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Acinic cell carcinoma originating in the nasal septum

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SUMMARY

Acinic cell carcinoma represents approximately 2.5% to 4% of all salivary gland tumors and 13% of all malignant parotid tumors. The nasal septum is a rare site of an acinic cell carcinoma. We have treated such a tumor in a 47-year old man who had been asymptomatic for the past 1.5 year. This is the second reported case of acinic cell carcinoma at the nasal septum.

Key words: acinic cell carcinoma, nasal septum.

INTRODUCTION

Acinic cell carcinoma (ACC) was first described by Nasse (1892), its malignant potential was first realized by Buxton et al., (1953). Foote and Frazell (1953) classified these tumors as acinic cell adenocarcinoma. There are no reliable clinical characteristics or histopathologic criteria to aid appropriate initial treatment or prognosis, although the clinical stage appears to be the most significant prognostic feature (Spiro et al., 1978).

ACC represents approximately 2.5% to 4% of all salivary gland tumors and 13% of all malignant parotid tumors (Batsakis, 1979). ACCs are rare in minor salivary glands and usually occur in the oral cavity. Acinic cell carcinomas arising in the nasal cavity are rare with few documented reports in the literature (Perzin et al., 1981; Takimoto et al., 1989; Ordonez et al., 1986; Hanada et al., 1988). We present the second reported case of ACC arising at the nasal septum. Finkelhor et al., (1987) reported another case.

CASE REPORT

A 47-year-old man was admitted with a one-month history of increasing difficulty in breathing through the left nostril. Rhinoscopic examination revealed that the left nasal cavity was obstructed by a well-defined reddish-gray mass having an easily bleeding surface measuring 1 x 1 x 0.5cm in greatest dimensions. It was based on the left anterior nasal septum. Initial biopsies resulted in a diagnosis ACC. Magnetic resonance imaging showed that the tumor was located on the left nasal septum (Figure 1). Computed tomographic scan showed thickening of the septal mucosa on the left side. There was no evidence of bone destruction or sinus involvement (Figure 2).

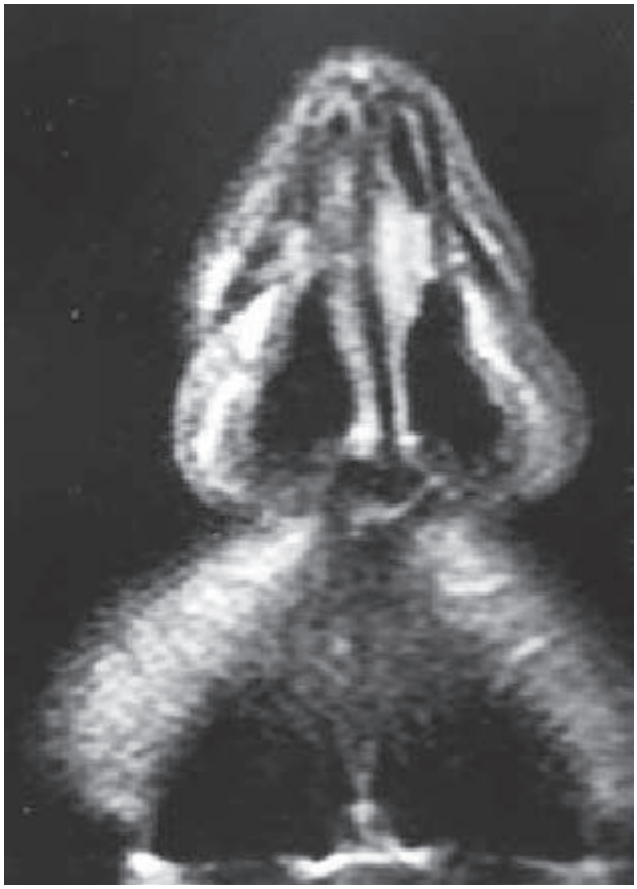

Figure 1a. T2-weighted coronal MRI showing tumor was located on the left nasal septum.

