Sebaceous Carcinoma of The Nasal Vestibule: A Case Report

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Abstract: Sebaceous carcinoma (SC) is a highly malignant tumour derived from the sebaceous glands. SCs have traditionally been divided into two categories, ocular and extraocular. Tumours arising in extraocular sites are rare and less aggressive than SCs of the eyelid. SC of the nasal vestibule is a very rare pathology, and for today there have been only a few reported cases worldwide. Here, we report a case of a 60-year-old man who presented with a 1-month history of a painful, recurrent bleeding tumour in the right nasal vestibule. Based on the histologic findings, a diagnosis of sebaceous carcinoma of the nasal vestibule was established. We discuss incidence as well as clinical, histologic, diagnostic, prognostic, and management issues of this aggressive neoplasm.

Keywords: Sebaceous carcinoma, nasal vestibule cancer, nose, sebaceous glands.

INTRODUCTION

Sebaceous carcinoma is a rare skin cancer, which originates from sebaceous glands, and its manifestation in the nasal vestibule is exceedingly uncommon [1]. Thus far, only 150 cases of extraocular SC have been reported worldwide [18]. Up to 75 percent of SCs occur in a periocular region and have lower mortality than extraocular SCs [3].

The mainstay of treatment is wide surgical resection. Neck dissection is performed for operable cases with regional lymph node metastases [4].

Here, the authors present a case of sebaceous carcinoma arising from the right nasal vestibule.

This study was conducted in compliance with the principles of the personal data protection law of the Republic of Latvia. The patient's medical records and photographs obtained during the implementation of the study were processed and published with the patient's consent.

CASE REPORT

A 60-year-old man was referred to the Pauls Stradiņš Clinical University Hospital with a one-month history of a recurrent bleeding mass in the right nasal vestibule which was also painful to the touch. His medical history revealed a middle cerebral artery (MCA) stroke, mixed-type encephalopathy, dementia, depression, and organic affective syndrome. The patient's ECOG PS was 3. On physical examination, there was an irregularly shaped mass measuring 4 cm in maximum diameter. Computed tomography (CT) scan revealed a 2.8 x 3.8 x 3.6 cm tumour that spread to the cutaneous and subcutaneous tissues of the upper lip, right anterior nasal spine, right nostril, and right nasal ala. It also showed pathological changes in right submandibular lymph node (Figure **1**).

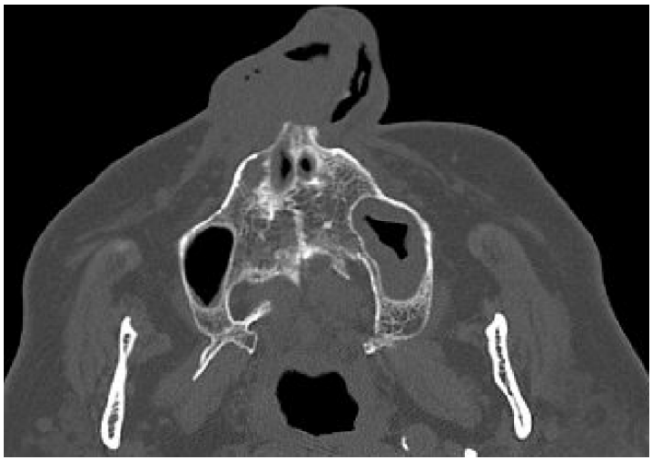

Figure 1: Radiographic Imaging. Axial contrast-enhanced CT demonstrated a mass measuring 2.8 x 3.8 x 3.6 cm that spread to the cutaneous and subcutaneous tissues of the upper lip, right anterior nasal spine, right nostril, and right nasal ala. Because bone involvement was not detected, radical surgery was indicated.

To confirm the diagnosis, an incisional biopsy was performed. Histologic examination revealed purulent detritus, mucous, and fragment of connective tissue with well-differentiated (Grade 2) SCC complexes with comedo-type necroses.

