The use of IPL has also been studied as a treatment option for patients with facial lentigines, such as those seen in PJS. Remington et al treated a single PJS patient with IPL (47 J/cm^2, 3.2 msec pulse duration, 591-1200 nm) after the administration of topical lidocaine and oral sedation. There was almost complete clearance of perioral, periorbital, and nasal areas after only one treatment, but 12 treatments on separate occasions were performed because of the patient's pain intolerance.

For this series, the recommended choices are: 6, e; 7, e; 8, e; 9, a; 10, a; and 11, b.

We thank Jordan Lema from Upstream BioMedia for taking photographs of the patient.

BIBLIOGRAPHY

Bandler M. Hemangiomas of the small intestine associated with mucocutaneous pigmentation. Gastroenterology 1960;38:641-5.

Bolognia JL, Lorizzo JL, Rapini R, editors. Dermatology. 1st ed. St. Louis: Mosby; 2003. p. 1719.

Buck JL, Harned RK, Lichtenstein JE, Sobin LH. Peutz-Jeghers syndrome. Radiographics 1992;12:365-78.

Dereure O. Drug-induced skin pigmentation. Epidemiology, diagnosis and treatment. Am J Clin Dermatol 2001;2:253-62.

Giardiello FM, Trimbath JD. Peutz-Jeghers syndrome and management recommendations. Clin Gastroenterol Hepatol 2006;4: 408-15.

Goldberg DJ. Laser treatment of pigmented lesions. Dermatol Clin 1997;15:397-407.

Kleinegger CL, Hammond HL, Finkelstein MW. Oral mucosal hyperpigmentation secondary to antimalarial drug therapy. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2000;90: 189-94.

Lenane P, Powell FC. Oral pigmentation. J Eur Acad Dermatol Venereol 2000;14:448-65.

Lenane P, Sullivan DO, Keane CO, Loughlint SO. The Laugier-Hunziker syndrome. J Eur Acad Dermatol Venereol 2001;15:574-7.

Meleti M, Vescovi P, Mooi WJ, van der Waal I. Pigmented lesions of the oral mucosa and perioral tissues: a flow-chart for the diagnosis and some recommendations for the management. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2008;105: 606-16.

Ozawa T, Fujiwara M, Harada T, Muraoka M, Ishii M. Q-switched alexandrite laser therapy for pigmentation of the lips owing to Laugier-Hunziker syndrome. Dermatol Surg 2005;31:709-12.

Remington BK, Remington TK. Treatment of facial lentigines in Peutz-Jeghers syndrome with an intense pulsed light source. Dermatol Surg 2002;28:1079-81.

Sabesan T, Ramchandani P, Peters WJ. Laugier-Hunziker syndrome: a rare cause of mucocutaneous pigmentation. Br J Oral Maxillofac Surg 2006;44:320-1.

Seoane Lestón JM, Vázquez García J, Cazenave Jiménez AM, de la Cruz Mera A, Aguado Santos A. Laugier Hunziker syndrome. A clinical and anatomopathologic study. Presentation of 13 cases [in French]. Rev Stomatol Chir Maxillofac 1998;99:44-8.

Taylor CR, Anderson RR. Treatment of benign pigmented epidermal lesions by Q-switched ruby laser. Int J Dermatol 1993;32: 908-12.

Veraldi S, Cavicchini S, Benelli C, Gasparini G. Laugier-Hunziker: a clinical, histopathologic, and ultrastructural study of four cases and review of the literature. J Am Acad Dermatol 1991;25:632-6.

A 70-year-old man with pruritic plaques

Kristen LoSicco, Andrew D. Lee, MD, and Gil Yosipovitch, MD
Winston-Salem, North Carolina

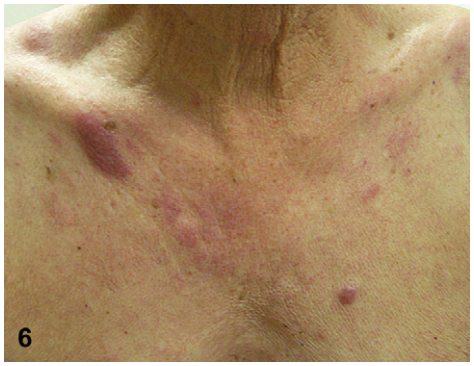
A 70-year-old male presented with a 5-year history of pruritic plaques arising on his neck, the upper portion of his chest, and back. These lesions had slowly increased in size and were unresponsive to topical corticosteroids. His medical history was significant for mucinous cystadenocarcinoma of the pancreas that was successfully resected several years earlier; the patient deferred chemotherapy and radiation therapy. He denied recent fevers, night sweats, or unintentional weight loss. The physical examination revealed violaceous, indurated plaques with excoriations on his neck, the upper portion of his chest, and back (Figs 6 and 7). He had no regional lymphadenopathy or hepatosplenomegaly.

