Perianal Paget's disease

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Perianal Paget's disease is a rare condition with fewer than 100 cases reported. We present a case and its treatment with wide local excision and cutaneous flap reconstruction.

CASE HISTORY

A 74-year-old man had a 34-year history of generalized psoriasis and a 4-year history of perianal rash that did not respond to anti-psoriasis medications. Biopsy showed the lesion to be perianal Paget's disease (Figure 1). There was no history of weight loss or rectal bleeding. He had no other medical conditions and there was no relevant family history.

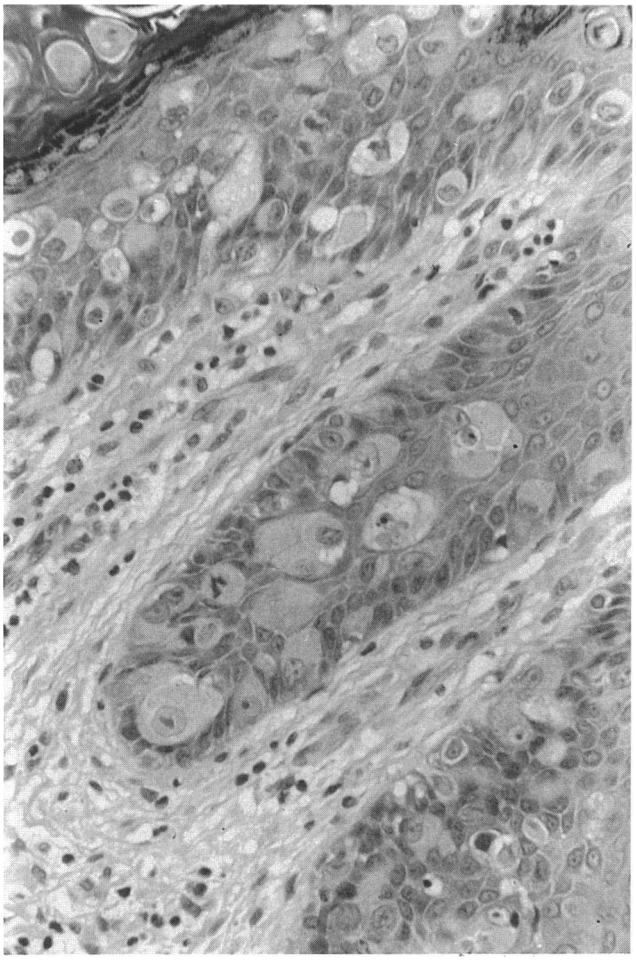

Figure 1 **Mucin laden Paget cells within dermis.** Haematoxylin and eosin

Examination under anaesthesia revealed an excoriated area extending circumferentially from the anus with a diameter of 7 cm into the anus, up to the dentate line. Subsequent flexible sigmoidoscopy and barium enema showed the remaining colon to be disease free except for a few sigmoid diverticula. A laparoscopic colostomy was fashioned as a preliminary to definitive surgical excision. Postoperatively the patient developed urinary retention secondary to benign prostatic hyperplasia. A transurethral resection of the prostate was performed and 2 weeks afterwards the perianal Paget's disease area was formally excised. The perianal skin was excised *en bloc* together with anal mucosa up to the level of the dentate line. The residual raw area was closed with bilateral buttock transposition flaps, which were sutured direct to rectal mucosa (Figure 2).

The patient made an uneventful recovery and was discharged home on the fifth postoperative day. Histology confirmed Paget's disease with clear resection margins and no evidence of invasion.