Consilium consisting of ENT surgeon, radiologist, and oncologist chemotherapist, decide on the future tactics of patient management. The treatment of choice was radical resection of the nasal vestibule tumour

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E-ISSN: 2310-998X/23

(Figure **2**). Soft tissue defect was closed by buccal skin graft (Figure **3**) and selective neck dissection was performed. Right submandibular salivary gland excision was done. The final pathologic review showed macroscopically 5 x 4.5 x 2.5 cm big mass, microscopically pT4aN1MxG2R0L(+)V(-)Pn(+). For cancer staging, the Eight Edition of the *AJCC Cancer Staging Manual* was used [5].

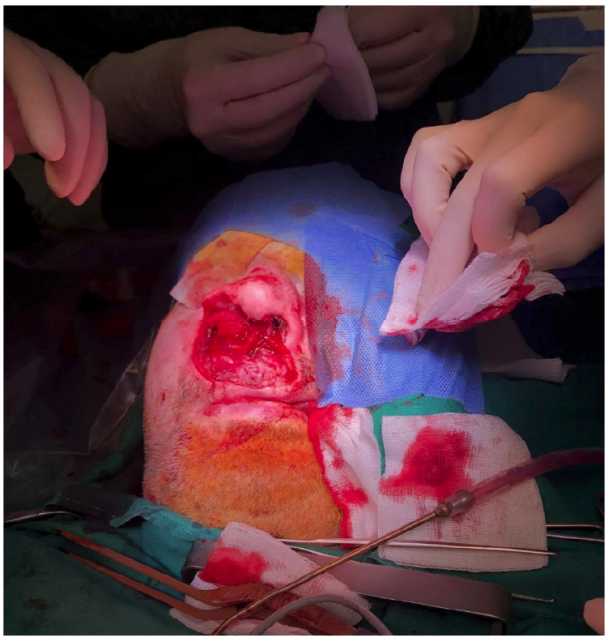

Figure 2: Clinical photography of soft tissue defect after radical tumour resection.

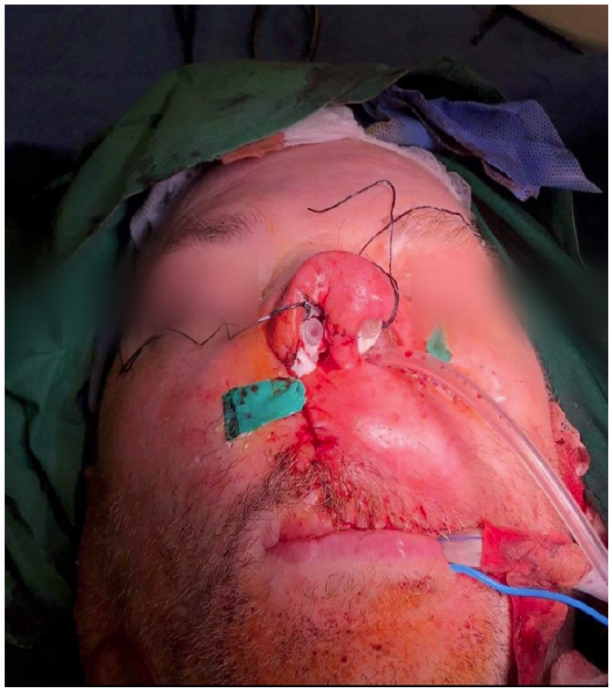

Figure 3: Clinical photography of soft tissue defect reconstruction by cheek rotation flap.

The patient did not receive any additional therapy after surgery because the resection margins were free of tumour and there were no signs of metastatic disease, and he was discharged with the diagnosis Ca vestibuli nasi T4aN1M0G2R0 stage IV.

