



## Polymorphous Low-Grade Adenocarcinoma in the Nasopharynx: A Case Study and Review of the Literature

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### Abstract

Polymorphous Low-Grade Adenocarcinoma (PLGA) is a neoplasm that frequently occurs in the minor salivary glands located in the mucosa of the soft and hard palates, the buccal mucosa, and the upper lip. It rarely occurs in the nasopharynx, and only six such cases have been reported. Nasopharyngeal PLGA can be treated by wide local excision with no additional treatment. We report a case of PLGA originating in the nasopharynx mucosa. We were able to completely resect the tumor using simple endoscopic surgery rather than an open approach, such as a transpalatal resection. Our patient had no local recurrence or distant metastasis at 24-months follow-up.

**Keywords:** Minor salivary gland; Polymorphous low-grade adenocarcinoma; Otorhinolaryngological surgery

### Introduction

Polymorphous Low-Grade Adenocarcinoma (PLGA) is a minor salivary gland neoplasm that primarily occurs at intraoral sites. An extra oral location of the tumor is rare, with nasal tumors representing less than 1% of PLGAs [1]. The tumor generally involves the palate, but has been described from the base of the tongue, upper lip, buccal mucosa, tonsils, and retromolar pad [2]. PLGA is a rare pathology that often occurs in the fifth and sixth decades of life with a female/male ratio of 2:1 [3,4].

To date, six cases of PLGA presenting as a nasopharyngeal mass have been reported [5-10]. Nasopharyngeal PLGA grows slowly, painlessly, and has only a few symptoms, including unilateral ear fullness and nasal obstruction. Here, we describe a case of nasopharyngeal PLGA including clinical characteristics, pathology, treatment, and prognosis with a review of the literature.

### Case Presentation

A 51-year-old male patient presented with right-side ear fullness with no nasal symptoms. No neck mass was noted. He did not smoke or drink alcohol and had no past history of acute disease with the exception of cataract surgery. Nasal endoscopy revealed an irregularly shaped mass on the right salpingopalatine fold (Figure 1). Paranasal sinus Computed Tomography (CT) showed a 20 mm × 23 mm × 30 mm polypoid mass in the lateral wall of the right nasopharynx with no surrounding bony destruction or intraorbital or skull base invasion (Figure 2). Right middle ear effusion was seen on otoscopy. Punch biopsy of the nasopharyngeal mass was performed in the outpatient department. Pathological biopsy revealed a benign salivary gland tumor consistent with pleomorphic adenoma (Figure 3).

The tumor was resected 2 months later using the endoscopic approach according to the preoperative biopsy [11]. Surgery was performed under general anesthesia with the patient in the supine position. We used the transnasal endoscopic approach with 0° and 30° 4 mm nasal endoscopes (Karl-Storz, Tuttlingen, Germany). The posterior portion of the right inferior turbinate was excised to create a working space for the endoscope and to expose the tumor. Then a circular incision of the mucosa was made around the mass using a sickle knife, and further dissection was performed using a freer elevator. The right pterygoid bone was exposed after the tumor was excised with a 2 mm resection margin. Frozen biopsy specimens revealed a carcinoma of undetermined type. Frozen sections prepared from the intraoperative margin tissue revealed a negative resection margin. We completely removed the nasopharyngeal tumor according to the preoperative CT scan, although we were unable to use an intraoperative navigator system to guide placement of instruments in the surgical field. The gross surgical specimen showed a pale brown to tan soft tissue mass measuring 2.0 cm × 1.3 cm × 1.0 cm. Finally, Tacho Comb was applied to the wound site, followed by Meroce

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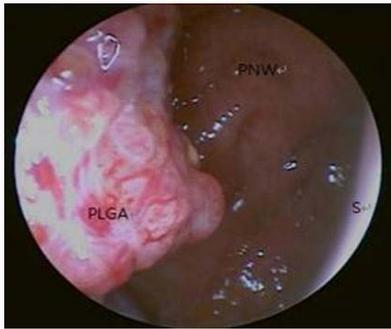
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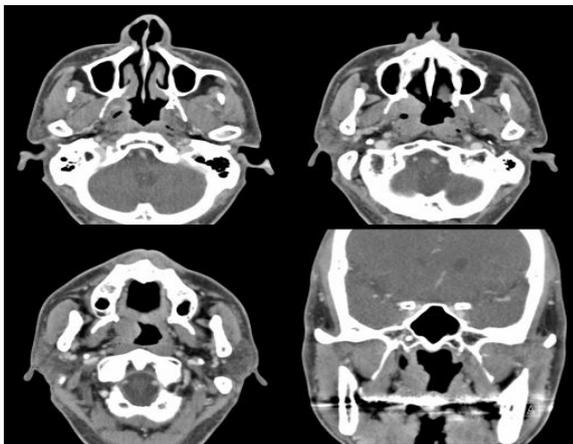
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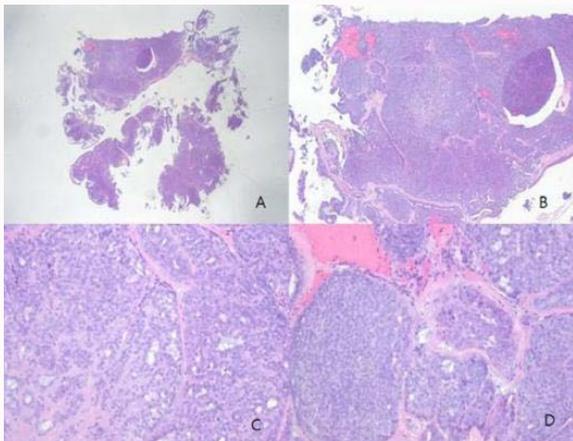
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**Figure 1:** Endoscopic image of a Polymorphous Low-Grade Adenocarcinoma (PLGA) in the right salpingopalatine fold. PNW: Posterior Nasopharyngeal Wall; S: Septum

