CASE REPORT

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FAMILIAL BILATERAL ACINIC CELL CARCINOMA OF THE PAROTID SYNCHRONOUS WITH PITUITARY ADENOMA: CASE REPORT

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Abstract: Background. Acinic cell carcinoma is a common neoplasm of the salivary glands that occurs predominately in the parotid. Only one case of a familial recurrence of such a neoplasm and 16 cases of bilateral tumors have been reported.

Methods. History files and histologic reports of a patient with bilateral multifocal acinic cell carcinoma of the parotid and a synchronous pituitary adenoma, and of the patient’s sister and his father, also treated for parotid tumors, were retrieved.

Results. There was one recurrence of acinic cell carcinoma in the family. A pituitary tumor was a chromophobe gonadotrophic adenoma.

Conclusions. This is the 17th case of bilateral acinic cell carcinoma of the parotid gland and the second reported case with a familial recurrence. It is the first with a synchronous pituitary adenoma. © 2005 Wiley Periodicals, Inc. Head Neck 27: 825–828, 2005

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Acinic cell carcinoma is a common neoplasm of the salivary glands occurring predominately in the parotid. It is the third most common epithelial malignancy of the salivary gland. It accounts for 17% of all primary salivary gland malignancies and for 6% of salivary gland neoplasms. Although it has been reported that 3% of the cases in children are bilateral, only 16 such cases in adults have been reported to date, one of which was an extraparotid localization in the submandibular gland.

The mean age of the patients is 44 years, with 12% younger than 20 years. The male-to-female distribution is 2:3.

Overall, the survival rate has been crudely estimated to be about 84%. The reported 5-year determinant (disease-specific) survival rate ranges from 76% to 90%. One should note that survival analyses should be interpreted with the understanding that 10 to 20 years of follow-up are
needed to identify the full impact of acinic cell carcinoma on extended survival.³

A familial occurrence of acinic cell carcinoma has been reported only once⁴ in a family in which the father and daughter were treated for acinic cell carcinoma of the parotid gland; however, several reports with a familial distribution of other neoplasms of the salivary glands, including pleomorphic adenoma, Warthin’s tumor, carcinoma of the submandibular gland, and malignant lymphoepithelial lesion, are present in the literature.⁴

CASE REPORT

A 64-year-old man was initially seen with bilateral parotid gland tumors that were noted by the patient 6 months before seeking medical advice. He had no pain, facial nerve impairment, local signs of inflammation, or lymph node enlargement. A CT scan (Figure 1) showed the tumors on both sides to be confined to the superficial lobe with defined borders. On the right side, a second, smaller foci was described. An MRI (Figure 2) was performed, and, apart from the parotid tumors, an incidental finding of a large (1.8 × 1.7 cm) pituitary adenoma was noted. Because of the size of the adenoma, it was decided to first treat the pituitary tumor, and the patient was referred for hypophysectomy. Two months later, a standard superficial parotidectomy approach was performed on the side with the larger tumor (right side). During the preceding period, no apparent change in the tumors’ size or clinical features was noted. Intraoperatively, two additional smaller lesions were discovered; these were determined by frozen section analysis to be malignant, so a near-total parotidectomy was performed, with preservation of the facial nerve, because it was not infiltrated by the tumor, at least macroscopically.

Histopathologic analysis showed the tumors to be acinic cell carcinoma, with typical features. Two intraparotid lymph nodes were found but were not infiltrated by the neoplasm.

Fine-needle aspiration cytology on the other side was suspicious for a similar diagnosis; therefore, in a second stage, a near-total parotidectomy was performed on the left side, with preservation of the facial nerve. Permanent section of the specimen also showed a multifocal acinic cell carcinoma.

The patient’s family history revealed that both his older sister and his father had undergone parotidectomy because of parotid tumors. A history file of his sister showed that she also was...
treated for bilateral parotid tumors. In 1971, at age 27, the sister underwent removal of an acinic cell carcinoma of the right parotid gland, and 9 years later, because of a recurrence, she underwent total parotidectomy with resection of the facial nerve. In 1994, when she was 50 years old, a tumor of the deep lobe of the left parotid (Figure 3) was treated with parotidectomy. This tumor was an oncocytic adenoma.

The father of both the aforementioned patients was also treated with right-sided superficial parotidectomy in 1989 (at age 89) because of a large Warthin’s tumor.

Apart from the histologic reports, slides of the specimens were not available for reviewing.

In the right-sided parotid gland, three encapsulated tumors of maximum diameters of 1, 2.5, and 3 cm were found, respectively. They consisted of sheets of polygonal cells with granular, lightly basophilic cytoplasm and uniform nuclei resembling serous acinar cells (Figure 4). Periodic acid–Schiff (PAS) stain was positive, and the neoplasm was infiltrating the capsule and surrounding tissue. The left-sided tumor, with a maximum diameter of 2.7 cm, had similar histologic findings, typical of acinic cell carcinoma. Two intraparotid lymph nodes were found on the right side and one on the left side, all with no neoplastic invasion.

Regarding the pituitary tumor, the specimen showed typical histologic findings of chromophobic adenoma by light microscopy (Figure 5). The PAS stain was negative, whereas immunostains using the standard avidin-biotin-peroxidase complex (ABC) method were positive for beta–follicle stimulating hormone (β-FSH) and α-subunit of glycoprotein hormones (α-SU), was focally positive for beta–luteinizing hormone (β-LH), and was negative for growth hormone (GH), prolactin (PRL), adrenocorticotropic hormone...
(ACTH), beta–thyroid-stimulating hormone (β-TSH) and β-FSH (Figure 6).

**DISCUSSION**

This case, the first such to be reported, is unique for three reasons. It regards a bilateral, multifocal acinic cell carcinoma of the parotid in a patient with synchronous pituitary tumor and a familial predisposition.

With the exception of adenolymphoma, which is bilateral in 5% to 10% of cases, bilateral parotid tumors are uncommon. Bilateral acinic cell carcinomas are reported to exist in 3% of acinic cell carcinomas in children. Published cases involving children are relatively few, but only 16 other cases in adults have been published before now. Acinic cell carcinoma is considered to present with multiple foci, leading to the conclusion that wide excision is mandatory for successful treatment.

Regarding the pituitary tumor, we believe that this is probably an incidental finding, because no histologic, genetic, or environmental features are known to connect the two tumors. A metastasis to the pituitary gland was once reported from an adenocarcinoma of the parotid gland 18 months after initial treatment, but no case has ever been reported of an adenoma of the pituitary synchronous with an acinic cell carcinoma of the salivary glands. Whether this could be the first reported case of a multiple endocrine-exocrine tumor syndrome is of course something that could be further studied.

This is the second case of familial recurring acinic cell carcinoma of the parotid worldwide. The first such case occurred in a 35-year-old father and his 16-year-old daughter, with an 8-year period between presentations. No chromosome analysis was conducted in the aforementioned or this report, even though genetic alterations have been observed in acinic cell carcinoma in one study. These include regions at chromosomes 4p, 5q, 6p, and 17p that were found to be more frequently altered than regions on chromosomes 1p and 1q, 4q, 5p, and 6q, which were investigated in the specific study.

It is remarkable that although other salivary gland neoplasms seem to have a familial distribution, there has been only one other similar case of familial acinic cell carcinoma.

One should not exclude a coincidental event, but nevertheless, the second such case raises the possibility of common genetic or environmental risk factors.

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**REFERENCES**