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Case Report

Sebaceous carcinoma of nasal ala: a case report

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ABSTRACT

Nasal sebaceous carcinoma is an extremely rare adnexal skin tumor representing 0.2%-4.6% of all malignant neoplastic cutaneous lesions. It is seen most affecting the meibomian glands in the periocular region. Here we share a case of 65-year-old female patient with nasal mass over the right alar region, diagnosed as sebaceous carcinoma and managed with wide local excision of the mass was done defect was reconstructed with a nasolabial flap. Sebaceous carcinoma being very rare in nasal region we want to share the case details.

Keywords: Nasal ala mass, Sebaceous carcinoma, Adnexal tumor, Nasolabial flap

INTRODUCTION

The skin malignancies are classified as nonmelanoma and melanoma. The nonmelanoma skin cancers are frequently diagnosed entity common in head and neck region. Of these 75% are basal cell carcinoma, 20% squamous cell carcinoma and 5% include other adnexal carcinoma, Merkel cell carcinoma, dermatofibrosarcoma and sebaceous carcinoma.[1] Sebaceous carcinoma is rare and aggressive cutaneous carcinoma.[3,10,11]. It is first described in 1891 by Allerire, cases of sebaceous carcinoma can be grouped as originating from the periorbital meibomian and zeis glands and those from extraocular sites.[1,9] Periorbital sebaceous carcinoma most common 38.7% comprising 1 to 5.5% of all malignant eyelid neoplasms.[1,5] Sebaceous carcinoma of eyelid known as The great Masquerade often mimics chalazion, intradermal nerve and blepharitis.[5] Extraocular sebaceous carcinoma of head and neck can involve the face, scalp and neck, ear and lip but rarely seen involving the nasal ala. Extraocular sebaceous carcinoma has been associated with 29% recurrence rate and 21% metastatic rate.[1,4] The exact cause of sebaceous carcinoma is unknown although number of pathways and genes such as PTEN, p53 are implicated.[1,5] The predisposing risk factor for sebaceous carcinoma include advancing age, any prior exposure to ionizing radiation in head and neck, immunosuppression, Muir Torre syndrome, familial retinoblastoma.[1,2,5,10] Patient presents most commonly with a gradually enlarging, painless, heterogenous lesion range from flesh coloured, umbilicate papules to subcutaneous nodules or tumour with exophytic growth.[1,4,10] The diagnosis of sebaceous carcinoma can be complicated by the ambiguous clinical presentation in that they resemble the most common skin malignancies such as basal cell carcinoma.[4]

CASE REPORT

A 65-year-old female presented with a complaint of growth over right ala region since, 4 months which was rapidly increasing in size. Patient is a known diabetic from past 14 years. She denied any history of pain, epistaxis, weight loss, nasal obstruction, loss of sensation over the face. On physical examination revealed an exophytic growth with crusting and necrotic debris, firm

lesion arising from the right alar rim (Figure 1). On anterior rhinoscopy examination nasal cavity was normal, devoid of any mass lesion (Figure 1). There is loss of sensation over the mass lesion and is not bleeding on touch. No palpable lymphadenopathy was noted. Patient was taken up for excision biopsy in view of any malignant lesions, after required fitness were attained. Intraoperative frozen section of the excised specimen was sent which was reported as basal margin shows infiltration of tumor cells around the skeletal muscle bundles up to resected margin. We proceeded with wide local excision of mass (Figure 2,3,4) and sent the complete specimen for histopathological examination (Figure 5,6). The defect in the surgical site was reconstructed using nasolabial rotation flap. The sutures of the reconstructed site were removed after 10 days (Figure 7) The nose was aesthetically normal on further follow up.

Figure 1: Right nasal alar exophytic mass with crusting.

Figure 2: Resultant surgical defect of the right nasal alar region.

Figure 3: Intraoperative picture showing the elevation of the nasolabial rotation flap along with its vascular continuity.

Figure 4: Intraoperative picture of the surgical site after reconstruction of the right nasal alar region.

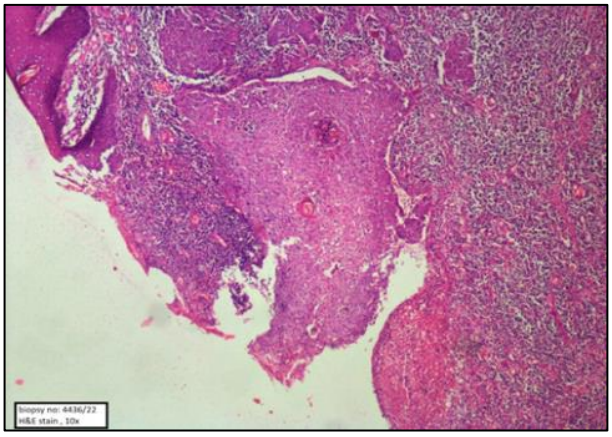

Figure 5: H and E stain in 4X showing hyperplastic stratified squamous epithelium with ulceration, tumour cell nests with lobules and sheets.

