

A case of Extra Mammary Paget's disease in a Geriatric Patient

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Background: Paget's disease is a rare form of adenocarcinoma of the breast. It involves apocrine gland-bearing skin that presents as a slowly expanding well-defined patch. It presents as an erythematous plaque on apocrine gland bearing areas including vulva, perineum, perianal region, scrotum, and penis. EMPD is a marginated plaque resembling Paget's disease and is very rare. We present a case with biopsy-proven EMPD in the left inguinal area which is an unusual area.

Case Report: A 67-year-old male with a history of renal cell cancer status post left nephrectomy, diabetes, HTN and COPD presented with complaint of fatigue and 40 lbs weight loss in 2 months. On physical examination, a suspicious erythematous plaque on his left inguinal region is noted. According to the patient, this lesion was present for several months and had recently increased in size. Skin biopsy was done that suggested invasive adenocarcinoma with pagetoid epidermal involvement of peripheral and deep margins. Surgery was consulted and a wide local excision of the left groin with sentinel lymph node biopsy was performed. The biopsy was positive for malignant cells. The patient is diagnosed with cutaneous invasive adenocarcinoma. Based on Ohara et al, his staging was pT1N1M0, stage IIIa. The patient underwent lymph node dissection. He was referred to medical oncology for discussion of the benefits and risks of systemic chemotherapy. Underlying malignancy was ruled out based on whole-body enhanced CT and colonoscopy. Since the patient was cancer free pathologically, it was recommended that he undergo routine follow-up every 6 months for 5 years postoperatively for surveillance.

Conclusion: EMPD is considered an adenocarcinoma originating from the skin or skin appendages in areas with apocrine glands. The primary location is the vulvar area, followed by the perianal region, scrotum, penis and axillae. Commonly confined to the epidermis, EMPD can be invasive, associated with contiguous extension or upward pagetoid spread of underlying malignancy or with distant synchronous malignancy. Because of its association with other cancers, formal evaluation is warranted. Surgical excision remains the mainstay of treatment. Conventional chemotherapies have been used for the treatment in patients with distant metastases, but the efficacy is not satisfactory, and the prognosis for such patients is poor.