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*Case Report*

# Sebaceous Carcinoma

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## ABSTRACT

Carcinoma is an extremely rare malignant tumour of sebaceous glands. Extra ocular variant is still more rare compared to its ocular counterpart accounting for only one fourth of all cases. Delay in diagnosis is common as it mimics a variety of tumours, both benign and malignant, however it is aggressive. Treatment is by surgery in most of the cases.

Here we present a case of 39 year old female with sebaceous carcinoma of four months history in the right arm. She underwent disarticulation of the joint for the same and recovered favorably. She had earlier underwent trans-hiatal oesophago-gastrectomy for squamous cell carcinoma involving lower third of oesophagus, one year back.

Both the tumours having the characteristic epidermal component is interesting.

***Key Words:*** Sebaceous carcinoma, epidermal carcinoma.

## INTRODUCTION

Sebaceous carcinoma is a rare tumour arising from adnexal epithelium of sebaceous glands. They are more common in the periocular region and mainly affect Meibomian glands of eyelids. Extra ocular incidence is rare and is associated with significant delay in diagnosis due to resemblance with other tumours like benign sebaceous neoplasms, other adnexal tumours and basal cell carcinoma. The tumour is multicentric and aggressive with a propensity for vascular and perineural invasion. Metastasis by lymphatics is seen and involves regional lymph nodes and distant spread to other organs.

Diagnosis is by biopsy. Surgery remains the primary modality of treatment with negative tumour margins. Advanced cases are treated with Radiation therapy, chemotherapy or combination therapy. Recurrence after negative control is common and is associated with poor prognosis.

## CASE REPORT

A 39-year-old female came with a swelling in the right arm since 4 months. It was insidious in onset and rapidly growing. There was no history of trauma or pus discharge. On 1examination the swelling was tendermeasuring20X15X8cms.

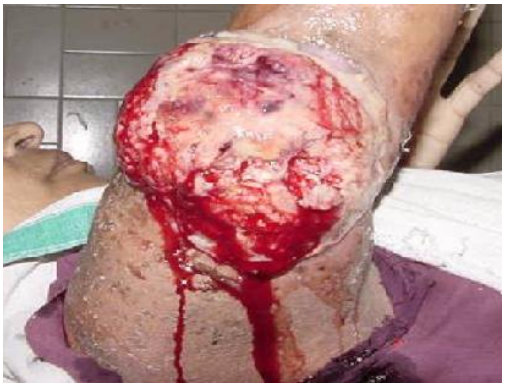

**FIGURE-1**

It was firm to hard in consistency and fixed to the underlying humerus. There was also bleeding from the swelling (Figure-1) However, there were no cervical

lymph nodes and no evidence of any secondaries.

Biopsy revealed clear cells, with clear cytoplasm and abundant mitotic figures along with lymph nodes and capsular integrity (figure 2). A diagnosis of sebaceous carcinoma was made and she underwent shoulder disarticulation for the same. The patient had uneventful recovery.

The same patient was admitted one year back with one month history of progressive dysphagia for solid food. Being diagnosed as squamous cell carcinoma lower third of oesophagus, with no secondaries, she underwent Trans hiatal oesophago-gastrectomy, with uneventful recovery. The then biopsy showed squamous cell carcinoma with epithelial pearls (figure 3)., lymph node metastasis and muscle invasion (figure 4).

