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Malignant Eccrine Spiradenoma (Spiradenocarcinoma) Of Face; Case Report of A Rare Adnexal Skin Tumor.

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Abstract*:Malignant eccrine spiradenoma (synonym, spiradenocarcinoma) is an extremely rare malignant adnexal tumor of the skin that originate in the sweat glands. It almost always arises from a preexisting benign eccrine spiradenoma, rarely it may arise de novo. We report a case of a 65-year-old lady with malignant eccrine spiradenoma of the face. The face mass was present for approximately 5 years, suddenly it became enlarged, ulcerated, tender, with a change in its color over a period of four months. The mass was excised & received in our department. Histopathological report confirmed it to be a case of malignant eccrine spiradenoma. The tumor has a poor prognosis due to multiple local recurrences & eventual widespread metastases. Wide local excision and close follow up are crucial in the management of malignant eccrinespiradenoma.*

Keywords: *adnexal, eccrine, malignant,spiradenoma,spiradenocarcinoma.*

I. Introduction

Malignant eccrine spiradenoma is an extremely rare adnexal tumor, mainly affecting middle aged population (mean age is 55 years) and its incidenceis similar in both the sexes. In contrast the benign counterpart i.e. eccrine spiradenoma is well recognized tumor, commonly affects young adults. These are sweat gland tumors arising from the intradermal straight part of the duct of eccrine sweat glands.[1] The presentation of the benign counterpart is often that of a single nodule that may or may not be tender.Malignant transformation is suggested by the typical history of a longstanding benign lesion (mean duration of preexisting lesion is 20 years), that suddenly becomes enlarged, ulcerated & tender with change in it's color.[2]

Diagnosis of the tumor is based on histopathological examination & malignancy is suggested by increased mitotic rate, nuclear atypia, pleomorphism & hyperchromasia, and absence of a dual cell population which is seen in benign eccrine spiradenoma.Here we have reviewed the clinical features, histopathological & immunohistochemical findings, treatment and prognostic factors in a patient of malignant eccrine spiradenoma of the face presented in our institution.

II. Case Presentation:

A 65-year-old lady presented in the surgery department with a mass in the right side of face, which had been present for last 5 years. Over the period of last 4 months she has noticed gradual increase in the size of the mass & it has become mildly painful. There was no palpable neck lymph nodes & also no other significant comorbidities. Subsequently wide localexcision of the mass was performed &specimen was received in our pathology department. Gross examination of the resected specimen showed a 1cm × 0.5cm × 0.5cm, grey brown soft nodular lesion.On microscopic examination, the dermis and subcutaneous tissue showed a tumor arranged in ragged sheets & solid masses along with occasional irregular glandular structures. (fig: 1&2). There is mild to moderate nuclear atypia, high mitotic figures (8/10 high power field), (fig: 2)prominentdesmoplastic inflammatory host response, focal area of tumor cell necrosiswith few small foci of squamous differentiation.Individual tumor cells show pleomorphic vesicular nuclei with variable prominent nucleoli and moderate amounts of cytoplasm. (fig: 3). This high-grade eccrine carcinoma was noted to be arising from a preexisting eccrine spiradenoma, seen as sharply demarcated lobules composed of 2 cell populations (peripheral basaloid cells & central pale cells) arranged in nests. The overlying epidermis was unremarkable.Immunohistochemical staining for cytokeratin and EMA showed positivity in tumor cells, (fig: 4&5). The patient has tumor free margin, no palpable lymph node in the neck & no evidence of distant metastasis; therefore, she wasnot put on any adjuvant therapy. The patient is under follow up since last three months & is doing good.

