

CASE REPORT**PAPILLARY SQUAMOUS CARCINOMA OF PARANASAL SINUS-AN UNCOMMON HISTOLOGICAL VARIANT**

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DOI URL- <https://doi.org/10.21176/ojohns.2018.12.1.10>**ABSTRACT:**

Papillary Squamous cell carcinoma is an uncommon histological variant Squamous cell carcinoma (SCC) of the upper aero-digestive tract (UADT). It rarely involves the paranasal sinuses. We report an aggressive case of papillary SCC of the left maxillo-ethmoid complex in a 59 year old male, who presented with rapidly progressive left cheek swelling and nose block. Biopsy revealed diagnosis of Papillary Squamous cell carcinoma arising in a transitional papilloma. The contrast enhance CT scan showed heterogeneously enhancing large lobulated soft tissue mass is seen involving the left maxillary sinus, left nasal cavity, left ethmoidal sinuses and left frontal sinus with extensions as.

Superiorly, there was destruction of inferior wall of left orbit with loss of fat plane between the inferior oblique and inferior rectus muscles possibly infiltrating and pushing the globe superolaterally. Inferiorly, with destruction of inferior wall of left maxillary sinus and extending along alveolar process of left maxilla is seen.

Patient underwent left subtotal maxillectomy (Pre-maxilla preservation) and orbital exenteration with endoscopic fronto-spheno-ethmoidal clearance. The resected margins were found to be free from tumour. The patient underwent postoperative Chemo-radiotherapy. Patient developed recurrence at orbital apex with extension to Middle cranial fossa after four months following post operative radiotherapy with a histopathological diagnosis of well differentiated carcinoma and was advised cyber knife treatment at another center. The patient did not undergo cyber knife treatment and came back after one month and was started on palliative chemotherapy, but expired after 2 months. The case is presented for its rarity and aggressive behavior.

Key words: Papillary, Squamous cell carcinoma, upper aero-digestive tract (UADT).

INTRODUCTION:

Squamous cell carcinoma (SCC) if found at the top of the list among the wide variety of malignancies of head and neck encountered with majority of them being of the conventional type. Other histological variants of SCC include spindle-cell (sarcomatoid), basaloid, verrucous, papillary, and adenosquamous carcinoma¹. These comprise up to 15% of all SCCs of UADT with Papillary Squamous cell carcinoma (PSCC) accounting to less than 1 % of all the cases¹. The PSCC of the head and neck regions conventionally has an excellent prognosis however that of paranasal sinus has the worst outcome. A case of papillary type SCC of the left maxilla-ethmoid complex with unusual clinical behavior is presented.

CASE REPORT:

A 57 year old male with no co-morbidities, presented with left cheek swelling of 20 days duration which was sudden in onset and rapidly progressive. Patient also had left sided nose block and hyposmia. On clinical examination, a 6x6 cm swelling was seen in the right cheek and malar area and the overlying skin was edematous and fixed to subcutaneous

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tissue. The eye was pushed upward and outward but the vision and extra ocular movements were normal. A diagnostic nasal endoscopy was done which revealed a granular friable mass in the left nasal cavity anterior to the middle turbinate and arising from the middle meatus. Contrast enhancement CT scan showed heterogeneously enhancing large lobulated soft tissue mass is seen involving the left maxillary sinus, left nasal cavity, left ethmoidal sinuses and left frontal sinus with involvement of orbit and orbital contents (Fig-1). Biopsy was taken from the mass and the histopathology revealed transitional type of epithelium thrown into papillae, with fibro-vascular core, focally exhibiting marked nuclear pleomorphism, hyperchromasia and squamous differentiation with atypical mitotic figures, bizarre cells and large areas of necrosis. A diagnosis of papillary type of SCC arising in a transitional papilloma was made. Patient underwent left sub-total maxillectomy with preservation of the pre-maxilla and left orbital exenteration via modified Weber Ferguson incision (sub-conjunctival) with Lynch extension (Fig-2). Fronto-ethmoido-sphenoidal clearance was done under endoscopic supervision. Histopathology showed papillae and anastomosing sheets of malignant squamous cells with pleomorphic nucleoli, coarse chromatin, atypical mitosis, with numerous keratin pearls and horn cysts consistent with papillary SCC (Fig-3). Resected margins, optic nerve, frontal and the sphenoid sinus were free of tumour infiltration. Patient was treated with adjuvant radiotherapy of 60Gy divided into 30 fractions over a period of 6 weeks. At 4 months post-surgery, nasal endoscopy revealed a polypoidal mass in the lateral wall of left sphenoid sinus. Biopsy sent for histopathology showed infiltrating nests and sheets of malignant cells, having pleomorphic nuclei, abundant

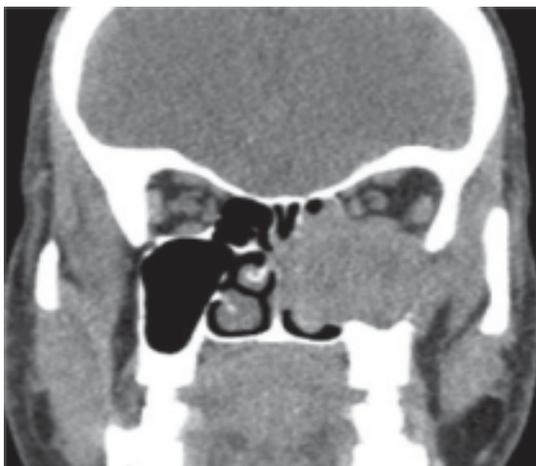


Fig-1. CECT showing heterogeneously enhancing large lobulated soft tissue mass is seen involving the left maxillary sinus, left nasal cavity, left ethmoidal sinuses and orbit.



