

also associated with less intraoperative bleeding and provides for better endoscopic follow-up of the remaining proximal intestine.⁵⁻⁸

In order to stage and make therapeutic decisions in duodenal polyposis, the Spigelman classification is used.^{5,8} This endoscopic-histological scale involves a scoring system with 4 items (number, size, histology and dysplasia of the polyps) that classifies duodenal involvement into 5 stages.¹ It is currently accepted that Spigelman grades IV and III with high-grade dysplasia require duodenal resection. Grades I, II and III without high-grade dysplasia are treated by endoscopy.⁵ The duodenal involvement of our patient corresponded with a Spigelman grade II. According to this classification, it could have been treated endoscopically, while preserving the duodenum. The gastric involvement could also be treated endoscopically, but the malignant degeneration of a gastric polyp in the context of a patient with this preneoplastic disease led us to the decision to perform gastrectomy. After gastrectomy and reconstruction of the tract with a Roux-en-Y anastomosis, the duodenal stump was excluded, which meant that posterior endoscopic monitoring of the duodenal polyps would not be possible, and duodenal resection was therefore justified. In a review of the literature, we found no similar cases.

In conclusion, total gastroduodenectomy with pancreatic preservation is a complex technique that could be considered an alternative to pancreaticoduodenectomy in cases of gastric polyposis, malignant degeneration and concomitant duodenal polyposis in GS, as it provides for endoscopic control of the proximal portion of the remaining small bowel.

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Anal Squamous Cell Carcinoma in Chronic Severe Perianal Crohn's Disease[☆]

Carcinoma escamoso de ano en enfermedad de Crohn perianal severa de larga evolución



Crohn's disease is a chronic, recurrent, inflammatory bowel disease. One of its most frequent manifestations is active perianal fistula, which can lead to severe perianal sepsis.

Although the appearance of squamous carcinoma over a fistula in perianal Crohn's is extremely rare, its development could be due to the cellular stimulus associated with

chronic suppuration, immunosuppression or the use of anti-TNF.¹

We present the case of a 59-year-old male, who was a smoker and had had Crohn's disease for 25 years, with no extraintestinal manifestations, and fistulising ileocolic involvement, associated with perianal disease. The patient's

progress was torpid and several hospitalisations were necessary: ileocecal resection (1990), perianal abscess (1996), right hemicolectomy (2000), perianal abscess and fistulotomy (2003). Since the onset of the disease, the patient had been intermittently treated with antibiotics, mesalazine, azathioprine and corticosteroids. Furthermore, in 2007 the patient presented a melanoma in the left thigh that was treated with interferon, surgery and radiotherapy.

After commencing treatment with interferon, the patient reported worsened abdominal and perineal symptoms. In 2008, he presented severe perianal recurrence that did not respond to conservative treatment and required drainage as well as debridement of 3 fistula tracts (Fig. 1). At this time, the patient was referred to our hospital.

Examination revealed inflammation, perianal and gluteal cellulitis, purulent drainage with compression of the abscess cavities and multiple fistulous openings, together with extensive perianal scar tissue.

Complimentary tests showed evidence of marked destructuring of the sphincter apparatus, a large perianal collection (2 cm×4.5 cm×10 cm) extending to the right ischio-rectal fossa, left pararectal and intergluteal folds, 2 large fistulous tracts on both sides of the anal canal to the supralelevator region, proctitis, and important inflammatory changes in the perirectal fat, perianal and intergluteal regions.

Given these findings, we decided to operate in order to control the perianal sepsis with extensive cutaneous debridement and immediate drainage of the abscess cavities. The fistulous tracts were identified and redirected, partial fistulectomy was done of the secondary tracts, followed by a lateral diverting colostomy (Fig. 2). Biopsies were taken. The pathology study reported keratinizing squamous-cell carcinoma over a fistulous tract in the left perianal region, although the precise invasion type could not be specified, so we decided to proceed with abdominoperineal amputation.

The pathology study reported transmural inflammation with mucosal ulcerations, moderate glandular epithelial dysplasia in the rectum and keratinizing squamous-cell carcinoma, excrecent and infiltrating the perianal tissue without affecting the rectal wall. The resection margins of the surgical specimen had a very high probability for involvement.

During this same hospitalization period, a recurrence of the cutaneous melanoma was once again removed by the Plastic Surgery Unit.



Fig. 1 – Perianal sepsis secondary to Crohn's disease.



Fig. 2 – Squamous carcinoma of the anus developed over perianal Crohn's disease.

Due to the high probability of residual tumour in the surgical margins and the inability for complete exeresis, palliative radiotherapy was initiated. Nonetheless, a bleeding, ulcerated and papilliform lesion developed in the perineum, which measured 8 cm×3 cm×6 cm. The pathology study of the lesion was compatible with carcinoma relapse. The lesion progressed and fistulised to the small bowel; the patient died a few days later.

The incidence of anorectal fistulas in Crohn's disease is from 20 to 25% and reaches 60% in rectal Crohn's. Due to the nature of the disease and its difficult treatment, these fistulas can become chronic.² Chronic disease is a predisposing factor for stenosis, sphincter destruction and even malignant transformation. Delayed diagnosis of malignancy is frequent, as the symptoms are usually attributed to the fistula and biopsy samples may be taken later on.^{1,3} In practice, anal examination under anaesthesia, endoanal ultrasound and magnetic resonance provide adequate assessment.

Perineal sepsis is the most severe complication of acute perianal disease. It requires intensive treatment using antibiotic therapy, immediate drainage of the abscess cavities and, frequently, prolonged use of drainage setons. If perianal sepsis progresses, a diverting stoma or even proctectomy can be required.

Anal canal carcinoma is an uncommon disease, and squamous carcinoma is the usual histological variety.⁴ Two very important aetiological factors involved in their development are human papilloma virus infection and chronic immunosuppression.

Symptoms are nonspecific; therefore, a high rate of suspicion is required, as well as a complete exploration and biopsy of any doubtful lesion.

Chemo-radiotherapy is the standard treatment of invasive carcinoma of the anus.⁵ Early-stage lesions (T1) should be evaluated for initial surgical treatment. The rate of complete response with chemoradiotherapy is 75%, and the overall 5-year survival rate is 66%.

The appearance of a squamous-cell carcinoma over a fistula due to perianal Crohn's disease, as in our patient, is extremely rare (0.7%).^{6,7} The causal relationship is unknown

(probable influence of constant cellular regeneration and the state of immunosuppression).^{1,8}

Treatment is the same as for squamous carcinoma of the anal canal, involving chemotherapy, radiotherapy or surgery, with a poorer prognosis as it develops on inflammatory tissue.^{6,9,10}

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