# A Rare Case of Low-grade Nasopharyngeal Papillary Adenocarcinoma in a Young Adult Woman: A Case Report from a Tertiary Hospital in Northwestern Nigeria

#### Abstract

Low-grade nasopharyngeal papillary adenocarcinoma (NPAC) is a rare nasopharyngeal malignancy that runs an indolent course. It presents with a variety of symptoms including but not limited to nasal obstruction, runny nose, postnasal drip, and nasal bleeding. The main stay of treatment is surgery, but other adjuvant treatments including radiotherapy and photodynamic therapy have been used with varying degrees of successes. Due to its low grade nature and absence of distant metastases, its prognosis is excellent. Here, we report the case of a 22-year-old young woman with low-grade NPAC who was treated in our hospital by simple surgical excision.

**Keywords:** Low-grade nasopharyngeal papillary adenocarcinoma, nasopharyngeal carcinoma, surgical excision, young lady

## Introduction

The World Health Organization (WHO) classifies malignant epithelial tumors of nasopharynx into nasopharyngeal carcinoma (NPC), nasopharyngeal papillary adenocarcinoma (NPAC), and salivary gland-type carcinomas (NPCs).<sup>[1]</sup> NPAC is said to originate from the mucosal surface lining epithelium of the nasopharynx.<sup>[2,3]</sup> Histologically, NPAC is very similar to thyroid papillary carcinoma, and both carcinomas are positive for TTF-1 on immunohistochemical staining.<sup>[4]</sup> While thyroid papillary carcinoma is also positive for thyroglobulin, NPAC is not<sup>[3,5]</sup>; hence it is used in differentiating the two. The biologic potential of NPAC was also that of a low-grade malignant tumour,<sup>[6]</sup> where perineural or vascular lymphatic invasion was not observed, and metastasis was not reported; hence, it is considered to have very good prognosis.<sup>[7]</sup>

Though controversy exists regarding the optimal treatment due to the rarity of this tumour,<sup>[8]</sup> complete surgical excision, with or without the aid of endoscope, has been considered the treatment of choice for most cases of NPAC.<sup>[1,4,6,8-11]</sup>

NPAC is a rare tumour and only few cases were reported in the literature. Most of the cases

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reported were from Southeast Asia where NPC is more prevalent.<sup>[4,5,8,10,12-15]</sup>

In Nigeria, NPAC was reported in a 20-yearold male student from the Southeast<sup>[16]</sup> as well as in a middle-aged woman from the Northwestern part of the country.<sup>[17]</sup>

To the best of our knowledge, this is the second case to be reported from the Northwestern part of the country. We herein report a case of primary low-grade NPAC in a 22-year-old woman who was treated in our hospital by simple surgical excision.

## **Case Report**

A 22-year-old young woman presented to our clinic with a 1-year history of progressive nasal blockage, nasal discharge, and snoring. She had no associated anosmia, epistaxis, facial pain, or headaches. There were no throat, otologic, or neuro-opthalmic symptoms. She had no neck swelling. Review of other systems was unremarkable. She had normal findings on general physical examination. Diagnostic nasoendoscopic examination revealed mucoid discharge in the nasal cavities with a smooth, polypoidal nasopharyngeal mass extending to and blocking the posterior choanae [Figure 1]. There was no contact bleeding. She had computerized tomographic (CT) scan of the nose and paranasal sinuses done, which

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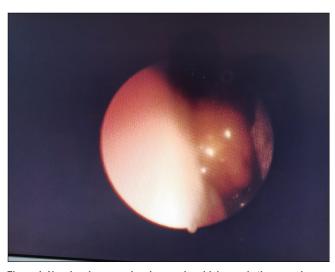


Figure 1: Nasal endoscopy showing a polypoidal mass in the nasopharynx extending into the nasal cavity



Figure 2: Sagittal view of CT scan showing a heterogeneously enhancing mass in the nasopharynx

showed an irregularly shaped heterogeneously enhancing mass lesion with finger-like projection in the nasopharynx extending into the posterior nares, measuring about 21.3 mm  $\times$  32.1 mm in its widest dimension [Figures 2 and 3]. There was no evidence of bony destruction. Full blood count with differentials, erythrocyte sedimentation rate, serum electrolyte urea, and creatinine were all within normal limits. She had an endoscopic biopsy taken from the nasopharyngeal mass which revealed features of low-grade papillary adenocarcinoma. She was counselled and prepared for excisional biopsy of the tumour by oropharyngeal route under direct vision. Sample of the nasopharyngeal mass [Figure 4] was sent for histology. The histology result confirmed the diagnosis of low-grade nasopharyngeal papillary carcinoma [Figures 5a and b]. She was reviewed by a clinical oncologist who advised close monitoring, and for 3 months after the surgery, she had no residual tumour in the nasopharynx or evidence of distant

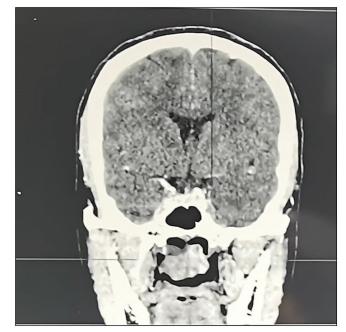


Figure 3: Coronal view of CT scan showing a heterogeneously enhancing mass in the nasopharynx



Figure 4: Sample of nasopharyngeal tissue

metastasis. She is currently on monthly follow-up in the Otorhinolaryngology Outpatient Department.

