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Primary mucinous adenocarcinoma of the scalp: A case report and literature review

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ABSTRACT

INTRODUCTION: Primary mucinous adenocarcinoma or mucinous eccrine carcinoma of the skin is a rare malignant neoplasm showing predilection to the head and neck. Distinguishing between these primary neoplasms and the more frequent metastatic mucinous deposits on the skin from primaries in the breast and gastrointestinal tract constitutes a diagnostic dilemma.
PRESENTATION OF CASE: We report a case of a 61-year-old lady who presented with a slow-growing, painless scalp nodule. Upon excision, it was diagnosed as "primary mucinous adenocarcinoma". An extensive work-up in search for another primary tumour failed to show a primary malignancy elsewhere and the diagnosis of a primary eccrine mucinous adenocarcinoma of the skin was rendered.
DISCUSSION: A review of the literature on this entity is presented, discussing diagnostic challenges and therapeutic options that of interest to surgeons, pathologists and dermatologists.
CONCLUSION: These tumours are indolent and low-grade, with a tendency for local, sometimes multiple, recurrences. Proper patient counselling and follow-up are important in treatment. Sound collaboration between clinicians and pathologists, for good therapeutic results, is of utmost importance.

1. Introduction

Mucinous sweat gland carcinoma is a rare malignant tumour of the skin. We report herein the case of a 61-year-old lady who presented with a scalp mass. After excision, histopathological examination of the mass revealed mucinous adenocarcinoma. Extensive metastatic work-up, including a total body computed tomographic and positron emission scan (PET/CT scan), showed the neoplasm to be a primary mucinous adenocarcinoma of the skin. Distinguishing this primary neoplasm from metastatic mucinous adenocarcinoma originating from other primary sites in particular from the breast presents a diagnostic dilemma. A review of the literature of this rare, yet challenging, entity follows.

2. Report of a case

A 61-year-old lady, of Caucasian origin, presented to our care with a slow growing, painless, spherical, scalp nodule overlying the right temporal region, measuring 1.5 × 1 cm. The mass was subcutaneous, being tethered to the skin, yet mobile with respect to the underlying skull. It was firm, non-compressible, non-pulsatile and showed no central punctum. The nodule was present for the last 5 years with no history of pain, secretions or overlying redness. Upon examination, no regional lymphadenopathy or abnormal findings in the head or neck region could be noticed. The patient had been medically followed up in the past with regular screening for breast cancer and gastrointestinal disease, namely colorectal cancer.

An excisional biopsy of the nodule was done under local anaesthesia with no adverse events and the skin defect was primarily closed. Histopathological examination of the mass showed hair-bearing skin with underlying lakes of mucin separated by delicate fibrous septae. Floating in between the mucinous lakes were nests, clusters, and cords of predominantly uniform round to cuboidal epithelial cells that exhibit mild to moderate nuclear variability. Focal areas show duct formation. Occasional tumour nests show prominent proliferation of cells with cribriforming. The tumour showed no connection to the overlying skin or adnexal structures (See Fig. 1). The neoplastic cells were strongly positive for cytokeratin 7 (CK7) and oestrogen and progesterone receptors. The cells were negative for cytokeratin 20 (CK20) and Her-2-neu (See Fig. 2).

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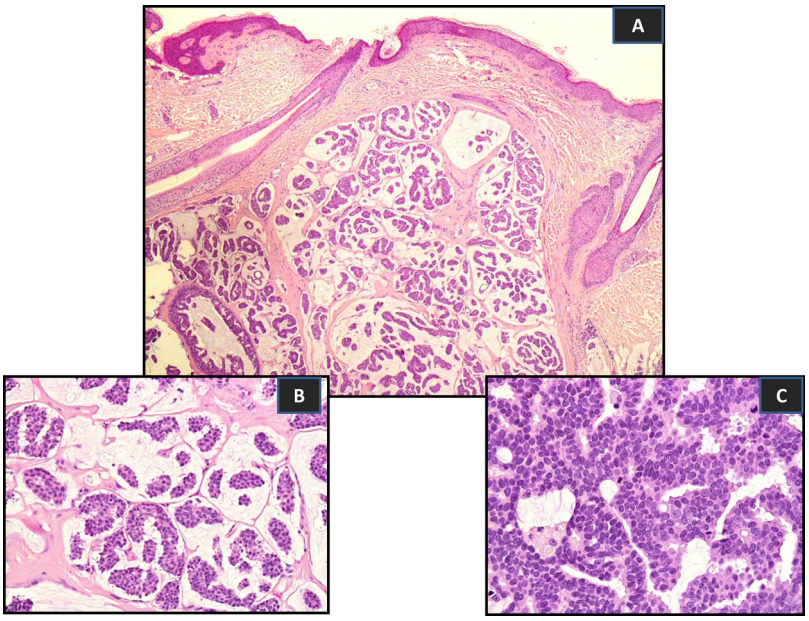

Fig. 1. A. Low power view of the skin nodule exhibiting skin with underlying lakes of mucin compartmentalised by delicate fibrous septae and floating nests and ducts of tumour cells B&C. Higher power showing variability in cellularity and proliferation of neoplastic cells ranging from less cellular nests/ducts to more cellular areas with cribriforming.

