Case Report

Perianal Paget's Disease Associated with Anorectal Adenocarcinoma – Is Abdominoperineal Resection Necessary?

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Key Words
Perianal Paget's disease;
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Case Report. A 65-year-old man was diagnosed as adenocarcinoma of anus and perianal Paget's disease (PPD) after biopsy of his anal tumor. The patient received neoadjuvant radiation therapy. Two months after radiation therapy, the patient received wide local excision under spinal anesthesia. The patient had no protective colostomy and no reconstructive flaps or skin graft. The patient discharged from our hospital 3 days later. We had followed the patient for one year and there was no recurrence or metastasis.

Conclusions. About 42% of patients with PPD may have concurrent malignancies, and it is an important prognostic factor. Because of limited cases, the treatment of PPD with underlying anorectal carcinoma is diverse. Most of the patients was treated with abdominoperineal resection according to the staging system for perianal Paget's disease proposed by Kyriazanos et al. Neoadjuvant radiation therapy and wide local excision maybe an appropriate treatment in patients with PPD and localized anorectal carcinoma. However, we still need more cases and longer follow-up.

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Paget's disease was first described by Sir James Paget in 1874. Paget's cell is an intraepithelial adenocarcinoma. It commonly appears in the nipple of breast. Extramammary Paget's disease was found mostly in the areas with increased density of apocrine glands, such as perianal region, vulva, scrotum, perineum, and axilla.

Perianal Paget's disease (PPD) is rare and less than 200 cases have been reported.[1] PPD is associated with nonspecific symptoms, mainly eczema-like symptoms, and frequently delaying diagnosis. Perianal Paget's disease is associated with underlying malignancy, especially bowel carcinomas. Because of limited cases, the treatment of PPD with underlying anorectal carcinoma is diverse.

Case Report

A 65-year-old man showed up at our outpatient department due to a mass lesion at right perianal region with pain and intermittent bleeding for 3 months. He has a family history of colon cancer. The patient's physical examination showed a 2-cm tumor lesion at 9

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o'clock direction and 1 cm distal to the anal verge with surrounding erythematous plaques, extending from anal verge to 4 cm distal to anal verge. His colonoscopy revealed negative finding. After excisional biopsy, the pathology revealed adenocarcinoma of anus and perianal Paget's disease. His CT scan of chest, abdomen and pelvis were negative. The tumor markers were within the normal range.

The patient refused our suggestion of abdominoperineal resection. After discussion with the patient and our multidisciplinary team, the patient received neoadjuvant radiation therapy with a dose of 59.4 Gy, divided in 7 weeks. Two months after radiation therapy, the patient received wide local excision under spinal anesthesia. We excised an area six centimeter distal from anal verge and up to one centimeter above the anal canal. The safe margin was at least one centimeter lateral to the affected area. We preserved the internal anal sphincter and attached the rectal mucosa to the internal anal sphincter (Fig. 1). The patient had no protective colostomy and no reconstructive flaps or skin graft. The pathology showed residual Paget's disease. The resection margins were negative for tumor. The patient discharged from our hospital 3 days later.

The patient had OPD follow-up every week after the surgery. The wound became shallow and the size of the wound shrank to 2 cm distal from anal verge one month after the surgery. The wound healed 3 months after the surgery. The patient had no fecal incontinence or anal stricture. He had defecation 2-3 times a day. We had followed the patient for one year and there was no recurrence or distant metastasis.

Discussion

Perianal Paget's disease accounts for 20% of all extramammary Paget's disease and 6.5% of all Paget's disease cases.[2] The age of onset is between 50 to 80. It affects male and female in average. The symptoms of PPD were non-specific, such as eczema, itching, pain and bleeding. As a result, delayed diagnosis was often.

The treatment of PPD depends on the depth of invasion, involvement of regional lymph nodes, and metastasis. Kyriazanos et al.[3] had proposed the staging system and management for perianal Paget's disease in 2011 (Table 1). The mainstay treatment of PPD treatment is surgical resection. The wide local excision needs a lateral margin extending from 1 to 3 cm beyond the clinically affected area.[4] Most of the time, the surgical wound defect needed reconstruction with flaps or skin grafts. If the patient was found to have underlying anorectal carcinoma, abdominoperineal resection and wide local excision were suggested.

