Annals of Case Reports
Wei K, et al. Ann Case Rep: 9: 102135
www.doi.org/10.29011/2574-7754.102135
www.gavinpublishers.com

OPEN ACCESS

Case Report

GAVIN PUBLISHERS

Extra-mammary Paget's Disease as a Cutaneous Manifestation of Recurrent Rectal Adenocarcinoma

Katherine Wei[1*], Aditya Joshi[2], Arti Gupta[1]

[1]Department of Radiological Sciences, University of California, Irvine, CA, USA

[2]Department of Molecular and Cell Biology, University of California, Berkeley, CA, USA

***Corresponding author**: Katherine Wei, Department of Radiological Sciences, University of California, Irvine, 101 The City Drive South, Orange, CA, 92868, USA

Citation: Wei K, Joshi A, Gupta A (2024) Extra-mammary Paget's Disease as a Cutaneous Manifestation of Recurrent Rectal Adenocarcinoma. Ann Case Report. 9: 2135. DOI:10.29011/2574-7754.102135

Received: 17 December 2024, **Accepted:** 19 December 2024, **Published:** 23 December 2024

Introduction

A 75-year-old male with history of moderately differentiated, invasive, stage T2N0M1a low rectal adenocarcinoma metastatic to the brain, status post laparoscopic abdominoperineal resection and left pelvic peritonectomy, presents with persistent perineal rash. Physical examination reveals pink plaques with overlying erosions and well-defined pink borders at the superior perianal region. Initial differential diagnoses included psoriasis, Paget's disease, contact dermatitis, and less likely fungal infection.

Recent FDG PET-CT performed for routine surveillance for recurrent or metastatic disease shows gluteal cleft skin thickening with associated increased radiotracer uptake measuring up to 5.11 SUV (Figure 1A). Subsequent contrast-enhanced MRI of the pelvis shows subtle T2 hypointense skin thickening and enhancement along the gluteal cleft corresponding to the area of FDG avidity on the PET/CT (Figures 1B and 1C). No other local or distant metastases were noted.

Shave biopsy was performed of the right gluteal cleft which revealed extra-mammary Paget's disease of the anoderm, compatible with a cutaneous manifestation of rectal adenocarcinoma. The shave biopsy was positive for CDX2, which is a sensitive and specific marker for colorectal adenocarcinoma, and not commonly expressed in extra-mammary Paget's disease [1]. The patient was initiated on a 16-week cycle of topical imiquimod (5-FU) treatment. Following treatment, repeat shave biopsy of the gluteal cleft was performed which again showed extra-mammary Paget's disease as a manifestation of either direct extension or metastases of rectal adenocarcinoma. Wide local excision was recommended; however, the patient declined surgery and instead chose to continue imiquimod therapy and routine surveillance.

In addition to the cutaneous metastases, the patient also had prior metastases to the left frontal lobe, for which he underwent left frontal pterional craniectomy for resection and stereotactic body radiation therapy (SBRT) to the left frontal lobe.Pathological analysis of the left frontal lobe mass revealed adenocarcinoma consistent with colorectal primary, with positive KRAS mutation, wild-type NRAS, BRAF, and no markers of microsatellite instability. Abdominoperineal resection and left pelvic peritonectomy was also performed which showed no local lymph node or peritoneal involvement. CEA levels were within normal limits (<2.5 ng/ml) at initial presentation and follow-up evaluations, and ctDNA (circulating tumor DNA) levels were also negative with the exception of one positive result.

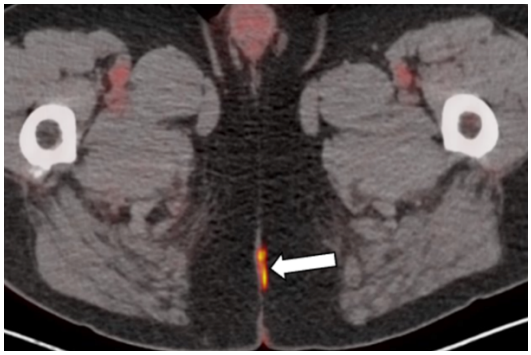

Figure 1A: A 75-year-old man with perianal cutaneous recurrence of rectal adenocarcinoma in the form of extra-mammary Paget's disease.

A. Axial FDG PET-CT shows increased activity at the gluteal cleft (white arrow) measuring up to 5.11 SUV corresponding to mild skin thickening on the accompanying nondiagnostic CT (not shown).