The morphologic and immunohistochemical features were consistent with mucinous adenocarcinoma. As the histopathological distinction between primary eccrine mucinous adenocarcinoma and metastatic mucinous tumour is almost impossible, it was recommended to do a complete workup to exclude the presence of a primary involving an extracutaneous site, especially the breast.

At this stage, the patient was referred to a cancer centre. She had an extensive oncological evaluation, including a detailed clinical assessment and a PET/CT scan, in search of a possible primary mucinous adenocarcinoma originating mainly in the breast and gastrointestinal tract, resulting in a negative metastatic work-up. Therefore, the diagnosis of a primary eccrine mucinous adenocarcinoma of the scalp was confirmed. She underwent Moh's micrographic surgery until negative margins were obtained. Having no evidence-based data regarding treatment, follow-up and recurrence rates of this tumour, the patient was put on a biannual follow-up programme for possible early detection of local recurrence or metastasis in relation to this unusual tumour.

3. Discussion

Primary eccrine mucinous adenocarcinoma of the skin is a rarely described pathological entity. This neoplasm was first described by Lennox et al. [1,2] in 1951, it affects more men than women in an approximately 2:1 ratio, mostly seen in 5–7th decade with an average age at occurrence of 61 years (range 8–84 years) [3,4]. There is an increased predilection to white rather than African American, East Asian or Indian individuals [5,21]. The majority of these tumours are located in the head and neck; however, less frequently, they may arise from other areas of the body [6]. More than 50% of reported cases are located in the peri-orbital region and the hair-bearing scalp [1,7].

Typically, these tumours present as slow growing, painless, soft, sometimes indurated, reddish or grey-blue, non-ulcerating nodules that have been present for several years [1,3], with longer courses for up to 20 years prior to presentation being reported [8]. The presentation of these neoplasms in the form of ulcers or cysts has also been described [3]. The tumours usually range in size between 1 and 8 cm [4], the mean diameter prior to excision being reported to be around 1.8 cm [21]. However, larger variants have been described in literature [4]. The nodules are well circumscribed, unencapsulated, and often fixed to the dermis making them unable to be "shelled out". The cut surface is mostly gelatinous.

These tumours are considered low-grade malignant neoplasms with an indolent course, having a tendency for local recurrence (19.6%) and metastasis (6.1%) after surgical treatment over a mean follow-up period of 37.4 months [21], with most metastases being to the regional lymph nodes [9,10]. Distant metastases have been reported in 2–7 percent of affected patients [6]. However, these tumours more frequently invade tissues by direct extension, due to the presence of satellite islands of tumour cells present around the main nodule, and via regional lymph node invasion [11,12]. Death due to mucinous adenocarcinoma of the skin is exceptional with only less than 5 cases reported thus far, most associated with multiple tumour recurrences and widespread metastatic disease [13].

The clinical presentation of mucinous carcinoma is nonspecific and the differential diagnosis includes epidermoid cyst, pyogenic granuloma, melanoma, sebaceous cyst, sebaceous carcinoma, cystic basal cell carcinoma, neuroma, lacrimal sac tumour, haemangioma, pilomatricoma, lipoma and metastatic adenocarcinoma [5,14].

Histogenesis of this tumour has not been clearly elucidated. There is strong evidence that these neoplasms arise from eccrine ducts, however, some studies support that at least a subset of these tumours to be originating from apocrine ducts [15].

The primary challenge in diagnosis lies in differentiating these rare primary skin neoplasms of sweat gland origin from the more frequent mucinous secondary deposits to the skin from primaries elsewhere. Mucin-producing primary tumours are known to originate in the breast, gastrointestinal tract, lung, kidney, ovaries, pancreas and prostate [3,16]. Metastatic lesions from the breast and colon are most likely to mimic mucinous carcinoma of the skin, knowing the fact that 19% of men with colon cancer and 6% of women with breast cancer have metastatic skin disease [14,17]. Differentiating secondary deposits, particularly from these two sites from primary mucinous skin carcinoma based on morphologic evaluation alone is almost impossible. Therefore, the