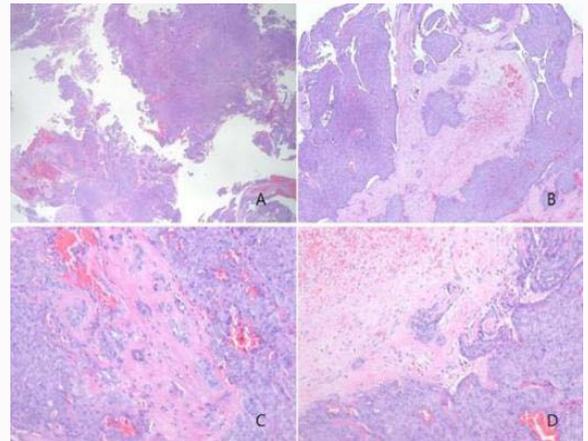


**Figure 2:** Paranasal sinus computed tomography scan showing a right nasopharyngeal mass. The surrounding bony structure was not affected.

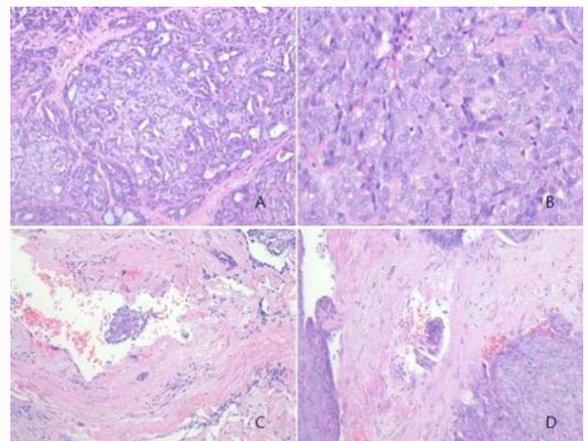


**Figure 3:** Histological findings obtained by punch biopsy performed in the outpatient department. Monotonous epithelial cells were arranged in well-marginated nests and along the tumor nests. Small hyperchromatic nuclei and spindle-shaped myoepithelial cells were present between and in the tumor cell nests, and the myxoid stromal matrix is seen focally. These findings are compatible with a pleomorphic adenoma. Hematoxylin & eosin stain: A (x10), B (x40), C (x100), and D (x200).

packing from the right posterior nasal cavity to the nasopharyngeal cavity. The Merocel was removed 2 days after surgery, and the patient left the hospital the following day with no significant complications, such as intranasal bleeding or postnasal drip. On day 3 after surgery,



**Figure 4:** Histological findings. Buffered formalin-fixed and paraffin-embedded tissue sections were prepared for examination by light microscopy and immunohistochemistry. **A:** Hypercellular and compact tumor cells were arranged in irregularly shaped nests. **B:** Multiple tumor cell clusters were scattered in the pinkish stromal matrix between tumor nests, suggestive of an invasive front. **C,D:** Multifocal hemorrhagic foci were visible in the tumor cell nests.



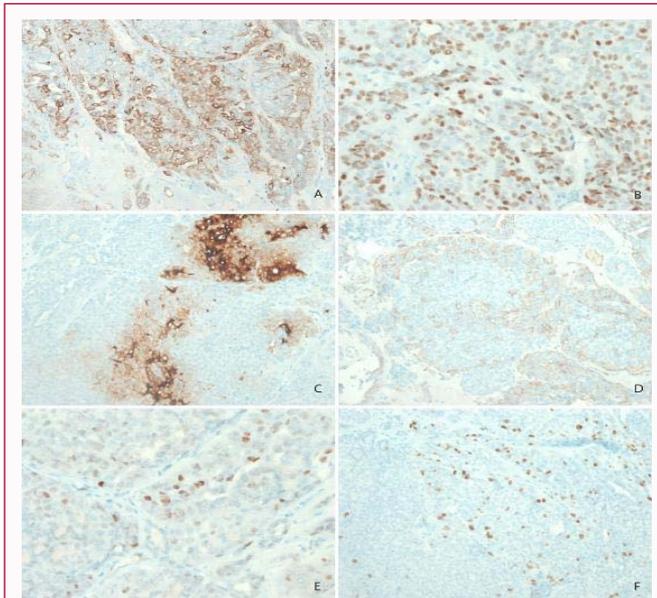
**Figure 5:** Hematoxylin & eosin staining results at x400 magnification. Tumor cells arranged in papillary and cribriform patterns were observed at higher magnification. **A:** Mitotic figures were rare but present. **B:** Small- to medium-sized tumor cells with mildly hyperchromatic and oval nuclei were observed. **C,D:** Two foci of lymphovascular invasion were seen.

