



SKIN CANCER (OTHER THAN MELANOMA)

AN ATYPICAL CASE OF EXTRAMAMMARY PAGET'S DISEASE PRESENTING ON THE LATERAL CERVICAL NECK

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Background: Extramammary Paget's Disease (EMPD) is a rare intraepithelial adenocarcinoma typically affecting elderly white female patients and is often associated with visceral carcinomas. EMPD frequently presents as well-demarcated, pruritic, persistent, scaly, erythematous plaques, often resulting in a misdiagnosis as an eczematous dermatitis and an average two-year delay to diagnosis. EMPD classically affects anogenital skin, less commonly the axillary skin, and has rarely been reported in the umbilicus, ear canal, and eyelids. We present an atypical case of EPMD, arising in an elderly male with a non-pruritic lesion on the lateral cervical neck.

Observation: A 59-year old white male presented with a 4.0 x 2.9 cm scaly plaque on his left lateral cervical neck. He reported pain when his clothing contacted the lesion, but denied pruritus and bleeding. He denied fever, chills, nausea, weight loss, night sweats, and other systemic symptoms. He had no personal or family history of cancer and reported a recent negative colonoscopy. His exam was negative for lymphadenopathy and oral, head, and neck lesions. Punch biopsy revealed an intra-epitheliomatous proliferation of neoplastic cells with immunostains positive for CAM5.2 and CK7, consistent with EPMD. The patient underwent Mohs surgery requiring two stages to clear the lesion with negative margins. He was recommended to undergo age appropriate cancer screening. There has been no evidence of recurrence, and he continues to be followed by Dermatology.

Key message: EMPD should be considered in the differential diagnosis for persistent scaly plaques, including those arising in the head and neck, regions typically unaffected by EMPD. CK20 positive and GCDFP negative variants suggest an underlying malignancy and warrant systemic workup. Treatments for primary lesions include phototherapy, topical immunomodulators, radiotherapy, and surgery. Mohs surgery results in lower recurrence rates compared to standard surgical management, and is therefore emerging as a leading treatment choice for EMPD.

