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Extraocular sebaceous carcinoma over lower back - A rare case report

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Abstract

Sebaceous carcinoma is a rare aggressive malignant tumor with an incidence of less than 1% of all cutaneous malignancies. These tumors are generally classified as ocular (75%) and extraocular (25%). Extraocular sebaceous carcinoma is infrequent, the most usual site being the head and neck region, followed by the trunk.

This condition is associated with a genetic disorder like Muir Torre syndrome with one or more visceral malignancies. Here we report a case of a 60-year-old male patient with swelling in the lower back of 6 years duration. Fine needle aspiration was suggestive of skin adnexal tumor. Histopathological examination was in favour of Sebaceous carcinoma, further confirmed by immunohistochemistry. Extraocular sebaceous carcinoma is rare and its clinical presentation may mimic other dermatological conditions which could delay their management.

Sebaceous carcinoma should be considered as one of the differential diagnoses in elderly patients with swelling and ulceration involving the skin.

Keywords: extraocular, sebaceous carcinoma, lower back mass

Introduction

Sebaceous carcinoma (SC) is a rare aggressive slow growing malignant adnexal tumor of the skin. Mostly

arise in the head and neck region predominantly in the periorbital area. It is rare to occur in extraocular sites.[1] It has local aggressive behaviour and also metastasizes to regional lymph nodes and distant organs.[2]

Case report

A 60-y old male presented with swelling over the left lower back for 6 years. Swelling is rapidly progressing since last 6 months. On examination, swelling measuring 8 x 7 cm on the left lower back, with an ovoid ulcer of size 2 x 2 cm. The clinical diagnosis was dermatofibrosarcoma.

USG scan shows a hyperechoic lesion of 7 x 5.5 x 3.5 cm in the subcutaneous plane likely suggestive of an infected chronic sebaceous cyst. Fine needle aspiration cytology was done and showed tumor cells arranged in clusters and sheets.

Individual cells are round to oval, with a round nucleus and scant cytoplasm. Few clusters and discretely arranged cells with abundant, vacuolated cytoplasm seen. Features suggested the possibility of skin adnexal tumor.

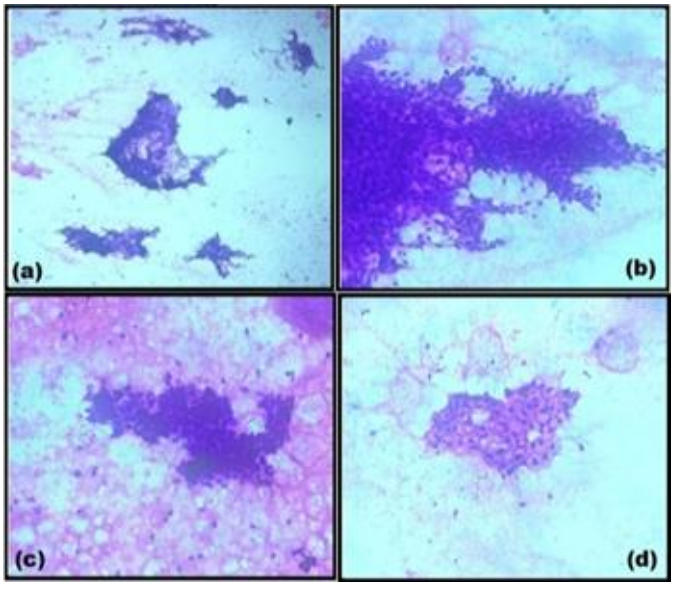

Figure 1: (a) Photomicrograph showing two populations of cells arranged in clusters and sheets. (H &E,100x) (b) Photomicrograph showing one population of cells arranged in sheets. Individual cells are round to oval in shape, with round to oval nucleus and scant cytoplasm. (H & E,400x) (c) Photomicrograph showing cells arranged in clusters. (H & E,400x) (d) Photomicrograph showing the second population of cells arranged in a cluster.

The individual cells are large and polygonal, with abundant, vacuolated cytoplasm and a central round nucleus. (H & E,400x).

An excision biopsy specimen was sent for histopathological examination. The cut section shows a grey-white lobulated mass measuring 7 x 5.5 x 3.5 cm. microscopic findings show skin with focal ulceration and underlying tumor.

Tumor mass is arranged in lobules, cords, nests separated by a thin fibrovascular stroma. Individual tumor cells are predominantly round to oval with scant cytoplasm, hyperchromatic nuclei, and coarse chromatin. Mitotic activity is raised.

Few of the cells at the center of these nests are large and polygonal with abundant foamy vacuolated cytoplasm, having round nuclei, fine chromatin, and prominent nucleoli suggesting sebaceous differentiation. Areas of necrosis seen. Histopathological diagnosis was suggestive of Sebaceous Carcinoma.

The final diagnosis consistent with Sebaceous carcinoma was given after immunohis tochemistry showed positivity for BerP4 (focal), AR, p40, Ki67 index is 40%, EMA (patchy) and negative for S100. Further investigations were done to detect any internal malignancies to rule out Muir Torres syndrome but unremarkable.