Fig-2 Showing (a) post resection defect and (b)resected specimen.

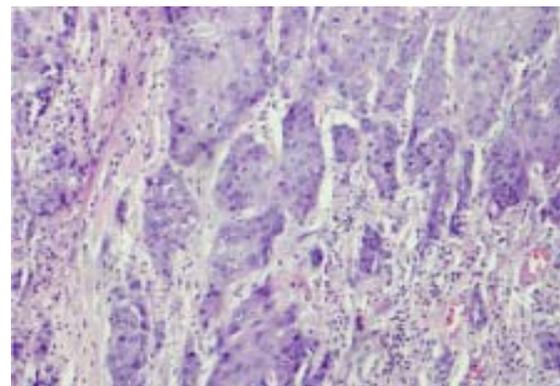


Fig-3. Histopathology showed papillae and anastomosing sheets of malignant squamous cells with pleomorphic nucleoli, coarse chromatin, atypical mitosis, with numerous keratin pearls and horn cysts.

eosinophilic cytoplasm, and suggestive of moderately differentiated SCC. PET-CT showed recurrent tumour in orbital apex extending into sphenoid sinus and middle cranial fossa, reaching till cavernous sinus. Patient was started on palliative chemotherapy with carboplatin but expired after 2 months of palliative CT.

DISCUSSION:

Crissman et al first proposed the term papillary carcinoma to this rare variant of SCC². PSCC are de novo malignancies which can arise from a pre- or a co-existing benign lesion occasionally can develop in benign squamous papillomas³. In view of their rare occurrence, the etiopathogenesis still remains undetermined. Papillary SCCs develop through different etiologies with a greater proportion probably secondary to Human Papilloma Virus of high risk types mainly 16 and 18. Most common sites of involvement

include the larynx and the sinonasal tract, which are also the common sites of benign HPV-related lesions like papillomas⁵. Papillary SCCs are common in men in 6th and 7th decade. But HPV related SCCs have early age at presentation of about 59.6 years compared with the HPV negative carcinomas which are about 68 years. Two-thirds of papillary SCCs were immunoreactive to antibody p16 and 68% of these immunopositive lesions harbor high-risk HPV. About 73% of HPV related papillary SCCs arise in oropharynx (base of tongue and palatine tonsils), with rare occurrence in nose, paranasal sinuses and larynx⁵.

Macroscopically papillary SCC appears as an exophytic tumour or polypoidal lesion which is usually friable, soft to firm in consistency, arising from a broad base. Most tumours present at early stage (T1 or T2), although multifocality has been described^{1,6}.

Microscopically PSCC have complex papillary fronds growing into the surrounding tissue, thereby making it difficult to assess the degree of infiltration into the surrounding tissue. These papillary projections consist of a central fibrovascular core layered with stratified squamous epithelium which exhibits surface keratinization, nuclear pleomorphism, increased nuclear-cytoplasmic ratio, loss of cellular polarity and increased mitotic figures which are all classical features of SCC^{1,6}.

On immunohistochemistry, papillary SCC shows increased expression of Ki-67, p53 and loss of heterozygosity for a microsatellite marker found characteristically on the long arm of chromosome 11⁵.

The treatment guidelines follow those of SCC. Wide excision with adequate margins with neck dissections in nodal positive patients in lesions involving larynx and oral cavity +/- radiotherapy and radiotherapy with/without chemotherapy in case of oropharyngeal lesions are the primary treatment modalities. Lesions can be locally aggressive and difficult to fully resect, especially when they involve the sinonasal tract because of the intricate anatomy and vicinity of vital structures like skull base and carotids which precludes adequate margins for clearance. Such lesions often eventually progress to invasive malignant neoplasm and have higher chances of recurrences⁴.

Owing to its minimal invasion and less aggressive behavior, patients with PSCC tend to have better results as compared to conventional SCC with same stage at presentation. Distant metastases are very rare in these cases.

About one-third of patients develop recurrence, which resemble the primary tumour histopathologically⁷, but in our case it was a conventional type of moderately differentiated SCC. The reason why the recurrence was different from the

primary remains unexplained. It probably would have been a radiation induced malignancy or a second primary.

CONCLUSION:

UADT tumors related to HPV infection seem to have a better prognosis than those related to alcohol and smoking. Papillary SCC is a low to moderate grade malignancy. It is presumed that distant metastasis of PSCC is rare. It has a better prognosis than conventional SCC of the similar clinical stage but worse than that for verrucous carcinoma. This report presents an unusual aggressive behavior of this histological type with recurrence despite surgery and post-operative radiotherapy. This case report enlightens the need for further studies on the behavior and treatment protocols of this papillary variant of SCC.

DISCLOSURES:

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- (d) Written consent of patient- taken
- (e) Animal rights- Not applicable

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