Indian Journal of Cancer Education and Research
Volume 1 Number 1, January - June 2013

Sebaceous Carcinoma of the Extremity: A Case Report

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(Recieved on 16.05.2013, Accepted on 28.06.2013)

Abstract

Sebaceous Carcinoma is an uncommon, aggressive malignant tumor derived from the adnexal epithelium of sebaceous gland either from ocular or extra ocular sites. Extra ocular sites being very uncommon and forms one-fourth of all sebaceous carcinomas. Patients presenting in extremity are rarest and have a high mortality rate. Here we present one case which presented with multiple orange colored nodular lesions over dorsum of right hand, it recurred and progressed very fast after surgery and no response was seen with chemotherapy.

Keywords: Upper extremity; Sebaceous carcinoma.

Introduction

Sebaceous carcinoma is an aggressive tumor derived from the adnexal epithelium of sebaceous glands and accounts for 1% of all cutaneous malignancies. These glands have a wide spread distribution in the skin but the tumor arises mainly in ocular adnexa and less commonly in extra ocular sites. Extra ocular sebaceous carcinoma comprises only 25% of all sebaceous carcinoma[1] involving mainly the head and neck region in which sebaceous glands are in plenty followed by external genitalia, parotid and submandibular glands, external auditory canal, trunk, upper extremity, sole and laryngeal and pharyngeal cavities in this order.[2] It may also occur in Muir Torre Syndrome (MTS) characterized by occurrence of sebaceous tumors in association with visceral malignancies.[3] A case of rapidly growing right upper extremity Sebaceous Carcinoma is presented here.

Case history

A 65 years old female presented with a non tender swelling over dorso-medial aspect of proximal interphalangeal joint of right Index finger. Excision of this swelling was done and the histology was inconclusive. She presented two months later with a recurrence and had developed multiple orange colored nodular lesions over dorsum of right hand and in the web between index and middle finger (Fig 1). These nodules were firm to hard and fixed to skin. Her past medical history was unremarkable and there was no family history of similar lesions or any other malignancy. Excision biopsy of one nodule was done which showed groups and strands of finely vacuolated and foamy cells showing clear to eosinophilic cytoplasm infiltrating in the stroma (Fig 2, 3). Based on the clinical picture and histology a diagnosis of sebaceous carcinoma of right upper extremity was established. The patient presented one month after excision with progressive disease and pain in arm. She has developed one 2x2 cm firm orange colored nodular growth over lateral aspect of right forearm and 3 to 4 other orange subcutaneous nodules over lower medial aspect of right arm. There was a 3x3 cm firm, mobile right axillary lymph node (Fig 4). Other lymph node was not palpable. A thorough investigation including CT Scan thorax and abdomen, triple endoscopy and colonoscopy

Fig 1: Multiple orange colored nodular lesions over dorsum of hand

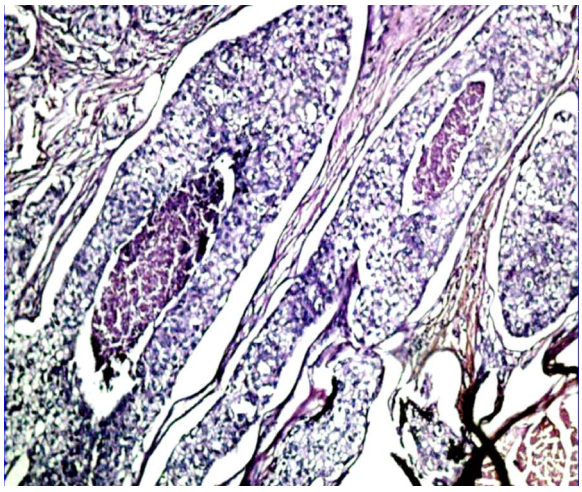
done were within normal limits. As patient denied surgery she was started on with palliative chemotherapy with which she had pain control but there was no response to chemotherapy after three cycles combination chemotherapy with cisplatin75mg/m^2 day 1 and Ifosphomide 1.2 gm/m^2 day 1 to 3 given every three weeks. The patient tolerated the chemotherapy well and did not have any haematologic or bladder toxicity. Later the patient was lost to follow-up.

