# Synchronous Squamous Cell Carcinoma of External Acoustic Meatus Following Pigmented Basal Cell Carcinoma of Cheek - A Rare Occurrence

*Debangshu Ghosh,[1] Rajarshi Sannigrahi,[1] Sumit Kumar Basu,[1] Parna Basu[2]*

**ABSTRACT**

***Introduction***

*A synchronous second primary malignancy as squamous cell carcinoma of external acoustic meatus following basal cell carcinoma of cheek is reported with their management and review of literature.*

***Case Report***

*Pigmented basal cell carcinoma of cheek was managed successfully by wide local excision followed by flap reconstruction and subsequently diagnosed squamous cell carcinoma of external acoustic meatus by concurrent chemoradiation after initial surgical debridement.*

***Conclusion***

*Second primary malignancy as squamous cell carcinoma of external ear canal is rare after basal cell carcinoma of cheek though there is anatomical vicinity. Surgery in case of basal cell carcinoma and concurrent chemoradiotherapy in case of external ear canal squamous cell carcinoma is the mainstay of treatment.*

***Keywords***

*Carcinoma, Basal Cell; Carcinoma, Squamous Cell; Ear Canal; Ear, Middle; Head and Neck Neoplasms; Neoplasms, Second Primary*

Second primary malignancy (SPM) is a tumour that presents either simultaneously or sometimes after diagnosis of an index tumour. The criteria used for the diagnosis of multiple primary cancers were first given by Warren and Gates(1932) and modified later by Moertel et al and National Cancer Institute for their surveillance, epidemiology and end results(SEER) program.[1,2,3,4] If the second cancer is of different histology or it develops in a different location then it is SPM. If the second cancer is of same histology and develop in the same region as the index cancer it can only be coded as SPM if greater than 60 months had passed since the diagnosis of an index cancer. SPM can be of two types. It is called synchronous lesion when the second primary lesion is detected within 2-6 months of diagnosis of first primary tumour or metachronous when this interval is at least 6 months or more after first primary.[2,3]

Patients with head and neck carcinoma are at increased risk of development of SPM. Second primary malignancies represent the second most common cause of death in patients with head and neck squamous cell carcinoma (HNSCC) contributing one-quarter to one-third of deaths in such patients highlighting the importance of SPM in head and neck cancers.[5]

## Case Report

A 56 year old female presented to the department of ENT with complaints of a black pigmented area over the right cheek for one and half years and foul smelling discharge from right ear for three months. The ear

*1 - Department of ENT, R G Kar Medical College, Kolkata*
*2 - Department of Radiotherapy, R G Kar Medical College, Kolkata*

**Corresponding author:**
Dr Debangshu Ghosh
email: ghoshdr.d777@ymail.com

discharge was occasionally mixed with blood and associated with severe earache and gradual decrease in hearing. On examination there was a black naevus measuring 1 cm x 1.5 cm in size and located 7 cm from midpoint of tragus over the right side of cheek. (Fig. 1) It was non-tender, firm in consistency and with smooth surface. Margins of the naevus were well delineated. No cervical lymphadenopathy was noted. On otoscopy there was right sided blood tinged purulent discharge. Provisionally we reached the diagnoses of black naevus right cheek and right active squamous chronic otitis media. The patient was referred to plastic surgery department and planned for incision biopsy from the naevus.

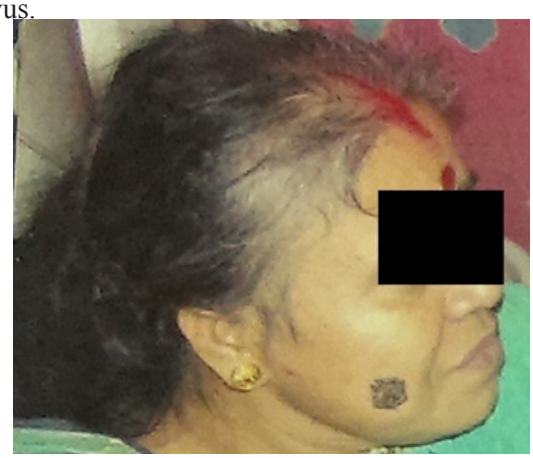

**Fig. 1 Appearance of the patient before treatment**

Histopathological examination (HPE) of the incision biopsy specimen revealed pigmented basal cell carcinoma (BCC). (Fig. 2) The patient underwent wide local excision of the nevus with 2 mm margin around it and reconstruction of the surgical defect was done with Limberg flap. After 3 days during her hospital stay the patient developed right mastoid subperiosteal abcess which was drained and pus sent for culture and sensitivity testing. As per culture sensitivity report Cefotaxime was administered in a dose of 1 gm. intravenous (IV) twice a day and continued for 10 days with regular antiseptic dressing.