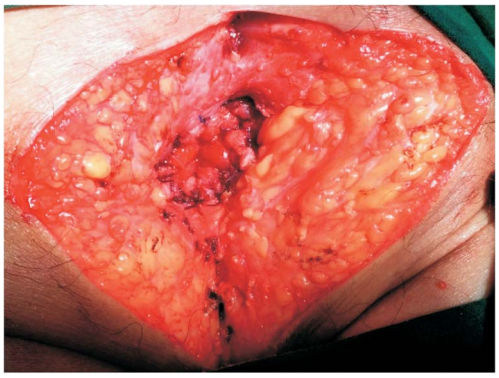

Fig. 1. Wide local excision deep to subcutaneous fat and up to the one centimeter above the anal canal.

Table 1. Perianal Paget's disease classification and accompanying suggested therapy

Stage	Description	Management
I	Paget's cells in the perianal epidermis and adnexa without primary carcinoma	Wide local excision
IIA	Cutaneous Paget's disease with associated adnexal carcinoma	Wide local excision
IIB	Cutaneous Paget's disease with associated anorectal carcinoma	Abdominoperineal resection
III	Paget's disease in which associated carcinoma has spread to regional nodes	Inguinal lymph nodes dissection and abdominoperineal resection/wide local excision
IV	Paget's disease with distant metastases of associated carcinoma	Chemotherapy, radiotherapy, local palliative management

Adapted from Kyriazanos ID, Stamos NP, Miliadis L, et al. Extra-mammary Paget's disease of the perianal region: a review of the literature emphasizing the operative management technique. *Surg Oncol* 2011;20:e61-71.

Chemotherapy and radiation therapy was suggested for patients presenting with distant metastases of associated carcinomas.

There are no definite management guidelines for PPD. Photodynamic therapy, Imiquimod 5% cream, and radiation therapy all had been discussed and used for treatment of PPD.[5] Most reported cases with PPD and underlying anorectal carcinoma were treated with abdominoperineal resection and wide local excision.

Neoadjuvant radiation therapy had become the standard treatment of anorectal carcinoma. For anorectal cancer, the suggested dose of radiation is 45-50 Gy in 25-28 fraction to the pelvis. And for resectable rectal cancer, after 45 Gy a tumor bed boost with a 2-cm margin of 5.4 Gy in 3 fractions could be considered for preoperative radiation; and 5.4-9.0 Gy in 3-5 fractions for postoperative radiation. Radiation therapy used as postoperative adjuvant therapy and in PPD with postoperative recurrence have been reported. The dose in these reported cases were between 40-50 Gy. Our case received a dose of 59.4 Gy for intention to reduce the affected area. The common side effects of perianal radiation therapy include acute dermatitis, moist desquamation and skin atrophy. These could be managed with moisturizing cream.

We preserved most of the internal anal sphincter and some of the external sphincter during wide local excision. The patient had normal defecation after the surgery. The defect of the surgical wound healed by secondary healing. The patient kept the wound clean with water flushing and soaking. These could shorten the surgical time, the hospital-stay and reduce the costs. However, it took more time for the defect to healed by secondary healing than by reconstruction with flaps or skin grafts.

Conclusions

According to the limited literatures, about 42% of patients with PPD may have concurrent malignancies, and it is an important prognostic factor. Although most patients with PPD and associated anorectal carcinoma were treated with abdominoperineal resection and wide local excision, neoadjuvant radiation therapy and wide local excision maybe an appropriate therapy in patients with PPD and localized anorectal carcinoma. Early detection of recurrence can be achieved by close postoperative follow-up. However, we still need more cases and longer follow-up.

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病例報告

肛門周圍佩吉特氏病合併肛門腺癌 – 腹部會陰切除手術是必要的嗎？

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大約 42% 的肛門周圍佩吉特氏病會同時合併其他惡性腫瘤，且是一個重要的預後因子。因肛門周圍佩吉特氏病合併肛門腺癌的病例罕見，目前治療此類病患的方法有許多種，最常見的方式為腹部會陰切除手術。術前放射治療加上廣泛性局部切除手術對於肛門周圍佩吉特氏病合併肛門腺癌可能是一有效的治療方式，但是我們仍需要更多的病例及更長久的後續追蹤來證明。

關鍵詞　肛門周圍佩吉特氏病、肛門腺癌、腹部會陰切除手術、術前放射治療、廣泛性局部切除手術。