Case Report

V-Y island flap to cover a defect in the perianal region from perianal Paget’s disease: case report

Kurt Y, Demirbas S, Akin ML, Balta AZ, Filiz AI

Gulhane Military Medical Academy, Haydarpasa Training Hospital, Department of General Surgery, Istanbul, Turkey

Abstract. Perianal Paget’s disease (PPD) is a rare entity which requires surgery with complete excision of the lesion leading a large skin defect in the perianal region. V-Y Island flap technique is a feasible way to cover up the large skin defect in perianal region. Care must be taken to the base of the graft which should be placed in a good shape into the anal canal. Perianal Paget’s disease was diagnosed in two patients after biopsy and they were treated with entire surgical excision. Both are alive at present. No important complication was experienced and there was no need to ostomy. No recurrent disease was experienced in a year follow up. Anal canal function and life quality of the patients are also in good condition. Despite of the fact that the treatment after wide local excision is challenging, we used V-Y island flap which is a simple and helpful surgical procedure to cover the wide skin defect in the lesion located around the perianal region.

Key words: perianal diseases, Paget’s disease, V-Y flaps

Perianal Paget’s Disease (PPD) is a rare condition which engages the whole circumference in perianal region. Paget’s disease is pathognomonic with Paget’s cells that contain pale-staining cytoplasm and large nucleus located peripherally [1,2]. These cells come from glandular basis. The lesion reveals a well-demarcated erythematous plaques which is often pruritic and eczematous. Paget’s disease in perianal region is frequently (about 40%) together with invasive carcinoma of the region [2-4]. Generally Paget’s disease in perianal region is secondary to prostate or colorectal cancers or transitional cell cancer [5]. On the other hand primary PPD has been appeared from intra-epidermal cells of the apocrine gland ducts (or from pluripotent keratinocyte stem cells) [6,9]. The potentially aggressive characteristics of invasive Paget’s disease has assessed distant metastasis; although, minimally invasive Paget’s disease with invasion less than 1mm deeper is similar to non-invasive Paget’s disease [6-8]. Then accepted treatment is wide local excision [2-5]. Significant rate of recurrence is the other challenging problem [4,5]. Several techniques have been described to cover the large perianal defect that can not be easily closed primarily [1,2,4,5].

Case

In the last 4 years, 2 patients with perianal Paget’s disease were evaluated for surgery after preoperative diagnostic biopsy. Both had entire physical and ano-rectal exam. Then flexible sigmoidoscopy and Guiac stool test were done for each. The day before surgery bowel cleaning utilized with fleet phospho-soda and fleet enema together, and prophylactic antimicrobial agent with ornidazole 2x500 mg per oral. (Nidazol®) were employed and continued 5 days postoperatively. Patients were instructed to ingest merely liquids the day before surgery. Two patients were taken to the operating room to perform wide local excision with V-Y flaps covering.

Operational technique

Under epidural anesthesia, patients were laid down on the table in prone Jack-Knife position. Rectum was cleaned by 10% Povidon- Iodine solution. Lesion (Fig 1) exposed clearly by using Lone-Star perianal retractor to perform multiple biopsies for frozen section and they all were 1cm from the edge of the lesion as suggested in the paper by Beck and Fazio [10].

Fig 1. The perianal Paget’s disease around the perianal region

At the dentate line biopsies were taken under care. Excision of the lesion was progressed deeper to the subcutaneous tissue until negative margins were confirmed by frozen section. A V-Y island flap was prepared on side of the perianal region with size matching to the defect (Fig 2).

Fig 2. A V-Y island flap on perianal region is displayed
The dissection for the flap was made deep down to the subcutaneous fat and continued till the island of the flap reached to the dentate line without tension. After the preparation of the inner side of the flap, it was slid to the dentate line and sutured the mucosa of the anal canal all around by using 3/0 vicryl which was used to suture the rest of the flap. The completed flap was in Y shape (Fig 3). Drain was not used in routine.