Pure tone audiometry showed profound mixed hearing loss in right ear. High resolution computed tomography (HRCT) of temporal bone was done which

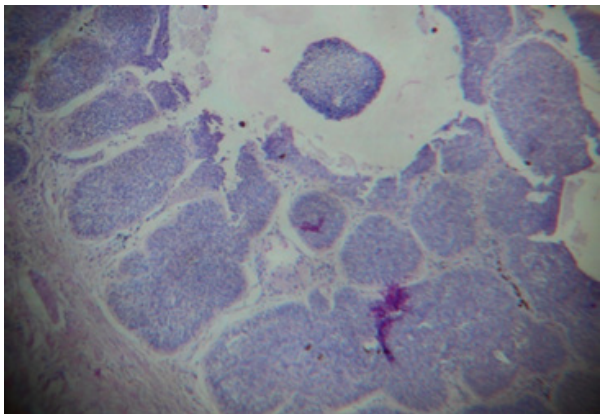

**Fig. 2 Histological photomicrograph (10x, H&E) showing pigmented BCC of cheek. Arrow showing nests of basaloid cells separated by thin clefts**

showed enhancing ill-defined soft tissue lesion in right external auditory canal and middle ear with erosion of bony inferior canal wall and absence of ear ossicles with probable involvement of labyrinth along with features of otomastoiditis. (Fig. 3) Based on this report biopsy was taken from post auricular region and sent for HPE. It was reported as pseudoepithelial hyperplasia with severe dysplasia along with extensive inflammation and necrosis in subepithelial tissue. No evidence of

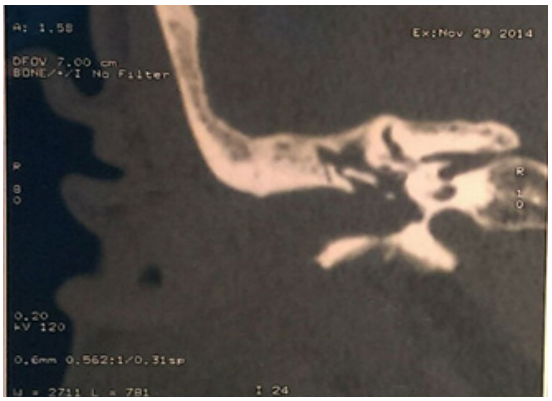


**Fig. 3 HRCT scan showing soft tissue density lesion causing bone erosion and widening of bony external meatus**

malignancy was found.

The patient underwent right canal wall down mastoidectomy with debridement of soft tissue mass from

the external auditory canal. There was gross adhesion with the surrounding tissues and part of parotid tissue was found to be attached with soft tissue mass that was excised. Facial nerve could not be identified separately from the mass. Postoperatively patient developed House Brackman grade IV facial palsy that could be due to intraoperative injury. HPE of the excised mass was reported as well differentiated infiltrating squamous cell carcinoma (SCC) of right external auditory canal. (Fig. 4) The patient was referred to Department of Radiotherapy where three dimensional conformal external beam radiation therapy was administered with

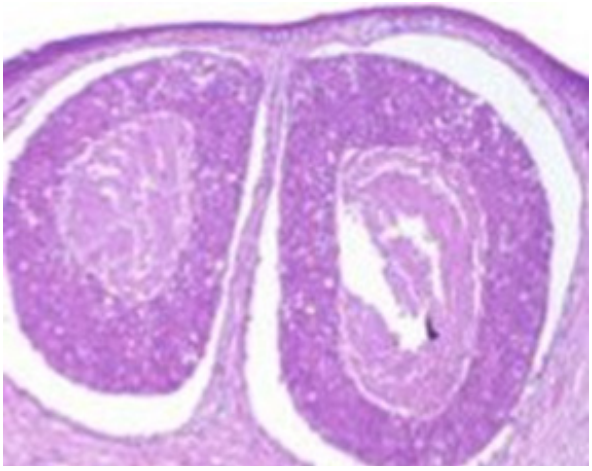

**Fig. 4 Histological photomicrograph (10x, H&E) showing SCC of external ear canal. Keratin pearl is shown by the arrow**

