

# 與佩吉特氏病相關之肛門腺癌：病例報告及文獻回顧

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## **Perianal Paget's disease Associated with Anal Adenocarcinoma: A Case Report and literature review**

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### **Case:**

The patient is a 65-year-old man with a family history of colon cancer. Three years ago, he came to our OPD because of anal bleeding for one week. The digital rectal examinations showed mixed hemorrhoids and small perianal nodules at 3 and 10 o'clock direction. He received colonoscopy and there was no abnormal mucosa or mass lesion up to the cecum. We suggested biopsy of the lesion on suspicion of Paget's disease, but the patient lost follow-up.

The patient showed up at our OPD due to a newly developed mass lesion of right perianal region with anal bleeding for 3 months. Plaques of perianal region and a 2-cm tumor lesion at 9 o'clock direction were found by physical examination. His follow-up colonoscopy revealed negative finding. We arranged wide excision of the anal tumor and biopsy of perianal lesions. The histopathologic reported in adenocarcinoma of anus, moderately differentiated with epidermal invasion and ulceration, and Paget's disease, CK7(+)/CK20(-).

### **Review:**

Perianal Paget's disease (PPD) is a rare intraepithelial adenocarcinoma. It accounts for 20% of extramammary Paget's disease (EMPD) and commonly develops in apocrine gland-bearing regions, such as the perineum, scrotum, penis, and perianal region. Perianal Paget's disease is associated with nonspecific symptoms, frequently delaying diagnosis. A biopsy should be taken to establish the diagnosis of EMPD and exclude other dermatologic diseases that may present similarly. Immunophenotype CK7(+)/CK20(-)/GCDFP-15(+) is common in primary EMPD, whereas immunophenotype CK7(+)/CK20(+)/GCDFP-15(-) may indicate underlying malignancies.

Nowadays, there is no definite guideline for the management of PPD since it is a rare disease and lack of large randomized controlled study for treatment. Surgery is the mainstay treatment for PPD. For noninvasive PPD, wide local excision with microscopically clear margins is preferred. In cases of synchronous or metachronous advanced anal or lower rectal malignancies, abdominoperineal resection with wide local excision of the cutaneous and subcutaneous tissues is the procedure of choice.

Other treatment options include photodynamic therapy, imiquimod, radiotherapy, chemotherapy, antiandrogen therapy, and carbon dioxide. But these modalities still lack of

long-term follow-up and comparison of treatment effectiveness. In addition, recurrence of Paget's disease is common. Multiple treatment modalities maybe necessary for invasive perianal Paget's disease.