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Xtramammary Paget’s disease (EMPD) is a neoplas
dsm affecting mainly the anogenital region in 
elderly. Because of the rarity of the disease, little is 
known about its prognostic factors and optimal treat-
ment. With regard to treatment, neither the method 
nor the extent of the surgical excision has been stan-
dardized because of the anatomical complexity of 
anogenital lesions. Although recent reports have sug-
gested the potential usefulness of lymph nodes bio-
opsy in EMPD, the management of enlarged regional 
lymph nodes in patients with invasive EMPD is con-
troversial. In this report, we presented a male patient 
who underwent hemorrhoidectomy and Paget’s dis-
ease in hemorrhoid was found incidentally.

\section{Case Report}

This 54-year-old male presented with difficult 
defecation associated with prolapsed anal mass in re-
cent one year. In addition, defecation bleeding was 
found off and on. Grossly, no perianal skin lesion, 
such as erythematous or eczematous change, was 
noted. No rectal mass was identified by the digital 
rectal examination. Mixed hemorrhoid was noted by 
the anoscopic examination. Before surgery, the labo-

datory report showed no anemia or other biochemis-
try abnormality. The patient underwent hemorrho-
idectomy on May 2nd, 2008 and then discharged 
smoothly.

However, the microscopic examination of the 
 specimen showed a picture of nests of tumor cells in 
the epithelia. The tumor cells have abundant pale cy-
toplasm, round to oval nuclei and positive for Mucic-
carmine stain. It proved the diagnosis of Paget’s dis-
ease (Figs. 1 and 2).

\section{Discussion}

Mammary Paget’s disease is caused by the epi-
dermotropic spread of underlying tumor cells, whe-
reas Extramammary Paget’s disease (EMPD) probably arises from intraepithelial cells of sweat gland origin. It was George Thin, in 1881, who was the first to describe the cytological features of Paget’s cells, which appeared microscopically as large rounded cells with abundant pale-staining cytoplasm and a large nucleus that is often displaced to the periphery of the cells. Grow et al. report a 76% association of perianal Paget’s disease with adjacent or bowel carcinoma.

Typically, the lesion is seen as a progressive, erythematous, eczematoid plaque. In addition, cell markers may enhance diagnostic accuracy. Most EMPD cases stain positive for CEA (carcinoembryonic antigen), EMA (epithelial membrane antigen), CK (cytokeratin). Cases associated with internal malignancy usually stain positively for CK20. In this reported patient, positive staining of mucicarmine, EMA (Fig. 3) and CK7 (Fig. 4) enhance the diagnosis of EMPD. In spite of the positive staining of CK20, no definite internal malignancy is identified. Furthermore, the negative staining of HMB-45 excludes the differential diagnosis of melanoma (Fig. 5).

The association of visceral malignancy with
EMPD is well established but the exact relationship is not well understood. It has been associated with carcinoma of the bladder, colon, kidney and prostate.\textsuperscript{11,12} The perianal disease is more likely associated with colorectal malignancy.

Surgical excision is the standard treatment. In the absence of invasive carcinoma, wide excision is the treatment of choice. For more advanced lesions, an abdominoperineal resection, along with inguinal lymph node dissection if positive lymph nodes are present, will be necessary. Whether preoperative chemoradiation is helpful remains unknown. Because of the commonly delayed diagnosis (average 4 years), around 25% of patients with Paget’s disease in this region already have metastases.

However, EMPD usually affects elderly patients and radiation therapy provides an alternative therapy, especially for the patients who may not tolerate surgery. The reported dose of radiotherapy used by Moreno-Arias \textit{et al} ranged from 40 to 50 Gy and was strongly recommended, as a lower radiotherapeutic dose was associated with a higher risk of recurrence.\textsuperscript{13-18}

For in-situ lesions, the prognosis is usually good. However, EMPD may become invasive and metastasize through lymphatic system. A patient with invasive perianal Paget’s disease has a poor prognosis even abdominoperineal resection performed, since distant metastasis has already occurred at the time of diagnosis. In order, the sites of the metastases are the inguinal and pelvic lymph nodes, liver, bone, lung, brain, bladder, prostate, and adrenal gland.\textsuperscript{19} Even after the standard treatment of surgical excision, the lesion may recur and that long-term check-ups may be necessary. The follow-up should be at half-year intervals. Greater vigilance would be recommended especially in those with evidence of dermal invasion, or younger age at presentation.\textsuperscript{20}

In summary, we present a case of prolapsed hemorrhoid containing Paget’s disease. In spite of no internal malignancy identified, long-term follow-up is essential to detect local recurrence and development of invasive Paget’s disease.

\textbf{References}


病例報告

肛門之 Paget’s disease：病例報告

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乳房外之 Paget’s disease 爲一罕見之皮膚癌症，尤其好發於年老病人的肛門或生殖部位。本病例為一 54 歲男性病人，於痔瘡切除手術後，病理檢查意外發現乳房外 Paget’s disease。本篇則進一步討論乳房外 Paget’s disease 的臨床及病理表現，以及治療方式。

關鍵詞  Paget’s disease、肛門或生殖部位。