

Recurrent Perianal Paget's Disease

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Abstract

Extra-mammary perianal Paget's disease (EPPD) is a rare skin disorder of unknown aetiology, which is frequently associated with malignancy. Here we report a case of recurrent perianal Paget's disease who had pruritus on that area for years which has no association with a malignancy and comments upon its diagnosis and treatment. The differential diagnosis of pruritus ani includes cutaneous candidiasis, tinea cruris, contact dermatitis, intertrigo, lichen simplex chronicus, psoriasis, Bowen disease and even basal cell carcinoma; most of which are seen commonly. PPD is a rare disorder, patients should be examined carefully not to miss the diagnosis.

Keywords: Extra-mammary perianal Paget's disease (EPPD); Multiple tubulovillous adenomas; Colonoscopy

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Introduction

Extra-mammary perianal Paget's disease (EPPD) is a rare skin disorder of unknown aetiology, which is frequently associated with malignancy. Here we report a case of recurrent perianal Paget's disease which has no association with a malignancy and comments upon its diagnosis and treatment.

Case

A 58 year-old man was referred to our dermatology department because of a perianal skin lesion that was recognized during a colonoscopy procedure which was performed for constipation. His lesion was a 4 cm round plaque which was erythematous at the center with a surrounding rim of whitish membranous appearance in the right perianal area (**Figure 1**). He had pruritus on that area for years. The patient told that he had a similar condition which was operated two years ago.

Histopathologic examination of his skin biopsy was reported as epidermal infiltration of polygonal atypical cells with round pleomorphic nucleus and prominent nucleoli and wide pale eosinophilic cytoplasm along the basal membrane (**Figure 2**).

These atypical cells were heavily stained by carcinoembryonic antigen (CEA) immunohistochemically (**Figure 3**). No other colonic, intestinal, or urogenital mass or sign of neoplasm was detected by imaging modalities. It was revealed that he had undergone the similar processes 2 years ago. The patient is diagnosed as primary recurrent perianal Paget's disease (PPD). An operation for a wide local excision was planned.

Discussion

Extramammary Paget's disease can be found in various locations, including the patients typically complain of anal pruritus, bleeding, and discharge [1]. Approximately 12% of patients with anal margin Paget's disease harbor a colorectal neoplasm mandating full colonoscopy and 24% of patients have associating cutaneous adnexal adenocarcinoma [2].

No malignancy was detected in our patient by abdominal ultrasound and computed tomography, a simple polyp was excised during colonoscopy in the rectum, which was reported as fibroid polyp histopathologically.

Pruritus ani was the main complaint of this patient which was present for many years. The patient had the same story 3 years



Figure 1 Erythematous at the center with a surrounding rim of whitish membranous appearance in the right perianal area.

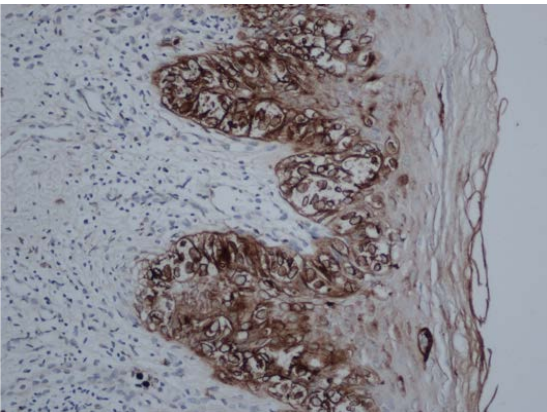


Figure 2 Skin biopsy was reported as epidermal infiltration of polygonal atypical cells with round pleomorphic nucleus and prominent nucleoli and wide pale eosinophilic cytoplasm along the basal membrane.

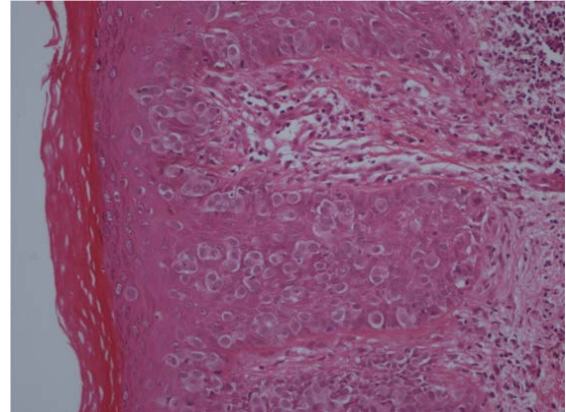


Figure 3 Atypical cells were heavily stained by carcinoembryonic antigen (CEA) immunohistochemically.

ago which was ended by excision of the lesion on the right perianal area. Multiple tubulovillous adenomas had been resected during work up at that time. Most authors recommend surgery as the treatment of choice [2-4]. Besa et al. recommends radiotherapy for patients not suitable for surgery, or post operatively [5,6]. Amin reported a successfully treated patient with radiotherapy who had a history as recurring more extensively four times after each excision [7]. A wide excision is planned for our patient.

The differential diagnosis of pruritus ani includes cutaneous candidiasis, tinea cruris, contact dermatitis, intertrigo, lichen simplex chronicus, psoriasis, Bowen disease and even basal cell carcinoma; most of which are seen commonly. PPD is a rare disorder, patients should be examined carefully not to miss the diagnosis. Wide local excision remains the treatment of choice with an evolving role for various adjuvant therapies. Despite adequate resection long term outlook remains guarded due to high recurrence rate.

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