Extraocular sebaceous carcinoma of neck: A case Report

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ABSTRACT

Context: *Sebaceous cell carcinoma is a malignant neoplasm, rarely recognized in extra ocular sites. This neoplasm is aggressive in 29% of patients, lymph node and visceral metastasis are common. Its prognosis depends on early diagnosis of lesion.*

Clinical &Diagnostic features: *An 49 year old female had slow growing nonulcerative subcutaneous nodule on the nape of neck of 9 months duration. On FNAC cytological findings favoured a benign skin adenexal tumor with focal nuclear atypia. On histological examination it was diagnosed as a poorly differentiated sebaceous carcinoma.*

Conclusion: *Extra ocular sebaceous carcinoma is a rare neoplasm. It is difficult to diagnose as it has diverse clinical presentations, histologic patterns and complicated nomenclature. We will discuss the incidence, clinical, and histological findings along with the prognosis of this aggressive neoplasm.*

Keywords: *Extra-ocular, Sebaceous cell carcinoma, Histopathology*

INTRODUCTION

Sebaceous cell carcinoma (SCC) is an aggressive malignant neoplasm derived from adenexal epithelium of the sebaceous glands [1]. It is known to occur in the peri orbital region due to an unusual abundance of sebaceous glands in the ocular region[2]. It can exhibit aggressive local behavior and can metastasize to regional lymph nodes and distant organs. Extra-ocular sebaceous carcinoma is very rare[3]. We report a case where tumor was found in the neck region, an extremely rare site.

CASEREPORT

A 49 year old female presented with multiple nodular swelling of 9 months duration at the back of neck, 2cm below the hairline. The rest of the physical examination was normal, no lymphadeno-pathy was found. Patient was advised FNAC, aspiration yielded scanty thick mildly mucoid tissue fragments from neck mass. Smear showed rich cellularity of several tightly cohesive clusters, few singly dispersed cells with uniform nuclei, focal nuclear atypia and moderate to scanty cytoplasm[Fig1,2], findings suggestive of a benign adenexal tumor. Excision of the nodule showed ask in covered nodular tissue measuring 4.6x3.6x3.1 cm having whitish, homogenous areas on cut section. Histological examination showed an intradermal proliferation of sheets of atypical enlarged undifferentiated basaloid cells with marked pleomorphism, prominent nucleoli, moderate to scanty cytoplasm and focal areas of sebaceous differentiation [Fig 3,4,5]. These cells were infiltrating into the skeletal muscle fibres[Fig 6]. Areas of haemorrhage & necrosis were also noted. A poorly differentiated sebaceous carcinoma was thus diagnosed.

Additional investigations were performed to detect any under lying malignancy to rule out Muir-Torre syndrome and they were unremarkable. The patient was received radiotherapy and chemotherapy despite of that, she developed recurrence of tumor at same site after 6 months of operation.

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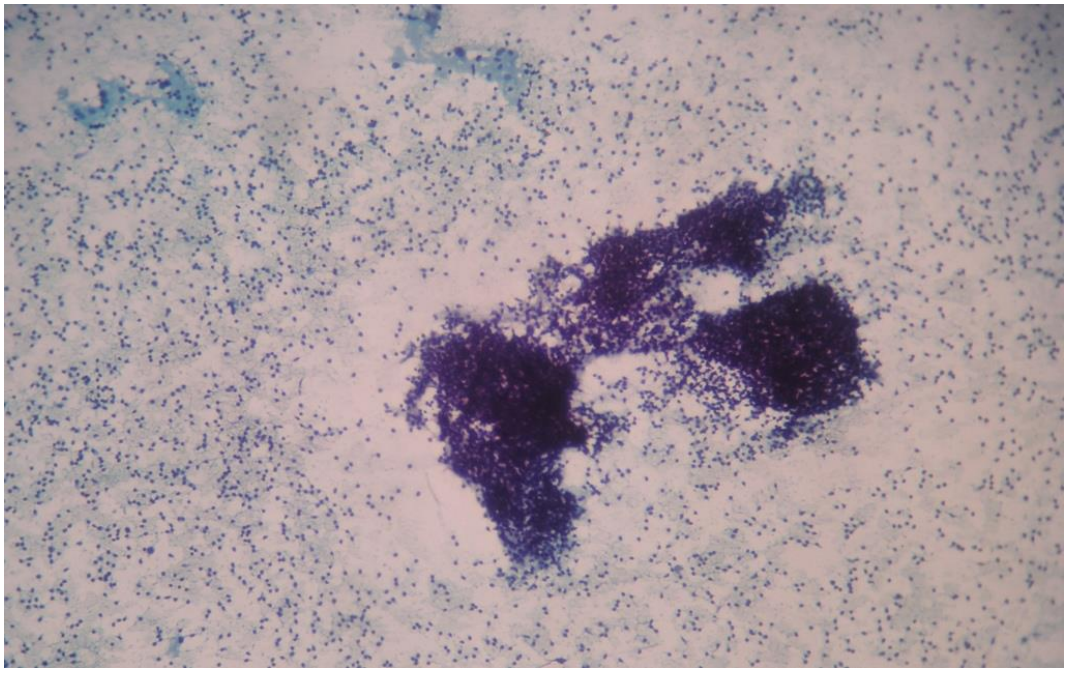