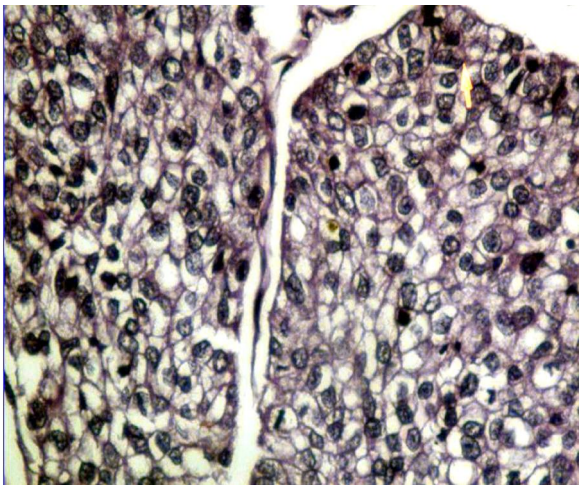
Discussion

Extra ocular sebaceous carcinoma involving the extremity is very uncommon, aggressive malignant tumor arising from sebaceous glands. Mean age of occurrence is 63 years involving both sexes in equal proportion. The prognosis depends on size, color and site of the lesion. Ocular lesions more than 1cm with red flags have unfavorable prognosis.[2] Our patient also presented at 65 years age and the larger lesions showed similar poor prognosis. The disease exhibits a variety of clinical presentation that the diagnosis is often delayed for months to years.[4] It may appear on the top of pre-existing dermatoses like naevus sebaceous and actinic keratosis or may follow radiation therapy received for other diseases. [5,6,7] It may also occur in MTS, which is characterized by occurrence of sebaceous tumor in association with visceral malignancies.[3] Although our patient did not

Fig 4: Orange colored nodular growth over right forearm and 3 to 4 other orange subcutaneous nodules over arm

Fig 2 and 3: Groups and strands of finely vacuolated and foamy cells showing clear to eosinophilic cytoplasm infiltrating in the stroma (Mag 10x & 40x)





have a positive family history or presence of any internal malignancy.

The lesions usually present as pink to red yellow nodular growth in skin and may clinically resemble pyogenic granuloma, haemangioma or squamous cell carcinoma. The draining lymph nodes may be involved in few cases only.[8] Ghosh et al also found a moist, yellowish pink cauliflower-like oval shaped growth on the pinna with mild bleeding and some purulent discharge.[4]

Regardless of the location this malignancy is highly aggressive with a potential for regional and distant metastasis. Our patient also presented with orange nodular lesions and progressed very fast involving the lymph nodes, although no distant metastasis was seen. There were no signs of internal visceral malignancy.

Criterion for the diagnosis of MTS includes the presence of at least one sebaceous adenoma, epithelioma or sebaceous carcinoma and at least one visceral cancer in the absence of other participating factors such as Radiotherapy and AIDS.[9] However Immunohistochemistry, an important aid to the diagnosis of MTS could not be performed on the tumor in our patient due to local non-availability of the facility and patients financial constraints.

Treatment of sebaceous carcinoma requires wide local excision with removal of involved regional lymph nodes. But Nelson showed that chances of local recurrence are very high as is seen in our patient also.[2] Bailet reported a review of 92 patients with extra ocular sebaceous carcinoma and found a recurrence rate of 28% and metastasis in 21% of cases after local excision.[10] Radiation therapy has been considered as an adjunctive or palliative treatment but is generally not recommended as a primary treatment. The role of chemotherapy has not been defined due to scarcity of these lesions.

Due to multicentricity of the lesion we started the treatment with chemotherapy, as wide local excision was not possible. The patient has a partial response with the first cycle of chemotherapy using cisplatin and Ifosphamide combination. After three cycles of chemotherapy we did not see any response in the tumor instead it progressed and involved the skin of chest wall although till last follow up there were no distant metastasis.

References

1. Al-Shobaili, AlGhamdi KM, Al-Ghamdi WA. Cystic Sebaceous carcinoma: is it a constant patognomonic marker for Muir- Torre Syndrome? *J Drugs Dermatol*. 2007; 6(5): 540-543.
2. Nelson BR, Hamlet KR, Gillard M, Johnson TM. Sabeceous carcinoma. *J Am Acad Dermatol*. 1995; 33(1): 1-15.
3. Graham RM, Mckee PH, Mcgibbon D. Sebaceous carcinoma. *Clinical Exp Dermatol*. 1984; 9: 466-471.
4. Sudip Ghosh, Debabrata Bandyopadhyay, Sandip Gupta. Rapidly growing extra ocular sebaceous carcinoma occurring during pregnancy: A case report. *Dermatology Online Journal*. 14(8): 8.
5. Mastuda K, Doi T, Kosaka H, Tasaki N, Yoshioka H, Kakibunchi M. Sebaceous Carcinoma arising in nevus sebaceous. *J Dermatol*. 2005; 32(8): 641-644.
6. Ansai S, Mihara I. Sebaceous carcinoma arising on actinic keratosis. *Eur J dermatol*. 2000; 10(5): 385-388.
7. Tchornobay AM, Levigne V, Favre J, Claudy AL. Extraocular sebaceous carcinoma. *Acta Derm Venereol*. 1992; 72(1): 20-21.
8. Jensen ML. Extra ocular sebaceous carcinoma of the skin with visceral metastasis: case report. *J Coetaneous Pathol*. 1990; 17: 117-121.
9. Pettey A, Walsh J. Muir-Torre Syndrome: a case report and review of the literature. *Cutis*. 2005; 75: 149-155.
10. Bailet JW, Zimmerman MC, Arnstein DP, Wollman JS, Mickel RA. Sebaceous carcinoma of the head and neck. Case report and literature review. *Arch Otolaryngol Head Neck Surg*. 1992; 118(11): 1245-1249.