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CASE REPORT Open Access

Sebaceous carcinoma of nasal ala: a case report

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Abstract

Background Sebaceous carcinoma is an uncommon non-melanoma skin cancer. Extraocular sites are extremely rare, with only a few cases reported in the medical literature. This article reviews the diagnostic challenges and treatment of a case of sebaceous carcinoma of nasal ala.

Case presentation A 75-year-old female presented with a slow-growing right nasal alar mass for the last 1.5 years. It was nodular in appearance and covered with crusts and concretions. An incisional biopsy confirmed the diagnosis of sebaceous carcinoma. The patient underwent a wide local excision of the mass followed by nasolabial flap reconstruction.

Conclusions Sebaceous Carcinoma should be included in the differential diagnosis of an elderly patient with nasal growth to avoid delaying treatment.

Keywords Sebaceous carcinoma, Nasal mass, Muir-Torre syndrome, Extraocular, Non-melanoma skin cancer, Nasolabial flap

Background

Skin cancer is classified as either melanoma or non-melanoma skin cancer (NMSC) [1]. Sebaceous carcinoma is a type of non-melanoma skin cancer involving the adnexa. It is a rare, slow-growing but aggressive neoplasm that accounts for 0.2–4.6% of all cutaneous malignancies [2]. More than 70% of sebaceous carcinoma is located in the head and neck region [3]. However, Recent data suggest the following distributions: 38.7% periocular; 40.8% extraocular skin of head and neck; 19.9% others; 0.6% overlapping sites [4, 5].

Allaire first reported it in 1891, but Dr. Straatsma described it as a meibomian gland tumor in 1956 [1, 6]. The clinical diagnosis of the entity is difficult because of its ambiguous presentation. It can present as a non-healing ulcerating mass, a flesh-colored umbilicated papule, or even a subcutaneous nodule [1].

Risk factors include advanced age, Asian race, female gender, previous irradiation, and an immunocompromised state. It can also be associated with Muir–Torre syndrome [1, 4]. Diagnosis is made on tissue biopsy. Prognostic factors include multicentricity, poor differentiation, and lymphovascular invasion [1, 5].

Extraocular sebaceous carcinoma is highly uncommon, with the nose as a subsite being even more infrequent. Only 19 cases have been reported in the region of nose [7, 8]. Since it is a rare subset of NMSC, sebaceous carcinoma is often missed as a differential diagnosis of nasal mass. It is essential to consider it since the treatment and follow-up protocols differ from other malignancies. Here, we present a rare case of sebaceous carcinoma of the right ala of the nose in an elderly female.

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Case presentation

A 75-year-old female presented to the hospital with a history of progressively increasing nodular mass on the right nasal ala since 1.5 years. The patient also complained of occasional spontaneous bleeding from the mass, which was accompanied by pain and itching. She also mentions a decrese in appetite. She visited a local doctor, who excised the lesion and prescribed oral medications, which the patient continued to take for another 8 months. Unfortunately, the lesion recurred. She is a recently diagnosed hypertensive and is on antihypertensive drugs. There is no history of radiation exposure or any intake of immune-modulatory or chemotherapeutic drugs. The patient does not smoke or drink. There is no history of direct trauma or pus discharge from the site of the lesion. The patient does not have a past or family history of malignancy.

The general physical examination was unremarkable. Local examination of the right external nose depicted a single well-defined nodular mass of around 3 cm×3 cm with hemorrhagic crusts, ulcerations, and yellow-white concretions. The lesion was seen to be partially obstructing the right anterior nares (Fig. 1). On examination of the neck, there were no palpable lymph nodes. The oral cavity and oropharynx were normal on examination. No such similar lesion was seen on the body. A provisional clinical diagnosis of nodular basal cell carcinoma was made.

The patient was subjected to an incisional biopsy of the lesion under local anesthesia, in which a significant chunk of the lesion was incised, preserving the alar framework, and the sample was sent for histopathological examination.

The biopsy revealed that the patient had sebaceous carcinoma (Fig. 2). The tissue was positive for EMA (Epithelial Membrane Antigen) and CK7 (cytokeratin) on immunohistochemistry which confirmed the diagnosis.

With the diagnosis of sebaceous carcinoma of the right nasal ala, the patient was further planned for a metastatic workup to look for the spread in the regional lymph nodes, lung, and colon. CECT Neck, Chest X-ray, and abdominal ultrasound were found to be normal.