Fig. 1: Cellular smear shows uniform round epithelial cells arranged in tightly cohesive clusters, discretely & few histiocytic cells (PAP stain X100)

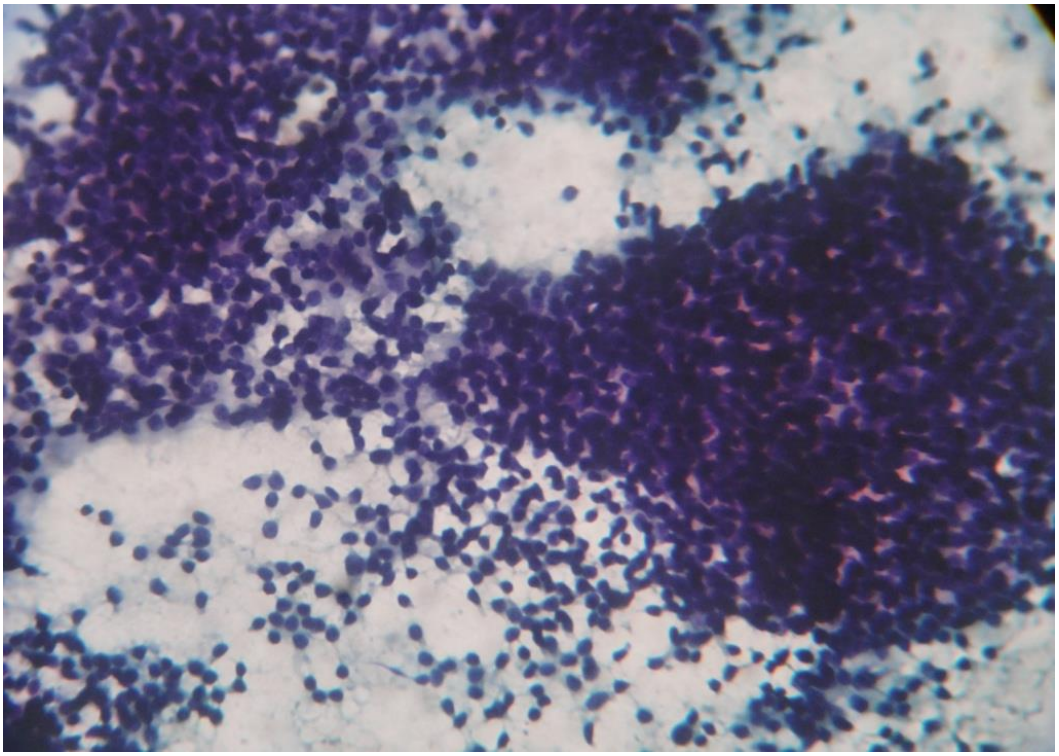

Fig. 2: High power image shows solid sheet of round to oval basaloid cells without nuclear atypia. (PAP Stain X 400)

