

Perianal Paget's Disease

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We report a rare case of Perianal Paget's disease. It presents with non-specific anal symptoms. The treatment of choice of Perianal Paget's disease is surgical excision. This patients due to refusal of any surgical intervention, managed successfully with radiotherapy. The patient had an eventful recovery after this conservative method.

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Perianal Paget's disease is a rare intraepithelial adenocarcinoma. The lesion was named after Sir James Paget, who described 15 patients with a characteristic breast lesion in 1874 while the first case of perianal disease was reported in 1893 by Darier and Couillaud^{1,2}. Since then, fewer than 120 cases of perianal Paget's disease have been described in the literature.

The average age of the patient with Paget's disease is usually 65 years of age and it affects male and female equally³. They present with non-specific complaints of anal itching, burning, or bleeding. Perianal lesions are well-demarcated eczematous plaques that are either ulcerative and crusty or papillary. Less commonly these lesions may have a gross appearance similar to other diseases as described in Bowen's disease.

Microscopically, the disease is characterized by large, faintly basophilic or vacuolated cells located in the epidermis. The nuclei are vesicular and demonstrate little mitotic activity. In contrast to Bowenoid cells, the Paget cells become highlighted with a periodic acid-Schiff (PAS) stain owing to their high mucin content⁴.

Treatment of perianal Paget's disease is usually regarded as surgical. Although most authors describe local recurrence even after extensive local resections. Local recurrence and the morbidity from surgery, especially in the elderly, can be high. Radiation therapy, as the primary therapy modality, is seldom used in this condition and the few reports that do include radiation poorly describe treatment selection, radiation dose, field size, treatment technique, beam energy or the outcome of treatment^{3,5}.

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We report eighty-one year old patient who presented with itching of perianal skin lesion of four months duration. The skin biopsy showed perianal Paget's disease and the patient had radiotherapy as the primary curative treatment. Relevant literature of perianal Paget's disease has been reviewed.

THE CASE

Eighty-one year old Bahraini male who is diagnosed with diabetes mellitus, hypertension, osteoarthritis of both knees and peptic ulcer, was seen in the surgical clinic as a referred case from dermatology clinic where he presented initially with a complaint of pruritis ani and perianal reddish scaly skin lesion of four months duration. In addition, the patient was complaining of constipation for the last eighteen months with occasional attacks of per rectum bleeding.

There was no history of nausea, vomiting, loss of appetite or decrease in weight. The patient had done proctosigmoidoscopy and band ligation.

On physical examination, the patient was afebrile, normotensive and had no pallor or jaundice. His abdomen was soft and non-tender, with no ascites or palpable organomegaly. His rectal examination showed a large erythematous plaque like lesion that extends three centimeters circumferentially around the anus and inward to the anal mucosa (see figure1). There was severe stenosis of the anal orifice and no palpable inguinal lymphadenopathy.

Figure 1

The laboratory values were all within normal limits. His liver function tests and CEA were within the normal limits, while his stool occult was negative.

Rigid proctosigmoidoscopy, barium enema and colonoscopy examinations were performed. No colorectal mucosal lesions were seen.

The patient was advised for local excision of his perianal lesion but unfortunately he refused the operation and asked for other modalities of treatment. He was referred to the oncology unit, where he was started on radiotherapy cycles for 8 weeks. He was treated with a combination of beam energy of 6mV(Photon) and 9meVelectron (Electron) beam, focal skin distance of 30 cm, using an applicator size 15x15cm. A total surface dose of 60Gy was prescribed in 30 fractions over 8 weeks period. The patient had responded very well with radiotherapy with no complications and the perianal skin lesion had disappeared completely.

Thereafter, the patient was seen regularly. Follow-up at six months showed good cosmetic result and no sign of local recurrence.

Figure 2

DISCUSSION

Extramammary Paget's disease can be found wherever apocrine glands are located and this can be in the axilla and anogenital area labia majora, penis, scrotum, groins, pubic area, perineum, thigh, and buttock.

The origin of Paget's cell has been debated for many years. Theories included primitive multipotential epidermal cells or elements of glandular origin. Recent work supports a glandular origin as these cells contain low molecular weight cytokeratins and carcinoembryonic antigen⁴.

In Paget's disease, progression of the lesion into an invasive carcinoma has been reported as high as 40% in untreated lesions¹¹. While, the incidence of associated malignancies with Paget's disease ranges from 38 to 86%^{4,5,6}.

The treatment of choice for non- invasive lesions is wide excision surgery, which is the standard treatment⁷. However, invasive lesions are managed by excision of the rectum or abdominoperineal resection.

Besa et al support the use of primary radiotherapy for patients not considered suitable for surgery and for the use of postoperative radiotherapy following resection⁸. They had 36-66% rate of regression of the margins of disease after surgical excision alone in their series of 65 patients with Paget's disease of the perianal skin. The ability of surgery adequately to control a multicentric widespread process is limited and likely to be associated with considerable morbidity or functional impairment.

Recurrences following radiation therapy occurred mainly in patient receiving less than 50Gy ; therefore Besta et al recommended doses greater than 50Gy^{8,9}.

There are reports advocating the use of chemotherapy in these patients⁹. Secco et al described eighty-five year old patient with perianal Paget's disease who refused surgery and was treated with combination of radiation therapy and chemotherapy using 5 fluorouracil and mitomycin C¹⁰.

Similarly, Thirbly et al successfully treated sixty-nine year old patient with extensive perianal Paget's disease with chemo-radiotherapy after he refused to undergo abdominoperineal resection¹¹.

In this patient, we found that the most suitable treatment was radiotherapy due his co - morbidities and his refusal of surgical management. Comparatively, the outcome of the treatment with radiotherapy was favorable for perianal Paget's disease. Although, the outcome was favorable, there are no set standard to this date which mode of treatment can be followed to guarantee success. It is still under continuous study in international centers. Brown et al reported very good outcome with radiotherapy in his patient who had perianal Paget's disease in 2002¹².

One of the larger series reported to date was 27 patients by McCarter et al in May 2003 which recounted the long-term outcome of perianal Paget's disease using standard surgical management of the disease¹³.

CONCLUSION

Perianal Paget's disease is a very rare disease with less than 120 cases reported worldwide. Although surgical excision is reported by many authors to be the treatment of choice, other modalities of treatment are used increasingly but are not standardized yet. Radiotherapy can be used as the primary therapy in patients not fit or refusing surgery.

In this paper we presented a case of perianal Paget's disease who was treated successfully with radiotherapy.

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Figure 1



Figure 2