DISCUSSION

Sebaceous carcinoma is a rare, aggressive adnexal carcinoma of sebaceous cells that often lacks clear cell features in poorly differentiated cases [6]. SCs are divided into two clinical types: extraocular and ocular [4,7]. The latter accounts for about 75% of all sebaceous carcinomas and comprise 1% of all eyelid neoplasms [4]. Extraocular tumours are more frequently associated with Muir-Torre syndrome than ocular sebaceous carcinomas [7]. Numerous studies have documented the presence of sebaceous carcinoma in the nasal area, however SC in the vestibulum nasi is extremely uncommon and has not been commonly reported in the literature [2]. The report presented here is an extremely rare skin cancer case.

Sebaceous carcinoma usually occurs in adults older than 60, with a mean age of 67.9 years [8]. There is a greater frequency of extraocular lesions in white ethnic group, and females tend to be affected more than males [8,9]. Additional risk factors for this cancer include UV exposure, pre-existing nevus sebaceous, Muir-Torre syndrome and immunosuppression [10].

The diagnosis of sebaceous carcinoma is established by incisional biopsy. The presence of sebaceous cells and fat in vacuolated tumour cells are the morphological indicators of sebaceous differentiation [2]. Common positive immunohistochemical markers include CK, EMA, CAM 5.2 and Anti-BCA-225 antibody [2,11,12].

Because sebaceous carcinoma can resemble benign growths, it can be difficult to identify in due course and diagnosis may be delayed, resulting in poor outcomes [13]. A clinical diagnosis of sebaceous carcinoma usually is delayed for approximately 15 months. Sebaceous carcinoma can present as a recurrent lesion previously misdiagnosed as BCC.

Cryotherapy, radiation therapy, surgical excision and Mohs micrographic surgery are available treatment modalities for sebaceous carcinoma [14]. Previous research found that patients who received radiotherapy as their main form of treatment had a higher mortality rate than patients who underwent extensive surgical

resection [9]. Individuals with this cancer have a 5 year overall survival rate of 78% for localized/regional disease and 50% for metastatic disease [15]. The poor prognostic factors have included a lesion size greater than 10 mm, delay in the diagnosis of longer than 6 months, multicentric origin, vascular, lymphatic, orbital, or pagetoid invasion, poor differentiation, and a highly infiltrative pattern [16,17].

It is common for the disease to reappear in the same area, and up to one-third of patients may encounter metastases in their regional lymph nodes [9,13]. The spreading of the SC to the draining nodes may indicate the disease's future systemic spread. Distant metastases are not uncommon, occasionally to the lungs, CNS, and visceral organs [13,17].

Owen *et al*. recommended a post-treatment clinical examination every 6 months for 3 years, which is complete skin and lymph node examination, to consider US of lymph node-positive patients [8].

CONCLUSIONS

Basal cell carcinoma with sebaceous differentiation is a very rare type of differentiation of a common tumor. This case highlights the diagnostic challenges of sebaceous gland carcinoma, especially in patients with extraocular disease. Pathologists should be aware of these histopathological variants to avoid misdiagnosis with clinical implications. Undoubtedly, histopathologic features remain the mainstay in differentiation and thus in selecting the correct treatment modality. Further investigation is needed to determine an appropriate long-term monitoring regimen for these patients.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

FUNDING SOURCES

None.

ACKNOWLEDGEMENTS

The authors would like to thank the Department of Otorhinolaryngology, Rīga Stradiņš University.

ABBREVIATIONS

SC	=	sebaceous carcinoma
MCA	=	middle cerebral artery
ECOG	=	Eastern Cooperative Oncology Group
PS	=	performance Status
CT	=	computed tomography
SCC	=	squamous cell carcinoma
UV	=	ultraviolet
CK	=	cytokeratin
EMA	=	epithelial membrane antigen
Anti-BCA-225	=	Anti-Breast Cancer Antigen 225
BCC	=	basal cell carcinoma
CNS	=	central nervous system
US	=	ultrasonography

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Received on 10-03-2023 Accepted on 05-04-2023 Published on 18-04-2023

https://doi.org/10.12970/2310-998X.2023.11.01