A Man with Progressive Swelling of the Face and Neck
(See pages 1506–7 for Photo Quiz)

Figure 1. Chronic swelling of the face and neck

Diagnosis: Lymphoepithelial cysts of the parotid glands associated with HIV infection.

The CT findings, as well as those of the percutaneous needle aspiration, are characteristic of lymphoepithelial cysts (LECs) of the parotid glands (figures 1 and 2). Because LECs of the parotid glands are associated with HIV infection, serologic testing for HIV was performed and yielded positive results. The CD4 cell count at the time of diagnosis was 427 cells/mm³ (25%), and the HIV load was 93,000 copies/mL.

LECs of the salivary glands were first reported by Mikulicz in 1888; their association with HIV infection was reported by Morris and Moore in 1987 [1]. The incidence of LECs in patients infected with HIV is 3%–6%. In HIV-infected patients, cysts are more likely to be bilateral, large, and multiloculated [2]. CD4 cell counts of patients with LECs range from 300 to 600 cells/mm³ [1]. Cervical lymphadenopathy is palpable in 50% of patients and is demonstrated by imaging studies (CT or MRI) in 100% of patients. HIV test results are positive for 66%–100% of patients with LECs [3]. Considering the rarity of LECs in HIV-negative patients, HIV testing should be con-
sidered for every patient who presents with this syndrome. The natural course of LECs is slow progression without malignant transformation or functional morbidity, although LECs ultimately can be a cause of significant cosmetic deformity.

Several theories have been proposed to explain the pathogenesis of LECs in HIV-infected patients. The parotids are the only salivary glands containing intracapsular lymph nodes; this may explain the selective tropism of LECs for the parotid glands. Encapsulation of the lymph nodes in the parotid glands is incomplete, and lymphocytes can affect the adjacent parenchyma by stimulating proliferation of ductal epithelium and replacement of acinar structures [3]. Alternatively, LECs may originate from proliferation of the glandular epithelium that is trapped within intraparotid lymph nodes [4]. Increased incidence of LECs in patients with HIV infection probably reflects the high concentration and rapid turnover of HIV in hyperplastic lymphoid tissue in and adjacent to parotid parenchyma causing lymphoid duct entrapment and cystic dilatation [5]. The presence of high levels of HIV-1 p24 antigen and high HIV load in cystic fluid probably reflects continuous shedding of HIV-infected cells from the surrounding lymphoid tissue. Some investigators have also suggested that the formation of LECs could result from an autoimmune disorder. There is no evidence that either cytomegalovirus or Epstein-Barr virus is involved in pathogenesis of LECs [6].

Therapy for LECs is aimed at correction of cosmetic deformities. Surgical excision, repeated percutaneous aspiration, sclerotherapy with doxycycline or tetracycline, low-dose radiation therapy, and HAART are being used with variable success [5–10]. Several small, nonrandomized, nonblinded studies have demonstrated the resolution of LECs with HAART and/or short courses of steroid therapy [2, 6–8]. These findings and the fact that most of the patients with LECs present with relatively high CD4 cell counts generate a new dilemma regarding timing of initiation of antiretroviral therapy.

Alexander T. Kessler, A. P. Kourtis, and R. Jurado
Division of Infectious Diseases, Department of Internal Medicine, Emory University School of Medicine, Atlanta, Georgia

References

Reprints or correspondence: Dr. Alexander T. Kessler, 540 Wembley Circle, Atlanta, GA 30328 (sandrok72@yahoo.com).

Clinical Infectious Diseases 2003; 37:1565–7
© 2003 by the Infectious Diseases Society of America. All rights reserved. 1058-4838/2003/3711-0020$15.00