AN EBV-ASSOCIATED SALIVARY GLAND CANCER

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SUMMARY

This is a case report of a 57-year-old Alaskan Native woman diagnosed with malignant lymphoepithelial lesion of the parotid gland. Extensive morphologic studies documented that the lesion was epithelial with a predominantly T-cell lymphocytic infiltrate. Tests for EBV demonstrated the tumor was positive for EBNA and EBV DNA, while adjacent non-malignant tissue was negative.
Eskimos of Alaska, Canada, and Greenland are at increased risk for cancers of both the nasopharynx and salivary gland (Lanier et al, 1980; Nielsen et al, 1977, 1978; Wallace et al, 1963; Mallen and Shandro, 1974). The excess risk of salivary gland cancer has been found to be largely due to the occurrence of unusual tumors classified as malignant lymphoepithelial lesions (Wallace et al, 1963; Arthaud, 1972; Nielsen et al, 1977). These malignant lymphoepithelial lesions (MLEL) are characterized by islands of anaplastic cells in a dense lymphocytic background.

Although the majority of tumors of this type have been reported to date among Eskimos, there have also been reports of malignant lymphoepithelial lesions in Caucasians and Blacks of Europe and America, and most recently, Japanese and Chinese (Ferlito and Donati, 1977; Gravanis and Giansanti, 1970; Nagao et al 1983; Redondo et al, 1981; Dong and Lo, 1983).

In Alaska during the time period 1966 through 1980, 16 Alaskan Natives (Eskimos, Indians and Aleuts) developed malignant tumors of the major salivary glands. The observed to expected ratio (based on U.S. white rates) was significantly high (4.7) in females and high (1.7), but not significantly high, in males. Approximately twice as many females as males developed the cancers. The age range was 17 to 70, however, all but one salivary gland cancer patient was diagnosed under age 60. Twelve of the 16 salivary gland cancers were MLEL. To date, tumor tissues from Eskimo patients with MLEL, (two from Alaska and one from Greenland) have been reported to be positive for EBV by DNA hybridization techniques (Lanier et. al, 1981; Saemundsen et. al., 1982).

The presentation in January, 1983, of a 57-year-old Alaskan Native woman with a salivary gland tumor which was classified as a malignant lymphoepithelial lesion, provided an opportunity to study the tumor for EBV and morphologic characteristics, and to evaluate the patient for evidence of autoimmune disease, such as primary and secondary Sjogren's syndrome.

Histologically the 1.5 cm tumor included islands of neoplastic cells within a dense lymphocytic background