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Carcinoma Ex Combined Inverted and Oncocytic Types Sinonasal Papilloma with Multifocal Severe Dysplasia: A Case Report

Case Report

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Abstract

Sinonasal papillomas, also called Schneiderian papillomas, are uncommon benign epithelial tumors arising from the pseudostratified ciliated sinonasal mucosa. Three subtypes of sinonasal papillomas have been described, namely exophytic papilloma, inverted papilloma, and oncocytic papilloma. These papillomas usually have a favorable prognosis, however, frequent recurrences and local aggressiveness has been observed. These may undergo malignant transformation in rare instances. We report a case of a 70-year-old male with multiple co-morbidities presenting with nasal obstruction and bleeding for 3 months. CT and MRI sinuses demonstrated a polypoid mass within the right nasal cavity extending into the nasopharynx, likely arising from the right maxillary sinus. Histopathology revealed a diagnosis of carcinoma (poorly differentiated, NOS) ex combined inverted and oncocytic types sinonasal papilloma (Schneiderian papilloma) with multifocal severe dysplasia (carcinoma in situ). Importance of correct diagnosis lies in the fact that these tumors show recurrence and malignant transformation, thus, requiring long-term follow-up.

Keywords: Schneiderian papilloma; Sinonasal papilloma; Malignant transformation; Radiotherapy; Oncocytic

Introduction

Sinonasal papillomas, also called Schneiderian papillomas, are uncommon benign epithelial tumors arising from the pseudostratified ciliated sinonasal mucosa [1]. Three subtypes of sinonasal papillomas have been described, namely exophytic papilloma (EP), inverted papilloma (IP), and oncocytic papilloma (OP). These papillomas usually have a favorable prognosis, however, frequent recurrences and local aggressiveness has been observed [2]. Additionally, malignant transformation of sinonasal papilloma has also been reported in literature. In a study, Nudell et al. [3] found a prevalence of 1.9% of carcinoma ex-sinonasal papilloma, however, previous literature shows an incidence ranging from of 2 to 27 % [3]. We report a rare case of carcinoma (poorly differentiated, NOS) ex combined inverted and oncocytic type sinonasal papilloma (Schneiderian papilloma) with multifocal severe dysplasia (carcinoma in situ). Our case is unique in that it represented a combination of inverted and oncocytic papilloma components along with severe dysplasia and malignancy. The malignant part of the tumor is poorly differentiated and does not fit into any well-defined categories of sinonasal carcinomas.

Case Presentation

A 70-year-old male with a history of chronic kidney disease, coronary artery disease and hypertension, presented with nasal obstruction and bleeding for 3 months. He had more than 50-pack-year smoking history. CT sinuses without contrast demonstrated a large polypoid soft tissue mass within the right nasal cavity extending into the nasopharynx, likely arising from the right maxillary sinus. Magnetic resonance imaging (MRI) of the face and neck without contrast revealed a large T2 hyperintense heterogeneous lesion near completely filling the right maxillary sinus and extending into the right aspect of the nasal cavity and the nasopharynx through the right osteomeatal unit. There was no evidence of extension into the orbit or intracranial compartment. The differential diagnoses on imaging included both benign and malignant sinonasal masses. A biopsy was obtained that was notable for a sinonasal undifferentiated carcinoma with a focus of oncocytic papilloma. Given the underlying malignancy, an endoscopic maxillectomy with a total ethmoidectomy, sphenoidotomy with orbital decompression and a dacryocystorhinostomy was performed to achieve appropriate margins. Grossly, multiple fragments of irregular pale-tan glistening tissue were identified.

Histopathology revealed an area of inverted papilloma with an endophytic polypoid growth pattern lined by hyperplastic squamous epithelium (Figure 1A) with neutrophilic infiltration into the epithelium forming microabscesses. Seromucinous glands were absent in this area. Areas of oncocytic papilloma with exophytic growth lined by oncocytic epithelium (Figure 1B) were also identified. The oncocytic papillomatous areas had mucous cells. Both types of papillomas showed severe dysplastic changes transitioning into poorly differentiated carcinoma. The malignant component demonstrated invasive nests of neoplastic cells showing nuclear pleomorphism and hyperchromasia (Figure 1C). A diagnosis of carcinoma (poorly differentiated, NOS) ex combined inverted and oncocytic types sinonasal papilloma (Schneiderian papilloma) with multifocal severe dysplasia (carcinoma in situ) was made. Immunohistochemical stains revealed immunoreactivity for CK5/6 (cytokeratin 5/6, focal and strong), and p63 in inverted papilloma and dysplastic area but not in invasive carcinoma, and mucicarmine in oncocytic papillomatous area (Figure 1D). Immunostains for HMB45, S-100, GATA, and pancytokeratin were negative, supporting our diagnosis and ruling out mucosal melanoma, and salivary type neoplasms. The patient's postoperative course was complicated by recurrent epistaxis requiring admission and transfusion. Nasal endoscopy with control of epistaxis via cauterization of posterior septal artery was performed. The course remained uneventful afterwards and the patient underwent radiotherapy and was on regular follow up. He later died of stroke of unknown etiology.