concurrent platinum based chemotherapy in 2 phases. Phase 1 consisted of 45 Gy in 25 fractions over 5 weeks (1.8 Gy per fraction). Phase 2 consisted of 18 Gy in 10 fractions over 2 weeks (1.8 Gy per fraction). The concurrent chemotherapy consisted of injection Carboplatin AUC 5 intravenous at three week intervals during the period of radiation. The patient was reviewed weekly for any treatment related complications.

The patient was reviewed after 2 weeks and thereafter monthly for next 6 months during which no recurrence was seen over the cheek or the external auditory canal. (Fig. 5) Now the patient is disease free but with grade-IV ipsilateral facial palsy. (Fig. 6)

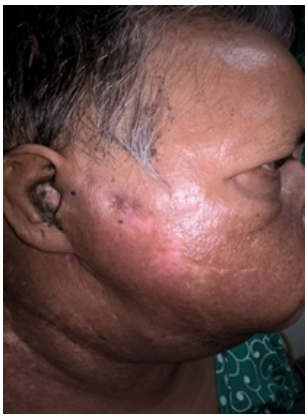

**Fig. 5 Appearance of the patient after 6 weeks of completion of chemoradiation**

## Discussion

Billroth first reported multiple primary tumours of different histology in separate organs at different time periods in same individuals in 1879.[2,4] Incidence of second primary and subsequent tumours are increasing due to (a) increased survival after cancer and (b) improved detection of cancers. Head and neck cancers are associated with a high likelihood of developing second primary malignancies. The standardized

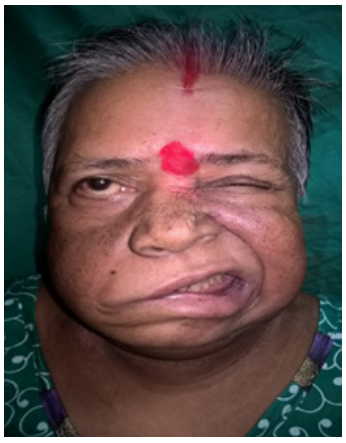

**Fig. 6 Facial palsy of the patient persisting after treatment**

incidence ratio (SIR) is approximately 2.18 with around 36% cumulative life time risk of developing SPM over 20 years after diagnosis of an index tumor for which the most common sites are the head and neck region, esophagus, and lungs.[6] This is explained to some extent by "field carcinogenesis theory" related to common risk factors like tobacco use and alcohol consumption on top of some genetic contribution. This concept was introduced by Slaughter et al, who discovered that in oral cancers the epithelium beyond the boundaries of tumor possessed histologic changes resulting in wide array of premalignant diseases that give rise to multiple independent primary tumor.[7] This is particularly true for skin cancers which have hereby increased in incidence by 20% over the last decade.[8]

BCC arises from pluripotent cells of epidermis or hair follicles. Usually these tumours take an indolent course and may take years to grow into significant size. BCC are most commonly seen in the sun exposed areas. The head and neck accounts for 85 to 93% of all skin carcinomas and nasal skin (31.5%) is the most common site of presentation followed by cheek (26.9%).[9] There are several types of BCC like nodular, superficial, pigmented and morphoeic. Among these nodular is the most common type accounting for 46.2% followed by pigmented variety (18.7%).[10] Surgery for BCC can achieve high cure rate. It has been reported that 94% cure rate can be achieved using a 2 mm excision margin and a 95% cure rate with a 4 mm margin for tumours less than 2cm. in size.[11] Pigmented basal cell carcinoma comprises about 6% of all BCC.[12] Pigmentations produced in this type of cancer make it necessary to rule out melanoma. Dermoscopy is a useful tool for this but immunomarkers are confirmatory. HMB-45 and S-100 are the most useful markers for melanoma.

