

# Successful treatment outcome of nasopharyngeal papillary adenocarcinoma following surgery: a case report

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Nasopharyngeal carcinoma (NPC) is a common head and neck malignancy, endemic in East Asia and South East Asia. The rate of incidence varied in different countries based on the relative exposure of the population to different risk factors. The majority of NPC cases are squamous cell carcinoma subtypes. Primary nasopharyngeal papillary adenocarcinoma (NPAC) is an extremely rare subtype of NPC with characteristic adenocarcinoma cell structure. We present a case of NPAC treated by an endoscopic transnasal nasopharyngectomy with a successful outcome.

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Nasopharyngeal carcinoma (NPC) is the most common malignancy in the nasopharynx.<sup>1</sup> The majority of NPC cases are non-keratinizing and keratinizing squamous cell carcinoma subtypes. Primary nasopharyngeal papillary adenocarcinoma (NPAC) is extremely rare, constituting only 0.38–0.48% of all malignant nasopharyngeal neoplasms, albeit a few cases have been reported.<sup>2–4</sup> We report the successful outcome of a case of NPAC treated by endoscopic nasopharyngectomy.

### CASE REPORT

A 40-year-old man presented with a complaint of epistaxis for several months. Endoscopic examination revealed a smooth surface, midline, pedunculated mass from the superior aspect of nasopharynx. A histopathologic examination of the nasopharyngeal mass demonstrated a NPAC. Unfortunately patient defaulted follow up due to financial constraints. After 3 years, he came back with left nasal blockage and blood-stained mucus exacerbated by sneezing. There was no tinnitus, otitis media or aural fullness. A repeat endoscopic examination showed a pedunculated mass from the roof of nasopharynx in continuity with the posterior septal margin and adenoid tissue. The mass was confined to the midline with bilateral fossa of Rosenmuller intact. There were no cervical lymph nodes or anterior neck mass palpable and other physical examination were unremarkable. Computed tomography (CT) of base of skull and neck reported a small lobulated soft tissue lesion confined to the nasopharynx, with no surrounding bony erosion nor lateral pharyngeal recess and parapharyngeal space involvement (Figure 1).

An endoscopic transnasal nasopharyngectomy with posterior septectomy was done. Intraoperatively a

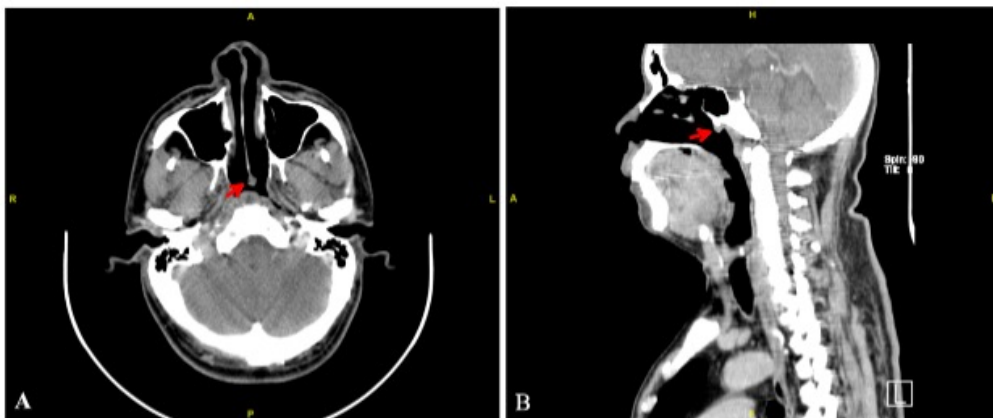
lobulated mass measuring 0.5cm x 0.3cm was resected with wide margin using monopolar diathermy. The mass was successfully dissected off from the nasopharyngeal vault with clear margin and no evidence of bony invasion. No further adjuvant therapy was necessary. At one-year follow-up following surgery, the patient remained well with no evidence of tumor recurrence.

### DISCUSSION

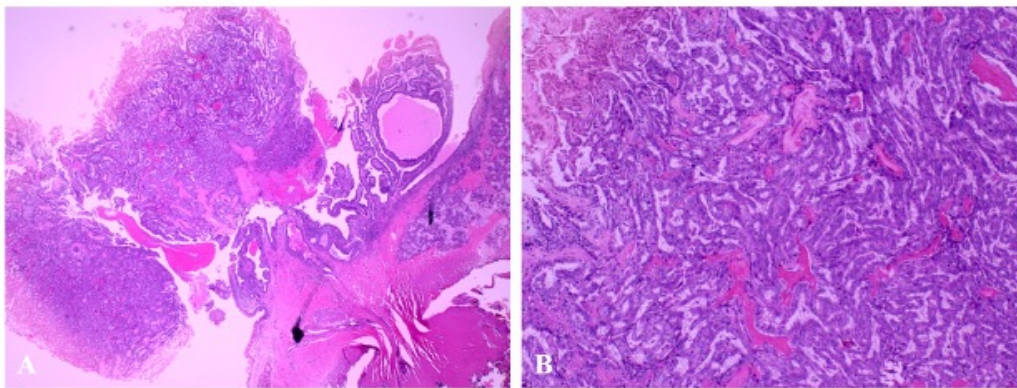
Primary nasopharyngeal malignant tumours can be divided into a few subtypes such as nasopharyngeal carcinoma (NPC), salivary gland tumours, soft tissue tumours, haematolymphoid tumours, notochord tumours, and a separate entity of nasopharyngeal papillary adenocarcinomas (NPAC).<sup>5</sup> The NPAC entity must not be confused with the closely-named entities of sinonasal adenocarcinomas, be it the intestinal type or non-intestinal type.

