Pleomorphic Adenoma of the Nasal Septum — A Case Report

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ABSTRACT

Pleomorphic adenomas are the most common benign tumor of the major salivary glands. In addition to the major salivary glands, they may also occur in the salivary glands of the hard and soft palate. Rare cases have been reported in the nasopharynx, oropharynx, hypopharynx, and larynx. Intranasal pleomorphic adenomas are quite rare and may be misdiagnosed because they have greater myoepithelial cellularity and few myxoid stromata compared to those elsewhere. We present a rare case of a pleomorphic adenoma of the nasal septum and discuss the pathologic findings and clinical management. The literature concerning the subject is reviewed. (***Tzu Chi Med J*** 2005; **17**:47-49)

Key words: pleomorphic adenoma, mixed tumor, nasal septum

INTRODUCTION

Pleomorphic adenomas (mixed tumors) are the most common benign tumor of the salivary glands. They appear, in decreasing frequency, in the parotid gland, submandibular gland, and palate [1]. They may occur in minor salivary glands, especially those in the hard and soft palate [2]. Intranasal pleomorphic adenomas are quite rare, and are frequently misdiagnosed because they are highly cellular and compared with pleomorphic adenomas of the major salivary glands, have few myxoid stromata [2]. We report a nasal septal pleomorphic adenoma in a 40-year-old male and discuss the literature on this rare entity.

CASE REPORT

A 40-year-old male complained of progressive right nasal obstruction with mild rhinorrhea for about 6 months. Occasionally, the rhinorrhea was tinged with blood. The patient had neither facial pain nor a postnasal dripping sensation. He came to our clinic in August 2003. Examination revealed a soft, gray mass with a broad base obstructing the right nasal cavity (Fig. 1), without cervical adenopathy. Other examination results were normal. Computed tomography showed a mass in the right nasal cavity, sized about $2 \times 1.1 \times 1$ cm (Fig. 2). Since the nature of the mass was uncertain, wide excision of the mass was performed under endoscopy. During surgery, the soft, gray tumor was found to be attached to the superior nasal septum and to be encroaching onto the lateral nasal wall. The tumor was removed with the attached perichondrium of the septal cartilage and the periosteum of the perpendicular plate of the ethmoid bone. Under sinuscopy, the underlying septal cartilage and perpendicular plate of the ethmoid bone were found to be intact. The final histopathologic result for the tumor was a pleomorphic adenoma (Fig. 3), and the resection margin was tumor-free. The patient was discharged on the same day, and the postoperative course was uneventful. After 4 months, the patient had experienced no further problems with the nasal airway, and a

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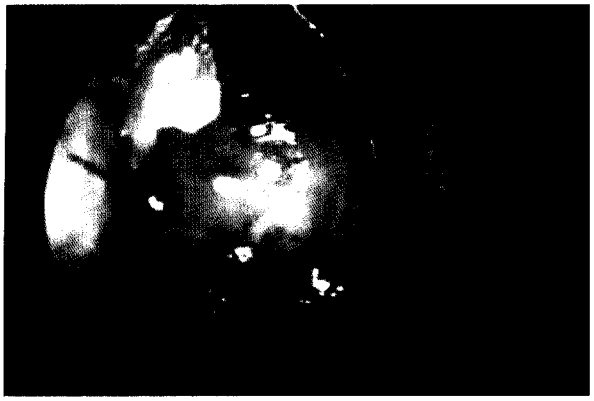

Fig. 1. A soft, gray mass with a broad base attached to the superior portion of the right nasal septum. It is obstructing the right nasal cavity.

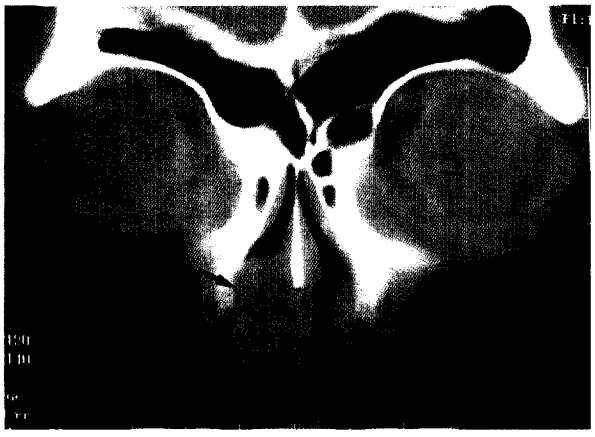

Fig. 2. Sinus CT scan (coronal section; window width, 2000; center, -100) showing a mass in the right nasal cavity, sized about 2 × 1.1 × 1 cm (arrow).