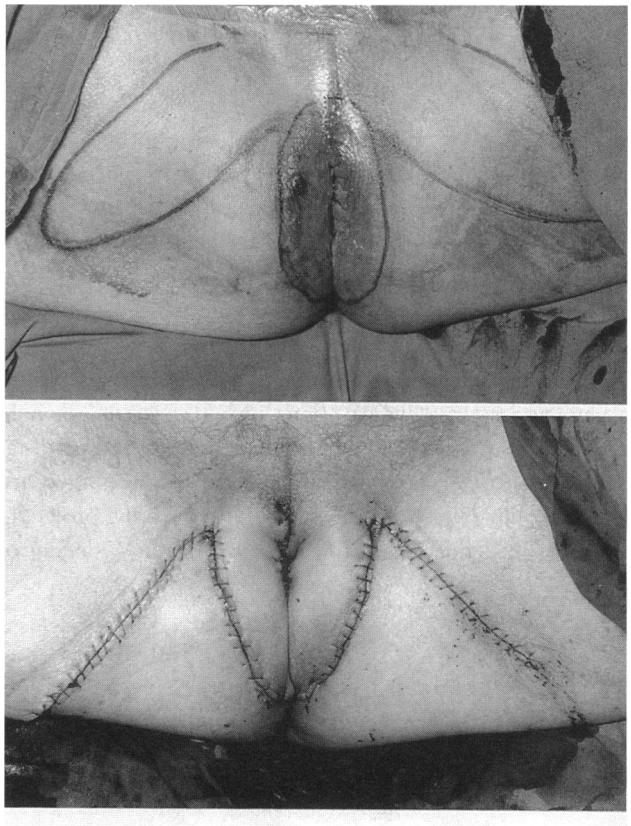

Figure 2 **Preoperative appearance (upper) and postoperative excision and cutaneous flap reconstruction (lower)**

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COMMENT

Perianal Paget's disease, first reported in 1893 by Darier and Coulillaud[1], is believed to correspond to an intraepithelial adenocarcinoma arising from dermal apocrine sweat glands.

About 100 cases have previously been reported[2]. Patients with this condition tend to present with non-specific symptoms such as anal itching, burning or bleeding. Macroscopically the lesion resembles an erythematous plaque which may be ulcerative and crusty or papillary. Microscopically perianal Paget's disease is characterized by large basophilic or vacuolated cells in the epidermis. With their high mucin content, the Paget cells stain heavily with periodic acid-Schiff. They also contain low-molecular-weight cytokeratins and carcinoembryonic antigen; the latter can be seen by immunofluorescence or immunohistochemistry[3]. Paget cells also express gross cystic disease fluid protein, which can be regarded as a specific marker for the cell[4]. Expression of c-ErbB-2 oncoprotein may play a role in promoting intraepithelial spread of adenocarcinoma cells[5].

Perianal Paget's disease may be associated with underlying malignant disease, so a search for synchronous or metachronous tumours is essential before treatment. When invasive growth is absent wide local excision is recommended[3]. For invasive carcinoma or lesions associated with synchronous malignancies, abdominoperineal resection is the treatment of choice. Radiotherapy has no place in treatment because of a high recurrence rate after its use[6].

Depending on the extent of perianal Paget's disease, a covering colostomy may be required before wide local excision. The surgical defect created may be repaired by cutaneous flaps (as in the case presented) or, if needed, bilateral myocutaneous flaps[7]. Preservation of bowel function is the aim. The covering colostomy is usually closed at six months.

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Clicking hyoid

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Abnormalities of the hyoid bone can be a cause of puzzling symptoms and signs.

CASE HISTORY

A woman aged 66 gave a six month history of clicking in her throat on swallowing. This was associated with a feeling of 'heaviness' in the same region. There was neither pain nor difficulty on swallowing. She did not have any respiratory symptoms or jaw-locking and denied any history of trauma.

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On clinical examination the hyoid bone was found to be large, with palpable greater horns that were abnormally long, extending all the way back to the cervical vertebrae. A click was detectable on palpation of the greater horns as they were caught and slipped off the spine on swallowing. Otolaryngological and head and neck examination was otherwise unremarkable. Plain X-ray of the neck showed the greater horns in contact with the lateral mass of the third cervical vertebra (Figure 1).

There were no associated abnormalities. The patient was offered the choice of conservative or surgical treatment. Because of the mild nature of the symptoms she chose the former. Reassurance was all that was required.

COMMENT

The hyoid bone is U-shaped, with greater horns extending posteriorly from each side. The lesser horns arise at the point of junction between the greater horns with the body. The base of the greater horn has a lateral boss of bone where a fibrous sling is attached through which the intermediate tendon of the digastric muscle glides freely. This is straddled by the insertion tendon of the stylohyoid