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Case Report

Nasopharyngeal Papillary Adenocarcinoma: Two Cases and Review of the Literature

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ABSTRACT

Nasopharyngeal papillary adenocarcinoma (NPAC) is a rare tumor that is usually detected in the nasopharynx. Its etiology has not been clearly elucidated. It is thought to be of surface epithelial origin. Histologically similar to thyriod papillary carcinoma (TPC), the tumor shows a unique expression pattern immunohistochemically and can be differentiated from TPC. Correct diagnosis of this tumor, in which local recurrence and metastasis are not reported with complete surgical treatment, is important for the patient to not receive more than necessary treatment and to maintain the comfort of life. In this article, the clinical, pathological, immunohistochemical and molecular findings of two NPAC cases were shared and attention was drawn to the prognostic feature of the tumor. Although it is a rare tumor, it is important to share two cases seen in our center and to keep in mind the existence of this tumor.

INTRODUCTION

Nasopharyngeal papillary adenocarcinoma (NPAC) was first described by Wenig et al.[1] and is a rare type of primary nasopharyngeal adenocarcinoma. It has been defined that NPAC, which has distinctive histological features, shows TTF-1 expression immunohistochemically.[1–10] No difference was found in the incidence between genders.[4,9] Although it is frequently seen on the posterior and lateral nasopharynx wall and roof of the nasopharynx, the nasal septum is also seen.[2–10] Patients usually present with nasal fullness, congestion, or nosebleeds.[5] NPAC has a good prognosis and local excision is considered sufficient in its treatment. No recurrence or metastasis has been reported in the literature. We reported the two young patients with low-grade NPAC and reviewed the literature to increase the awareness of this tumor and prevent it from receiving aggressive surgical and medical treatment.

CASE REPORT

Case 1– A 29-year-old female patient applied to the ear-nose-throat clinic with the complaint of sore throat. Imaging with diffusion magnetic resonance revealed a 5 mm diameter nodular thickening in the posterior of the nasal septum (Fig. 1a). In the examination, a well-circumscribed mass was found in the posterior nasal septum and on the free edge (Fig. 1b). Moreover, punch biopsy was performed. The biopsy material was 0.8×0.5×0.5 cm in size and had an off-white polypoid structure. In the microscopic examination; diffuse papillary structures were surrounded by cells with prominent nucleoli, with rounded, irregular membranes, nuclear nicks, clear-vesicular nuclei

with chromatin (Fig. 1g). Cells lining prominent papillary surrounding and few glandular structures with hyalinized fibrovascular core; it had large eosinophilic cytoplasm, single row, mostly cuboidal, columnar, and occasionally hobnail character. Mitosis, cellular atypia, and necrosis were not observed. Immunohistochemically (Ventana Medical System Inc., USA), TTF-1, and CK7 were positive, and thyroglobulin, CK20, CDX2, S100, p40, and p63 were negative and Ki-67 index was <1%. It was diagnosed as NPAC (Fig. 1h and i). Afterward, the mass was completely resected. Macroscopically, the white and papillary lesion was 0.5 cm in diameter and was diagnosed as having no tumor in the surgical margins (Fig. 1c and d). The case has been disease free for 14 months.

Case 2– A 22-year-old male patient presented with the complaint of epistaxis. On examination, a papillamatous lesion was detected on the posterior free edge of the septum and an incisional biopsy was performed. The material was 1 cm in diameter and had a white polypoid character. In microscopic examination, cuboidal or columnar cells with extensive cytoplasm, vesicular nuclei, and prominent nucleoli were arranged as single layered around fibrovascular cores to form papillary structures (Fig. 1e and f). In addition, the same cells formed diffuse glandular structures (Fig. 1f). Mitosis, cellular atypia, and necrosis were not observed. Cells were positive with TTF-1, CK7 by immunohistochemical staining (Ventana Medical System Inc., USA). Ki-67 proliferative index was <1% and focal positivity of S100, p40, and p63 was observed, and thyroglobulin was negative. The case was diagnosed as NPAC. There was a tumor of the same character, 2 cm in diameter, in the excision material, and the surgical margins were free and the case has been disase free for 12 months.