## Discussion

NPAC is a rare, low-grade, malignant tumour, constituting <1% of all nasopharyngeal malignancies.<sup>[6]</sup> Although NPAC can occur at any age irrespective of the gender,<sup>[12]</sup> patients with this tumour tend to be young (mean age: fourth decade) at presentation.<sup>[3,5,10,11,14]</sup>

It expresses a variety of symptoms including nasal obstruction, runny nose, postnasal drip, nasal bleeding, otitis media, and hearing loss.<sup>[2,3,5,10,16]</sup> Though our patient had only nasal obstruction and nasal discharge at presentation, the features mentioned in the literature are typical of most nasopharyngeal tumours.

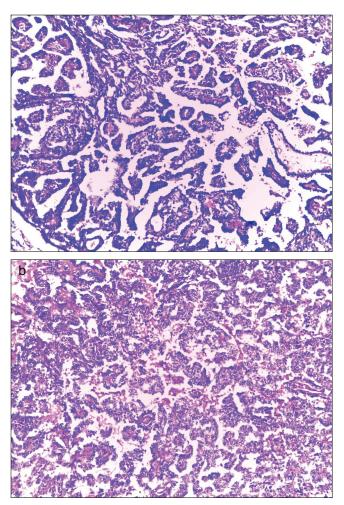


Figure 5: (a and b) Histopathological finding of the nasopharyngeal mass confirming nasopharyngeal papillary adenocarcinoma

Grossly, NPAC may appear as an exophytic, nodular, or polypoid mass, commonly arising from the roof, lateral, and posterior walls of the nasopharynx.<sup>[1,3,6,10,15,18]</sup> Sometimes, the tumour appears gritty if psammoma bodies are present.<sup>[3]</sup> In our index case, the tumour appeared to be smooth, polypoidal mass arising from the posterior wall of the nasopharynx extending to and blocking the posterior choanae, hence presenting with features of nasal obstruction. We performed an endoscopic transnasal incisional biopsy from the mass before embarking on definitive surgery. This is in keeping with findings from another study in which a biopsy was taken before a definitive treatment was performed.<sup>[10]</sup>

Histologically, NPAC typically exhibits papillary architecture with fibrovascular cores, overlapping nuclei with clear chromatin, psammoma bodies, and prominent nucleoli attached to the nuclear membrane.<sup>[5,10,11,18]</sup> These features are in keeping with our case, which revealed an infiltrating tumour growing in papillary fronds and glandular pattern. The papillae are lined by columnar epithelial cells with hyperchromatic oval nuclei and central fibrovascular cores exhibiting oedema and infiltrates of plasma cells.

Complete surgical excision has been considered the treatment of choice for most cases of NPAC.<sup>[1,4,6,8-11]</sup> In their study, Takakura *et al.*<sup>[10]</sup> found out that among the 28 case reports of NPAC they reviewed, all cases were treated with complete resection and without adjuvant treatment. Endoscopic surgery was performed in 16 cases (57.1%). Four cases had secondary resection due to an insufficient surgical margin after the first operation. No patients received adjuvant therapy after complete resection. None of them had distant metastasis or recurrence over the follow-up period of at least 15 years (mean follow-up period was 31.0 months).<sup>[10]</sup>

Low-grade NPACs are generally shown to have low sensitivity to conventional radiotherapy or chemoradiation, and it became a challenge to deal with partially removed tumours.<sup>[8]</sup> Despite that, postoperative radiotherapy has been recommended as an adjuvant treatment, especially after an incomplete surgical resection to prevent tumour recurrence.<sup>[19]</sup>

In a recent report from the Middle East, Mohammed *et al.*<sup>[20]</sup> described the case of a young man diagnosed with low-grade NPAC with positive cervical lymph node who was treated with endoscopic resection of the lesion and concomitant left neck dissection followed by planned chemoradiation.

Other adjuvant therapies for the treatment of NPAC including photodynamic therapy combined with topical 5-aminolevulinic acid have been suggested as an effective modality of the postoperative adjuvant therapy to successfully eradicate the residual disease, without compromising the quality of life of the patient.<sup>[8,21,22]</sup>

Due to the excellent prognosis and rarity of recurrence of NPAC following an appropriate surgical management, we offered our patient surgical excision of the nasopharyngeal mass via the transoral route with no any adjuvant treatment. She had no residual tumour postoperatively and has remained symptoms-free at 3-month follow-up. She is currently on follow-up and has no features of recurrence or any evidence of distant metastasis.

## Conclusion

Low-grade NPAC is an extremely rare tumour. It runs an indolent course and has an excellent prognosis. Although there is no standard protocol for its management, surgical excision is considered to be the treatment of choice. While the role of radiotherapy and chemotherapy has been mentioned in several studies, adjuvant radiotherapy is suggested for patients with incompletely resected tumours.

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### **Conflicts of interest**

There are no conflicts of interest.

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