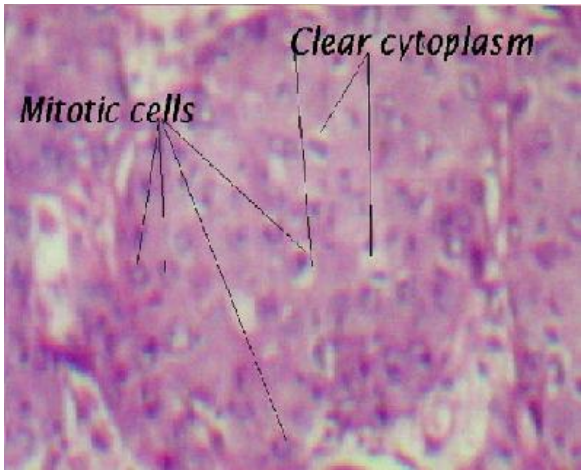


**FIGURE-2**

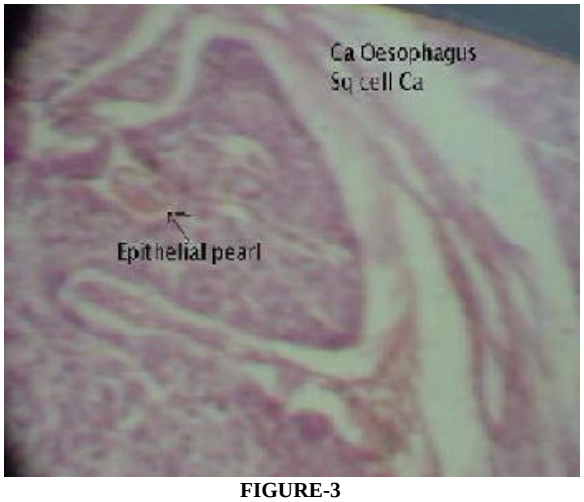


**FIGURE-3**

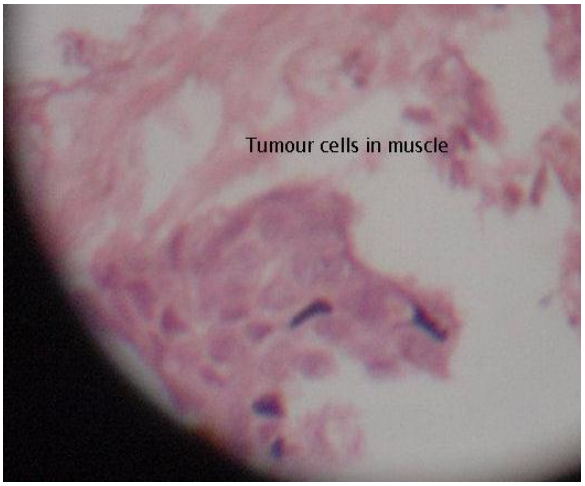


**FIGURE-4**

## DISCUSSION

Sebaceous glands occur throughout the hair bearing regions of the body and is closely associated with hair follicles, forming a component of pilosebaceous apparatus. Sebaceous glands are derived from embryonal stratum germinatum, the one which gives rise to epidermis and epidermal appendages. They secrete sebum.

Sebaceous carcinoma is a rare, aggressive and malignant tumor derived from the adnexal epithelium of sebaceous glands. The prevalence ranges from 0.05% to 0.7% of all skin cancers. [1] Majority of tumours are seen in ocular region and extra ocular sebaceous carcinoma contribute only 25% of the cases, 15% affecting the torso and 10% involving the extremities. [2]

All aire documented the first case report of ocular sebaceous carcinoma in 1891 though cases were noted as early as 1865 by thiersch. [3,4] Earlier it was considered a variant of basal cell carcinoma until Straatsma described 16 patients with detailed histological and clinical presentations in 1956. [5]

Extra ocular sebaceous carcinomas are very uncommon and usually arise from head and neck owing to abundance of sebaceous glands [6] constituting about 25 % of all sebaceous carcinoma cases. [7] Other reported sites include external genitalia, parotid and sub mandibular glands, external auditory canal, the trunk and upper extremity, sole and pharyngeal cavities. [8]

Sebaceous carcinoma appear to have a higher incidence in Asian population. [10] Males and females are equally affected with extra ocular sebaceous carcinoma unlike the ocular variety which has a slight female preponderance, though both have the mean age of occurrence at 63 years. [11]

The cause of most cases of sebaceous carcinoma is unknown. A few have been found to be associated with Muir-Torre syndrome, an autosomal dominant genodermatosis in which patients have low grade internal malignancies and at least one sebaceous tumour, [12] history of previous radiation in the area of tumour [13] and diuretic use has weak associations in sebaceous carcinoma. [14]

Clinically the size ranges from 6mm to 20 cm. [9] Clinical appearance of the tumour is not pathognomonic, bleeding has been reported in approximately one third of cases and the tumour is multicentric. Histologically, Sebaceous carcinoma is dermally based and non encapsulated. [15,16] Diagnosis is by biopsy.

About 14-25% of cases involve regional lymph nodes followed by spread to liver, lung, brain and bones. [17]

Several studies have demonstrated correlation between histological pattern and prognosis. The characteristic four features associated with poor prognosis include multi-centric origin, poor sebaceous differentiation, intra epithelial pagetoid distortion of the overlying epithelium and highly infiltrative growth pattern apart from vascular invasion.

The primary modality of treatment is surgery i.e. surgical excision with wide margins. Multicentricity of the tumour and pagetoid spread stands in the way to achieve a negative margin. Radiotherapy has been tried with curative intent in post operative management of metastatic disease, palliative care and has been giving good results in local control of disease though studies are in sufficient to use as primary treatment. [18]

Chemotherapy has been tried for the treatment of recurrent sebaceous carcinoma with intralesional 5-flourouracil and intravenous doxorubicin and 5-flourouracil with subsequent cisplatin and vinblastine for three months. [19] Radical neck dissection is done for regional node metastasis. Cryotherapy has been tried as a new modality for intraepithelial neoplasia with inadequate studies. [20]

## CONCLUSION

Sebaceous carcinoma is an extremely rare aggressive tumour and needs high index of suspicion in diagnosis. Early treatment can benefit the patient and reduce significant morbidity. Here in this patient it was managed surgically by disarticulation of the proximal joint to achieve tumour free margin.

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