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Microcystic adnexal carcinoma of the breast – A Case Report

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Abstract

Microcystic adnexal carcinoma of breast is a rare, slowly progressing, malignant tumour of sweat gland origin. This tumour is most commonly seen on the skin of the upper lip, chin, nasolabial fold and cheek particularly the central face. It is a locally aggressive neoplasm with rare lymph node metastasis. We reported a case of 39 years old female presented with lump in right breast. The lump was soft to firm, slightly mobile, measuring 3x2 cm in size. The lump was fixed to the skin and underlying breast tissue. FNAC done was inconclusive. However, an incisional biopsy revealed the nature of the tumour as microcystic adnexal carcinoma of the breast skin. The patient underwent excision for the lump. Very few reports on adnexal tumour of breast are described in the literature. This case demonstrates the importance of biopsy which plays an important role to decide mode of surgery and also explains that microcystic adnexal carcinoma can occur on the breast skin and should be treated with wide excision due to its locally aggressive behaviour and thus mastectomy can be avoided for such cases.

Keywords: Microcystic adnexal tumour, locally aggressive.

Introduction

Microcystic adnexal carcinoma (MAC) of breast is a rare infiltrative cutaneous tumour with a confusing historical nomenclature including malignant syringoma, sweat gland carcinoma with syringomatous features, and sclerosing sweat duct carcinoma.[1]

MAC usually involve centrofacial region. It has been reported on the scalp and axilla. Breast is rarely involved and very few cases have been reported. No significant sexual predilection is reported, although some studies suggest a slight female predominance[2].

Middle-aged or older adults are generally affected. The lesions are infiltrative and perineural invasion is frequent. Locoregional metastasis is an infrequent occurrence, and widespread metastasis has been described in only one case.[3] MAC of breast are frequently misdiagnosed based on superficial biopsy alone. Treatment requires complete surgical excision.

Case report

A 39 years old female presented with history of lump located in right breast. It gradually increased in size. On examination the lump was soft to firm, slightly mobile and tender, It was appearing to be fixed to overlying skin and underlying breast tissue, measuring 3x2cms. Overlying skin was slightly ulcerated.

No lymphadenopathy was present on physical examination.

FNAC done was inconclusive.

The patient had no other significant medical history and she reported no prior radiation therapy.

Incisional biopsy results revealed a microcystic adnexal carcinoma (MAC).

Gross specimen we received was irregular partly skin covered tissue mass, soft to firm in consistency, colour was greyish -brown along with attached fibrofatty tissue. It measured 3 x 2 cm. Sections were taken.

Histopathology

On microscopic examination, sections revealed irregular tissue fragment composed of nests and cords of tumour cells. The cells were small round to oval with round hyperchromatic nuclei with or without prominent nucleoli and moderate cytoplasm. There was anisonucleosis and pleomorphism. These neoplastic cells showing microcystic pattern in the subcutaneous plane. Microcysts were lined by single cells with bland nuclei and scanty cytoplasm. Mitotic figures were not seen. (Figure 1-3)

Underlying dermis was free from tumour and overlying skin was seen. Areas of sclerosis and keratinization was also seen. Immunohistochemistry was advised.

Discussion

Microcystic adnexal carcinoma is a type of cancer that either arises from the sweat or sebaceous glands. MAC is a slowly progressing tumour, which is associated with the skin, and not the breast duct tissues. It either arises from the sweat or sebaceous gland. It is not really considered as a breast cancer but rather a skin cancer which has grown in the skin of breast. It is often **misdiagnosed** both the clinically and histologically.

Clinically it presents as a **firm subcutaneous nodule**, which is often pale yellow and with overlying telangiectasias. Ulceration can be seen rarely. An adnexal carcinoma of the breast can also present as a cyst, and frequently can be present for many years. They are usually asymptomatic and thus they tend to be ignored. They may create symptoms of numbness and burning. Due to its slow growing nature, tumours lasting for decades have been reported. After a search of the literature, only 1 case of MAC has been reported in the nipple.[4]

Although MACs can arise in virtually any age group, most MACs occur in older individuals, with a reported average age of incidence between 44 and 64 years and an overall range of 11 to 90 years. Although a slight female predominance has been noted in some case series, over all the sexes are equally affected.

Fewer than 300 cases have been reported worldwide, according to an analysis by Yu et al[5] using the National Cancer Institute's Surveillance, Epidemiology, and End Results (SEER) database[5]

William C. et al reported a case of 41 year old male with history of Microcystic adnexal carcinoma on scalp and also described that it was the first case of MAC reported on scalp.[6]

A case of metastasis Breast Carcinoma on Lip had been reported by Cotton D.W.K et al in an 72yrs female who presented with a 2 months history of an asymptomatic lesion on the upper lip which was later confirm to be a

metastasis from breast by using immune-peroxidase markers.[7]

Histological features of MAC vary at different depths of the tumor.

Tumours With Histologic Features Similar to Microcystic Adnexal Carcinoma

Benign Tumors

Syringoma Desmoplastic trichoepithelioma Trichoadenoma

Malignant Tumors

Basal cell carcinoma (infiltrative and morpheic types) Squamous cell carcinoma (poorly differentiated) Metastatic adenocarcinoma

A Trichoepithelioma is a benign epithelial tumour which develops from the **hair follicles**. It is commonly seen on the face, but could develop on the breast as well. A Syringoma is another type of sweat duct tumour, which is commonly found on the **eyelids** but can also occur on the breast. They are usually skin-coloured firm rounded bumps, and very small. Hidradenocarcinoma is derived from the **epithelial cells of sweat glands.**

Microcystic adnexal tumour is also known as Infiltrating syringomatous adenoma of the breast. The lesion is also referred to as a **well-differentiated** adeno-squamous carcinoma, or as a sclerosing sweat duct.