DISCUSSION

NPAC is defined as low-grade adenocarcinoma characterized by papillary and glandular configurations and exophytic growth, originating from the nasopharynx. Although the tumor classically has a papillary microscopic structure, it may be glandular and spindle in character.[3,4,9,10] Its etiology has not been clearly elucidated.[2,4] Although the tumor is continuous with the surface epithelium, the absence of a precursor lesion and its absence in our cases supports that the tumor originates from the surface epithelium.[3,5] No difference was found in the incidence between genders.[1,3–5] Patients usually present with nasal fullness, congestion, or nosebleeds.[5] NPAC has a good prognosis and local excision is considered sufficient in its treatment.[3–5] Although it is frequently seen on the posterior and lateral nasopharynx wall and roof of the nasopharynx, the nasal septum is also seen,[1,3,5,9,10] and both of our cases were located in the posterior of the nasal septum.

In its differential diagnosis, there are tumors with worse prognosis, primarily metastatic thyriod papillary carcinoma, intestinal adenocarcinoma, and polymorphous adenocarcinoma. The tumor shows the extensive expression of TTF-1, CK (AE1-3), CK19, vimentin and focal expression of S100.[1,3–5] It is clearly differentiated from NPAC and other salivary gland-type tumors by the expression of TTF-1. Although the tumor is histomorphologically very similar to thyriod papillary carcinoma (TPC) and expresses TTF-1, it can be clearly differentiated from TPC immunohistochemically by being thyroglobulin and Napsin-A negative.[1,3,4,6] Our cases showed strong TTF-1 and CK7 expression, and in addition, Case 2 also showed focal S100, p63, and p40 expression. Although thyroglobulin is almost