the final diagnosis showed PLGA and the resection margin was free of tumor (Figure 4-6). A Positron Emission Tomography (PET)-CT scan performed the same day confirmed no remnant nasopharyngeal mass or neck metastasis; however, an exophytic mass seen in the left kidney was resected and diagnosed as clear-cell renal cell carcinoma. Follow-up PET-CT 7 months after nasal endoscopic surgery revealed no definite abnormal lesion suggestive of recurrence.

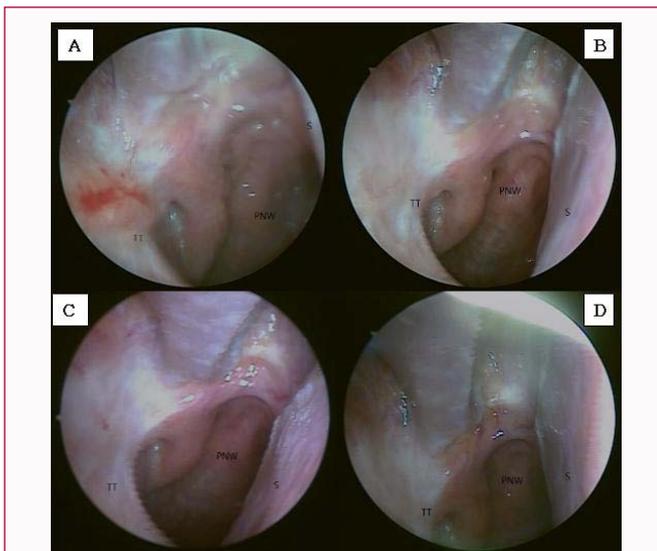
At the 24-month follow-up, the patient received no additional treatment and showed no signs of local recurrence or distant metastasis (Figure 7).

**Discussion**

PLGA has a low tendency for aggressive behavior; thus, the recommended treatment is surgical excision, and the results are generally excellent. No cases of lymphatic spread have been described; therefore, neck dissection is not necessary unless there is clinical evidence suggestive of lymph node involvement. If the tumor



**Figure 6:** Immunohistochemical findings. A: The tumor cells were positive for CK and B: p63; C: focal positive for EMA, D: SMA, and E: p53; and F: 20% positive for Ki-67. CK: Cytokeratins; EMA: Epithelial Membrane Antigen; SMA: Smooth Muscle Antibody.



**Figure 7:** Endoscopic images showing no local recurrence at 3 months. A: 8 months, B: 12 months, C: 24 months, D: after nasal endoscopic surgery. TT: Torus Tubaris; PNW; Posterior Nasopharyngeal Wall; S: Septum.

has not been completely resected, adjuvant radiotherapy should be performed to prevent tumor recurrence [6]. PLGA rarely occurs in the nasopharynx, and there is no definite treatment of choice. En-bloc mass excision using the transpalatal or nasal endoscopic approaches are options; however, the transpalatal approach may cause several complications, including pain, bleeding, and oronasal fistulas. Exclusive endoscopic resection can avoid potential complications and shorten the hospital stay.

To date, six cases of nasopharynx PLGAs have been reported. Lengyel et al. [7] reported a case with skull base invasion and intracranial spread confirmed by CT. Radiotherapy was performed. Follow-up CT scans revealed a partial response to radiotherapy, and the patient was still living after 51 months post-radiotherapy. Wenig

et al. [5] reported a case of a patient with PLGA who underwent transpalatal tumor resection and recovered uneventfully for 9 months after operation. Furthermore, four previous studies found that patients with PLGA who underwent exclusive transnasal endoscopic tumor resection fully recovered with no additional treatment [6,8-10]. Our case is the fifth report of a patient with nasopharyngeal PLGA who underwent nasal endoscopic tumor resection with no additional therapy. We found no local recurrence or evidence of metastasis at the 24-month follow-up. Our findings, together with those of previous studies, suggest that the simple endoscopic approach is a feasible treatment for nasopharyngeal PLGA. However, during endoscopic surgery, the surgeon must be able to resect the mass with sufficient margins to ensure they are free of tumor. In cases where the tumor extends beyond the nasopharynx or endoscopic view, we suggest that open surgery such as the transpalatal approach should be used rather than the endoscopic approach. In addition, open surgery should also be performed in cases where local recurrence is detected after endoscopic surgery.

## Conclusion

In cases of nasopharyngeal PLGA which is limited to nasopharynx mucosa, if surgeon is able to resect it with sufficient margins, the endoscopic approach will be feasible choice. This procedure is curative, cuts down the length of operation, decreases wound-related complications and shortens hospital stay.

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