CASE STUDY

Undifferentiated Carcinoma in Oncocytic Schneiderian Papilloma. A Rarity

Carcinoma indiferenciado en el papiloma oncocítico Schneideriano. Una rareza

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The term Schneiderian mucosa refers to the ectodermally derived lining of the nasal cavity and paranasal sinuses, composed generally of stratified ciliated columnar cells, loose abundant lamina propria, and minor salivary glands and their ducts. This unique mucosa may give rise to three distinct histomorphologic papillomas collectively called Schneiderian papillomas. Hyams† has described these variants and categorized them as inverted, fungiform (exophytic, squamous) and oncocytic Schneiderian papilloma (cylindrical cell papilloma) based on their pattern of growth. Schneiderian papillomas are uncommon, representing only 0.4%–4.7% of all sinonasal tumors.2–4 Oncocytic Schneiderian papillomas are the rarest of the three morphological variants, accounting for only 3 per cent of all sinonasal papillomas.3,4 Sinonasal papillomas have a small but distinct risk of malignant transformation.5–10 Here, we report a case of undifferentiated carcinoma arising from oncocytic Schneiderian papilloma (OSP). To the best of our knowledge, the current patient is the fourth reported case of undifferentiated carcinoma arising in OSP in English literature.

Case Report

A 75-year-old lady presented with recurrent unilateral epistaxis associated with progressive nasal blockage and right nasal purulent discharge with a foul odor of eight months duration. Clinical examination revealed a fleshy, polypoid mass arising from right lateral nasal mucosa obstructing the right nostril with diffuse swelling on the right side of face. Computed tomography confirmed the presence of soft tissue mass in the right lateral nasal cavity and opacification of maxillary and frontal and ethmoid sinuses (Fig. 1). No bony destruction was noted. Incisional biopsy through Caldwell Luc approach revealed it to be a case of OSP.

The patient was subjected to general anaesthesia under nasoendotracheal intubation on left side for lateral rhinotomy with medial maxillectomy and frontoethmoidosphenoidectomy. Intraoperatively, multiple polypoidal masses were found involving right lateral wall of the nasal cavity with extension into the all paranasal sinuses on the right side. The entire tumor mass was delivered en masse and subjected to histopathological examination after staining with hematoxylin–eosin. Other confirmatory tests like immunohistochemical analysis were not carried out for lack of sufficient resources. The patient had an uneventful postoperative recovery. She was last seen six months post operatively and there was no recurrence noted. The patient is being followed periodically for any evidence of

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recurrence. This study followed the Declaration of Helsinki on medical protocol and ethics and the regional Ethical Review Board of Meenakshi Ammal Dental College & Hospital approved the study.

On histological examination, the multiple polyoid lesions showed a central core of connective tissue composed of multilayered columnar type of epithelium. The epithelium was thrown into numerous papillary projections exhibiting both endophytic and exophytic growth patterns indicative of OSP (Fig. 2). The epithelium was made of tall columnar cells with small dark nuclei and eosinophilic granular cytoplasm. Connective tissue stroma is myxomatous and edematous showing minimal chronic inflammatory cell infiltrate predominantly of lymphocytes and plasma cells. The specimen also showed malignant cells arranged in sheets, islands and in papillary pattern. The tumor cells were pleomorphic with enlarged and hyperchromatic nuclei (Fig. 3). In continuing areas between the benign papilloma and the obvious malignant tumor, the papillary epithelium showed a gradual change from a low-grade to a high grade dysplasia (Fig. 4).

Discussion

Fungiform papilloma, inverted papilloma, and OSP are three morphologically distinct lesions arising from the Schneiderian membrane.1,3,4,6,7 Fungiform papilloma, which is usually located on the nasal septum, has a risk of recurrence but is not associated with malignant potential. By contrast, both OSP and inverted papillomas involve the lateral wall of the nose and the paranasal sinuses with high potential for local invasion and malignant changes especially when incompletely excised.1,2,11 Inverted papilloma has a definite association with malignancy. However, OSP has been distinguished from other papillomas by its histological as well as cytologic features.

Barnes and Bedetti3 demonstrated that the epithelial cells of oncocytic Schneiderian papilloma are true oncocytes which arise from the sinonasal respiratory epithelium- hence the term oncocytic Schneiderian papilloma. The oncocytoid appearance of OSP is due to mitochondrial hyperplasia, as
they are in oncocyes of varied tissues, such as oncocytoma, and Hurthle cell or Warthin’s tumor.