Fig 3. A V-Y Flap slid in to the anal canal and sutured to the anal mucosa of the anal canal, Note Y shape at completion

Postoperative period

Patients were started on elemental diet and clear liquid diet was advanced with return of bowel function, if necessary, Lomotil for 5 days were also engaged to prevent defecation. There was no complication disturbing the patients’ daily life.

Results

Graft survival was full in both patients. Dressing was kept 3-4 days postoperatively. Sit baths were prescribed after first defecation. The wound completely healed after 3 weeks using simple water baths, soap-water baths and dry dressing after defecation. After 12 weeks anal function was evaluated using fecal incontinence severity index (FISI) and the SF-36, quality of life questionnaire was performed to determine the patient’s anal canal function. While FISI scores [1-3] for two patients were not challenging in the preoperative period, FISI scores for both were also same as in the previous ones. SF-36 questionnaires stated anal canal function was well and there was no restriction at each patient’s daily life.

Discussion

It is not easy to make estimation about the true incidence of PPD because of the rare nature of this disease. It typically presents with a reddish elevated, crusty and scaly lesion which appears in 6th or 7th decade [5,11,12]. Characteristic presenting symptoms are pruritus, irritation, rash and sometimes maceration in the perianal region. It happens to be treated by dermatologists as a benign dermatologic condition in the first place. If the lesion is not healed by any treatment biopsy is performed (12, 13). PPD could be associated with an underlying gastrointestinal cancer, in 12-15% [4,5,14]. Two patients certainly had no underlying carcinoma. It is reported by Goldblum and Hart that PPD with rectal adenocarcinoma have a tendency of endodermal differentiation with gastrointestinal-type glands, frequently positive for CK20 and negative for GCDFP15. In the other type primary intraepidermal neoplasm, Paget’s cells display sweat gland differentiation with GCDFP15 positivity [5,6,9]. It is also reported about the different Paget’s disease that the sialomucins were present in normal anal ducts but they were not present in transitional epithelium of the anal canal. A patient with perianal Paget’s disease showed strongly positive staining, both in the underlying mucinous adenocarcinoma and in Paget’s cells of the affected anal and perianal region. In contrast, stains of other forms of Paget’s disease were completely negative, as well as malignant melanoma and Bowen disease [12].

The treatment of PPD is surgical; wide local excision is the accepted approach and adjuvant therapy has remained controversial [2,4,15,16]. Four-quadrant biopsy mapping is advocated preoperatively, because of the existence of Paget’s cell beyond surgical margins of the lesion [4,10]. In the study, the surgical margins was found positive in 53% of 30 patients with primary PPD [17], and another literature reported microscopically positive margins in more than 50% of cases [18]. On the other hand, the report stated that the use of frozen on punch biopsy has limited value and is so demanding for the pathologist. St. Peter et al. declared that recurrent disease developed in spite of negative margins in their series [20]. Wide local excision has been carried out by large tissue loss which should be covered by using local muscle and/or myocutaneous flaps or skin grafts [1,2,19,20]. As in here, two cases underwent local wide excision covered by V-Y flaps after multiple biopsies on all 4 quadrants taken preoperatively. Frozen section gave no involvement on surgical margins then the flaps were run to the ano-cutaneous line. In this study our technique of V-Y island perianal flaps mentioned above, was found very useful. It was associated with almost none flap-related complications but in just two patients. In the literature the complication about flaps which are frequently seen as anal stenosis and ectropion, are in wide range from 12 % to 65 % [20,21]. In our experience cases are very limited and the flaps were small and they involved only one side of perianal region. Thus strong confirmation is not available at now.

Conclusion

Because coverage after wide local excision is challenging, several different surgical procedures have been described and employed. V-Y island flap is simple and valuable method to cover the wide skin defect in the lesions located in perianal region.

References

6. Choi YD, Cho NH, Park YS, Cho SH, Lee G. Lymphovascular and marginal invasion as useful prognostic indicators and the role of e-erb


