Sclerosing Mucoepidermoid Carcinoma with Eosinophilia: Characterization of a Rare Distinct Entity

Erik Ames, Ph.D.1, Michael J. Campbell, M.D.2, Alaa Afify, M.D.1, Jeffrey F. Krane M.D., Ph.D.3 and Eric C. Huang, M.D., Ph.D.1

1University of California, Davis Medical Center, Department of Pathology and Laboratory Medicine, Sacramento, CA 95817, USA; 2University of California, Davis Medical Center, Department of Surgery, Sacramento, CA 95817, USA; 3Brigham and Women’s Hospital and Harvard Medical School, Department of Pathology, Boston, MA 02115, USA

ABSTRACT

Sclerosing mucoepidermoid carcinoma with eosinophilia (SMECE) is an extremely rare thyroid carcinoma with limited cytologic descriptions in the literature. Here we present a 52-year-old woman with a 3.9 cm thyroid nodule. Fine needle aspiration smears consisted of a highly cellular specimen with tumor cells in isolated patterns and solid squamoid nests. Tumor cells had round to oval nuclei, prominent nucleoli, smooth nuclear contours and moderate amounts of dense cytoplasm. In addition to the polymorphous population of lymphocytes, the background contained a striking abundance of eosinophils. The subsequent right thyroidectomy showed histologic features diagnostic for SMECE.

INTRODUCTION

Sclerosing mucoepidermoid carcinoma with eosinophilia (SMECE) of the thyroid was first described by Chan and colleagues in 1991 as a distinct entity which occurs in a background of chronic lymphocytic (Hashimoto’s) thyroiditis. It is an exceedingly uncommon entity with fewer than 20 articles available in the English literature to date. SMECE occurs predominantly in middle-aged women typically as a single nodule confined to a unilateral thyroid lobe. The defining histologic features of SMECE include squamoid and mucoid cells with eosinophilic inclusions in a setting of chronic lymphocytic thyroiditis. A recent study demonstrated that these tumors are distinct from mucoepidermoid carcinoma of the salivary gland, based on both histologic appearance and genetic analysis: SMECE uniformly lacks the MAML2 rearrangement which is seen in roughly two-thirds of mucoepidermoid carcinomas. While these tumors typically follow an indolent course, they can be locally aggressive. In addition, extrathyroidal spread and metastasis have been reported. Given the uncommon nature of SMECE, the majority of studies have described the histologic features of this unique tumor, though limited cytomorphic descriptions exist in the literature3,11. Here we report the cytologic characteristics from fine-needle aspiration (FNA) smears and correlate with the histologic findings of this rare disease.

CASE AND RESULTS (continued)

Gloss and Histologic Findings

During the surgery, the thyroid was noted to be extremely inflamed with numerous heterogenous nodules. The 2.1 x 0.9 x 0.6 cm right nodule contained a 3.9 cm dominant nodule occupying the entire upper mid pole. This nodule was firm, well-circumscribed, tan-white and homogenous in appearance (Fig 3A). The nodule abutted the surgical margins without gross involvement.

Histologically, the tumor consisted of variably-sized nests within a dense, fibrous stroma (Fig. 3B). Higher magnification revealed numerous eosinophils within the stroma surrounding well-circumscribed clusters of tumor cells (Fig. 3C). Squamous differentiation with keratin pearls was present (Fig. 3D) as well as goblet cells and mucous pools within the nests (Figs. 3E and 3F). The uninvolved thyroid demonstrated chronic lymphocytic thyroiditis. A definitive diagnosis of SMECE was established.

CONCLUSIONS

Sclerosing mucoepidermoid carcinoma with eosinophilia is an exceedingly unusual malignancy of the thyroid. Histologically, the tumors consist of variably-sized nests of squamoid and mucin secreting cells in a fibrous stroma rich in eosinophils and in a background of chronic lymphocytic (Hashimoto’s) thyroiditis. The cytologic features of SMECE recapitulate its histologic features quite well, though cytology alone may be difficult to make a definitive diagnosis. The main cytologic characteristics of SMECE in our case included hypercellularity, oval to round nuclei, prominent nucleoli and rare nuclear grooves. Some clusters showed squamous differentiation with scattered keratin pearls. The background contained abundant eosinophils and lymphocytes.

It is important to cytologically differentiate SMECE from other thyroid neoplasms for prognostic and therapeutic purposes. While SMECE shows occasional grooves and has been reported to contain intranuclear inclusions, nuclear contours in SMECE are quite smooth and lack the pale chromatin typical of papillary carcinoma. Architecturally, SMECE consists of single isolated cells and nests of tumor cells, rather than microfollicles: thus, follicular neoplasms can be excluded. However, this architectural pattern may be suggestive of medullary carcinoma. Fortunately, calcitonin staining is absent in SMECE. Conventional mucoepidermoid carcinoma (MEC) of the thyroid should also be considered along with SMECE in the presence of squamoid and mucous cells. However, MEC lacks the dense eosinophilic infiltrate and typically displays more mucinous cells. Additionally, MEC is less likely to occur in a background of Hashimoto’s thyroiditis. SCC may also be considered in cases where mucous cells are rare. Here, SCC can be identified by its more prominent atypia and mitotic activity. Again, SMECE would not be as pronounced.

Adapted from “Papillary Thyroid Carcinoma”. University of Iowa. https://medschool.ucdavis.edu/depts/papillary_thyroid_carcinoma