Squamous cell carcinoma arises from basal layer of epidermis. It has a poorer prognosis than BCC because of its aggressive local invasion and metastatic potential. It usually occurs within altered skin such as within an actinic keratosis, by malignant change in a chronic ulcer or sinus. The risk of metastatic spread is 2% to 5% and occurs usually via lymphatics. There are no variants of SCC but they are graded histologically to indicate the aggressive nature of individual tumour. Prognosis of SCC depends upon depth of tumour, degree of differentiation and mitotic index. Squamous cell carcinoma of the temporal bone and external auditory canal are extremely rare with reported incidence of 1 to 6 cases per million population per year.[13]

The preferred treatment for external auditory canal SCC consists of en-bloc surgical resection with postoperative chemo and radiotherapy. For well differentiated carcinoma of less than 2cm diameter, a minimum of 4 mm margin and for ≥2 cm diameter tumour, a minimum of 10 mm margin is required.[14] Surgery, that is most commonly performed, is lateral temporal bone resection (LTBR) or a subtotal temporal bone resection (STBR). Poor prognostic factors are wide extent of disease at presentation, positive margins, dural and cranial nerve involvement.

Patients with primary head and neck squamous cell carcinomas are also at elevated risk of second primary malignancies, most commonly of the head and neck, lung, and esophagus. In patients with HNSCC, the risk and distribution of SPM differs significantly according to subsite of the index cancer.[3] Before the 1990s, hypopharyngeal and oropharyngeal cancers carried the highest excess risk of SPM. Since then during the human papilloma virus (HPV) era SPM risk associated with oropharyngeal SCC has declined to the lowest risk level for any subsite though the exact risk ratio is unknown. Data regarding subsite-specific risks and trends over time may be helpful in the rational application of surveillance to patients with HNSCC after treatment of the index tumor. As in our case it's difficult to suspect an SPM in a hidden location like external auditory canal, in a patient presenting with ipsilateral mastoiditis following long standing BCC of cheek.

Most of the synchronously diagnosed second tumors are incidentally diagnosed. They are often detected during the staging evaluation of the primary tumor. Metastatic disease has to be aggressively ruled out to stamp it as SPM. Any unusual site of metastasis should be thoroughly evaluated to rule out the rare possibility of second primary. A baseline positron emission tomography scan coupled with CT (PET-CT) may aid in the diagnosis of such multiple tumors and in some cases also helps in therapeutic planning. We treated both the tumours surgically while the patient remained admitted in our hospital and subsequent chemoradiation regime

was administered on ambulatory basis.

When multiple tumors are pathologically confirmed at the time of presentation itself, each tumor should be evaluated and staged as independent tumors. They should be treated aggressively with the curative intent depending on the stage of each disease to achieve maximum therapeutic benefit. If surgery is needed for both the tumors, it can be done in a single stage in majority of the cases with low rates of morbidity and mortality. We offered two surgeries to our patient in two stages as we could not diagnose squamous cell carcinoma of external ear canal till the first surgery for BCC was over.

Surgery and/or radiotherapy were the standard modalities used to achieve locoregional control, but since the publication of the 1st meta-analysis on chemotherapy in head and neck cancer (MACH-NC), Platinum based concurrent chemoradiotherapy has largely replaced radiotherapy alone in the treatment of unresectable squamous cell carcinoma of head and neck. Despite this therapeutic approach the prognosis of HNSCC patients remains poor. The 5 year survival rate in USA in the period 1996-2003 was around 50% compared to 32% in the control group.[15] Chemotherapy can be administered in 3 ways in the treatment of locally advanced HNSCC: as induction chemotherapy, concurrent with radiotherapy and as an adjuvant after radiotherapy and/or surgery. The absolute benefit in 5 year survival was seen as 2%, 8% and 1% respectively.[16] Taking this into account concurrent chemoradiotherapy has become the standard treatment for locally advanced HNSCC. We treated our case with the same.

## Conclusion

The occurrence of two concurrent non melanoma cutaneous malignancies of head neck region is very rare. Second primaries in head neck carcinoma are a predominant cause of morbidity and mortality. Surveillance for a second primary malignancy following the diagnosis of an index malignancy could aid in early diagnosis of another life threatening condition that might still be in a curable stage and could be operated in the same setting with the index tumour. Till date India lacks site-specific and histology-specific registries of SPM that might guide us in such surveillance procedures. Future studies are necessary in this direction.

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