

INTRODUCTION

- Extramammary Paget's disease (EMPD) is a rare cutaneous malignancy most commonly affecting the genitals, perineum, and perianal area.
- EMPD has cellular similarities to Paget's disease of the breast.
- The incidence of Perianal Paget's Disease (PPD) is 1.3% of all cases of Paget's disease.

CASE DISCUSSION

- A 75 year old Caucasian male underwent a colonoscopy for surveillance due to a history of tubular adenomatous polyps.
- Eleven sub-centimeter polyps were resected, none of which showed evidence of colorectal adenocarcinoma.
- During the procedure, a perianal exam revealed irregular circumferential erythematous rash (Figure 1 and 2)
- He was referred to colorectal surgery who performed a perianal skin biopsy which revealed extramammary Paget's disease.
- A computerized tomography of the chest, abdomen and pelvis was performed due to concern for secondary Paget's disease which did not show evidence of neoplasms.
- The patient was given the choice to undergo local wide excision or radiation treatment but given the extensive skin involvement of the PPD, he opted for radiation.

IMAGES



Figure 1 and 2 Perianal erythematous rash and hemorrhoids

DISCUSSION

- There are two types of PPD; primary PPD represents carcinoma in situ of the apocrine gland ducts, whereas secondary PPD is thought to occur from intraepithelial spread of a separate underlying carcinoma.
- Clinically patients may present with anal pain or pruritus, but some may be asymptomatic, as shown in our case. On examination, it appears as a slow-growing, erythematous plaque in the perianal region.
- PPD has been associated with synchronous or metachronous genitourinary and/or gastrointestinal malignancies.
- When confirmed on biopsy, clinicians should closely monitor PPD for local progression and screen all patients for distant and local neoplasms however the appropriate frequency of surveillance remains unknown.

TAKE AWAY POINTS

- Extramammary Paget's disease should be considered as a differential in the diagnosis for perianal dermatitis.
- EMPD is associated with primary skin cancer, GI and GU cancers.
- Consider screening for secondary malignancies once the diagnosis of Perianal Paget's Disease is made.