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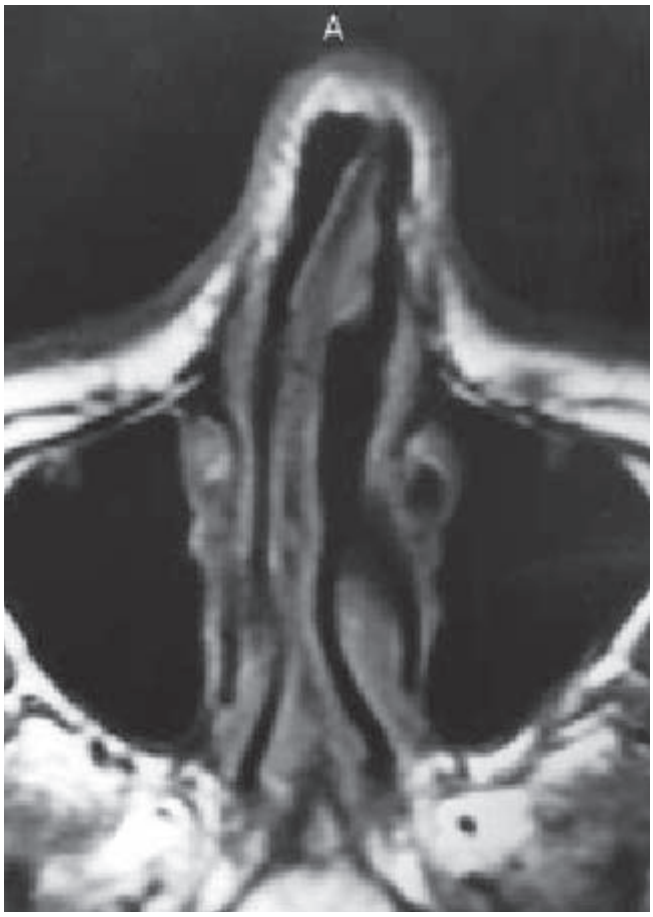

Figure 1b. T1-weighted axial MRI showing tumor was located on the left nasal septum.

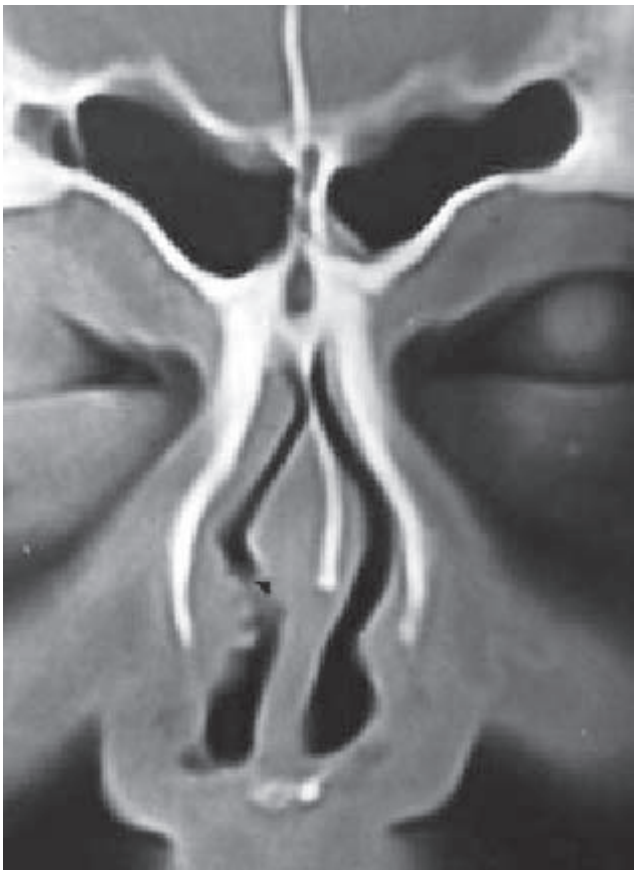

Figure 2. CT scan shows thickening of the septal mucosa on the left side (arrowhead).

Surgical removal of the tumor was performed by left lateral rhinotomy, under general anesthesia. The macroscopic findings during the operation suggested that the primary site of the tumor was the left anterior nasal septum. The tumor was dissected with nasal septal cartilage, and the defect was covered with a free skin flap from the thigh.

The patient has since done well and to date has shown no local tumor recurrence or metastasis.

Histopathological findings

Gross examination of the excised specimen showed a well-delineated mass located on the left anterior nasal septum measuring 1.2 x 1.2x 0.7 in diameter (Figure 3). The mass was soft to rubbery, pale pink in color and had a smooth easily bleeding surface. The cut section was homogeneous and hyperemic. The hematoxylin and eosin (H&E) stained sections exhibit a well-circumscribed tumor with pure solid growth pattern (Figure 4a, 4b). The tumor is composed of a mixed cell population in which well-differentiated acinar cells predominate. Acinar cells are round to polygonal in shape, have hyperchromatic round nuclei and large basophilic to amphophilic cytoplasm which contains dark, periodic acid Schiff (PAS) positive granules. Also noted are few numbers of intercalated ducts like cells and scattered clear cells. Necrosis and mitoses were absent. All surgical resection margins were free of tumor.

