

Sebaceous Cell Carcinoma of Face- A Rare Presentation

Ruchi Khadayate¹, Reeta Dhar², Siddhi Karale¹

¹Second year Post- Graduate student, ²Professor,
Department of Pathology, MGM Medical College & Hospital, Kamothe, Navi Mumbai

Corresponding Author: Reeta Dhar

ABSTRACT

Sebaceous carcinoma is a very uncommon malignancy of Sebaceous glands which forms the dermal adnexal unit. They usually occur singly, in and around orbits. Hereby, presenting a case of 80 yr old male with two swellings, one at forehead and other at the jawline, both on the left side of the face, swellings were painless, firm, fixed, globular, exophytic. They were surgically resected under local anesthesia.

Keywords- Sebaceous carcinomas, Face, Appendageal malignancy, Extraocular Sebaceous carcinomas

INTRODUCTION

Sebaceous carcinoma (SC) is one of the rare dermal appendageal malignancies. It is characterized by aggressive behavior of the tumour ranging from local invasion, recurrence, to metastasis to draining lymph nodes and distant organs. ⁽¹⁾ Frequency of metastasis and mortality has been reported to be as high as 25% in some series. ⁽²⁾

SC is most commonly seen in the periocular region commonly eyelid. Extra ocular SC is very rare. The periocular region consists of 5 different kinds of sebaceous glands. So far only 150 cases of extra orbital SC were reported in the literature. ⁽³⁾ The extra ocular SC commonly occurs on head and scalp due to the abundant sebaceous glands ⁽⁴⁻⁵⁾ the other sites involved by SC are external genitalia ⁽⁶⁾ and extremities. ⁽⁷⁾

The most common clinical presentation is the painless, round subcutaneous nodule or as an irregular, ill defined mass with extensive tissue invasion. ⁽¹⁾ Due to benign behaviour and varied presentation of the lesion, it leads to misdiagnosis or late diagnosis. ⁽⁸⁾

CASE REPORT

A 80 year old, male, came to surgery OPD of the MGM hospital complaining of swelling on the left side of forehead, above the eyebrows and another swelling on jawline of left side since 2 years, and it was small to start with and gradually increased to present size.

On Inspection-

The swelling on the forehead was single, dense, firm, brownish, exophytic measuring approximately 3 x 3x 2cm with irregular surface. No ulceration seen. Another swelling on the jawline was soft to firm, reddish, globular measuring 2x2x1.5 cm with relatively smooth surface.

On Palpation-

The swellings were non-tender, local temperature was not raised, fixed to underlying tissue, non-compressible, non-fluctuant, non-pulsatile. After examination, clinical diagnosis of sebaceous horn was made and workup for excisional biopsy under local anaesthesia was planned.



Figure 1. Photograph shows the hard, protruding mass on the forehead and relatively softer looking, fleshy mass on the chin on the left side of the face.

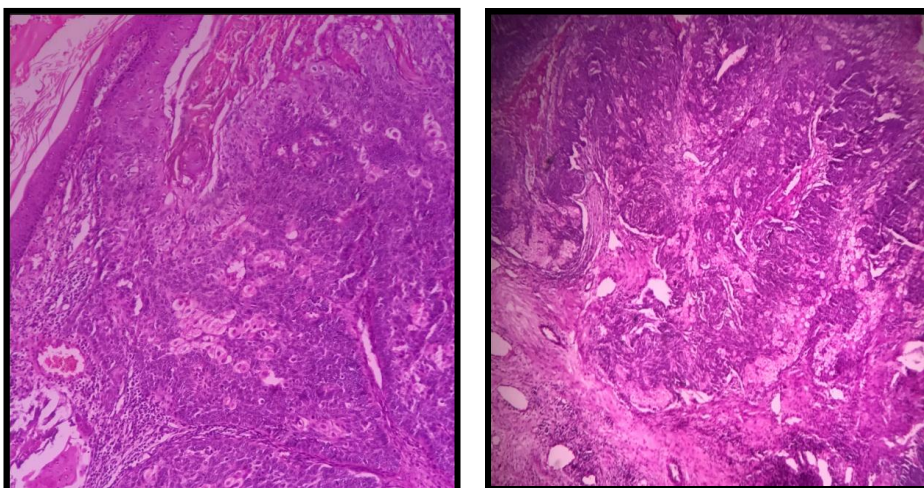


Figure-2- Micrographs show lesion with epidermis and dermis. The dermis shows extensive infiltration by the tumour cells forming lobules(H&E stain-4x).

The excision of the tumour was done and the resected tissue was sent to the department of pathology for histopathological examination

Gross findings-

Received two grey-brown, soft to firm, skin covered tissue bits largest measuring 4x3x2 cm and smaller measuring 3x2x2cm.

Microscopic Findings-

Sections studied show tumour comprising of irregular, lobulated masses of

round to oval to basaloid cells with round nuclei infiltrating the dermis as well as epidermis. Numerous atypical cells with sebaceous differentiation showing vacuolated cytoplasm with centrally placed nuclei. Tumour cells showed nesting, pearly and papillaroid arrangement. Few cells were spindle shaped with variable eosinophilic cytoplasm. Few lobules revealed central areas of necrosis (comedo necrosis).