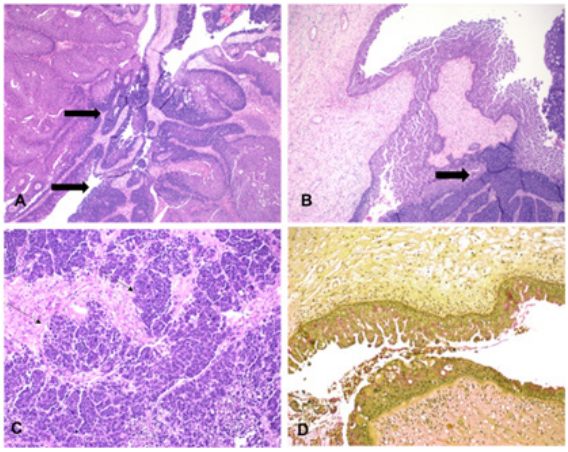

Figure 1: Hematoxylin and eosin staining shows an inverted papilloma with an endophytic polypoid growth pattern lined by hyperplastic squamous epithelium (A: 20x) with areas of dysplastic epithelium (arrows) mixed with areas of oncocytic papilloma (B: 20x) with exophytic growth pattern lined by oncocytic epithelium with a focus of dysplastic epithelium (arrows). Poorly differentiated carcinoma demonstrated invasive nests of neoplastic cells showing nuclear pleomorphism and hyperchromasia (C: 40x). Mucicarmine highlighting mucous cells in oncocytic papillomatous area (D: 20x).

Discussion

Sinonasal papillomas also called Schneiderian papillomas are benign epithelial tumors. These originate from sinonasal or Schneiderian mucosa, which has pseudostratified ciliated columnar epithelial cells, variable mingled goblet cells, and inconspicuous basal cells [1]. Sinonasal papillomas are divided into three different histopathological subtypes-oncocytic (OP), exophytic, and inverted (IP). Inverted sinonasal papilloma is the most common type of sinonasal papilloma accounting for 0.5-4% of primary nasal tumors [2-4]. Almost all sinonasal papillomas are IP or exophytic papillomas. Oncocytic papilloma is the least common, seen in only 3-5% of sinonasal papillomas [5]. OP has not shown any sex preference, however, a 3:1-8:1 male/female preponderance is observed with exophytic papillomas and IP. Most sinonasal papillomas occur between 40 to 60 years of age [5]. The pathogenesis of sinonasal papilloma is still not completely understood. Human papilloma virus (HPV) types 6, 11, 16, and 18 have been thought of as an accountable risk factor for incidence of exophytic and IP, though OP does not have such a connection [1]. Epithelial growth factor receptor (EGFR) mutations also play a significant part in the pathogenesis of IP and IP-associated sinonasal squamous cell carcinoma. However, this mutation is not identified in exophytic or OP related sinonasal carcinomas [6]. Recent data demonstrate role of activating Kirsten rat sarcoma virus (KRAS) mutations in the occurrence of OP [7].

Sinonasal papillomas may go through a malignant transformation into a sinonasal carcinoma that could be concurrently present with the papilloma or may develop at the previous resection site [1,3,8]. The mechanism of carcinoma development from sinonasal papilloma has not been elucidated yet. Increased expression of EGFR and tumor growth factor α (TGF-α) are implicated in early carcinogenesis of inverted papilloma [8]. Some studies supported the role of HPV infection [5,8].

