

Nasopharyngeal carcinoma ex-pleomorphic adenoma (noninvasive): A report of a rare case

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Abstract

Pleomorphic adenoma (PA) is the most common type of benign tumors of minor salivary glands. A carcinoma ex-pleomorphic adenoma (CXPA) is a malignant epithelial neoplasm originating from either a primary or recurrent benign PA. The nasopharynx is an extremely uncommon location for this tumor. A 32-year-old male had complaints of nasal blockage. In noncontrast computed tomography, a soft-tissue mass was present in the nasopharynx. Histopathological and immunohistochemical examination of the endoscopically excised mass revealed features of CXPA, noninvasive in nature. Careful histopathological examination is the key to identify this uncommon entity. To the best of our knowledge, <20 cases have been published so far.

Keywords: Carcinoma ex-pleomorphic adenoma, minor salivary gland tumors, nasopharynx, pleomorphic adenoma

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INTRODUCTION

Salivary gland neoplasms are common in major salivary glands but less frequently occur in minor salivary glands.^[1] Salivary gland tumors are rare in the head and neck region, accounting for <5% of all tumors.^[1,2] Pleomorphic adenoma (PA) is the most common type among benign tumors of salivary glands. PA is also known as a benign mixed tumor. PA rarely occurs in the minor salivary glands, though it is the most common minor salivary gland tumor. Minor salivary gland PAs are mostly located in the hard and soft palates, upper lip, floor of the mouth, lacrimal gland, larynx and trachea. PA in the nasopharynx is very rare, with only very few cases reported in English literature.^[1-3] Malignant mixed tumor of the salivary gland is uncommon. Malignant mixed tumor of the salivary gland

comprises three different entities. They are carcinoma arising in a benign mixed tumor (carcinoma ex-pleomorphic adenoma [CXPA]), carcinosarcoma and metastasizing mixed malignant tumor.^[4] A CXPA is a malignant epithelial neoplasm taking origin from a primary or recurrent benign PA.^[2] There are <20 reported cases of nasopharyngeal CXPA in the world literature to the best of our knowledge.^[1] Hereby, we report one such extremely rare case.

CASE REPORT

A 32-year-old nonsmoker, nondiabetic male presented with the complaints of nasal blockage more on the left side nostril. These symptoms were present for the last 1 year and were associated with fullness of the head intermittently. They were aggravated on supine position. There was no history of nasal

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discharge or epistaxis. History of allergy was not elicited. Otoloscopic examination revealed an intact bilateral tympanic membrane. The nasal mucosa was normal, and the throat was clear. Noncontrast computed tomography revealed a soft-tissue mass in the nasopharyngeal region extending up to choana with partial blockage (left > right), reported as likely adenoid. The patient was kept on follow-up. After 11 months, he complained again of aggravating symptoms. Contrast-enhanced computed tomography (CECT) revealed a well-defined lobulated homogeneously enhancing soft-tissue lesion located in the midline extending to the left lateral wall to the choana and causing luminal narrowing measuring 2.2 cm × 1.9 cm × 2.3 cm (AP × TR × CC). Few foci of calcification were noted within the lesion. There was no evidence of parapharyngeal extension, bony erosion or lymphadenopathy [Figure 1a]. Diagnostic nasal endoscopy displayed a smooth pinkish large nasopharyngeal mass completely obstructing the left side choana [Figure 1b].

Intraoperatively, the mass was seen attached to the superior and lateral wall of the nasopharynx. It was excised endoscopically and sent for histopathological evaluation. Macroscopically, a single piece of tan white globular tissue measuring 3.2 × 2.2 × 1.8 cm was received, the outer

surface of which was smooth with glistening white areas in cut surface [Figure 1c]. Microscopic examination revealed pseudostratified ciliated columnar lining epithelium with an ill circumscribed lesion showing features of PA. There was presence of epithelial, myoepithelial and chondromyxoid areas. Squamous and adipocytic metaplasia was noted. Focally, there was evidence of malignancy in the form of myoepithelial carcinoma [Figure 1d and e]. The surgical resected margins were free of tumor. Immunohistochemical studies with CK7, CK5/6 and vimentin showed intense cytoplasmic positivity and focal positivity for p63. S-100 showed diffuse positivity. Smooth muscle actin (SMA) and HER-2 were negative. Ki-67 proliferative index was 5% in the malignant area [Figure 1f and g]. Based on the characteristic histopathological findings, supported by immunohistochemistry, the final diagnosis given was noninvasive CXPA. Postoperative CT, magnetic resonance imaging (MRI) and positron emission tomography did not reveal any residual tumor. He is under follow-up for the last 1 year and is currently doing well.