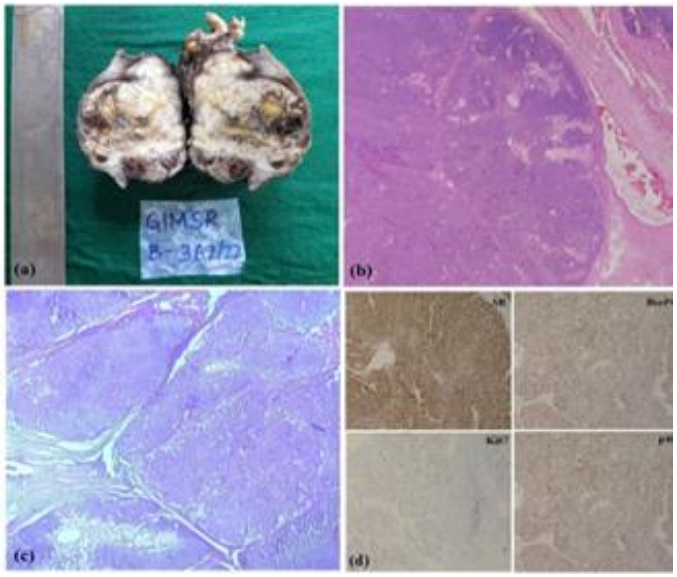

Figure 2: (a) Gross specimen showing grey white lobulated tumor mass which is situated beneath the skin. Dark brown areas of necrosis and hemorrhage are seen. (b) Photomicrograph shows a nodule composed of a tumor arranged in solid nests, lobules and sheets separated by fibrovascular septa. (H & E,100x) (c) Photomicrograph showing tumor cells exhibit moderate atypia having vesicular nuclei, multiple nucleoli and moderate amount of eosinophilic cytoplasm. Brisk mitosis and necrosis seen. Few multivacuolated cells noted. (H & E,400x) (d) IHC showing tumor cells positive for AR, BerEP4 (focal), P40 and Ki67 is 40%.

Discussion

Sebaceous carcinoma (SC) is a rare aggressive malignant adnexal tumor that accounts for less than 1% of all cutaneous malignancies. It was first described by Allaire in 1891. [3,4] Based on the location they are divided into ocular and extraocular with the former accounting for about 75%. Extraocular SC presentation is rare and also only 200 cases have been reported in the literature, that too very few cases over the trunk. Extraocular SC occurs mostly in the head and neck region followed by the trunk, extremities, genitalia, breast, [3,4,5] They may occur sporadically or in association with familial autosomal dominant inherited disorder Muir Torre syndrome.[4] Extraocular SC presents as a nodular, exophytic, or ulcerative lesion. Due to this varied presentation which may mimic benign tumors or inflammatory conditions that lead to delay in diagnosis and management.[5,6] Muir Torre syndrome is characterized by the presence of any one of sebaceous neoplasms along with one or more visceral malignancies such as colorectal, mammary or gastric. Almost 15% of primary SC is diagnosed after 5 years. [7,8] In our case, the swelling was present for the last 6 years. Sebaceous carcinoma is mostly a locally aggressive tumor but may exhibit metastasis. Recurrence and distant metastases are more commonly seen with ocular type than extraocular type but many other studies suggest that mortality and metastasis are equal in both. [2,5,9] In our case, there are no regional or distant metastases at the time of presentation. Extraocular SC occurs mostly in elder patients with slight male predominance. [4,5] In our case the patient is a male patient aged 60yr which is in accordance with previous studies. The present case shows lobules of tumor cells separated by thin fibrovascular septa with basaloid cells, nuclear pleomorphism, an increase in mitosis, necrosis, and also a few cells with foamy vacuolization of cytoplasm. Mature sebaceous cells show scalloped nuclei and abundant pale vacuolated cytoplasm. The presence of these features is regarded as the hallmark of sebaceous differentiation.Differentiated sebaceous carcinoma has abundant sebaceous cells at the center of the nests, whereas in poorly differentiated SC these cells may be scanty or absent. [3,7] In our case, SC is a basaloid variant with moderate sebaceous differentiation. Immunohistochemistry markers of SC are cytokeratin (CK), EMA, BerEP4, androgen receptor (AR), adipophilin, and also increased p53 and Ki67 levels.

[3,4]Present case is positive for BerEP4 (focal), AR, P40, EMA (patchy), Ki67 index is 40%, and negative for S100. Considering both his topathological and immunohistochemical findings, the final diagnosis was given as sebaceous carcinoma.

WLE with regional nodal dissection is the most common treatment.[9] In our case, wide local excision was done. There are no specific systemic therapies but adjuvant radiation or chemoradiotherapy should be performed for incompletely resected extraocular SC. [4] The mortality rate is equal in both ocular and extraocular types ranging from 9% to 50%. Mohs micrographic surgery (MMS) is done cosmetically in sensitive areas, such as the eyelids and face. [6,9]

In our case lesion is located in the lower back and wide local excision was done. Extraocular SC is a rare tumor and because of its varied clinical presentation that mimics other benign tumors may cause a delay in diagnosis.

Also, an additional diagnostic challenge of these tumors is their risk for association with Muir Torres syndrome.[8,10] Thus the presence of this tumor should suggest the possibility of the development of visceral malignancies and alert clinicians to search for internal malignancies.[7]

Regular follow-up for local recurrence and monitoring for internal malignancies is needed even with a single sebaceous neoplasm irrespective of family history.[5,9] Our patient was advised to do regular follow-ups for early detection of any new lesions.

Conclusion

Extraocular sebaceous carcinoma is a rare and aggressive malignancy. Sebaceous carcinoma should be considered as a differential diagnosis in any ulceroproliferative growth over the skin.

A sebaceous neoplasm should suggest the possibility of Muir Torre syndrome and elicit a search for occult internal malignancy. Regular follow-up of the patient for a long period is required as late recurrence and metastasis are known.

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