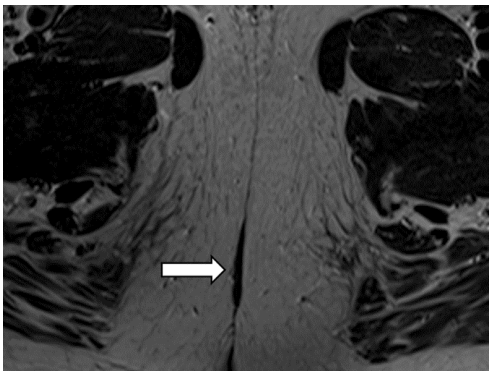

Figure 1B: A 75-year-old man with perianal cutaneous recurrence of rectal adenocarcinoma in the form of extra-mammary Paget's disease.

B. Axial T2 sequence of the patient's pelvic MRI showed subtle T2 hypointense skin/soft tissue thickening (white arrow) along the gluteal cleft which corresponds to FDG avidity on the PET/CT, compatible with patient's known cutaneous manifestation of rectal adenocarcinoma.

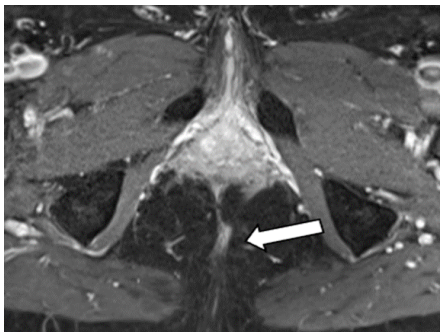

Figure 1C: A 75-year-old man with perianal cutaneous recurrence of rectal adenocarcinoma in the form of extra-mammary Paget's disease.

C. Axial post-contrast T1 sequence of the patient's pelvic MRI showed subtle skin/soft tissue thickening and enhancement (white arrow) along the gluteal cleft, compatible with patient's known cutaneous manifestation of rectal adenocarcinoma.

Discussion

Extra-mammary Paget's disease (EMPD) is a rare, slow-growing adenocarcinoma of the apocrine glands of the axillary and anogenital region [2]. Primary EMPD is a primary intraepithelial adenocarcinoma of the epidermis, while secondary EMPD typically occurs with a primary malignancy of the urinary or gastrointestinal system. The true incidence of EMPD remains uncertain, with estimates as low as 0.12 per 100,000 [3]. Perianal Paget's disease is even more uncommon, with an estimated 200 cases reported in the literature [4].

Of patients with perianal Paget's disease, one cohort analysis estimated 8% of them developed the condition as a manifestation of metastatic rectal adenocarcinoma [6]. Due to the limited number of reported cases, there is no standardized approach to treatment. There have been reported cases of complete remission with topical imiquimod therapy alone [5], while other studies emphasize surgical management for perianal Paget's disease due to its overall poor prognosis [4].

Surgical treatment generally involves wide local excision [3]. Previously reported less invasive management strategies include topical chemotherapy such as imiquimod, photoreactive drugs such as aminolaevulinic acid, radiation therapy, and laser ablation [2,3]. Due to the rarity of this disease and variable treatment methods, further research into the most efficacious therapeutic options is needed to establish a more standardized approach to treatment. Awareness of this uncommon disease entity is also needed on the part of the radiologist when interpreting rectal cancer patient imaging.

References

1. Saad RS, Ghorab Z, Khalifa MA, Xu M. (2011) CDX2 as a marker for intestinal differentiation: Its utility and limitations. World J Gastrointest Surg. 3:159-66.
2. Apikotoa S, Stein J. (2022) Stage 3 perianal extra mammary Paget's disease: A case report of a non-operative approach. Int J Surg Case Rep. 91:106796.
3. Ishizuki S, Nakamura Y. (2021) Extramammary Paget's Disease: Diagnosis, Pathogenesis, and Treatment with Focus on Recent Developments. Curr Oncol. 28:2969-2986.
4. Kyriazanos ID, Stamos NP, Miliadis L, Noussis G, Stoidis CN. (2011) Extra-mammary Paget's disease of the perianal region: a review of the literature emphasizing the operative management technique. Surg Oncol. 20:e61-71.
5. Isik O, Aytac E, Brainard J, Valente MA, Abbas MA, Gorgun E. (2016) Perianal Paget's disease: three decades experience of a single institution. Int J Colorectal Dis. 31:29-34.
6. Dos Santos JS, Bonafé GA, Pereira JA, Kanno DT, Martinez CAR, et al (2018) Rare perianal extramammary Paget disease successfully treated using topical Imiquimod therapy. BMC Cancer. 18:921.