Summary

We present a series of 16 salivary gland tumors with histomorphological and immunohistochemical features reminiscent of secretory carcinoma of the breast. This is a hitherto undescribed and distinctive salivary gland neoplasm, with features resembling both salivary acinic cell carcinoma and low grade cystadenocarcinoma, as well as displaying strong similarities to breast secretory carcinoma. Microscopically, the tumors have a lobulated growth pattern and are composed of microcystic and glandular spaces with abundant eosinophilic homogenous or bubbly secretory material positive for PAS, mucicarmine, MUC1, MUC4 and mammaglobin. The neoplasms also show strong vimentin, S-100 protein, and STAT5a positivity. For this tumor we propose a designation mammary analogue secretory carcinoma of salivary glands (MASC).

The 16 patients comprised 9 men and 7 women, with a mean age of 46 years (range 21-75). Thirteen cases occurred in the parotid gland, and one each in the minor salivary glands of the buccal mucosa, upper lip, and palate. The mean size of the tumors was 2.1 cm (range 0.7 to 5.5 cm). The duration of symptoms was recorded in 11 cases and ranged from 2 months to 30 years. Clinical follow-up was available in 13 cases, and ranged from 3 months to 10 years. Four patients suffered local recurrences. Two patients died, one of them due to multiple local recurrences with extension to the temporal bone, and another due to metastatic dissemination to cervical lymph nodes, pleura, pericardium and lungs.

We have demonstrated a t(12;15) (p13;q25) *ETV6-NTRK3* translocation in all but one case of MASC suitable for analysis. One case was not analyzable and another was not available for testing. This translocation was not found in any conventional salivary acinic cell carcinoma (12 cases), nor in other tumor types including pleomorphic adenoma (1 case) and low grade cribriform cystadenocarcinoma (1 case), whereas *ETV6-NTRK3* gene rearrangements were proven in all three tested cases of mammary secretory carcinoma. Thus, our results strongly support the concept that MASC and acinic cell carcinoma are different entities.