Primary Mucinous Eccrine Adenocarcinoma: A Rare Skin Adnexal Tumor at an Unusual Site

Primary mucinous eccrine carcinoma (PMEC) is a rare skin adnexal neoplasm. Most common sites for this tumor are eyelid and periocular tissues. Periumbilical region is a rare site for this neoplasm. The main differential diagnosis for this neoplasm is metastatic mucinous adenocarcinoma. It is important to correctly diagnose this tumor because the prognosis of this neoplasm is better than metastatic disease. To differentiate it from metastasis complete clinical, radiological, histopathological, and immunohistochemical work up is required.[1]

A 64-year-old male presented with a slow growing painless mass at the periumbilical region. He had no other complaints or swelling. On examination, there was a 5 × 5 cm subcutaneous mass at the periumbilical region on abdominal wall. The surface of the mass was crusted, and it was firm, nonfluctuant, and fixed to the skin. On fine needle aspiration cytology (FNAC), the smears were cellular with cells present in clusters forming tubules and cords in abundant pale mucinous background. The cells were monomorphic with round to oval nuclei, fine chromatin, and inconspicuous nucleoli. Few cells were plasmacytoid in shape with abundant cytoplasm and minimal nuclear atypia. [Figure 1a and b]. On FNAC, two possibilities were considered – metastatic mucinous carcinoma and primary mucinous eccrine carcinoma of the skin. Ultrasound abdomen, upper gastrointestinal endoscopy, computed tomography of the chest and abdomen, and barium swallow did not reveal any site of primary malignancy. An excision biopsy was performed to confirm the diagnosis.

On histopathological evaluation, the tumor was in the dermis. Tumor cells were floating in mucin pools separated by fibrous septae [Figure 1c and d]. The cells were monomorphc and lacked nuclear atypia. They were arranged in nests, cords, and tubules. The extracellular mucin was periodic acid Schiff (PAS) and Alcian blue positive [Figure 2a and b]. On immunohistochemistry, the tumor cells were positive for CK 7 and negative for CK 20 which supported the eccrine origin of the tumor [Figure 2c and d]. The final histopathological diagnosis of primary eccrine mucinous adenocarcinoma was given.

PMEC most commonly affects elderly male patients. Most common sites are eyelid and periorbital sites. Other sites include scalp, face, cheek, chin, nose, foot, vulva, and chest/ abdominal wall. This tumor is a low grade malignant neoplasm with an indolent course and a tendency for local recurrence.

The most important differential diagnosis is metastatic carcinoma. Mucin producing primary tumors are known to occur in the breast, colon, lung, kidney, ovaries, pancreas, and prostate. Metastatic tumors from breast and colon most closely mimic primary eccrine mucinous carcinoma of the skin. To rule out metastasis it is important to perform an extensive clinicoradiological and histological evaluation.

Only few case reports have described the cytological features of this rare tumor.[2,3] Usually cytological smears of this

Figure 1: (a) Epithelial cells in small clusters with fine chromatin and inconspicuous nucleioli. (Papanicolaou stain x400) (b) Cell clusters floating in abundant mucinous material. (MGG stain x400) (c) Excision biopsy, Tumor located in the dermis with tumor cells floating in mucin pools separated by fine septae. (H and E stain x100) (d) Excision biopsy, The cells had minimal nuclear atypia with few cells showing intracytoplasmic mucin (H and E stain x400)

Figure 2: The mucin stained pink with periodic acid Schiff (PAS) with diastase (a), (PAS diastase stain x400) and blue with Alcian Blue stain at pH 2.5 (b), (Alcian Blue pH 2.5 x100). The tumor cells were immunopositive for CK-7 (c), (IHC CK7 x 400) and negative for CK-20 (d), (IHC CK20 x 400)
neoplasm are cellular with monomorphic cells in a mucinous background. Cells are monomorphic with minimal nuclear atypia.

On immunohistochemistry, primary mucinous carcinoma of the skin is positive for CK 7, epithelial membrane antigen (EMA), CEA, p63, and occasionally ER and PR and negative for CK 20. CK 20 negativity helps to differentiate from lower gastrointestinal malignancies which are positive for the same. The mucin in this primary skin adnexal tumor is sialomucin which is PAS positive and diastase resistant and Alcian blue positive at pH 2.5. [3]

Surgical excision with wide margins is the main stay of treatment due to the tendency of local recurrence. [4] Older and Asian patients have better postsurgical outcomes. Larger tumors, those persisting for longer duration prior to the surgery, and those located on the trunk have poorer outcomes. [5] The overall prognosis is good with a tendency for recurrence and only very rare distant metastasis.

To conclude, primary mucinous eccrine carcinoma of the skin should be considered in the differential diagnosis of mucinous carcinoma on aspiration cytology of subcutaneous masses even at rare sites such as periumbilical region.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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