NPACs of the mucosal surface origin are low grade malignancies with a papillary configuration.<sup>2</sup> They are a rare type of epithelial tumor but may uncommonly present at the nasopharynx. Unlike its other malignant sinonasal tumour, NPAC is not associated with wood exposure or other known factors such as EBV infection. Histologically this type of papillary configuration is also seen in metastatic papillary thyroid carcinoma, hence the literature term of 'thyroid-like nasopharyngeal papillary adenocarcinoma' is not uncommonly-encountered. Thyroid primary was excluded in this case in the light of negative stains of thyroglobulin and normal thyroid gland on CT scan. Despite the three years lapse, owing to the slow growing low-grade tumor, we were able to remove the tumor with clear margins.

NPAC has no known etiological factors and has been reported to affect patients ranging from age 9 – 64 years with no sex predominance.<sup>5</sup> The most common



**Figure 2** (A) Polypoidal tumor tissue formed by the compact and complex papillary configurations with hyalinized fibrovascular core. (B) These papillae are lined by columnar and cuboidal cells, that display round to oval nuclei, fine chromatin pattern and small nucleoli. The intervening stroma shows pinkish collagenous appearance



**Figure 2** (A) Polypoidal tumor tissue formed by the compact and complex papillary configurations with hyalinized fibrovascular core. (B) These papillae are lined by columnar and cuboidal cells, that display round to oval nuclei, fine chromatin pattern and small nucleoli. The intervening stroma shows pinkish collagenous appearance

presentations are nasal blockage and epistaxis. Other reported presenting symptoms include rhinorrhea, aural fullness, otitis media, blood stained sputum and reduced hearing.<sup>2,5</sup> NPAC is a remarkably well-behaved tumor with low percentage of local recurrence and no cervical lymphatic spread been described.<sup>6</sup> This is evidenced in our patient where there was no lymphatic involvement despite the three years delay in treatment. Endoscopically there was a smooth surface, midline, pedunculated mass arising from the roof of nasopharynx in continuity with the posterior septal margin. The differential diagnosis of a tumor in this location is the oft encountered NPC, keratinizing or non-keratinizing squamous cell carcinoma, which usually arises in the lateral nasopharynx as opposed to the midline and virtually never pedunculated.

In the case of NPAC, it can be challenging for pathologist to arrive at a diagnosis due to the similarity of the histological features with other tumors arising from mucosal surface, salivary gland or thyroid gland. Histological assessment of our specimen showed the tumor consists of compact and complex papillary configurations with hyalinized fibrovascular cores. The papillae are lined by cuboidal to columnar cells that have round to oval nuclei with generally fine chromatin pattern and small nucleoli (Figure 2). Some interpapillary areas form gland like structures that have bubbly appearance and contain periodic acid-schiff-diastase positive material. Varying amount of spindle cells were seen within the stroma, which partly merge imperceptibly with the epithelial component. Immunohistochemistry disclosed positivity for pan-cytokeratin, vimentin and thyroid transcription factor-1 (TTF-1), and negativity for thyroglobulin, glial fibrillary acidic protein (GFAP) and S-100 protein. In the event of ambiguous clinical presentations, given the similarities between NPAC and papillary thyroid carcinoma (PTC), a molecular

genetic study can be performed to determine the mutational status of the BRAF-gene.<sup>7</sup>

Due to the rarity NPAC cases, the assessment and treatment options must be highlighted. A review by Xu et al<sup>8</sup> suggested that cranial nerve invasion, skull base erosion and the presence of positive cervical nodes should be considered on tumor staging. It is advocated that surgery with clear resection margins is adequate especially in early-stage tumor. An adjuvant radiotherapy (RT) is reserved for patients who are at high risk of developing recurrence.<sup>8</sup> The use of a more precise radiation techniques can help improve local control and spare radiation-induced damage to the surrounding structures. The low grade nature of NPAC has a good prognosis and allows less aggressive treatment measures to avoid unnecessary morbidities.

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## CONCLUSION

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NPAC is a rare subtype nasopharyngeal malignancy with adenocarcinoma cell structure in papillary configuration that differs from NPC of squamous cell type in its propensity for a slow growth, low incidence of neck nodes metastasis and good prognosis if identified early. Therefore the use of a proper staging system and further deliberation of treatment options between surgical and oncological intervention will give better insight to help establish an optimal treatment strategy in the future.

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