Abstract
Extramammary Paget’s disease is a rare cutaneous neoplasia, sometimes associated with internal visceral malignancies. It is very rarely encountered in “ectopic” areas that are not associated with the presence of apocrine glands. Treatment with wide surgical excision can be curative although recurrences are not uncommon. We present a case of ectopic extramammary Paget’s disease found on the forearm of a male.

Case
A 67 year old male presented with an asymptomatic lesion on his left dorsal forearm that had been present for about 5 months. Surgical history was significant for a normal colonoscopy 4 years prior to presentation. His only medication was daily aspirin.

On examination, the patient was found to have an erythematous, eczematous-appearing patch on his left dorsal forearm, measuring about 3x1 centimeters. A shave biopsy was obtained which showed intraepidermal proliferation of large, atypical, epithelioid cells arranged both as small nests and single cells. The tumor cells were described as having ample cytoplasm, prominent nuclei, and conspicuous nucleoli while displaying pagetoid spread and involving adnexal epithelium. The intraepidermal proliferation was positive for CK-7. These findings were interpreted as being consistent with extramammary Paget’s disease. The patient returned to the office for wide local excision. Pathology showed residual extramammary Paget’s disease with clear margins. The patient was referred to oncology to rule out occult malignancy. Work-up, including blood tests, radiological scans, and colonoscopy were negative.

Discussion
Extra-mammary Paget’s disease (EMPD) is a cutaneous, intraepithelial adenocarcinoma found outside of the breast. There are rare cases in which EMPD has been found in areas of the body that do not typically contain apocrine glands. These cases have been referred to as “ectopic” EMPD. Ectopic EMPD cannot be distinguished from EMPD other than by its location in a non-apocrine bearing area. While it remains unclear how Paget’s disease arises in non-apocrine bearing skin, it is hypothesized that an epidermal multipotent stem cell may eventually gain an apocrine phenotype and result in EMPD occurring in non-apocrine gland bearing areas.

Clinically, EMPD presents as red to brown plaques with secondary changes of scale or ulceration. The location of the lesion on apocrine bearing skin may provide a clue in the diagnostic process. For this reason, ectopic lesions require a high level of clinical suspicion and necessitate a biopsy.

Treatment of EMPD is with wide local excision, although there are high rates of recurrence. Moh’s micrographic surgery and 5-fluorouracil have also been used successfully.

References