III. Figures:

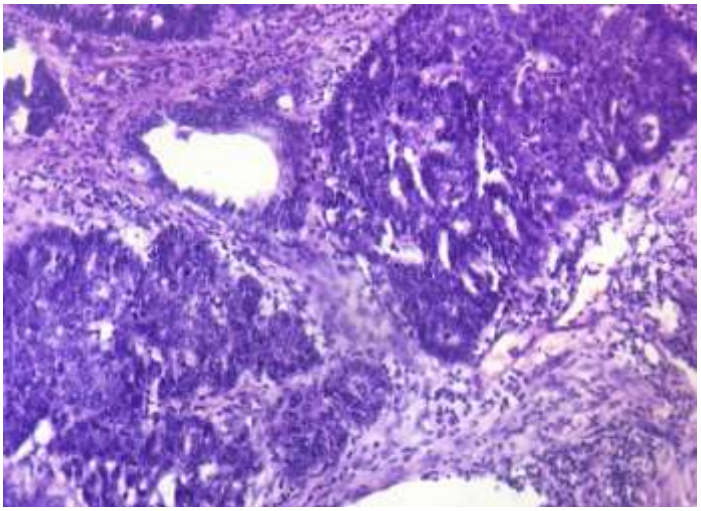

Fig 1: Solid areas of the tumor cellswith occasionalirregular glandular structures& prominent desmoplastic inflammatory host response. (H&E, 20X).

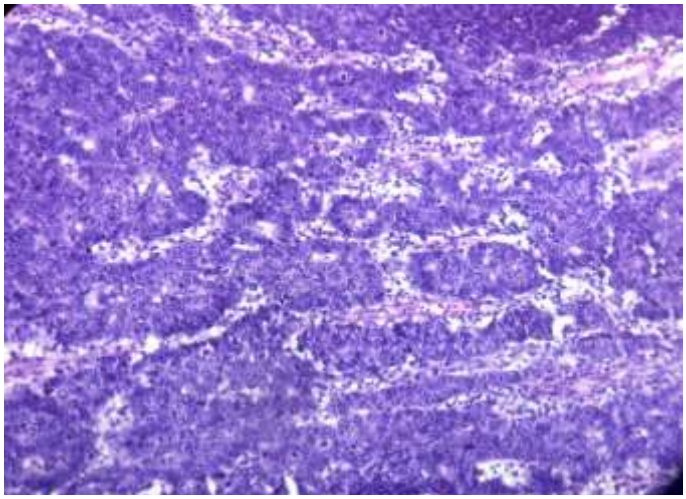

Fig 2: Solid areas of the tumor composed of malignantcells with nuclear atypia, high mitotic figures. (H&E ,20X).

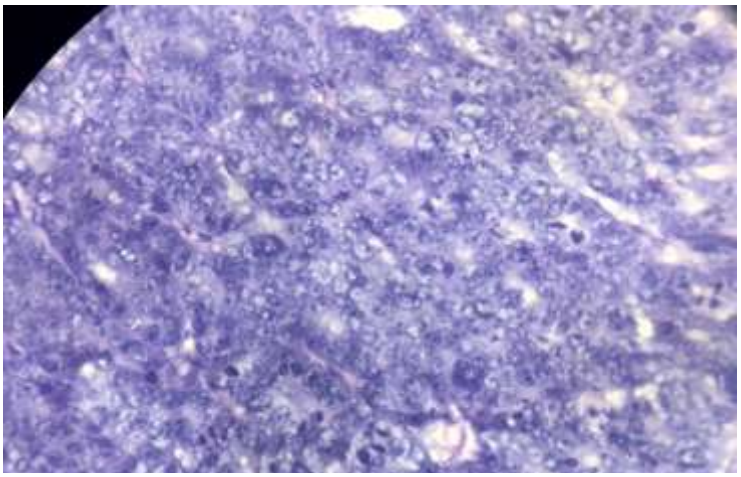

Fig 3: Individual tumor cells show pleomorphic vesicularnuclei with variable prominent nucleoli. (H&E, 40X).

