Rapidly progressing necrotic ulcerations and sinuses in specific cutaneous Hodgkin's disease

Sir,

A 25-year-old gentleman presented with a progressively enlarging swelling over the right side of the neck for seven months. He also had a large fungating ulcer over the right supraclavicular region with extensive edema of the chest wall and right upper limb for three months. He complained of high spiking fever, cough and significant weight loss. Examination revealed a large, soft-to-firm, non-tender, swelling encompassing the neck, right shoulder, arm and upper chest, with visible dilated veins. There were large ulcers with everted margins and a necrotic floor over the supraclavicular region and multiple smaller ulcers over the chest wall [Figure 1]. He had hard and matted cervical lymph nodes.

The clinical differential diagnoses of nocardiosis, cervicothoracic actinomycosis, mycobacterial or atypical mycobacterial infection, and lymphoma were considered. The histopathology of the skin and lymph nodes were consistent with a diagnosis of classical Hodgkin's lymphoma, nodular sclerosis type. The skin biopsy revealed dense infiltrates of lymphocytes, plasma cells, histiocytes, neutrophils, eosinophils, and a few scattered medium-sized cells with vesicular nuclei, visible nucleoli, and moderate amounts of cytoplasm [Figure 2]. On immunohistochemistry, the medium-sized cells stained strongly for CD30 [Figure 3] and weakly for Pax-5. A submental lymph node biopsy showed many mononucleate cells and occasional binucleate (Reed–Sternberg) cells with prominent nucleoli [Figure 4]; the mononuclear and lacunar cells were positive for CD15 and CD30. Pus culture grew *Pseudomonas aeruginosa* resistant to all drugs other than aztreonam. A computed tomography scan of the chest revealed suppurative cervical and axillary lymphadenitis along with enlarged paratracheal, subcarinal, and anterior mediastinal lymph nodes. The patient was administered 12 cycles of chemotherapy with adriamycin, bleomycin, vinblastine, and dacarbazine (ABVD) regimen as suggested by the hematologists, with which the ulcers healed [Figure 5], but the disease relapsed 6 months later. Radical dissection of cervical lymph nodes was not done. The patient refused further palliative chemotherapy or radiotherapy and succumbed to the disease six months later.

Hodgkin's lymphoma was described by Sir Thomas Hodgkin in 1932. It commonly presents as painless lymphadenopathy, mostly involving lymph nodes above the diaphragm.[1] Cutaneous involvement can be classified as nonspecific skin involvement, specific cutaneous Hodgkin's disease, and primary cutaneous Hodgkin's disease.

Primary cutaneous Hodgkin's disease includes histologically proven skin involvement without disease in any other site.[2] In our patient, the rapid progression, the massive swelling with the presence of dilated veins and large necrotic ulcerations overlying hard lymph nodes made the diagnosis of specific cutaneous Hodgkin's lymphoma the most likely. Specific cutaneous Hodgkin's disease was described by Grosz in 1906. It signifies advanced disease, and is a poor prognostic sign heralding the need for more aggressive therapy. The lesions could be papules, nodules, plaques, tumors, ulcerative lesions, or erythroderma.[3] The Grosz–Hirschfeld type is the presence of painless, erythematous papules and nodules that frequently ulcerate.[4] Our patient had an ulcerative type of specific Hodgkin's disease. The skin over the chest and axilla is the most common site of involvement, as was seen in our patient. There are three mechanisms for spread of the

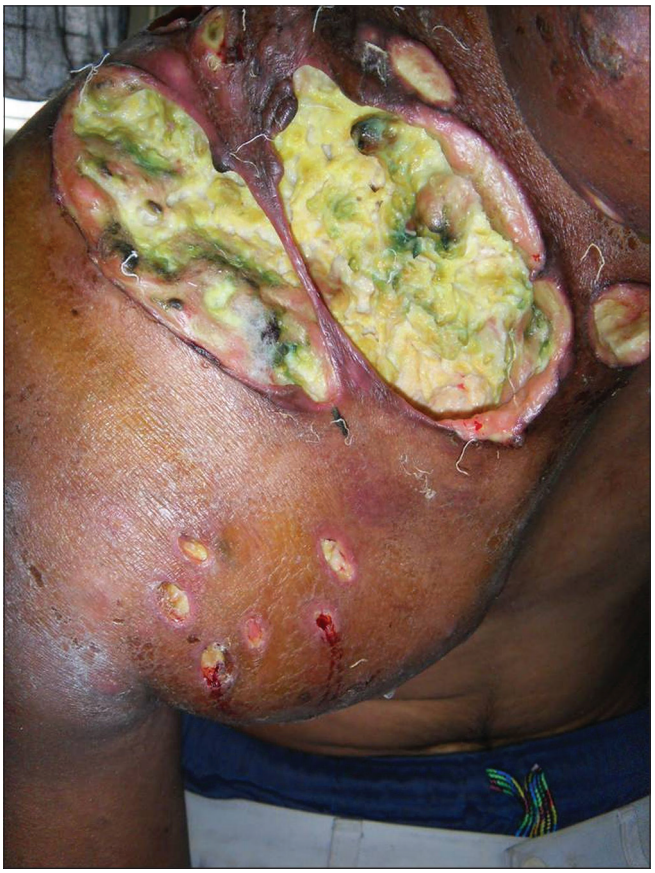

Figure 1: Right-sided neck, shoulder, and chest wall swelling with ulcerations and sinuses

