11. Sciot R, Dal Cin P, de Vos R, van Damme B, de Wever I, van der Berghe H, et al. Alveolar soft part sarcoma: evidence for its myogenic origin and for the involvement of 17q25. Histopathology 1993;23:43-4.
12. Ladanyi M, Lui MY, Antonescu CR, Krause-Boehm A, Meindl A, Argani P, et al. The der(17)t(X;17)(p11;q25) of human alveolar soft part sarcoma fuses the TFE3 transcription factor gene to ASPL, a novel gene at 17q25. Oncogene 2001;20:48-57.
13. Argani P, Lal P, Hutchinson B, Lui MY, Reuter V, Ladanyi M. Aberrant nuclear immunoreactivity for TFE3 in neoplasms with TFE3 gene fusions: a sensitive and specific immunohistochemical assay. Am J Surg Pathol 2003;27:750-61.
14. Argani P, Antonescu CR, Illei PB, Lui MY, Timmons CF, Newbury R, et al. Primary renal neoplasms with the ASPL-TFE3 gene fusion of alveolar soft part sarcoma: a distinctive tumor entity previously included among renal cell carcinomas of children and adolescents. Am J Pathol 2001;159:179-92.
15. Vistica DT, Krosky PM, Kenney S, Raffeld M, Shoemaker RH. Immunohistochemical discrimination between the ASPL-TFE3 fusion proteins of alveolar soft part sarcoma. J Pediatr Hematol Oncol 2008;30:46-52.
16. Deyrup AT, Althof P, Zhou M, Morgan M, Solomon AR, Bridge JA, et al. Paraganglioma-like dermal melanocytic tumor: a unique entity distinct from cellular blue nevus, clear cell sarcoma, and cutaneous melanoma. Am J Surg Pathol 2004;28:1579-86.
17. Swetter SM, Ecker PM, Johnson DL, Harvell JD. Primary dermal melanoma: a distinct subtype of melanoma. Arch Dermatol 2004;140:99-103.
18. Volmar KE, Cummings TJ, Wang WH, Creager AJ, Tyler DS, Xie HB. Clear cell hidradenoma: a mimic of metastatic clear cell tumors. Arch Pathol Lab Med 2005;129:e113-6.
19. Ohta M, Hiramoto M, Fujii M, Togo T. Nodular hidradenocarcinoma on the scalp of a young woman: case report and review of literature. Dermatol Surg 2004;30:1265-8.
20. Saadat P, Cesnorek S, Ram R, Kelly L, Vadmal M. Primary cutaneous paraganglioma of the scalp. J Am Acad Dermatol 2006;54(Suppl):S220-3.
21. Liegl B, Hornick JL, Fletcher CDM. Primary cutaneous PEComa: distinctive clear cell lesions of skin. Am J Surg Pathol 2008;32:608-14.
22. Mentzel T, Reisshauer S, Rütten A, Hantschke M, Soares de Almeida LM, Kutzner H. Cutaneous clear cell myomelanocytic tumor: a new member of the growing family of perivascular epithelioid cell tumors (PEComas): clinicopathological and immunohistochemical analysis of seven cases. Histopathology 2005;46:498-504.

Localized pagetoid reticulosis (Woringer-Kolopp disease) in early childhood

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Localized pagetoid reticulosis, also known as Woringer-Kolopp disease, is an uncommon cutaneous lymphoproliferative disorder classified as a solitary variant of mycosis fungoides. We describe a case of localized pagetoid reticulosis that occurred in early childhood. A 6-year-old boy presented with an erythematous plaque on the left thigh. His parents had first noted brownish erythema of his thigh a few months after birth. The size and elevation of the plaque gradually increased. Physical examination showed well-demarcated reddish plaque, 6.2 × 2.3 cm, with central erosion. Histopathological examination showed an epidermotropic infiltrate of medium atypical lymphocytes with hyperchromatic and pleomorphic nuclei. The atypical cells infiltrated as individual cells or clusters in the epidermis. Immunohistologically, the phenotype of the atypical cells was $CD3^+$, $CD4^-$, $CD8^+$, $CD45RO^+$, $CD20^-$, $CD30^-$, and $CD79a^-$. We discuss the characteristics of this rare disease, including the differential diagnoses. (J Am Acad Dermatol 2009;61:120-3.)

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Localized pagetoid reticulosis (LPR), also known as Woringer-Kolopp disease, is an uncommon cutaneous lymphoproliferative disorder clinically characterized by a unilesional erythematous plaque on the extremities.[1,2] The lesions histopathologically show a markedly epidermotropic infiltrate of atypical T lymphocytes.[3] In the new World Health Organization (WHO)-European Organization for Research and Treatment of Cancer (EORTC) classification for cutaneous lymphomas, it

Abbreviations used:

APACHE:	acral pseudolymphomatous angiokeratoma of children
EORTC:	European Organization for Research and Treatment of Cancer
LPR:	localized pagetoid reticulosis
MF:	mycosis fungoides
WHO:	World Health Organization

is classified as a solitary variant of mycosis fungoides (MF).[4,5] Despite the presence of atypical lymphocytes, lesions enlarge very slowly and local recurrence after treatment has rarely been reported.[4] We report LPR that presented as an erythematous plaque a few months after birth.