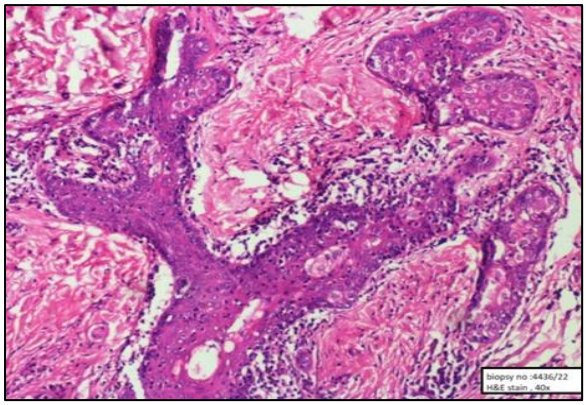

Figure 6: H and E stain 40X showing cells with vacuolated cytoplasm, large nucleus and nucleoi with granular cytoplasm.

Figure 7: The postoperative nasal alar region after suture removal.

DISCUSSION

Sebaceous gland malignancies represent one of the rare and potentially aggressive types of skin neoplasms. Sebaceous carcinoma originates in the holocrine adnexal epithelium of sebaceous glands.[10] Extraocular sebaceous carcinoma forms less than 25% of all sebaceous carcinoma and occurs predominantly in the head and neck area where the concentration of sebaceous glands is higher. Nasal sebaceous carcinoma is an extremely rare adnexal tumour with few cases reported so far.[4] Dasgupta et al reported a case of sebaceous carcinoma of nasal vestibule out of 1349 cases of sebaceous carcinoma in the head and neck region. Murphy et al reported another case of right nasal vestibule sebaceous carcinoma.[3,6,10]

The risk factors for sebaceous carcinoma include advancing age, previous radiation exposure and Muir-Torre syndrome.[5] Muir-Torre syndrome is an autosomal dominant condition with variable penetrance, consists of sebaceous carcinoma with colon cancer, adenomas and epitheliomas as well as multiple or early onset keratoacanthomas. In the present case its unlikely to be associated with Muir-Torre syndrome due to absence of family history and lesions in other locations like colon.[2,11]

Sebaceous carcinoma is a rare entity of nose, usually presenting as gradually enlarging, firm nodule, mimics common dermatological conditions like basal cell carcinoma.[6] Histopathologically, the cells show vacuolated cytoplasmic cells with PAS negative.[10] Immunohistochemical staining with EMA and Cytokeratin, helps distinguish sebaceous carcinoma from other similar appearing lesions.[2,4,5] Treatment modalities for sebaceous carcinoma include cryotherapy, radiotherapy and surgical excision.[3] Standard wide local surgical excision or Mohs micrographic surgery are common treatment.[1,6,9] Post surgical excision, the defect can be reconstructed using facial flaps like nasolabial rotation flap as done in our case. The face and nasal area have rich vascular supplies with various anastomoses, hence are good areas for flap reconstruction.[4,7]

Sebaceous carcinoma spreads either through direct extension, perineural involvement or lymphovascular invasion6. The most common sites of metastasis are the preauricular and parotid lymph nodes.[1,4,5] Reports show that the extraocular type of sebaceous carcinoma has metastatic rate of 21%. Sawyer et al recommended sentinel lymph node biopsy due to the aggressive behaviour of extraocular sebaceous carcinoma.[8] Prognostic factors include lymphovascular invasion, multicentric diseases and tumour differentiation.[5] Radiation therapies hold high mortality rates than those treated surgically with free wide margin. Hence surgical excision with clear margins of normal tissue in all directions with frozen section of margins is the treatment of choice.[3] In a retrospective study of 1349 cases Dasgupta et al found no difference in survival of patients with periorbital and nonorbital disease.[6] Local recurrence affects 30% to 40% patients while distant metastasis occurs in 20% to 25% patients. The tumour has a high tendency for local recurrence postoperatively and metastasize, needs to be treated in an aggressive manner with careful follow up.[2,6]

CONCLUSION

Sebaceous carcinoma in the head and neck region is not infrequent presentation, but as nasal mass is rare. Sebaceous carcinoma presenting as nasal ala should be treated with wide local excision of the lesion with free margins. Regular follow up of the patient should be done as the tumour has high propensity for recurrence and metastasis.

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