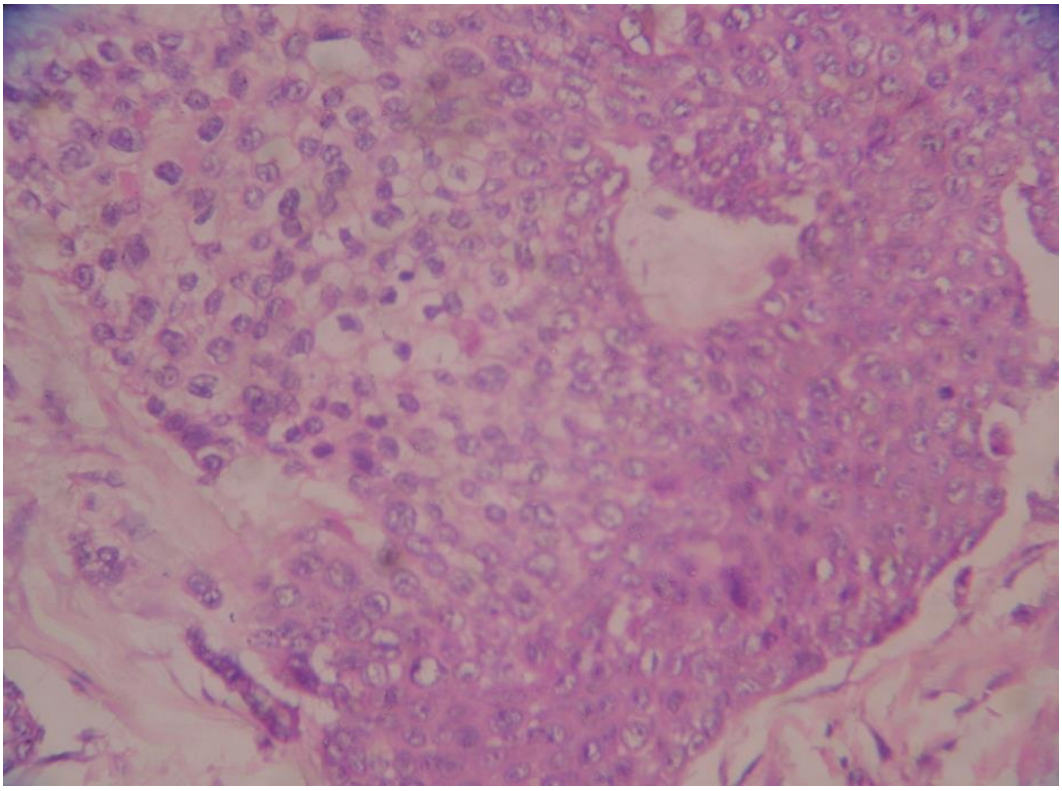

Fig. 3: Dermis shows pleomorphic sebaceous and undifferentiated cells arrangedin irregular lobules. (H&E Stain x100)

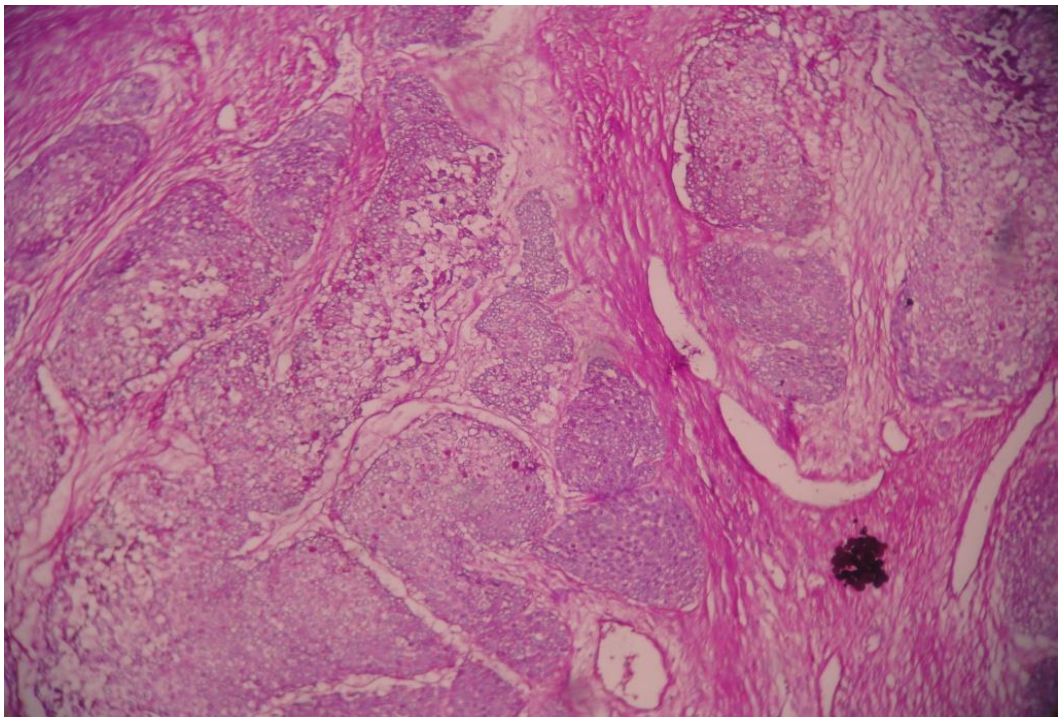

Fig. 4: Undifferentiated cells with marked cytologic atypia, eosinophilic cytoplasm and focal sebaceous differentiation with foamy/clear cytoplasm.(H&E x 400)

