Pleomorphic adenoma of the nasal septum – A rare entity

ABSTRACT

Pleomorphic adenoma (PA) is the most common benign tumor of major salivary glands, and it rarely arises in minor salivary glands. PA has been reported at various sites where minor salivary glands are found and include nasal cavity, nasopharynx, pharynx, larynx, hypopharynx, and even lacrimal glands. We report one such a case in the nasal cavity in a middle-aged woman. The patient presented with nasal mass and symptoms of obstruction and epistaxis. After clinical and radiological evaluation, the mass was excised endoscopically, and histopathological examination revealed a biphasic benign cellular tumor with scant hyalinized stroma. On immunohistochemistry, the tumor was diffusely positive for CK7 and focally positive for P40 and smooth muscle actin (SMA). Proliferation index was 2%–3%. Finally, the diagnosis of PA was rendered. PA is generally restricted to the cavity due to paucity of space and presents early in the course. Surgical excision with wide margins is the treatment of choice. There are chances of recurrences and malignant transformation of the tumor. To conclude, although rare, one should consider a possibility of PA in the differentials of nasal masses, and accurate diagnosis of this entity is essential to ensure follow-up as recurrence and malignant transformation are reported in PA.

Keywords: Minor salivary gland, mixed tumor, P40, smooth muscle actin

INTRODUCTION

Pleomorphic adenoma (PA) is the most common benign salivary gland tumor. Parotid gland accounts for 60% of these tumors, whereas only 8% are reported in minor salivary glands. They have been reported at locations where minor salivary glands are seen, and these include nasal cavity, nasopharynx, pharynx, larynx, hypopharynx, and lacrimal glands.

We present a rare case of PA of the nasal septum in a middle-aged female. Intranasal PAs are rare, present early due to nasal obstruction, and usually arise from the septum, although minor salivary glands are predominant in the lateral wall of the nose.

CASE REPORT

A 48-year-old female presented with a mass in the left nasal cavity with symptoms of nasal obstruction and recurrent epistaxis. She first noticed the mass a year back. Although nasal obstruction was progressive, she complained of epistaxis 3 months back, and it was recurrent thereafter. There was no history of previous trauma to the nose or face, fever, weight loss, visual defect, or pain. Anterior rhinoscopic examination showed a fleshy nontender mass arising from the anterior cartilaginous septum on the left side, and it measured 1 cm × 0.5 cm × 0.5 cm. The mucosa overlying it was normal, and there was no active bleed. Endoscopic examination confirmed the findings of anterior rhinoscopy and showed a right-sided spur additionally. There was no evidence of rhinosinusitis, and postnasal space was normal. No neck nodes were palpable.

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Radiological examination (noncontrast computed tomography scan of the nose and paranasal sinus [PNS]) showed well-pneumatized PNSs with patent drainage and a soft-tissue mass in the anterior part of the left nasal cavity. A spur was seen inferiorly toward the right side, and the mass arising from the septum anterior to the inferior turbinate was also noted [Figure 1].

After obtaining consent and ascertaining fitness for general anesthesia, a submucous resection with spurectomy was done. A segment of cartilage attached to the mass was removed. Mucosa and the underlying perichondrium for 1 cm around the mass were also excised. Mucosa on the opposite side was preserved.

Histological evaluation of the mass showed a biphasic cellular tumor composed of sheets of epithelial cells punctuated by a few glandular structures and stellate cells merging in hyalinized areas [Figure 2]. Plasmacytoid morphology was evident at places, and hence, the possibility of myoepithelial tumor was considered initially along with PA. Chondroid and myxoid areas were not seen in the tissue submitted. On immunohistochemistry, tumor cells were positive for CK7, Pan-CK (AE1/AE3), P40, and focally expressed S-100 and smooth muscle actin (SMA) [Figures 3-5]. Proliferation index was 2%-3%. There were no areas of necrosis, anaplasia, or carcinomatous transformation.

Postoperative period was uneventful. The patient and the attenders were counseled after the receipt of histopathology and immunohistochemistry reports about the benign nature of the disease. They were explained about the chance of recurrence and were also told about the risk of the tumor to have turned malignant if left without excising. The patient was followed up after a week of surgery when clots were suctioned out and the nasal cavities inspected. Repeated nasal endoscopies during subsequent follow-ups showed no sign of recurrence.

DISCUSSION

Although PAs are the most common tumors of the major
salivary glands, they may present in minor salivary glands also. When they do, they can occur at any site where the minor salivary glands exist. Cases have been reported in the hard palate and tongue in the oral cavity and nasal cavity, PNSs, nasopharynx, oropharynx, hypopharynx, trachea, and even lacrimal glands.

In the upper respiratory tract, the most common locations for these tumors are nasal cavity, maxillary sinuses, and nasopharynx. Although the majority of minor salivary glands occur in the lateral wall of the nose and turbinates, PA usually arises from the septum.

They present in the middle age, with a slight female preponderance. The predominant symptoms are unilateral nasal obstruction and bleeding. Other symptoms include mass in the nose, swelling, mucopurulent rhinorrhea, and epiphora.

Diagnosis usually relies on histological examination of the resected mass. PAs show epithelial tissue mixed with tissues of myoid, chondroid, or mucoid appearance. Nasal PAs have high cellularity with low stromal component and absent capsule. This feature differs from major salivary gland tumors and can resemble malignant epithelial tumors. Immunohistochemistry assessment usually confirms the diagnosis.

In our case, immunohistochemistry was positive for CK7 which is a marker for salivary gland tumors, negative for SMA which rules out adenoid cystic carcinoma and other myoepithelial tumors of the salivary glands. Ki67 was positive in only 2%-3% of cells, which rules out high-grade tumor. Ki67 is an antibody which binds to the antigen expressed in the nucleus of the dividing cell, and the density of staining gives an indication of proliferating cells in growth. It is a useful prognostic marker in adenoid cystic carcinoma, acinic cell carcinoma, carcinoma ex PA, and other salivary duct cancers. P40 is a marker of basal cells which is positive in epithelial-myoepithelial carcinoma, mucoepidermoid carcinoma, myoepithelial carcinoma, clear-cell carcinoma, oncocytoma, and oncocytic carcinoma but negative in acinic cell carcinoma and salivary duct carcinoma. Cytokeratin pan (AE1/AE3) is positive in ductal, acinar, myoepithelial, and basal cells.

Treatment involves surgical excision with wide margins all around to prevent recurrence. Approaches depend on the size of tumor. They include intranasal, transnasal endoscopic, lateral rhinotomy, external rhinoplasty, and midfacial degloving.

In this case, we used transnasal endoscopic approach and removed 1 cm of the mucosa and perichondrium of the septum and curetted the cartilage where the mass was attached.

The recurrence rate for intranasal PAs after full excision with clear margins is less than the rate for major salivary gland mixed tumors. However, regular follow-up with nasal endoscopies is recommended.

Due to the lack of space for the growth of the tumor inside the nose, the patients present early in these cases leading to an early diagnosis. Involvement of adjacent bone is rare due to the space constraint. In a few cases, the tumors can turn malignant carcinoma ex PA.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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