An enlarging, ulcerated scalp nodule



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A 77-year-old woman with no previous medical history presented with an 18-month history of an enlarging painless scalp nodule, intermittent fevers, fatigue, and 2-kg unintentional weight loss. The patient reported progressive enlargement and ulceration of the skin lesion despite 2 injections of intralesional triamcinolone. Her physical examination was notable for a 10-cm ulcerating occipital scalp nodule with multiple smaller adjacent dermal nodules (Fig 1). Bilateral anterior cervical and right supraclavicular lymphadenopathy was palpable. Skin biopsy found a diffuse infiltrate of lymphocytes, eosinophils, and plasma cells with scattered cells characterized by large vesicular nuclei and prominent nucleoli (black arrows) and binucleate cells (red arrow) (Fig 2). Large atypical cells were positive for CD30, CD15, and MUM1 and weakly positive for PAX-5, whereas ALK, OCT-2, and BOB.1 were all negative.

Question 1: What is the most likely diagnosis?

- A. Squamous cell carcinoma (SCC)
- **B.** Pyoderma gangrenosum (PG)
- C. Hodgkin lymphoma (HL)
- D. Angiosarcoma
- E. Kerion

Answers:

A. SCC – Incorrect. SCC is a progressive ulcerating keratinocytic neoplasm that does not involve atypical mononuclear cells staining positive for CD30 and CD15.

B. PG – Incorrect. PG commonly presents as a primary ulcer rather than an ulcerating nodule; the insidious course and failure to respond to

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corticosteroids are features inconsistent with PG. Dermal neutrophilic infiltrate progressing to ulceration is a central histopathologic finding of PG.

C. HL – Correct. The clinical presentation is consistent with classical cutaneous HL, which generally presents with painless erythematous papulonodules and plaques that progressively ulcerate. Atypical lymphocytes are the hallmark histopathologic finding in classical cutaneous HL. The atypical cells stain positive for CD30 and most are CD15 positive (75%-85% of cases).¹ They are usually negative for CD45, CD20, ALK-1 protein kinase, and EMA.¹ Cutaneous HL may be a primary process, originating in skin, characterized by the absence of lymph node involvement and minimal systemic symptoms. Secondary cutaneous HL typically features lymphadenopathy with constitutional symptoms (ie, fever, fatigue, weight loss), suggesting cutaneous involvement caused by retrograde lymphatic, hematogenous, or direct spread from involved lymph nodes.

D. Angiosarcoma – Incorrect. Angiosarcoma is an endothelial malignancy that may present as an ulcerating lesion; however, atypical mononuclear cells staining positive for CD30 and CD15 are not observed.

E. Kerion – Incorrect. Kerion is caused by a fungal infection of the hair follicle and is diagnosed by mycologic culture of involved hair. Fungal elements are typically visualized on biopsy, and atypical mononuclear cells are not a common feature.

Question 2: Which of the following is true regarding this case?

A. The lesion is most likely a primary cutaneous lesion.

B. The most commonly affected site for this disease is the upper extremity.

C. The patient has a poor prognosis.

D. The cutaneous presentation of this disease is common.

E. Metastatic calcifications are a common histologic finding in this disease.

Answers:

A. The lesion is most likely a primary cutaneous lesion – Incorrect. This patient likely has secondary cutaneous HL consistent with the presence of lymphadenopathy and B symptoms.^{2,3}

B. The most commonly affected site for this disease is the upper extremity – Incorrect. The most common reported site of involvement for cutaneous HL is the trunk followed by the scalp.²

C. The patient has a poor prognosis – Correct. Case studies have found a poor prognosis associated with retrograde lymphatic and hematogenous spread to skin seen in secondary cutaneous HL. Cases of primary cutaneous HL are characterized by a more benign course and improved survival.^{2,4}

D. The cutaneous presentation of this disease is common – Incorrect. Cutaneous HL is very uncommon with a prevalence of 0.5% to 3.4% among patients with HL.^{3,5} Common cutaneous symptoms and findings in patients with HL include pruritus, ichthyosis, xeroderma, urticaria, hyperpigmentation, and eczematous and psoriasiform eruptions.

E. Metastatic calcifications are a common histologic finding in this disease - Incorrect. Metastatic calcifications are not commonly observed in cutaneous HL. However, Reed-Sternberg cells are common histologic findings in cutaneous HL characterized by multinucleated or bilobed nucleus cells with prominent eosinophilic inclusion-like nucleoli ("owl's eye" appearance). The presence of Reed-Sternberg cells is not a sensitive finding for HL, as other conditions in the differential diagnosis, including lymphomatoid papulosis and anaplastic large cell lymphoma, exhibit cells that resemble Reed-Sternberg cells making immunohistochemistry a critical diagnostic tool.

Question 3: What is the next best step in management?

- A. Excision
- **B.** Oral corticosteroids
- C. Intralesional methotrexate
- **D.** Referral to hematology/oncology
- **E.** Skin tissue cultures

Answers:

A. Excision – Incorrect. The treatment for secondary cutaneous HL involves chemotherapy such as the ABVD regimen (Adriamycin, bleomycin, vinblastine, and dacarbazine) to treat this systemic disease.

B. Oral corticosteroids – Incorrect. The use of oral and topical corticosteroids is inappropriate for treatment of cutaneous HL.

C. Intralesional methotrexate – Incorrect. The use of intralesional methotrexate for a patient with signs of secondary cutaneous HL is inadequate to treat this systemic disease.

D. Referral to hematology/oncology – Correct. The patient's biopsy results suggest classical cutaneous HL. Given there is clinical evidence of lymph node involvement with a clinical history of B symptoms, the patient is likely to have secondary cutaneous HL and requires referral to hematology/oncology for further evaluation for staging and to begin chemotherapy. After referral to hematology/oncology, the patient underwent a 6-month course of systemic chemotherapy and 1-month radiation treatment and is awaiting restaging results.

E. Skin tissue cultures – Incorrect. Cultures are appropriate in the initial evaluation when the diagnosis is unknown; however, the skin biopsy has provided a diagnosis of HL; thus, subsequent

tissue cultures would not be the next best step in management.

Abbreviations used:

HL: Hodgkin Lymphoma SCC: Squamous cell carcinoma PG: Pyoderma gangrenosum

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