After careful consideration, the patient was managed surgically with wide local excision of the mass, leaving a full-thickness defect of the entire right ala and a part of the nasal dorsum (Fig 3). The defect was reconstructed with the help of a nasolabial flap (fascio-cutaneous flap)

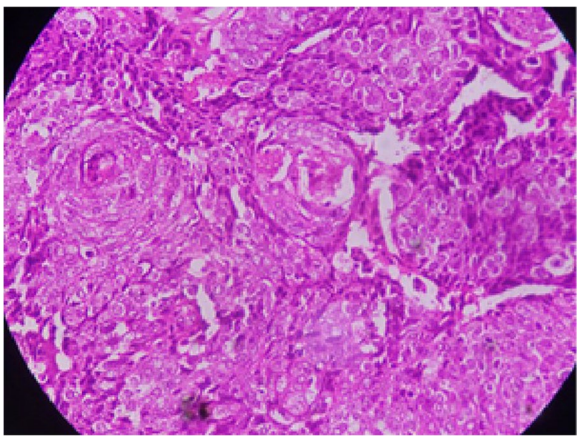

Fig. 2 Microspic picture showing sheets of multivacuolated cells separated by fibrovascularsepta, mitotic activity, nuclear pleomorphism, prominent nucleolus, and minimal necrosis

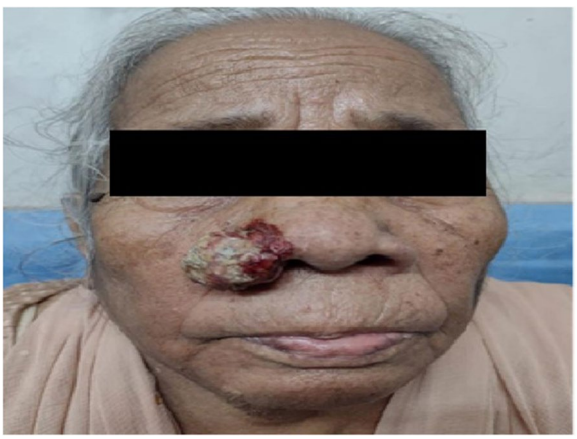

Fig. 1 Nodular mass (3 cm × 3 cm) with haemorrhagic crusts, ulcerations, on the surface, lesion seen to partially obstruct the right anterior nares

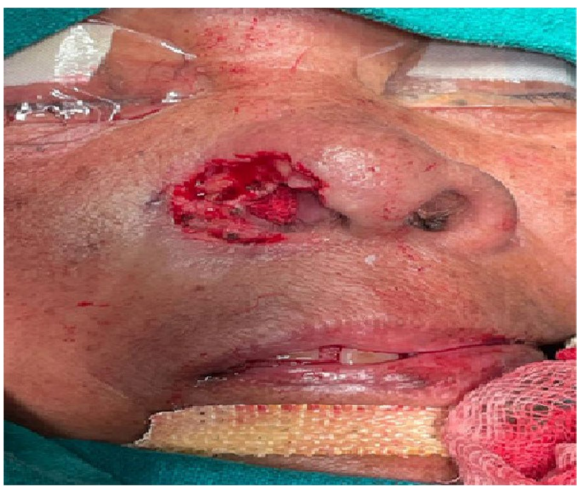

Fig. 3 Wide local excision of the mass, leaving a full-thickness defect of the entire right ala and a part of the nasal dorsum

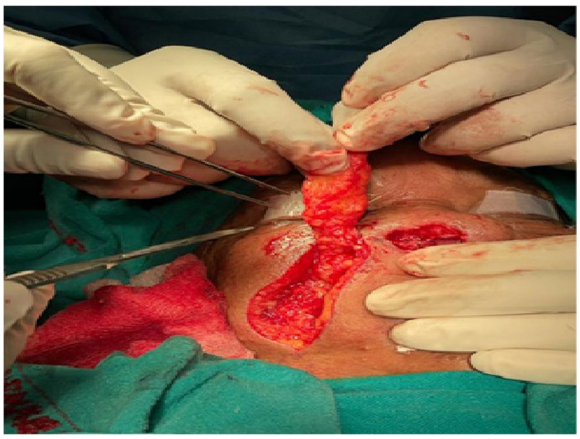

Fig. 4 Reconstruction with nasolabial flap

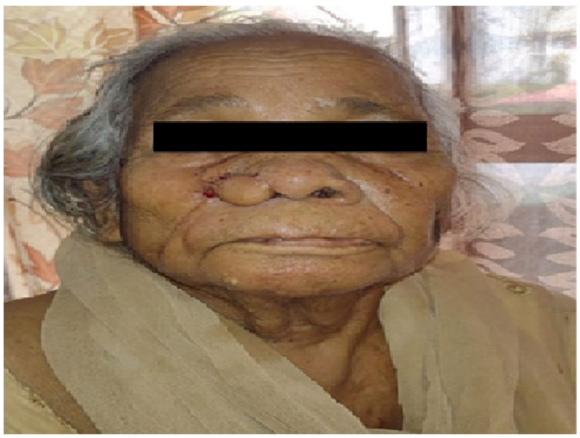

Fig. 5 Post operative picture–post-flap division

under general anesthesia (Fig. 4). The post-operative recovery was without any complications. The patient has been disease-free and under close follow-up for the last 1 year (Fig. 5).