It is always confused with a well-differentiated tubular carcinoma of the breast. These tumours can invading the surrounding nerves and smooth muscle tissue. It can be differentiated from tubular breast cancer on basis of presence of squamous metaplasia.

Lesley K. Wong et al reported a case of microcystic adnexal carcinoma of nipple. They demonstrate the importance of having high index of suspicion to obtain diagnosis and concluded that MAC has a confusing nomenclature and inadequacy of superficial biopsy to achieve an accurate diagnosis. Its treatment consist of wide local excision, sometimes requiring Mohs surgery to achieve clear margin. [8]

Robert N. Page et al also conducted a study on multiple microcystic adnexal carcinomas and concluded the importance of close follow up and difficulties in diagnosis of the patient.[9]

Conclusion

Microcystic adnexal tumour of breast is an uncommon clinical entity with locally aggressive behaviour and rare metastatic potential. This case demonstrates the importance wide range of suspicion in order to obtain a diagnosis, extend of disease and to achieve adequate resection. As microcystic adnexal tumour is locally aggressive it requires local excision and hence mastectomy can be avoided. Due to possibility of future recurrences close clinical observation is recommended.

MAC is considered to be a tumour with excellent overall prognosis. This case also emphasises the importance of tissue biopsy which helps clinician to decide mode of treatment. However microcystic adnexal carcinoma of breast is rare but possibility of it should be kept in mind as the mode of management varies.

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Legends Figure

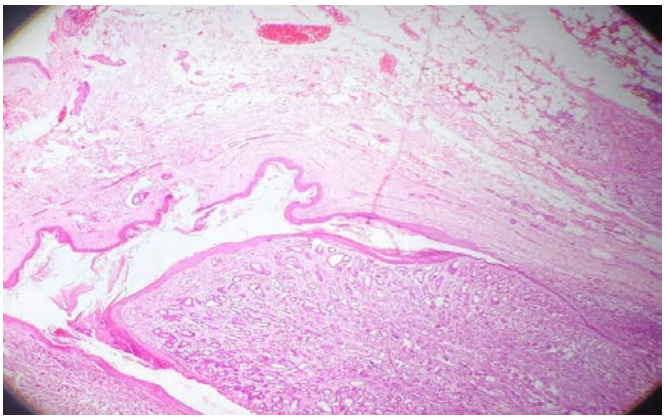

Figure 1: tumour cells are seen in microcystic pattern in the subcutaneous plane. Few nests and cords are also seen. Areas of sclerosis and keratinization was also seen.

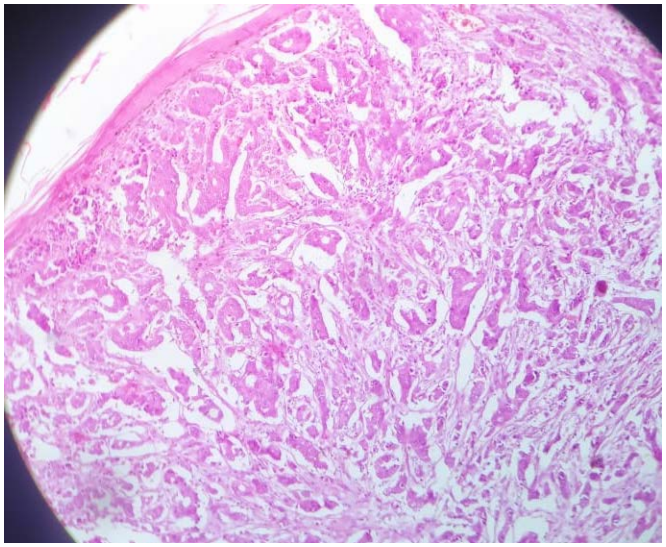

Figure 2

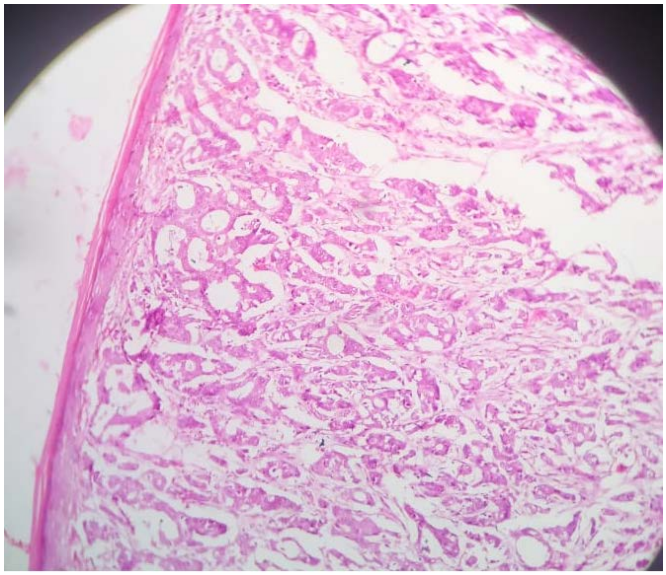

Figure 3

Figure 2& 3: Shows round to oval tumour cells with hyperchromatic nuclei and Microcysts were lined by single cells with bland nuclei and scanty cytoplasm.