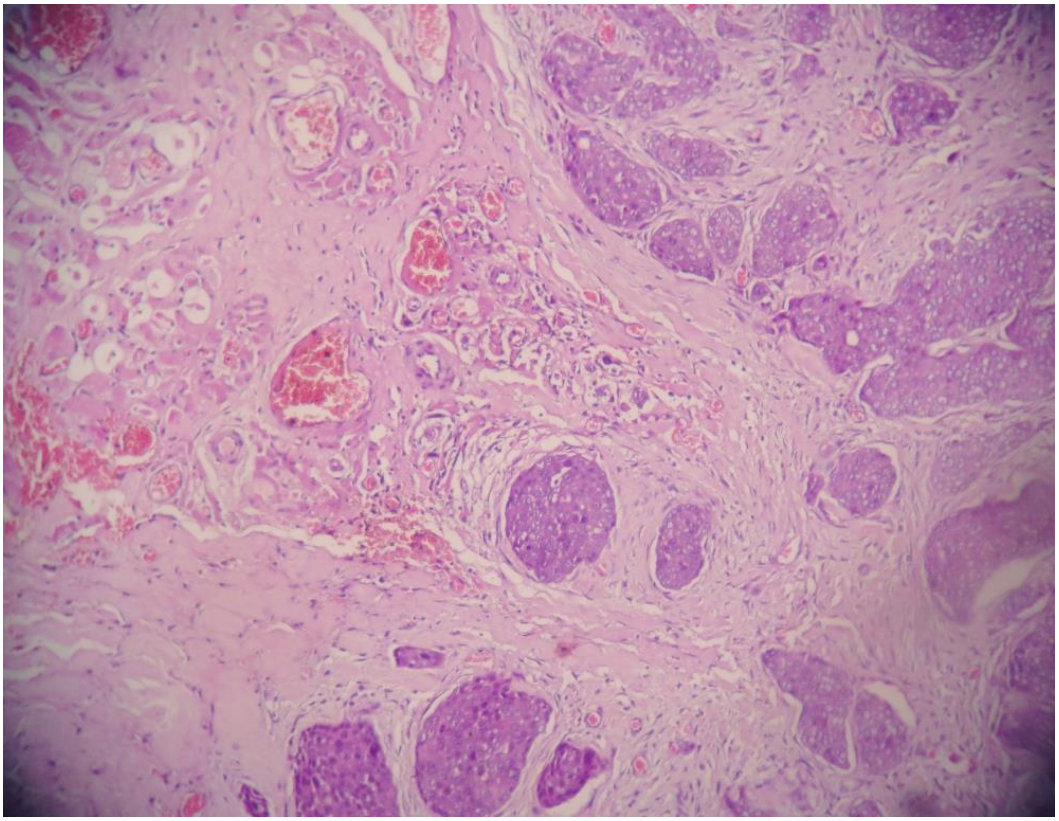

Fig. 5: Sebaceous cells does not react with PAS stain due to lack of intracellular glycogen despite presence of fine lipid globules.(PAS x 100)