DISCUSSION

ACC is an uncommon, low-grade, slow-growing malignancy composed of cells that resemble normal acinar cells (Spafford et al., 1991). ACC is believed to be derived from the pluripotent intercalated duct cell itself or anywhere along its line of differentiation to the mature serous acinar cell. Abrams et al., (1965) studied the histomorphological features of ACCs and examined the cell types and growth patterns present. The architectural growth patterns are categorized as solid, microcystic, papillary-cystic, and follicular. Cellular features are identified as acinar, intercalated ductal, vacuolated, clear, and nonspecific glandular. These descriptive categories do not define specific subtypes of

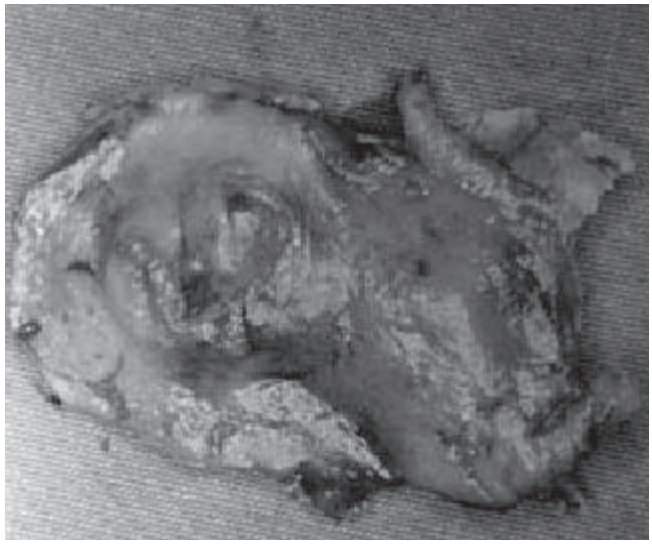

Figure 3. Excised tumor with nasal septal cartilage.

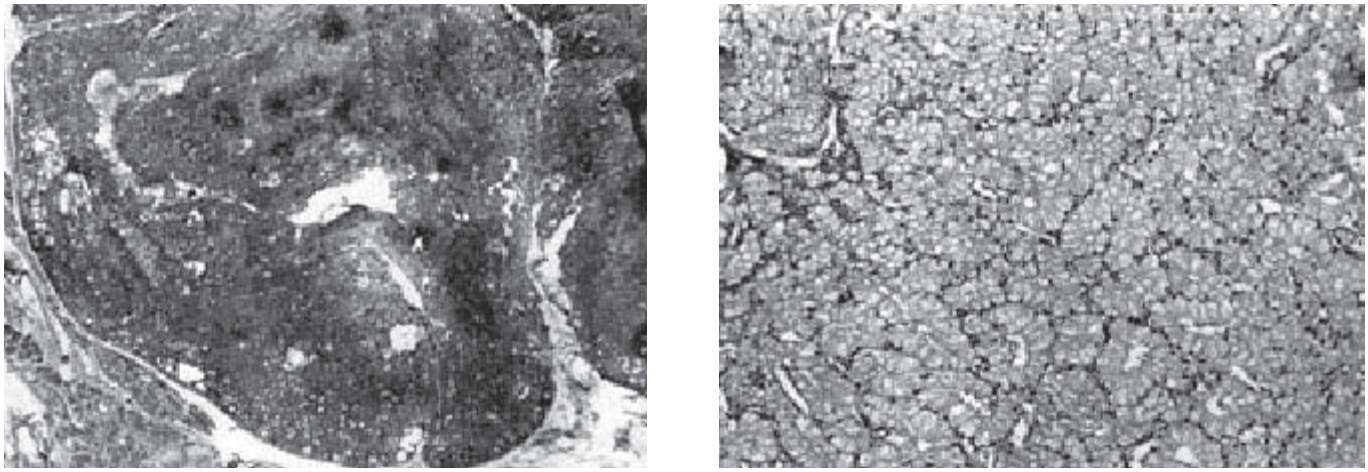

Figure 4a, 4b. The section demonstrates a solid growth pattern in which well-differentiated acinar cells predominate (Figure 4a: H&E stain, ×40, Figure 4b: H&E stain, ×200).

ACC; the patterns are not exclusive of one another but rather help to define the spectrum of histologic features of these tumors. Although an individual tumor usually has a predominance of one pattern or cell type, many tumors have a mixture of features (Ellis and Auclair, 1996). Batsakis et al., (1979) divided ACC histologically into high-and low-grade lesions. High-grade lesions demonstrate aggressive local invasion, extraparenchymal invasion, medullary growth architecture, and prominence of undifferentiated cells.