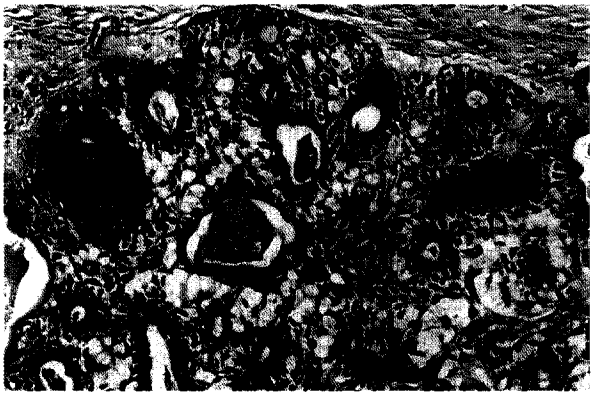

Fig. 3. Histopathologic section demonstrating a pleomorphic adenoma with increased myoepithelial cellularity and a relatively small stromal component (hematoxylin-eosin, × 200).

nasal endoscopic examination revealed no recurrence of the disease. Because of the known propensity of pleomorphic adenomas to recur, follow-up of this patient is continuing.

DISCUSSION

Pleomorphic adenomas are the most common tumor of the major salivary glands, but are unusual in the respiratory tract. Rare cases have been reported in the nasal cavity, paranasal sinus, nasopharynx, oropharynx, hypopharynx, and larynx [3]. Batsakis stated that within the upper respiratory tract, the most favored site of origin is the nasal cavity, followed by the maxillary sinus and the nasopharynx [4]. The first reported case in the literature of a pleomorphic adenoma of the nasal cavity was by Denker and Kahler [5]. Although the vast majority of minor mucous and serous glands are located in the lateral nasal wall, pleomorphic adenomas in the nasal cavity mostly originate from the nasal septum. Large series studies of nasal pleomorphic adenomas include 40 cases reported by Compagno and Wong, 41 cases by Suzuki et al, and 59 cases reported by Wakami et al [6-8]. Fewer than 20% of the intranasal pleomorphic adenomas were noted to have arisen from the lateral nasal wall, with the rest originating from the nasal septum [6-8]. Demographically, the majority of tumors are noted in the third through sixth decades of life. Patients typically present with the complaints of unilateral nasal obstruction (71%) and epistaxis (56%). Other complaints include nasal swelling, a mass in the nose, epiphora, and mucopurulent rhinorrhea [7].

Pleomorphic adenomas are characterized by epithelial tissue mixed with tissues of mucoid, myxoid, or chondroid appearance. The features of pleomorphic adenomas in the aerodigestive tract are somewhat similar to those of mixed tumors of the salivary glands. Nevertheless, some differences are recognized. Myoepithelial cellularity is unusually increased in these tumors compared with major salivary gland tumors. Therefore, epithelial elements rather than the stromal elements predominate. Occasionally, pleomorphic adenomas are composed almost entirely of epithelial cells with few or no stromata [6]. Because of the high cellularity and lack of a stromal component, histologically, they resemble aggressive epithelial tumors. Haberman and Stanley reported a case of a nasal septal pleomorphic adenoma misdiagnosed as an adenoid cystic carcinoma on the basis of tissue biopsy [9]. This difficulty is reflected in a study by Compagno and Wong, in which 55% of cases were initially misdiagnosed [6].

Many authors have speculated as to the etiology of these tumors. Stevenson suggested that mixed tumors in the nasal septum originate from the remnants of the vomeronasal (Jacobson's) organ [10]. However, mixed tumors also occur in the lateral wall, where no such areas exist. Matthew believed that the origin of these tumors was from displaced embryonic ectodermal epithelial cells which are carried via the nasal pits into the septum [11]. Evans and Cruickshank contradicted the previous 2 theories and claimed that these tumors are entirely epithelial tumors that arise in fully developed gland tissue [12].

Surgical resection by local or wide excision via a lateral rhinotomic approach and with histologically clear margins is the treatment of choice. Batsakis stated that "for practical purposes, all minor salivary gland neoplasms of the sinonasal tract, regardless of their histologic composition, behave in an aggressive manner" and advocated the need for a more-radical resection [4].