CASE REPORT

A 6-year-old Japanese boy presented with an erythematous plaque on the left thigh. His parents had first noted brownish erythema of his thigh a few months after birth. The size and elevation of the plaque increased gradually, and local corticosteroid treatment was not effective. Physical examination showed a well-demarcated reddish plaque, 6.2 × 2.3 cm, with central erosion (Fig 1). The clinical appearance was similar to a hypertrophic scar. An excisional biopsy specimen was taken from the involved skin with a surgical margin of 5 mm. Histopathological examination showed a markedly epidermotropic infiltrate of medium atypical lymphocytes with hyperchromatic and pleomorphic nuclei, and hyperkeratotic acanthosis (Fig 2, *A*). The atypical cells infiltrated as individual cells or clusters into the epidermis at the dermoepidermal junction (Fig 2, *B*). Immunohistologically, the phenotype of the atypical cells in the epidermis was $CD3^+$, $CD4^-$, $CD8^+$, $CD45RO^+$, $CD20^-$, $CD30^-$, and $CD79a^-$ (Fig 2, *C* and *D*). In the papillary dermis, the bandlike lymphocytic infiltrate was $CD4^+$ and $CD8^+$. T-cell receptor gene rearrangement was not detected by genotypic analysis. General examination revealed no evidence of extracutaneous involvement. There was no recurrence during a 5-year follow-up period after the excisional biopsy.

DISCUSSION

LPR, originally reported in 1939, is a rare T-lymphocyte-proliferative disorder, characterized by a favorable clinical course, despite a histologic appearance similar to epidermotropic MF.[1,2] LPR manifests as a solitary, slowly growing erythematous and verrucous plaque on the extremities.[6] The histologic features include hyperkeratotic acanthosis with striking epidermotropism of T lymphocytes.[6] In the new WHO-EORTC classification for cutaneous lymphomas, pagetoid reticulosis is classified as one of the indolent forms of MF[4,5]; however, the relationship to unilesional MF is poorly defined. Unilesional MF is a solitary eczematous lesion, histopathologically identical to classic MF.[7] LPR and MF have some distinct histopathological and immunophenotypical features. LPR is distinguished histologically from MF by the presence of marked hyperkeratosis, prominent pagetoid epidermotropism of atypical lymphocytes with hyperchromatic and hyperconvoluted nuclei, and a dermal infiltrate lacking eosinophils.[6] In contrast, MF cells are present both above and below the dermoepidermal junction.[7] Immunophenotypically, MF cells are $CD4^+$ T-helper cells. In contrast, the immunophenotype of atypical T lymphocytes in LPR is more variable than in MF.[6] Lesional immunohistochemical analysis has revealed that a $CD4^+$, $CD8^+$, or $CD4^-/CD8^-$ phenotype may at times predominate.[8]

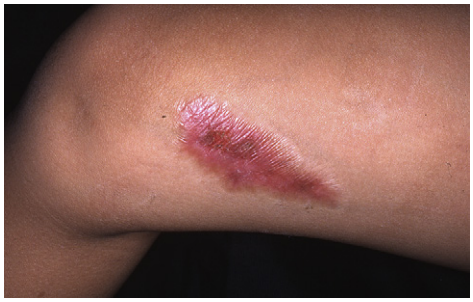

Fig 1. Physical examination revealed 6.2- × 2.3-cm well-demarcated reddish plaque with erosion in the center on the left thigh.

Our case is unusual because the disease presented in a 6-year-old child in whom the lesion had been present since a few months after birth. Histopathological features included striking epidermotropism of medium lymphocytes surrounded by a clear halo. Atypical lymphocytes were located below the dermoepidermal junction and in the epidermis; however, there was no eosinophil infiltration in the dermis. Furthermore, the bandlike infiltrate of lymphocytes expressed both CD4 and CD8 antigens and there was no T-cell receptor gene rearrangement, suggesting that most cells infiltrating the papillary dermis were reactive lymphocytes. Atypical lymphocytes with marked epidermotropism were $CD8^+$, which is distinct from unilesional MF. In contrast to classic MF, extracutaneous dissemination or disease-related deaths have rarely been reported.[4] Moreover, no recurrence for 5 years after excision supported this diagnosis. Most cases of LPR occur in adolescents.[3,8] In our patient, the lesion appeared on the thigh a few months after birth and progressed slowly, suggesting benign indolent behavior.