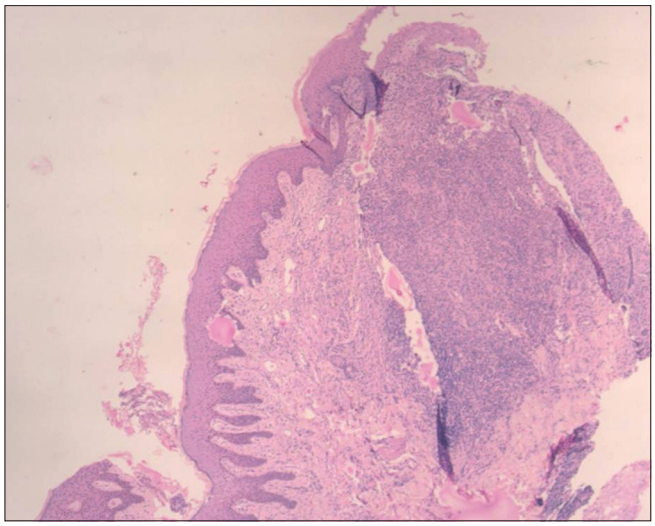

Figure 2: Dense infiltrate of cells in the dermis (hematoxylin and eosin, original magnification ×100)

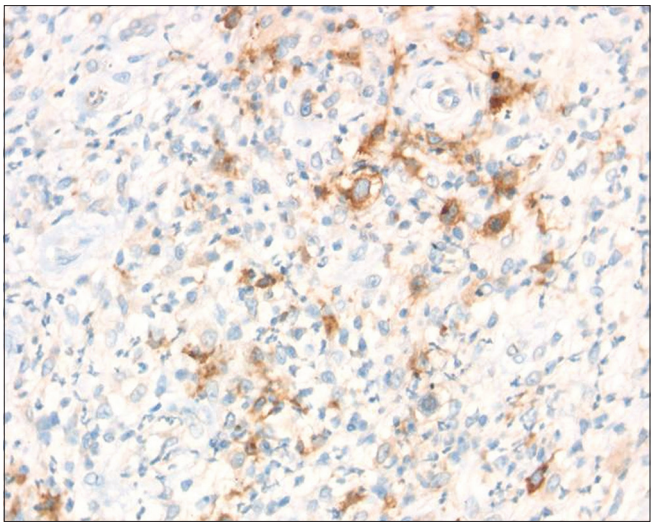

Figure 3: The large cells are positive for CD30 (original magnification × 40)

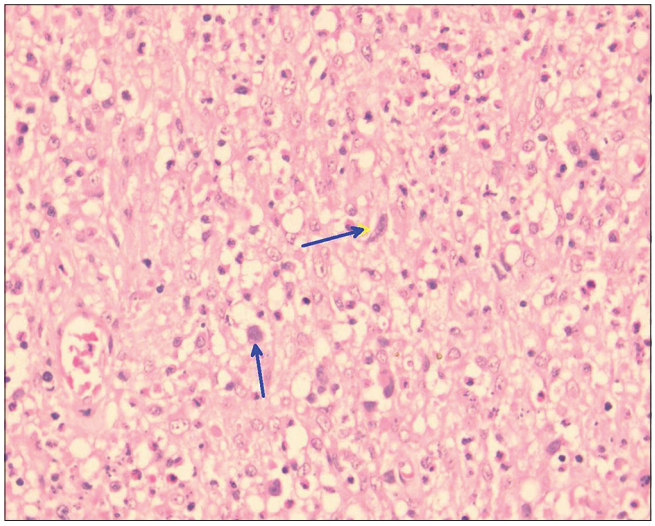

Figure 4: Mononucleate and binucleate large Reed–Sternberg cells (arrows) (hematoxylin and eosin, original magnification × 400)

disease; hematogenous dissemination, direct extension from involved lymph nodes and retrograde spread from proximal involved lymph nodes via lymphatics; which is the most common mechanism, and the presumed mode of spread in our patient as well.[4]

Identification of characteristic binucleated tumor cells (Reed–Sternberg cells) or mononuclear cells (Hodgkin's cells) within an inflammatory milieu, comprising 0.1%–10% of all cells in the biopsy suggests the diagnosis, which can be confirmed by immunohistochemistry.

Our patient was treated with the ABVD regimen, which is the standard protocol used for this disease. The cause of death in our patient was relapse of Hodgkin's lymphoma.

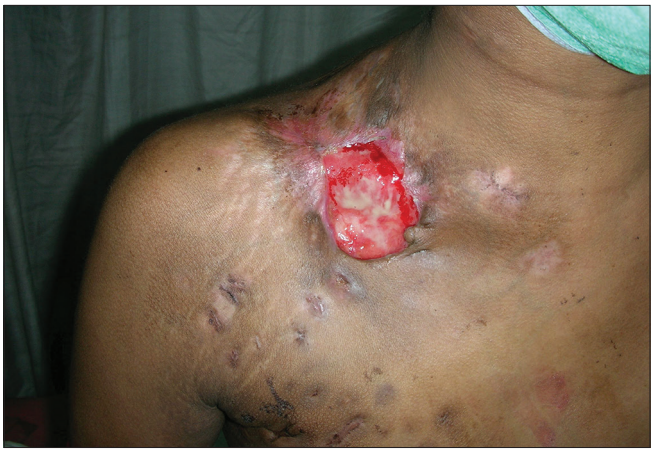

Figure 5: Reduction in size of swelling with resolving ulcers after ABVD chemotherapy

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Conflicts of interest

There are no conflicts of interest.

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