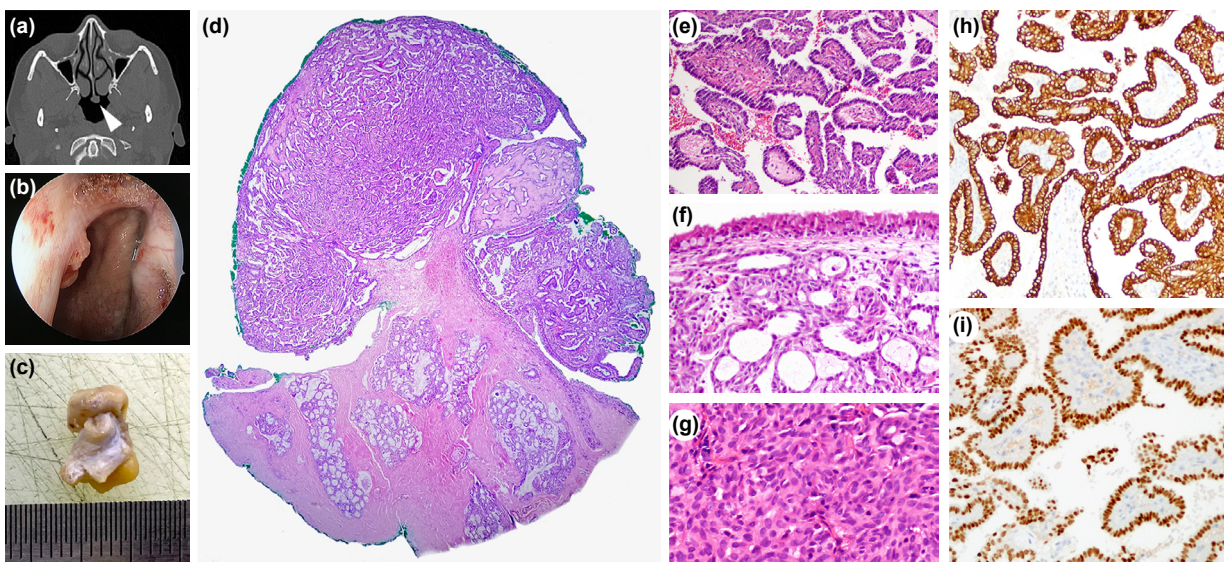

Figure 1. (a) Magnetic resonance imaging image of the mass posterior to the nasal septum (white arrowhead). **(b)** The appearance of the mass in the posterior nasal septum with rigid endoscopy. **(c)** Excision material containing papillary tumor of Case 1. **(d)** Panoramic histological image of the papillary structure in the excision material (H&E, original magnification ×40, software-assisted panoramic view). **(e)** Papillary structures with a row of columnar cells surrounding the fibrovascular cores (H&E, ×100). **(f)** Tumor area with prominent glandular structures (H&E, ×200). **(g)** Tumor area consisting of spindle cells (H&E, ×200). **(h)** Diffuse staining with CK7 and **(i)** TTF-1 in tumor cells (×200).

always reported as negative, focal positivity has been reported in the literature; therefore, attention should be paid to NPAC in cases with focal positivity.[8] In addition to the positivity of TTF-1, the negativity of CK20 and CDX2 distinguishes it from intestinal-type adenocarcinoma.[6,7]

The molecular genetic studies showed no mutation in the BRAF and KIT gene, RT-PCR and FISH assays were negative for SYT-SSX1/2 fusion transcripts and SYT (18q11.2) gene rearrangement.[1,3] Similar to the examples in the literature.[1,6] No results regarding the relationship of the tumor with EBV and HPV have been reported.[5,6] In situ hybridization of EBV and HPV and FISH for ALK and ROS1 was negative in both of our cases. The lack of a genetically distinct signature does not seem promising for different modalities for treatment, and the primary treatment of the tumor remains primarily surgical. In accordance with the fact that no recurrence or metastasis has been reported in the literature, both of our cases have been recurrence and metastasis-free for 12–14 months.[1–10]

CONCLUSION

NPAC is a rare and low-grade tumor located in the nasopharynx. The exclusive morphological and immunohistochemical character of NPAC is important to correct diagnosis and prevent the patient from receiving more aggressive treatment modalities.

Peer-review

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Authorship Contributions

Concept: G.K.A., M.Ç., G.A.Y., Z.E.Ç., B.D., K.B.; Design: G.K.A., M.Ç., G.A.Y., Z.E.Ç., B.D., K.B.; Supervision: G.K.A., M.Ç., G.A.Y., Z.E.Ç., B.D., K.B.; Fundings: G.K.A., M.Ç.; Materials: Z.E.Ç., B.D., K.B.; Data: K.B., M.Ç., Z.E.Ç.; Analysis: G.A.Y., Z.E.Ç., K.B.; Literature search: G.K.A., G.A.Y., K.B.; Writing: G.K.A., K.B., B.D.; Critical revision: G.K.A., K.B., B.D.

Conflict of Interest

None declared.

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Nazofaringeal Papiller Adenokarsinom: İki Olgu ve Literatürün Gözden Geçirilmesi

Nazofaringeal papiller adenokarsinom (NPAK), genelde nazofarinkste saptanan nadir bir tümördür. Etiyolojisi net olarak aydınlatılamamıştır. Yüzey epitel kaynaklı olduğu düşülmektedir. Histolojik olarak papiller tiroid kanserine (PTK) benzer, immünhistokimyasal olarak karakteristik bir ekspresyon patterni gösterir ve bu sayede PTK'den ayırt edilebilir. Tümörün tam olarak çıkartıldığı cerrahi tedavi sonrası lokal rekürrens ve metastazın bildirilmemektedir. Bu tümörün doğru tanısı, hastanın gereğinden fazla tedavi almaması ve yaşam kalitesini idame ettirebilmesi için önemlidir. Bu makalede iki NPAK vakasının klinik, patolojik, immünhistokimyasal ve moleküler bulguları paylaşılmış ve tümörün prognostik özelliğine dikkat çekilmiştir. Nadir bir tümör olsa da merkezimizde görülen iki olgunun paylaşılması ve bu tümörün varlığının akılda tutulması önemlidir.

Anahtar Sözcükler: Ayırıcı tanı; immünohistokimya; nazofaringial papiller adenokarsinom; nazofarinks.