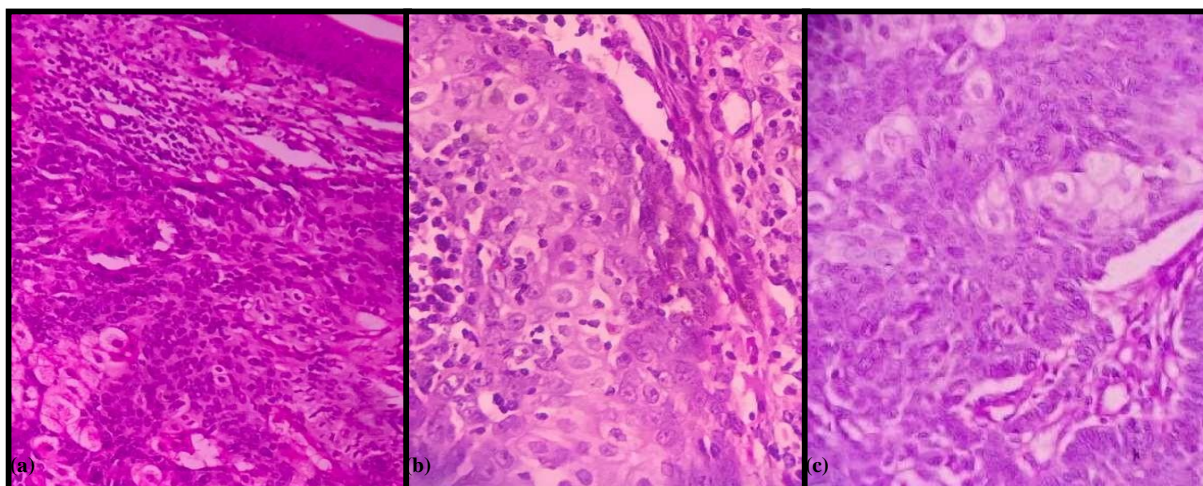


Figure-3- (a)Micrographs shows compactly arranged tumour cells in lobules infiltrating dermis as well as epidermis.(H&E stain 10x)
(b)Micrographs (40X) shows tumor cells with round to oval to basaloid cells with round nuclei. Numerous atypical cells with sebaceous differentiation showing vacuolated cytoplasm with centrally placed nuclei. Lobule shows central areas of necrosis (comedonecrosis).(H&E stain 40x)
(c)Micrographs (40X) shows moderate pleomorphism and nuclear atypia, high N:C ratio and hyperchromasia. Abundant mitotic figures were seen.(H&E stain 40x)

Individual cells had moderate pleomorphism and nuclear atypia, high N:C ratio and hyperchromasia. Abundant mitotic figures were seen. Stroma was infiltrated with sheets of lymphocytes, plasma cells, neutrophils with extensive areas of necrosis. The skin appendageal structures were seen to be infiltrated by the tumor. Based on the aforementioned findings, a pathological diagnosis of Malignant Skin appendageal tumour suggestive of Sebaceous Carcinoma was made.

DISCUSSION

Sebaceous carcinoma is a disease of the sixth and seventh decade of life. It presents as a firm, painless slow growing mass. The most frequent extra-orbital site for this tumour is skin in the head and neck region because of abundance of sebaceous glands. ⁽⁹⁾ The tumours of sebaceous glands consist of three major categories: a) Sebaceous Adenoma b) Basal Cell Carcinoma with sebaceous differentiation c) Sebaceous Carcinoma. ⁽¹⁰⁾ It is known to be associated with Muir-Torre syndrome, a clinical subset of hereditary non polyposis colorectal cancer syndrome. Muir-Torre syndrome is an autosomal dominant disorder characterized by multiple sebaceous gland neoplasm viz. sebaceous adenoma, sebaceous carcinoma, sebaceous

epithelioma along with multiple low grade visceral malignancy. ⁽¹¹⁾

Histologically, the most characteristic feature is sebaceous differentiation. The cells show high mitotic activity, nuclear pleomorphism, lobular architecture and foamy vacuolization of the cytoplasm. ⁽¹²⁾

Tumor cells in sebaceous carcinoma are often large and may show squamoid changes and that is why it is needed to be differentiated from squamous cell carcinoma with hydropic changes. Sometimes, tumor cells show basaloid differentiation with inconspicuous lipidization, and the tumor must be distinguished from basal cell carcinoma with sebaceous differentiation. ⁽¹³⁾ The sebaceous carcinoma gives immunoreactivity for keratin, cytokeratin, EMA, Leu-M1(CD15), androgen receptor and adipophilin whereas CEA and S-100 are negative. ^(14,15) Poor differentiation, infiltrative growth pattern, multicentric origin of tumour, delay in diagnosis of over 6 months and size of tumour more than 10mm have been postulated as poor prognostic indicators. ⁽⁹⁾

Various case reports have mentioned local recurrence, aggressive nature of the tumour with nodal, meningeal, bone and lung infiltration. ^(9,11,12,16) Ocular Sebaceous

Carcinoma (eyelids, caruncle and orbits being the common locations) exhibit more aggressive and unfavorable clinical course as compared to the tumours located elsewhere. (5,6,12)

With regard to present case, no previous history, family history or history of any visceral malignancy was noted.

The treatment of choice is complete surgical excision with wide margins. The role of adjuvant radiotherapy and chemotherapy is not clear in extraocular SC due to rarity of cases. As only 150 cases being reported in the literature so far, unanimous opinion regarding aggressive nature of the Extraorbital SC tumour is not well established. (16)

CONCLUSION

We conclude that Extraocular Sebaceous carcinoma is as rare aggressive neoplasm which needs to be followed up closely to assess the recurrence and distant metastasis after the surgery and also for additional treatment to be given on the basis of various prognostic features.

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