Burkitt lymphoma with cutaneous involvement

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Clinical synopsis

A 25-year-old HIV-infected man, on highly active antiretroviral therapy with CD4 count of 198 cells/μL and viral load of 13,800 copies/mL, presented with a 2-month history of progressive, painful left axillary lymphadenopathy, which was 11 cm. Lymph node biopsy revealed a malignant lymphoid population of cells with intermediate size, mature chromatin, several small eccentrically located nucleoli, a high mitotic rate, and a clonal CD19+, CD20+, CD10+ B-cell immunophenotype. There was effacement of lymph node architecture, necrosis, and many tangible-body macrophages imparting a starry-sky appearance. Burkitt lymphoma was diagnosed. Peripheral blood, cerebrospinal fluid, and bone marrow biopsy were negative for leukemic cells. Clinically and radiographically, the lymphoma increased in size over the next 10 months, extending to overlying muscle and skin, despite receiving 3 cycles of fractionated cyclophosphamide, vincristine, adriamycin, and dexamethasone, palliative radiation therapy, 4 cycles of rituximab, ifosfamide, carboplatin, and etoposide, and prophylactic intrathecal methotrexate.

Approximately 10 months after being diagnosed with Burkitt lymphoma, he was referred to the dermatology service with a 2-month history of an enlarging, erosive plaque over his left axilla, chest, and abdomen. A computed tomography (CT) 1 month earlier had revealed multiple large, enhancing, necrotic, soft tissue masses involving the left axilla, chest, and abdomen, with overlying skin thickening and nodularity, as well as an exo-enteric, ulcerating gastric fundus mass. The radiology service interpreted the masses as Burkitt lymphoma although infection could not be excluded. The oncologists treated him empirically for cellulitis, but wound and blood cultures were negative and he did not respond to antibiotics.

Physical examination revealed a large, erythematous, slightly indurated, centrally verrucous plaque along the left chest wall and abdomen with erosions and bullae (Fig. 1). The remaining physical examination was normal. Skin biopsy from the abdomen revealed full-thickness abnormalities. The epidermis was acanthotic and spongiotic with scattered dyskeratotic keratinocytes. There was prominent extravasation of erythrocytes and an infiltrate filling the dermis and the sampled subcutis consistent with Burkitt lymphoma. The tumor is seen here in the subcutis with mononuclear cells with minimal cytoplasm and multiple small nucleoli. Abundant apoptotic forms are present. The classic "starry-sky" appearance that is imparted by histiocytes with ingested cellular debris is absent from this tumor sample. Stains were not performed because of the poor preservation of the tumor cells. Previous diagnosis of Burkitt's lymphoma had been made on nodal tissue from the axilla (hematoxylin-eosin, 40X).
nelarabine and high-dose methotrexate but expired 6 weeks later from progressive disease and sepsis from Candida albicans.

Discussion
Burkitt lymphoma is an aggressive non-Hodgkin lymphoma which can be classified into endemic, sporadic, and immunodeficiency variants [1]. Although each variant frequently involves extranodal sites, cutaneous involvement with Burkitt lymphoma is very rare [2, 3, 4, 5, 6, 7]. In most lymphomas and leukemias, cutaneous involvement occurs through hematogenous dissemination. However, cases of cutaneous Burkitt lymphoma have typically resulted from tumor seeding after surgical procedures[4] or local invasion of rapidly enlarging underlying tumors [2, 3]. In our case, cutaneous involvement likely resulted from the former, in the context of widespread multiorgan disease. Our patient's cutaneous Burkitt lymphoma was initially misdiagnosed as cellulitis. Persistent skin lesions in patients with Burkitt lymphoma, especially in proximity to known tumor foci, should raise concern for cutaneous Burkitt lymphoma.

References

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