Compagno and Wong found a 10% recurrence rate following surgical excision in their patients who had 1 to 41 years of follow-up [6]. Krolls and Boyers suggested the histologic characteristic most frequently associated with recurrent pleomorphic adenomas was the myxoid stromata, which might easily spill into the surgical field, providing a focus for recurrence [13]. Thus, Compagno and Wong attributed the low recurrence rate to the histologic nature of high cellularity and few myxoid stromata. Prognosis of intranasal mixed tumors is better than for those in other ectopic sites, because they show early symptoms leading to an early diagnosis, and because involvement of the surrounding structures occurs at a rather late stage since the tumors have sufficient space to expand before involving the bone [14]. Malignant transformation and distant metastasis are extremely unusual [1,15]. Freeman reported a case of a histologically benign recurrent pleomorphic adenoma of the nasal septum with cervical metastasis. Seeding of tumor cells during the initial excision of the nasal mass 17 years prior to the recurrence was thought to have been the possible cause [1]. The 2 nasal septal neoplasms reported by Cho et al represent the first confirmed examples of a carcinoma ex-pleomorphic adenoma of the nasal mucosa. No distant metastasis occurred in either case [16]. Freeman et al reported the third case of a carcinoma ex-pleomorphic adenoma of the nasal cavity [17]. The pathological diagnosis of the case was a carcinoma ex-pleomorphic adenoma with adenoid cystic and squamous carcinomatous differentiation. After surgical excision and radiotherapy, the patient still perished due to brain, liver, and bone metastases.

In summary, pleomorphic adenomas are rare tumors of the nasal cavity. They have a higher epithelial and lower stromal component compared to their major salivary gland counterparts. Although the recurrence rate is low under adequate excision, cases should be kept under observation due to the recurrence potential.

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一位小孩之非典型的細胞性神經鞘瘤

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摘要

在皮膚的良性腫瘤中，細胞性神經鞘瘤並不常見，它是神經鞘瘤的一種變形。細胞性神經鞘瘤的組織病理形態通常表現輕微的細胞變形與偶發的細胞分裂，而有非典型表現的細胞性神經鞘瘤則非常少見(包括腫瘤較大、深部穿透性、明顯的細胞多形性、高細胞分裂、廣泛的邊緣浸潤性、與血管侵犯等非典型表現)，這些非典型表現可能讓它很難與軟組織肉瘤做區分而誤判。我們報告一位9歲的小女孩在左大腿皮膚表面長了一個在6個月內逐漸長大成1.5 × 1.3 × 1公分的腫瘤，在腫瘤切除後，病理檢查發現是一個細胞性神經鞘瘤但有很明顯的非典型表現：包括深部穿透到皮下脂肪組織、明顯的細胞多形性、高細胞分裂比率(5/10高倍視野)、及廣泛邊緣浸潤。免疫組織化學染色顯示這些細胞的α-smooth muscle actin是陽性，但cytokeratin和S-100是陰性。她術後恢復良好，在一年半的追蹤期間，腫瘤沒有復發。(**慈濟醫學** 2005; **17**:43-45)

關鍵語：細胞性神經鞘瘤，非典型表現，腿部腫瘤

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鼻中隔之多形性腺瘤一病例報告

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摘要

多形性腺瘤為唾液腺常見之良性腫瘤，但少見於唾液腺以外之部位，原發於鼻中隔之多形性腺瘤實為罕見。我們報告一例40歲男性病人，近六個月來有右側鼻塞及間歇性鼻分泌物中帶血絲之表徵，檢查發現右側鼻腔內有一腫塊，電腦斷層掃描顯示此腫塊侷限於鼻腔內，於是安排在內視鏡下切除此腫塊，於手術過程中確認此腫塊原發自鼻中隔。病理切片結果顯示此腫塊為一多形性腺瘤，術後病人追蹤四週，預後良好，無復發或其它併發症。原發於鼻中隔之多形性腺瘤實為罕見，特此提出報告。(**慈濟醫學** 2005; **17**:47-49)

關鍵語：多形性腺瘤，混合瘤，鼻中隔

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因蛛網膜囊腫造成中耳鼓室頂缺損所導致的自發性腦脊髓液耳漏一病例報告

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摘要

自發性腦脊髓液耳漏是臨床少見的情況。成年發作的腦脊髓液耳漏多與腦部手術或外傷所造成的腦組織脫垂有關；因蛛網膜囊腫造成中耳鼓室頂缺損所導致的自發性腦脊髓液耳漏過去在文獻中未曾被報告過。本文報告一因顳下部蛛網膜囊腫侵蝕中耳鼓室頂而造成傳導性聽力喪失、腦脊髓液耳漏以及反覆性腦膜炎發作症狀的病例；手術採中顱窩路徑進入，將蛛網膜囊腫造口並封閉硬腦膜缺損，同時完整的保存了聽小骨。(**慈濟醫學** 2005; **17**:51-54)

關鍵語：蛛網膜囊腫，鼓室頂，自發性腦脊髓液耳漏

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