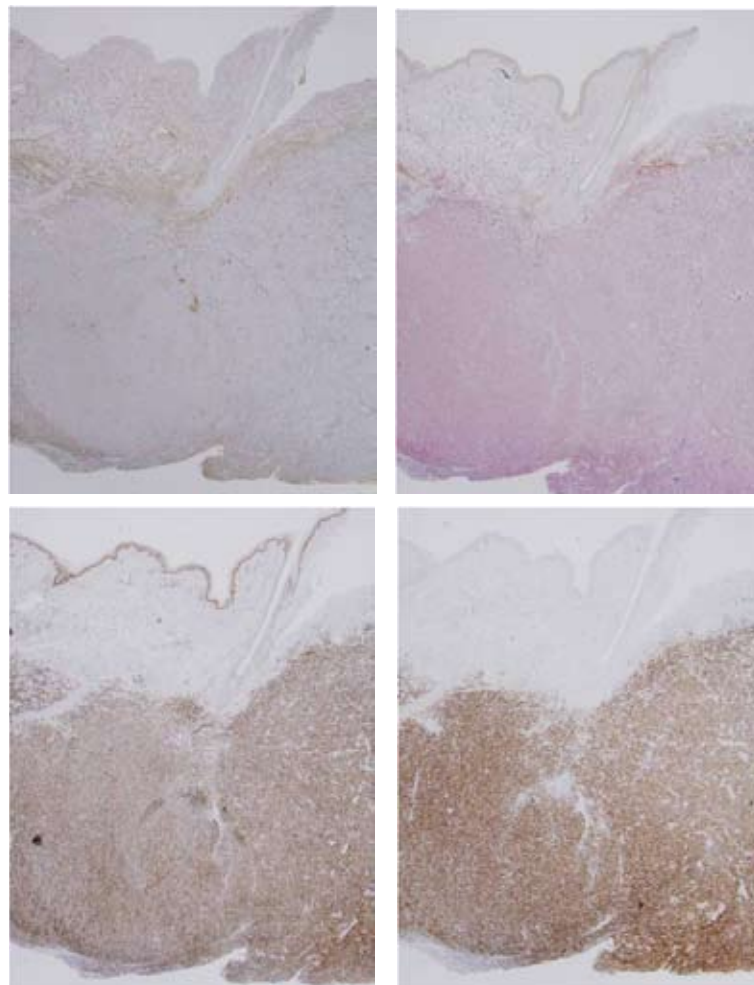


Figure 4. The neoplastic cells expressed IgG (top left), IgG λ light chain (top right), bcl-2 (bottom left) and MUM-1 (bottom right).

aggressive clinical course, drug resistance, and a decreased survival rate (5). Tumor expression of bcl-2 and the presence of multiple tumors in the present case were associated with a poor prognosis.

References

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**Hideki Maejima¹, Kensei Katsuoka¹,
Tomiteru Togano²**

*Departments of ¹Dermatology and ²Haematology,
Kitasato University School of Medicine, 1-15 Kitasato,
Sagamihara, Japan*

Corresponding author:

Hideki Maejima, MD
Department of Dermatology
Kitasato University School of Medicine
1-15 Kitasato
Sagamihara 228-8885
Japan
hm4765@yahoo.co.jp

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