DISCUSSION

In the event of detection of nasopharyngeal mass in an

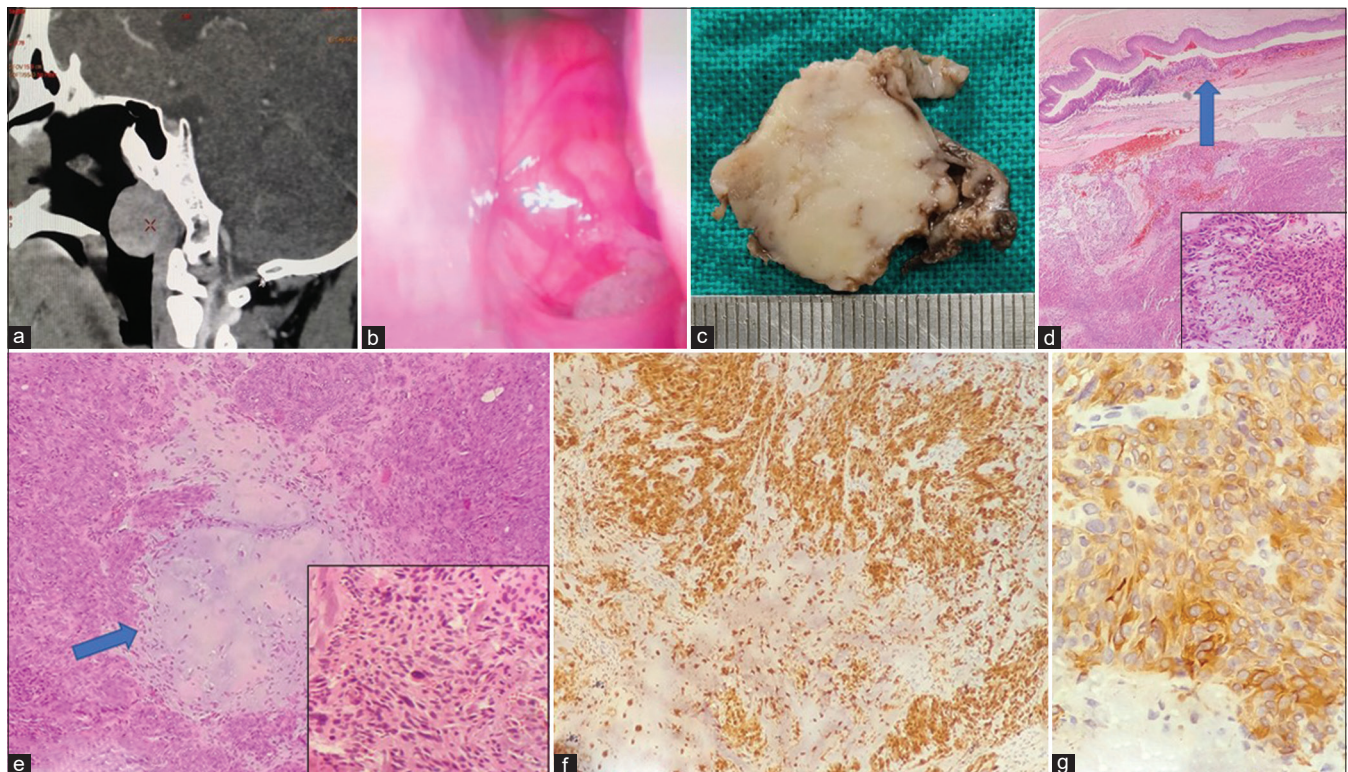


Figure 1: (a) Contrast-enhanced computed tomography showing a well-defined lobulated homogeneously enhancing soft-tissue lesion. (b) Diagnostic nasal endoscopy shows a smooth pinkish large nasopharyngeal mass. (c) Cut surface of the mass with glistening white areas. (d) Pseudostratified ciliated columnar lining epithelium (blue arrow) with the lesion (H&E, ×40) Inset: Pleomorphic adenoma component. (e) Pleomorphic adenoma with chondroid area (blue arrow) (H&E, ×100) Inset: Myoepithelial carcinoma. (f) S-100 positivity in tumor (IHC, ×40). (g) CK5/6 positivity in epithelial component (IHC, ×100)

adult patient, suspicion of nasopharyngeal malignancy arises. Common nasopharyngeal cancers are keratinizing squamous cell carcinoma (SCC), nonkeratinizing SCC and basaloid SCC.^[1,5]

A CXPA is a malignant epithelial neoplasm which takes origin from either a primary or a recurrent benign PA. CXPA is extremely rare in the nasopharyngeal regions, with <20 reported cases in the English literature.^[1,3,6,7]

There is usually a delay in the diagnosis of malignant nasopharyngeal lesions as most of them remain asymptomatic at an early stage. They are often confused with other more common benign lesions.^[1,5] The diagnosis of CXPA in the nasopharyngeal regions is difficult as the symptoms and radiologic findings are usually nonspecific. The presence of osteolysis in CT or MRI may be an indicator for malignancy.^[1]

The important clinical manifestations of CXPA are recent rapid growth and malignant transformation following repeated resection of PA.^[2]

CXPA usually appears in PA in three-fourth of surgical specimens. However, the proportion of malignant components is variable. CXPA is divided into noninvasive, minimally invasive and invasive categories according to the degree of invasion of carcinoma beyond PA. This distinction is important for determining prognosis and appropriate management. Noninvasive and minimally invasive CXPA does not behave like a malignant lesion.^[1,6,7] In our case, the malignant focus was confined within the PA. Hence, we considered it as a noninvasive CXPA.

Various immunohistochemical markers such as cytokeratins, vimentin, S100 protein, SMA and glial fibrillary acidic protein are used to demonstrate the mixed nature of PA.^[8] In addition, overexpression of the p53 protein, HER-2 and proliferation marker Ki-67 (MIB-1) helps to identify malignant areas in PA.^[9,10]

Although any salivary carcinoma can arise from a PA, poorly differentiated or undifferentiated adenocarcinoma, not otherwise specified is the most common type. The adenoid cystic carcinoma subtype of CXPA is more common than the adenocarcinoma subtype in the nasopharynx and nasal regions.^[1,2] In our case, there was presence of myoepithelial carcinoma.

CXPA should be treated as early as possible. For nasopharyngeal CXPA, several surgical approaches are available to achieve extensive local removal. These include endoscopic surgery, external rhinoplasty, lateral rhinotomy and facial degloving.^[2]

Noninvasive and minimally invasive CXPA behave in a benign fashion. Invasive CXPA has a 5-year survival rate of approximately 30%.^[2,6]

CONCLUSION

CXPA in the nasopharynx being an extremely rare entity, awareness and careful histopathological examination is essential for correct diagnosis.

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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