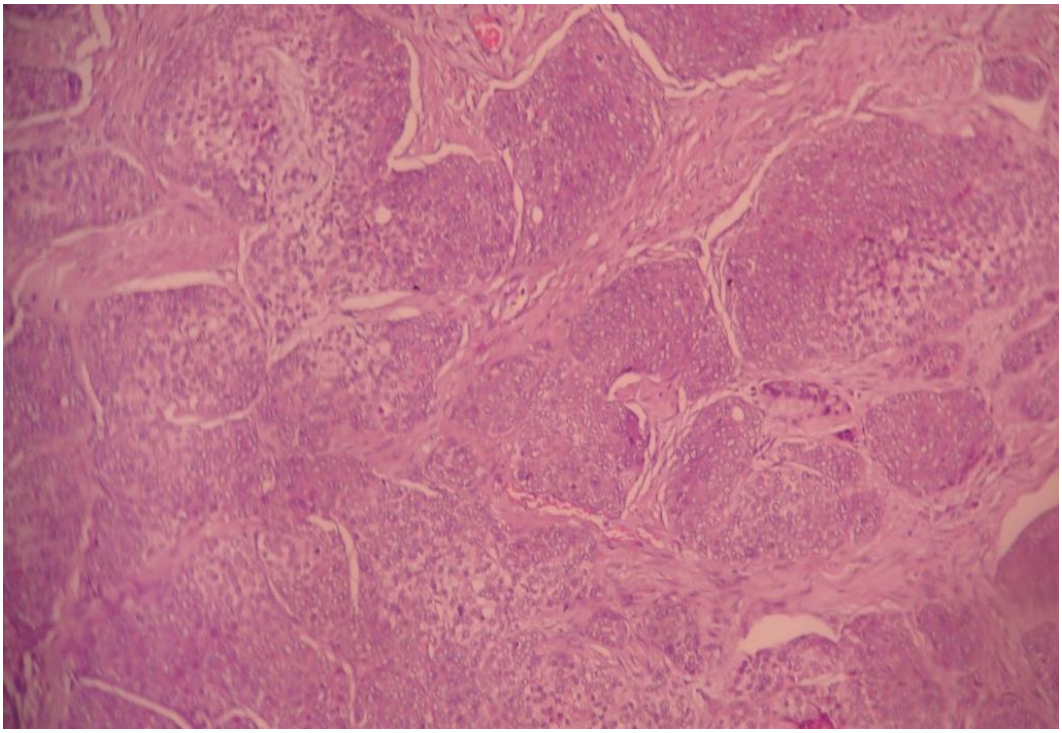

Fig. 6: Shows infiltrative growth pattern & invaded into skeletal muscle fibres. (H&E x 100)

DISCUSSION

SCC accounts for less than 1 percent of all cutaneous malignancies. It is a rare, but aggressive neoplasm. It's more common in ocular region, particularly in Asian countries[1]. SCC is a potential marker of Muir - Torre syndrome (MTS), alerting to search for other occult internal malignancies, most commonly colorectal carcinoma[4]. In this syndrome, the skin lesions may precede the presentation of internal malignancies, but often develop later. Our case had no internal malignancy observed.

The extra ocular sebaceous carcinoma is rare, constituting 25% of all sebaceous carcinomas[5]. Most cases (75%) are encountered in the skin of the head and neck[5], followed by the trunk, salivary glands, genitals, and extremities. It usually occurs in adults, average patient age 62years, with a slight male predominance[5]. Clinically, extra ocular sebaceous carcinoma presents as a nodular erythaematous, exophytic/ulcerative lesion. In our case of a middle aged female presented with an exophytic, subcutaneous nodular lesion.

Cytologically, the cells have vacuolated, bubbly cytoplasm and malignant nuclear morphology including mitotic figures and prominent nucleoli. They may have an admixture with basaloid and squamous cells with evidence of necrosis[6,7].

In our case smears uniform basaloid cells with only focal nuclear atypia was noted. There were no increase in mitosis or necrosis. Hence was reported as a benign skin adnexal tumor. Excisional biopsy showed a proliferation of atypical enlarged basaloid and epithelioid cells, intermixed with focal mature sebaceous cells. Marked nuclear atypia, mitotic activity 3-4/10HPF, focal necrosis, the tumor infiltrating the hypodermis and skeletal muscle fibres was noted. In contrast to peri ocular carcinoma, pagetoid intra-epithelial migration is uncommon. Histology consistent with a poorly sebaceous differentiated carcinoma, according to Wolfe et al [8] can be divided into four grades based on degree of differentiation, namely extremely well, well, moderately and poorly differentiated sebaceous carcinoma.

The differential diagnosis includedsquamous cell carcinomas with hydropic changes[9,10], basal cell carcinomas with sebaceous differentiation &metastatic clear cell carcinoma of the kidney[11]. However evidence of keratinization in the form of dyskeratotic cells, keratin pearls & basaloid differentiation and peripheral palisading was not seen. The neoplastic cells in our case showed scalloped centrally situated nuclei with micro vacuolated cytoplasm compared to the eccentric nuclei with clear cytoplasm in metastatic clear cell carcinoma. Sebaceous carcinoma can exhibit aggressive local behavior and metastasize to regional lymph nodes and distant organs. It seems that extra ocular SCC has a poor prognosis than peri ocular form, but other studies have indicated that the pattern of metastasis and mortality are similar[12,13].Our pateint developed tumor recurrence at local site after receiving treatment in the form of wide local excision along with radiotherapy.

CONCLUSION

In summary, SCC is a rare entity with unfavourable prognostic behaviour. This tumor should suggest the possibility of MTS and alert to search an occult internal malignancy.

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