Many investigators have found that the biologic behaviour of ACC cannot be reliably predicted on the basis of histomorphologic features (Ellis and Auclair, 1996).

In the histopathological differential diagnosis of these tumors, ACCs are unlikely to be confused with other neoplasms as long as the basophilia of the cytoplasm is not overlooked. Since a typical acinic cell tumor is reported to be generally PAS-positive and not to react to mucicarmine staining, the diagnosis of a well-differentiated acinic cell tumor is relatively easy to make (Hanada et al., 1988).

ACC is usually confined to the parotid gland; although occasionally it may arise in the submandibular, sublingual, and minor salivary glands (Batsakis, 1979). The male-female ratio is approximately 1.5:1 in most studies (Spiro et al., 1978; Lewis et al., 1991). The diagnosis is usually made during the fifth decade of life, although the tumor may occur at any age. After mucoepidermoid carcinoma, ACC is the second most common salivary gland malignancy in childhood (Batsakis, 1979).

Reported incidences of recurrence, metastases, and patient deaths for ACC vary widely. Metastasizes to regional lymph nodes occur in 10% to 16% of patients, and distant metastases occur in approximately 10% (Spiro et al., 1978, Batsakis et al., 1979). Local recurrence rates of 33% to 85% have been reported (Spiro et al., 1978, Batsakis et al., 1979). Distant metastases are primarily to lung and bone (Levin et al., 1975). ACC has the longest disease-free interval and the highest survival of all salivary malignancies, with 83% to 94% and 78% to 85% of patients alive at 5 and 10 years, respectively (Spiro et al., 1978; Batsakis et al., 1979; Spafford et al., 1991; Spiro et al., 1989). Recurrences are often delayed, and a follow-up of 20 years or more may be necessary to evaluate treatment outcome (Spafford et al., 1991). As with other head and neck cancers, the clinical stage determines prognosis (Batsakis et al., 1979).

Although ACC is relatively common in the parotid gland, it is extremely rare in extra parotid sites, with less than 90 cases reported in the literature. Minor salivary gland tumors com-prise approximately 23% of all salivary tumors, and up to 88% of minor salivary tumors are malignant (Spiro et al., 1973). A number of studies of ACC arising in minor salivary glands indicate that these tumors are much less aggressive than those that occur in the parotid and submandibular glands (Abrams and Melrose, 1978; Castellanos and Lally; 1982; Chen et al., 1978; Gardner et al., 1980; Zbaeren et al., 1991). Tumors in the minor salivary glands rarely metastasize and rarely lead to death (Ellis and Auclair, 1996).

ACCs of minor salivary origin have been observed in a variety of sites, including the palate, buccal mucosa, lip, tongue, retromolar or gingival region, mandible, paranasal sinuses, nasal cavity, and nasal septum, as in this report (Zbaeren et al., 1991). The incidence of malignant tumors originating from the nasal septum is very low, representing approximately 1% of human malignancies (Beatty et al., 1982). Most commonly, squamous cell carcinoma is seen, but adenocarcinoma, malignant melanoma, reticulum cell sarcoma, basal cell carcinoma, histiocytic lymphoma, transitional cell carcinoma, chondrosarcoma, adenoid cystic carcinoma, small-cell carcinoma, lymphoepithelioma, mucoepidermoid carcinoma, and plasmacytoma have been reported (Finkelhor et al., 1987).

Treatment modalities for nasal cancer have evolved over the years. Because of the high recurrence rate, the best chance for cure lies in complete surgical excision of the tumor at the time of initial treatment. More recent series incorporate endoscopic sinus surgery, microscopic surgical techniques, lateral rhinotomy, and combined cranial resection for advanced disease.

Finkelhor et al., (1987) reported the first case of ACC arising at the nasal septum. The present case is the second report of ACC occurring at the nasal septum. The cytomorphology findings of our case are similar to those described in cases of ACC arising from major salivary glands. In our case, the tumor was dissected

with nasal septal cartilage, and the defect was covered with a free skin flap from the thigh. There was no evidence of residual cancer present in the final definitive surgical procedure. Because there was also no evidence of regional or distant metastasis, and the tumor was a low-grade lesion, no further treatment was given. Our patient has been disease-free for the past 1.5 years. We think it is too early to comment on the success of the treatment.

In summary, ACC is a low-grade malignant salivary neoplasm that is rarely diagnosed in minor salivary glands, especially minor salivary glands of the nasal septum. Prognosis is directly related to the extent of lesion and the adequacy of the initial resection.

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