Discussion

We presented a case of sebaceous carcinoma of nasal ala. It is a rare, slow-growing, and aggressive neoplasm. It is generally seen in old age with the median age of presentation at 72 years [4]. Clinically, it is divided into two types based on location: periocular and extraocular. The latter constitutes almost 1/4th of all sebaceous carcinomas [9]. Periorbital variety occurs mainly in Meibomian and Zeis glands.

Risk factors like old age, radiation exposure, or depressed immunity contribute to this neoplasm. Sebaceous carcinoma may be associated with Muir-Torre syndrome. It is an autosomal dominant condition with mutations in MSH2 and MLH1 genes and is characterized by the presence of internal malignancy (e.g., colon cancer), sebaceous neoplasms, and keratoacanthomas. It is critical to rule out this syndrome in patients with sebaceous carcinoma [9]. This is ideally done by screening for visceral cancers [1].

The tumor is difficult to diagnose, given its rarity and the extra-orbital location. The delayed diagnosis often results in increased morbidity and mortality. On average, the delay is about 1–2.9 years from the point of disease onset [10]. It was 1.5 years in our case.

The carcinoma presents as a slowly progressing painful nodular mass with a history of episodic bleeding. Sebaceous carcinomas often mimic benign conditions like pyogenic granuloma and molluscum. Other conditions include sebaceous adenoma, basal cell carcinoma with sebaceous differentiation, and clear cell squamous cell carcinoma [11]. Therefore, tissue biopsy is mandatory for a conclusive diagnosis.

On microscopy, the basaloid neoplasm was visible in lobules separated by a fibrovascular stroma [11]. The morphological hallmark of sebaceous carcinoma are sebaceous cells, which are epithelial cells with multiple clear vacuoles containing fat which is easily stained using oil red O stain [11, 12]. Immunohistochemical staining for EMA (epithelial membrane antigen), CK (cytokeratin), adipophilin, and androgen receptor will be positive for cells with sebaceous differentiation. There is also increased expression of p53 and Ki67 [11].

Extraocular sebaceous carcinomas have a recurrence rate of 4–29 % and a metastatic rate of 21% [11]. There is no difference in survival patterns of both periocular and extraocular types [1]. Factor like multifocality, lymphovascular invasion, tumor size > 10 mm, and pagetoid spread have bad prognoses. Furthermore, tumor thickness > 2 mm, Clark level ≥ IV, perineural invasion, the primary site of the ear or lip, and poorly differentiated or undifferentiated tumors are all high-risk factors [13].

Treatment choices for the management of sebaceous carcinoma include wide local excision with a 5–6 mm margin or MMS (Moh's micrographic surgery) [11]. MMS is supposed to have a low recurrence rate [11]. Reconstruction after excision is tricky in the case of the large nasal lesion as we have to cater to both functional and aesthetic aspects, and it can be done using a nasolabial or forehead flap [14]. Because extraocular sebaceous carcinomas have a 1.4% risk of regional lymph nodal metastasis, sentinel lymph node biopsies can be done to detect early spread, but it is more useful in the periocular variety [11]. Adjuvant therapy like radiation and chemotherapy can also be helpful in case of surgical contraindication [11]. Close follow-up is necessary given the high local recurrence. It is essential to consider this rare tumor while investigating a nasal mass to prevent any delay in treatment. Early diagnosis and prompt treatment are

shown to have reasonable survival rates and excellent outcomes.

Conclusion

Although a rare malignancy, as discussed above, sebaceous carcinoma should be included in the differential diagnosis of non healing ulcerating mass. Cases should be investigated with tissue biopsy and local imaging, for assessing the extend and spread of the tumour. Primary modality of treatment is surgical excision with reconstruction of the defect.

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Authors' contributions
AC, PN, PS, and SMS were involved in management of the case. SMS and PN wrote the 1st draft of paper. PS and AC did review and made the final draft of paper. KA was responsible for he pathology slides and its images. All authors take responsibility for the authenticity of case reported.

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Availability of data and materials
All images have been submitted with the manuscript.

Declarations

Ethics approval and consent to partcipate
Ethics approval is not applicable for the case report.

Consent for publication
A signed consent was taken by the patient for use of the image and her case history for publication.

Competing interests
The authors declare that they have no competing interests.

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