CASE REPORT

Perianal Paget's Disease—a Case Report and a Review of Current Diagnosis and Management

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Abstract Paget's disease is an intraepithelial adenocarcinoma arising from the apocrine gland component of the skin. Paget's disease is most common in the breast but extra mammary disease is also seen. Perianal Paget's disease is a rare form of extramammary disease with few cases reported in literature. It can be primary—arising from the skin or secondary—cutaneous metastases of anorectal or genitourinary malignancy. We hereby wish to report a case of perianal Paget's disease that presented as an eczematous lesion and was diagnosed incidentally on biopsy. After appropriate staging, the patient underwent wide local excision till negative margins were obtained. The resultant tissue defect was successfully covered by split-thickness skin grafting.

Keywords Perianal Paget's disease · Split-thickness skin grafting · Extramammary Paget's disease

Introduction

Paget's disease is a term used to describe an intraepithelial adenocarcinoma arising from apocrine glands. The most common site of involvement is the breast; however, extramammary Paget's disease [EMPD] is also seen. Perianal Paget's disease

[PPD] is a rare entity comprising about 20 % of EMPD with relatively small number of cases reported in literature [1]. Unlike Paget's disease of the breast which is invariably associated with ductal carcinoma, this may not be the case with PPD. The most common associated malignancies are of colorectal and tuboovarian origin [2].

Case Report

A 65-year-old female patient had perianal itching for 6 months which persisted despite treatment with topical antifungals and steroids. She developed a skin tag in the region for which she took surgical consultation. She was treated for fissure in ano with topical creams and the skin tag was excised. The histopathological report showed Paget's disease following which she was referred to us.

On examination, perianal region showed eczematous erythematous lesion along three fourth of the circumference (Fig. 1). Inguinal lymph nodes were not palpable. Per rectal and breast examination were normal. Staging MRI pelvis, CT scan [abdomen + pelvis], colposcopy, cystoscopy, and colonoscopy were normal.

Surgical treatment was planned after discussion with the patient. Wide excision of the diseased area was done with visible clear margin and three-fourth circumference was left open. Histology showed positive mucosa margin between 3 o'clock and 12 o'clock. Redo wide excision was done where entire 360 degree circumferential skin was widely excised along with the mucosa up to the dentate line. Rectal mucosal flap was advanced and mucocutaneous sutures were taken. Histology showed negative margins. Two weeks later, split-thickness skin grafting was done to cover the raw area with 90 % graft acceptance Fig. 1.

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Fig. 1 Shows the eczematous lesion (a), the wound after wide local excision with the mucocutaneous sutures (b) and the result after skin grafting (c)

During follow-up of 2 months, patient had stricture at anal verge for which dilator was used. At a follow-up of 2 years now, the patient is free of disease and is continent to flatus and feces.

Discussion

Etiology and Clinical Features

Based on etiology, PPD is of two types—primary and secondary. Primary PPD is of cutaneous origin and is a primary adenocarcinoma of epidermis. Secondary PPD is of extracutaneous origin

and represents metastases to the skin from underlying visceral or adnexal malignancy [3, 4]. PPD presents as a perianal eczematous lesion with associated symptoms of itching, burning, and oozing. These are often treated as benign dermatological conditions but show no response to local therapy. Therefore, any such perianal lesion that persists despite medical treatment given for eczema should undergo a biopsy.

Investigations and Diagnosis

Investigations aim at histopathological diagnosis of the skin lesion, local and systemic staging of the disease, and to rule out the

Fig. 2 The various treatment options for invasive and non-invasive perianal Paget's disease

presence of underlying anorectal, genitourinary, or breast malignancy.

Adequate histological examination requires full-thickness samples of the affected skin. The microscopic presence of Paget's cells is diagnostic which appear as large cells with hyperchromatic nuclei and pale clear cytoplasm [5]. Investigation modalities for staging include a CT scan of abdomen and pelvis, ultrasonography of the pelvis, sigmoidoscopy, cystoscopy, mammogram, colposcopy, and tumor markers. A hysteroscopy and laparoscopy may be added if clinically relevant. A MRI pelvis may be considered in case of anorectal malignancy for better local staging [6].

Staging

Stage I—Paget's cell in perianal epidermis and adnexa without primary carcinoma

Stage IIA—cutaneous Paget's disease with associated adnexal carcinoma

Stage IIB—cutaneous Paget's disease with associated anorectal carcinoma

Stage III—Paget's disease associated carcinoma has spread to regional lymph nodes

Stage IV—Paget's disease with distant metastasis of associated carcinoma

Stage I and II are termed as non-invasive whereas stage III and IV are called invasive [7].

Treatment

Treatment is decided by the local extent of disease, lymph node involvement, and systemic involvement. Surgery forms the mainstay of the treatment but numerous non-surgical options are available. Due to the rarity of this disease, controlled comparative studies between then surgical and non-surgical treatments are not available.

Surgical options comprise of local excision [macroscopic clearance of margins], wide local excision [>1-cm microscopic clearance of margins], and surgeries for removal of underlying carcinoma such as abdomino-perianal resection. Attaining adequate margins can be aided by frozen section of margins and Mohs micrographic surgery. Resection aims at achieving three-dimensional disease free resection margins. Management of the tissue defect can be done by vacuum-assisted closure devices, split-thickness skin grafts, or local reconstructive flaps [6].

Non-surgical modalities include radiotherapy, photodynamic therapy, topical 5-fluorouracil, bleomycin or imiquimod [an immunomodulator], systemic chemotherapy, argon beam laser therapy, or cryotherapy. These modalities may be considered in patients who are medically unfit for surgery, those who wish to

avoid surgery and those who have a multifocal widespread disease precluding resection Fig. 2 [8].

Prognosis

Even after obtaining wide and negative margins, recurrences are very common because of tumor multifocality and noncontinuity. Overall recurrence rates are reported around 40 to 50 % with a higher incidence in invasive than non-invasive disease. Despite higher recurrence rates, with appropriate treatment, long-term survival can be expected with some case series demonstrating 33 % 10-year survival rates [9].

Conclusion

PPD should always be considered as a differential in perianal eczematous lesions that are refractory to treatment. The use of investigations for pre-treatment staging is essential before planning therapy. Surgery forms the mainstay of treatment but new non-surgical modalities are being developed. There is a need for controlled trials comparing various therapies.

Compliance with Ethical Standards

Conflict of Interest The authors hereby declare that they have no conflict of interest.

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