Oncocytic Schneiderian papilloma are equally distributed between the sexes, and the majority of patients are more than 50 years of age at the time of diagnosis. There is no sex predilection, unlike in inverted and fungiform types which occur predominantly in males. Unilateral nasal obstruction is the most common presenting symptom for OSP. Intermittent rhinorrhea, sinusitis and allergic symptoms are rarely described. The duration of symptoms is usually months to years. All cases known to date have been unilateral. 

A gross shaggy or papillary configuration is most typical. However, smooth, edematous, polypoid lesions are not uncommon as evidenced in our patient. Routine sinus radiographs reveal abnormalities confined to the ipsilateral sinonasal passages. Sinus opacification associated with an intranasal soft tissue density is the predominant finding.

The radiographic findings vary with the extent of the disease. Early on, there may be only a soft tissue density within the nasal cavity and/or paranasal sinuses. Later, with more extensive disease, unilateral opacification and thickening of one or more of the sinuses are common, as are expansion and displacement of adjacent structures. Pressure erosion of bone may also be apparent, and must be distinguished from osseous invasion associated with malignancy.

Grossly, OCP appears as a fleshy pink papilloma or gray polypoidal growth. The classic histologic description of OCP is the presence of both exophytic and inverted growth patterns, composed of multilayers of columnar cells with eosinophilic cytoplasm and small uniform dark nuclei. The epithelial cells of OCP are true oncocyes from the sinonasal respiratory epithelium and hence the term oncocytic Schneiderian papilloma was used. The epithelium characteristically contains small cysts filled with mucon or neutrophils (microabscesses).

Although the clinical behavior of OSP parallels that of inverted papilloma, it appears to have a higher frequency of association with malignancy (10% to 17%) compared to the later (5% to 10%). Fortunately, the incidence of OSP is much lower than that of inverted papilloma. Of the 178 cases of OSP previously reported in the English literature which constitute 3% to 10% of Schneiderian papillomas, only 13 cases were found to be associated with carcinoma. But Maitra et al. study reported 16 cases of malignancies arising in OSP. Hyams reported the first case of OSP (which he named cylindrical cell papilloma) associated with invasive carcinoma among 10 cases of OSP. Barnes and Bedetti described a case associated with mucoepidermoid carcinoma among six cases of OSP. However, the relationship between carcinoma and the epithelium of OSP is unclear. The evidence that carcinomas can arise in an OSP came when Ward et al. reported two cases of OSP, associated with poorly differentiated squamous cell carcinoma in one case, but poorly differentiated nonkeratinizing carcinoma and papillary intermediate (transitional) cell carcinoma in the other. Their study is the first to document dysplastic epithelium in OSP, which they claimed can be the site of origin for invasive carcinoma. Kapadia et al. reported nine cases of carcinoma in OSP, among which six cases were associated with squamous cell carcinoma, two cases with high grade mucoepidermoid carcinoma, and one case with sinonasal undifferentiated carcinoma. This was the first reported case of undifferentiated carcinoma arising from OSP. Moreover, Yang and Abraham also reported a case of sinonasal undifferentiated carcinoma. Using morphological, histochemical and immunohistochemical analysis, they were able to demonstrate the malignancy associated with the OSP. The third case of sinonasal undifferentiated carcinoma arising in OSP was reported by Maitra et al. In our case, the oncocytic features of this papilloma were clearly demonstrated histomorphologically. It is of utmost importance to determine the origin of carcinoma in papilloma. It could be either arising from within the papilloma or merely a coexisting disease. In our case, the epithelium of papilloma showed areas of transition from low-grade to high-grade dysplasia, thereby validating the origin of carcinoma from within the OSP. The malignancy associated with this OSP resembles an undifferentiated carcinoma histomorphologically.

This article emphasizes the association of OSP with malignancy and not on discussing the modalities of treatment of OSP. In our case, carcinoma was present in a small proportion of each papilloma and could easily have been missed if only a few tissue sections were taken. Moreover, if such lesions (as this case) were to be treated by a conservative approach, that is, polypectomy, foci of malignant cells will be undoubtedly left behind. Hence, the authors suggest a more radical approach at a time when there is minimal tumor burden.

The purpose of this case report in highlighting this subtype of sinonasal papilloma in general and OSP, in particular lies in its rarity, tendency to be misdiagnosed as ordinary polyp, well-differentiated adenocarcinoma, or rhinosporidiosis and the necessity of advocating an extensive operative approach due to its high recurrence rate, local destructive ness and a relatively high malignant potential.

Conflict of Interest
The authors have no conflict of interest to declare.

References