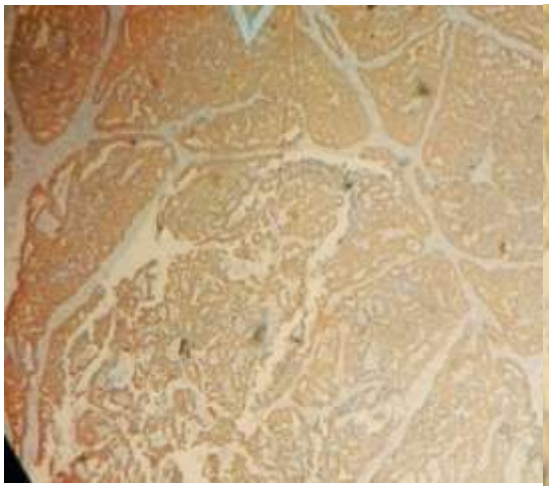

Fig 4: IHC staining by cytokeratin showing positivity by tumor cells. (20X).

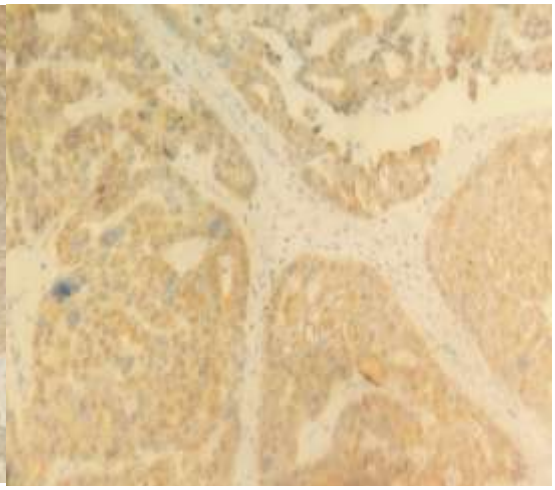

Fig 5: IHC staining by EMA showing positivity by tumor cells. (40X).

IV. Discussion

Malignant eccrine spiradenoma (MES) can arise from a preexisting benign eccrine spiradenoma or occur as a primary malignant tumor. [3]Clinical features of these tumors may include ahistory of sudden enlargement in a previously stable lesion. The tumor usually presents as a solitary firm round dermal nodule& can affect any body site, but the most common locations are extremities, trunk and the head and neck areas.[4]The average size of MES at presentation is 3.9 cm (range, 0.5-15 cm). [4]On histopathology, there will be focus of benign spiradenoma within or adjacent to the malignant tumor.[4] The malignant transformation is usually into a carcinoma;[4,5] however, in few cases carcinosarcomatous transformation has also been reported.[6] Histological hallmarks of malignant eccrine spiradenomas are proliferation of solid masses of tumor cells with large hyperchromatic nuclei, nuclear atypia, frequent mitosis, invasion of surrounding connective tissue, loss of basement membrane and squamous differentiation.

Immunohistochemically the tumor is variably positive for majority of the cytokeratins, CEA, EMA & shows a spotty reaction for S-100 protein.[7,8,9] Overexpression of p53 protein has also been reported in cases with malignant transformation.[7]Recently estrogen receptor positivity has been shown in some cases of MES, which may result in potential therapeutic options.[8]Metastasis of regional lymph nodes, lungs, brain, liver and bone have been reported.[4]The mainstay of therapy is surgical excision with wide excision margin. Regional lymph nodes should be dissected if tumor metastases are suspectedclinically. [10] Role of prophylactic lymph node dissection, postoperative adjuvant radiotherapy, or chemotherapy is uncertain because of limited data and follow up time. [10]Hormonal receptor status (estrogen receptor) should be evaluated in the tumor for potential therapeutic options.

Conclusion

Although malignant eccrine spiradenoma is an extremely rare adnexal tumor of the skin, it is thought to have the high capacity of metastasis and lethal potentiality. Rapid changes of appearance of the benign skin lesions, rapid growthand ulcerations are clues indicating malignant transformation. Also as the malignant changes may be focal in a benign spiradenoma, the specimen must be adequately sampled & there should be high index of suspicion.All eccrine spiradenomas must be carefully examined in view of malignant transformation. Due to the high risk of developing metastases& recurrences, close follow up of these patients should be carried out postoperatively.

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