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Extramammary Paget's Disease

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SUMMARY: Surgical treatment of perianal Paget' disease using the gracillis musculocutaneous flap was reported on an 83-year-old patient' with some discussions.

INTRODUCTION

The term, extramammary Paget disease refers to a disease which is neoplastic lesion like eczema around genitalien and perianus regions. The extramammary Paget diseases occurred so frequently in heavily growing sites of the apocrine glands that the genitalien and the perianal regions are well known as a predominant sites.

The best way to cure is complete resection before infiltrative extension via lymphatic channels and bloodbore metastasis of cancer cells. However, a great deal of difficulty in wide resection is how to manage the tissue defect. Recently advancement in plastic surgey have made it possible to reconstruct well, functionally and cosmetically. Pedicled musculocutaneous flap is suitable for reconstruction of tissue defect.

Surgical experience with a case suffering from extramammary disease in the perianal region is herein reported in terms of the use of musculocutaneous flap to cover the tissue defevt after the wide resection.

PATIENTS

A 83-years-old man has complained of erosion in the perianal region since December 1987.

As this lesion was gradually extending in its extent, he consulted a dermatist and it was diagnosed as extramammary Paget' disease by biopsy in October 1988. Therefore, he was admitted in our hospital for surgical treatment in February 1989.

His general condition was good, and the values of CEA and AFP were within normal limit.

In the perianal region, erosion and redness were severe, and the lesions of depigmentation and pigmentaion coexisted. It was 9×6cm in size with slight degree of elevation as compared with the normal as shown in **Fig. 1.** Digital examination was free of any induration and stenosis. Colonoscopy revealed no abnormal finding in the proximal portion of the ampulla

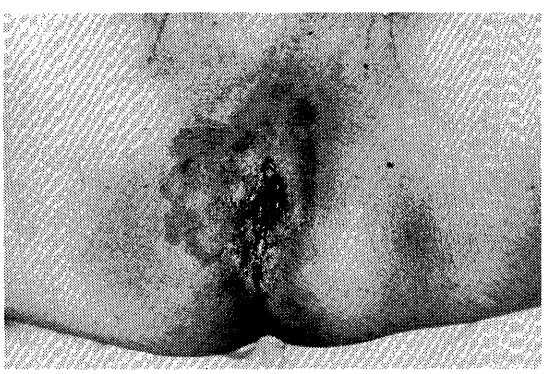

Fig. 1. Preoperative macroscopic findings around the anus, showing erosidon and depigmentation in part.

recti. Surgical finding: The skin, maximun 3cm to minimum 1cm apart from the margin of the lesion, including subcutaneous fatty tissues was widely excised to expose the muscular fascia. In the anus the external sphincter muscles were preserved as much as possible and the anal canal was removed 2 cm proximal to the dented line. The surgically cut edge of the skin and the anal mucosa were histologically examined to exclude Paget cells by using frozen section.

Pedicled musculocutaneous flap was prepared from the gracillis muscles for the tissue defect as shown in **Fig. 2.** In the middle of the pedicled musculocutaneous flap, the anus plasty was made by suturing the anal mucosa to the hole of the skin flap as shown in **Fig. 3.**

The surgical specimen showed the skin of 12.0×8.5cm in size and there were small polypoid lesions with erosion around the anus.

There were numerous large Paget cells with foamy vacuolation cytoplasms and vesicular nuclei. The surgical margin was free of a presence of paget's cells.

Postoperative course: although oral intake was inhibited by IVH control, postoperative infection on day 6 in the anastomotic line between the skin and the anal mucosa led to hasten a diverting colostomy.

Thereafter, infection subsided under the control of drugs, and it was possible to take food orally. However, ensuing stenosis required to make it distended and widended by Hegar dilator. He is alive well, enjoying his daily life until now 1year after surgery.