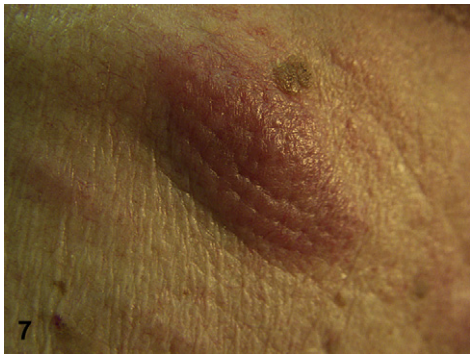
12. The differential diagnosis includes all of the following *except*:
    a. Primary cutaneous B-cell lymphoma
    b. Mycosis fungoides
    c. Metastatic adenocarcinoma
    d. Granuloma annulare
    e. Pseudolymphoma

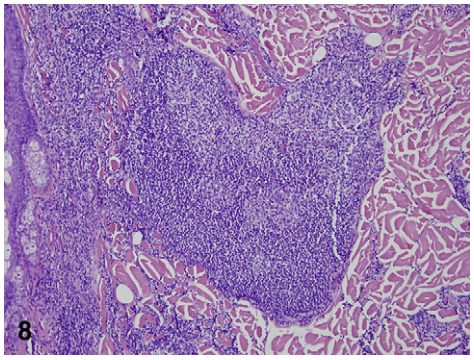
A punch biopsy specimen revealed a nodular dermal infiltrate of small and medium lymphocytes with oval nuclei, coarse chromatin, and scant cytoplasm, along with germinal centers of variable size and shape, with sparing of the epidermis (Figs 8 and 9). Immunohistochemical studies showed positive staining of lymphocytes for CD20 and CD79a, with bcl-2 highlighting the mantle zones. Bcl-6 and CD10 stains were negative. A polymerase chain reaction study for B-cell gene rearrangement revealed positive monoclonality.

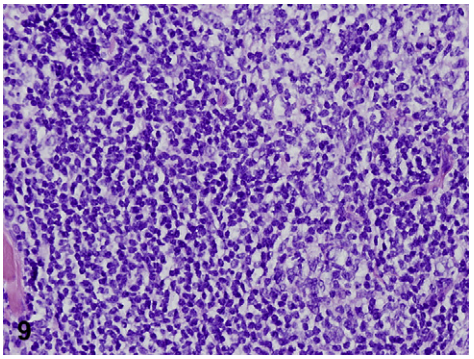
13. An appropriate laboratory work-up includes which of the following?
    a. Biopsy specimen
    b. Computed tomograpic scan of the chest, abdomen, and pelvis
    c. Complete blood cell count, blood chemistries, and lactate dehydrogenase
    d. Bone marrow aspirate
    e. All of the above

The patient was referred to our hematology/oncology service, and subsequent complete blood cell count, chemistries, lactate dehydrogenase, and serum protein electrophoresis values were normal. Prostate-specific antigen, carcinogenic embryonic antigen, and carbohydrate antigen 19-9 values were









also within normal limits. Computed tomographic scans of the patient's chest, abdomen, and pelvis were unremarkable. A bone marrow biopsy was also performed, revealing normal cellularity.

14. What is the most likely diagnosis?
    a. Primary cutaneous B-cell lymphoma
    b. Mycosis fungoides
    c. Metastatic adenocarcinoma
    d. Granuloma annulare
    e. Pseudolymphoma

15. Among the following primary cutaneous B-cell lymphomas, which variety is associated with a good prognosis (5-year survival rates >90%)?
    a. Follicle center lymphoma
    b. Marginal zone lymphoma
    c. Large B-cell lymphoma, leg type
    d. Intravascular large B-cell lymphoma
    e. a and b

16. Which of the following has been shown to be an effective treatment for this disease?
    a. Surgical excision with wide margins
    b. Cyclophosphamide, doxorubicin, vincristine, and prednisolone chemotherapy
    c. Rituximab
    d. Local radiation therapy
    e. All of the above

Discussion

Cutaneous B-cell lymphoma (BCL) is a type of non-Hodgkin lymphoma (NHL) that involves the skin as either the sole site of involvement or as the result of cutaneous spread from systemic disease. Up to 10% of patients with various grades of systemic B-cell NHL will develop skin involvement during the course of the disease and, with the exception of medium grade BCL, is associated with a poor prognosis. Primary cutaneous BCLs (PCBCLs) are defined as malignant B-cell proliferations presenting with cutaneous involvement alone, without evidence of extracutaneous manifestations when complete staging has been performed. The overall incidence of primary cutaneous lymphomas is estimated at one case per 100,000 annually, with 20% representing PCBCL. There is equal incidence in men and women, and the average age at onset is in the late fifties. Lesions present as red to violaceous, painless papules, plaques, or nodules on the head and neck, with the trunk and extremities involved to a lesser extent.

Recently, the World Health Organization and the European Organization for Research and Treatment of Cancer (WHO-EORTC) have collaborated on a classification for cutaneous lymphomas. Among the cutaneous BCLs are primary cutaneous follicle center lymphoma; primary cutaneous marginal zone lymphoma; primary cutaneous diffuse large BCL, leg type; and primary cutaneous diffuse large BCL, other, which includes the rare intravascular large BCL. Each