Perianal Paget’s Disease Treated With a Wide Excision and Gluteal Fold Flap Reconstruction Guided by Photodynamic Diagnosis

Report of a Case

Yasumi Araki, M.D., Toshihiro Noake, M.D., Hirohumi Hata, M.D., Kazuya Momosaki, M.D., Kazuo Shirouzu, M.D.

From the Department of Surgery, Kurume University School of Medicine, Kurume, Japan

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Perianal extramammary Paget’s disease is a malignant tumor of the skin invading the epidermis, and the first-choice treatment is a wide excision of the skin. In many cases, however, it is very difficult to know whether the disease is localized or extends to the anus. Photodynamic diagnosis (PDD) is a method more convenient and less invasive than biopsy for the detection of cutaneous infiltration before surgical incision.1–3 We present here a case of perianal Paget’s disease in which PDD was used to delineate the skin to be excised before preservation of the anal sphincter, wide skin excision, and reconstruction with gluteal fold flap and a full-thickness pedicle flap and which had a favorable postoperative course without any anal stenosis.

REPORT OF A CASE

A 72-year-old female was admitted to our hospital complaining of perianal itching sensation that had started in February 2001, and she was diagnosed as having perianal Paget’s disease on biopsy and hospitalized in April 2001. An eczematous erythema was measured and found to be 10 cm × 20 cm around the perianal skin, involving the anal verge. Biopsy of the perianal lesion revealed typical Paget cells, which were large and pale with foamy cytoplasm. Paget cells could not be detected in rectal mucosa and dentate line by biopsies performed by endoscopy. A 20 percent 5-aminolevulinic acid (ALA) of hydrophilic ointment was applied to and around the lesion extensively, which was then protected from light. Six hours later, it was examined for red fluorescence by means of fluorescent lamp (FL 205BLB, Toshiba Corp., Tokyo, Japan) under black light (Fig. 1). The area involving the anal region up to the vicinity of the dentate line was found to be fluorescent. These findings allowed us to place markings for an operative safety margin.

The patient was placed in the lithotomy position, and surgery was conducted under general anesthesia. A temporary stoma was constructed with the sigmoid colon, and then a wide excision of the subcutaneous tissue was made with a safety margin of approximately 3 cm, while the anal sphincter was preserved up to the dentate line. A rhombic gluteal fold flap measuring 4 cm × 8 cm was prepared with the perineal artery, and this served as a branched subcutaneous pedicle for a full-thickness pedicle flap and which had a favorable postoperative course without any anal stenosis.

Address reprint requests to Dr. Araki: Department of Surgery, Kurume University School of Medicine, 67 Asahi, Kurume 830-0011, Japan.
DISCUSSION

Perianal extramammary Paget’s disease is a slow-growing intraepidermal cancer characterized by well-defined erythematous erosive lesions. In 1991, Misas et al. used PDD in patients with perianal extramammary Paget’s disease. This is a noninvasive diagnostic tool that exploits the phenomenon that when cancer cells are exposed to ALA, they accumulate intrinsic protoporphyrin IX, which emits red fluorescence under ultraviolet light.

Surgical excision is the treatment of choice for perianal extramammary Paget’s disease; however, an inadequate surgical margin will cause local recurrence. To determine the extent of tumor infiltration, several mapping biopsies are required, which causes false-negative problems and increases the patient’s pain. In the present study, we conducted PDD in conjunction with mapping biopsy for comparison purposes. The location of Paget’s cells was almost exactly the same between red fluorescence and histopathological biopsy specimens. As for ALA, intravenous administration requires the patient to be protected from light, whereas topical application has no adverse effects and is safe and suitable for patients with malignant skin diseases.

Although perianal extramammary Paget’s disease has been treated with topical ALA, PDD, and CO₂ laser, the long-term outcomes of this therapy are not known, and surgery remains the treatment of choice. A wide skin excision that leaves a circumferential loss of skin all around the anus, which is the case with our patient, requires a skin graft for reconstruction. A gluteal fold flap, which induces anal stenosis less frequently than does a split-thickness graft, is a pedicle flap with good mobility and good blood circulation taken from the area adjacent to the excision. The graft donor site looks like a gluteal furrow and does not show. Our patient has no evidence of local recurrence 24 months postoperatively, with good anal function.

CONCLUSION

We report a case of perianal Paget’s disease in which PDD was used to delineate the skin to be excised before preservation of the anal sphincter. The