A histopathological examination is required to distinguish various types of sinonasal papillomas. Grossly, IP is usually firm, polypoid, and multinodular gray colored mass with an uneven surface like mulberry [8]. On microscopic examination, IP shows endophytic growth of thick nonkeratinizing transitional cell epithelium that undergoes squamous maturation and inverts into the edematous stroma [5]. A definite basement membrane separates the epithelium from the underlying stroma [8]. The epithelium lacks mucus-secreting cells and eosinophils. As opposed to IP, OP appears as a pink, fleshy, and soft papillary lesion. On microscopic evaluation, it displays both exophytic and endophytic growth with multiple layers of pseudostratified columnar epithelium having small round nuclei, and eosinophilic cytoplasm, with negligible epidermoid component [9]. Intraepithelial mucus filled microcysts can be identified. The oncocytic nature of the epithelium differentiates OP from the other types of sinonasal papillomas [9].

Complete surgical excision is the main treatment modality for sinonasal papillomas which can be accomplished either via external approaches or endoscopic resection depending on the tumor location and extension. Endoscopic approach is preferred due to decreased morbidity without impacting recurrence rates [8,9]. Radiation therapy is usually not indicated for the treatment of sinonasal papillomas unless associated with malignancy. Some previously reported literature suggests radiation therapy to treat locally advanced papillomas or those showing multiple recurrences [9]. Recurrence following surgical removal broadly varies in the prior reports suggesting incomplete resection. 5-60% of IP cases and 25-35% of OP cases may show recurrence within 5 years of surgical resection. Recurrences have been reported from first 3 years to 10 years postoperatively suggesting long-term follow-up [8,9].

Conclusion

Sinonasal papillomas are unusual tumors of sinonasal tract which may undergo malignant transformation in rare instances and usually show an evolution from dysplasia to carcinoma in malignant cases. Thus, it is crucial to mention the presence of dysplasia in pathology reports. Importance of correct diagnosis lies in the fact that these tumors show recurrence and malignant transformation and thus, long-term follow-up of these patients is warranted. More studies are needed to determine response to radiation therapy and rate of recurrence after complete excision.

References

1. Von Bueren AO, Karremann M, Gielen G H, Benesch M, Fouladi M, et al. (2018) A suggestion to introduce the diagnosis of "diffuse midline glioma of the pons, H3 K27 wildtype (WHO grade IV)". Acta Neuropathol 136(1): 171-173.

2. Solomon DA, Wood MD, Tihan T, Bollen AW, Gupta N, et al. (2016) Diffuse Midline Gliomas with Histone H3-K27M Mutation: A Series of 47 Cases Assessing the Spectrum of Morphologic Variation and Associated Genetic Alterations. Brain Pathol 26(5): 569-580.

3. Funata N, Nobusawa S, Nakata S, Yamazaki T, Takabagake K, et al. (2018) A case report of adult cerebellar high-grade glioma with H3.1 K27M mutation: a rare example of an H3 K27M mutant cerebellar tumor. Brain Tumor Pathol 35(1): 29-35.

4. Louis D, Giannini C, Capper D, Paulus W, Figarella Branger D, et al. (2018) cIMPACT NOW update 2: diagnostic clarifications for diffuse midline glioma, H3 K27M mutant and diffuse astrocytoma/anaplastic astrocytoma, IDH mutant. Acta Neuropathologica 135(4): 639-642.

5. Dono A, Takayasu T, Ballester L, Esquenazi Y (2020) Adult diffuse midline gliomas: Clinical, radiological, and genetic characteristics. J Clin Neurosci 82(Pt A): 1-8.

6. Funato K, Major T, Lewis P, Allis CD, Tabar V (2014) Use of human embryonic stem cells to model pediatric gliomas with H3.3K27M histone mutation. Science 346(6216): 1529-1533.

7. Ebrahimi A, Skardelly M, Schuhmann M, Ebinger M, Reuss D, et al. (2019) High frequency of H3 K27M mutations in adult midline gliomas. J Cancer Res Clin Oncol 145(4): 839-850.

8. He P, Chen W, Xi Qiu X, Bin XY, Guan H, et al. (2019) A Rare High-Grade Glioma with a Histone H3 K27M Mutation in the Hypothalamus of an Adult Patient. World Neurosurg 128: 527-531.

9. Wang L, Li Z, Zhang M, Piao Y, Chen L, et al. (2018) H3 K27M-mutant diffuse midline gliomas in different anatomical locations. Hum Pathol 78: 89-96.

10. Himes B, Zhang L, Daniels D (2019) Treatment Strategies in Diffuse Midline Gliomas With the H3K27M Mutation: The Role of Convection-Enhanced Delivery in Overcoming Anatomic Challenges. Front Oncol 9: 31.