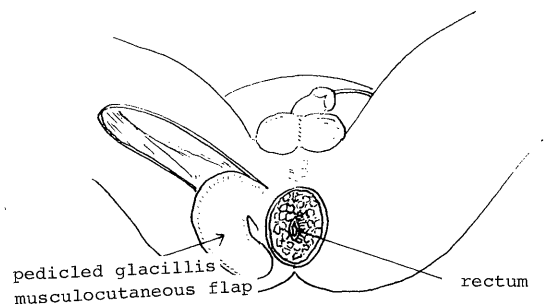

Fig. 2. Schema of the operation method. Wide resection of the skin around the anus and transferred pedicled gracillis musculocutaneous flap.

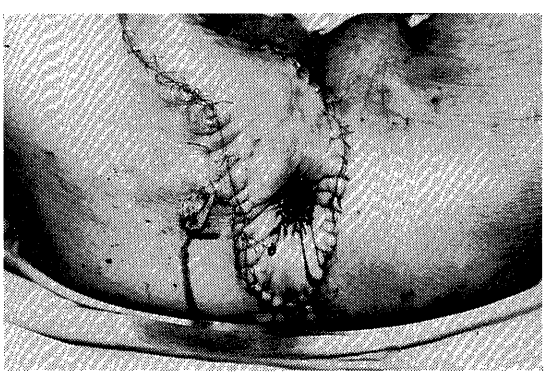

Fig. 3. Macroscopic finding at completion of reconstruction for skin defect around anus.

DISCUSSION

Extramammary Paget's disease predominates in the genitalien, axilla and perianal regions in which the apocrine glands were densely growing.

It is common that this lesion discloses slow extension of cancer infiltration within the epidermis. However, when it is extending to the basal membrane, it becomes infiltrative and cancer spreads widely metastasizing via lymphatic channels.

It is required in the treatment that it should be dealt with a wide resection of carcinoma, less than precussor in carcinoma and/or carcinoma in the epidermis as reported by Ohara[1].

The questions regarding surgical operartion are how to treat metastatic lymphnodes[2], how to determine the resection line[3] how to reconstruct the tissue as Soeba emphasized[2]. Nodal involvement has become manifest in infiltrative carcinoma although carcinoma in the epidermis frequently involved neighboring nodes.

When nodal involvement is suspected, the superficial inguinal nodes should be dissected. If definite nodal involvement exists, the surgical inguinal node as well as the internal iliac nodes must be dissected.

In this case there was no nodal involvement although the lesion was widely extended, as large as 9×6cm in size. Therefore, node dissection was not attempted for the reasons of absence of nodal involvement and the old age of the patient.

As to the resection line, it is recommended

that the resection line should be 3-5cm apart from the diseased margin[2,4]. It is emphasized that the diseased margin is to the margin of depigmentaion[1].

A wide resection is essential to carry out a complete removal and to prevent recurrence.

As it is limited for achivement of wide resection, so intraoperative histology is needed for adequate and minimal resections for this disease.

When cancer extension reaches the rectum, urethra, and vagina, it is not easy to determine the resection line apropriately. In particular, according to the rule of the resection extending to the rectum, amputation of the rectum should be selected as the operative procedure of choice. Soeda insisted that the resection line should be 5cm, at least 3cm, apart from the visible margin of the lesion.

In this case, the resection line in the rectum was kept within 2cm because it was non-infiltrative carcinoma and no residue of carcinoma was found by intraoperatively histologic examination.

As for reconstruction, it is recommeded that pedicled skin graft should be used for prevention of scar constriction of suture line. It is generally accepted that pedicled gracillis musculocutaneous flap is most favorable because it enables us to design easily primary suture and satisfactory wrap for the tissue defect.

It is also emphasized[5] that the pedicled musculocutaneous flap has rich blood flow and high resistance to infection.

Extramammary Paget's disease in the perianal region occurs frequently. When the lesion extends to the rectal mucosa, it is difficult to determine an ideal resection line. As far as we can apply frozen section for this purpose, adequate surgery would be promised.

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