Moderately differentiated squamous cell carcinoma associated with pilonidal cyst

ABSTRACT

Background: Malignant degeneration of pilonidal sinus is a rare complication observed mainly in recurrent chronic cases of the disease and is associated with a poor prognosis. In the international literature, it is an infrequent entity. In Spain, 367 patients with simple and complicated pilonidal cysts were reviewed, finding three cases (0.81%) of epidermoid carcinoma.

Clinical case: We present the case of a 60-year-old male with a clinical picture of 50 years of evolution of the presence of a sacral tumor. The tumor was resected and the histopathological report was pilonidal cyst. Evolution of healing showed necrotic edges that were resected, reporting squamous cell carcinoma.

Conclusions: Epidermoid carcinoma of the pilonidal sinus is an infrequent disease with an unfavorable prognosis.

Key words: Pilonidal sinus, epidermoid carcinoma.
INTRODUCTION

Pilonidal sinus is a common condition, especially in males, and often becomes complicated by local infections. It is usually located in the sacrococcygeal area and also in the umbilicus, armpit, sole of the foot, penis, clitoris and anal canal. Controversy continues whether these pilonidal cysts are congenital or acquired. Asymptomatic cases are usually treated conservatively and those that are symptomatic are surgically removed. Despite the technique used, there is a high rate of postoperative recurrence. Malignant degeneration is a rare complication observed mainly in cases of chronic recurrent disease. It is associated with a poor prognosis compared with cutaneous neoplasms of the same nature. Its rate of recurrence after surgical treatment is high. So far, in the international literature, only 62 cases have been published.1 A new case is reported in the General Surgery Service of Hospital Juárez of Mexico.

CLINICAL CASE

A 60-year-old male patient presented with 50 years of clinical evolution of a tumor in the sacral region of ~3 cm in diameter (Figure 1). During the last 5 years, the patient noted increased tumor volume and growth to the right side of the sacrum; growth was slow and without symptoms. During the prior 6 months, the patient noted more rapid tumor growth, leading him to seek medical consultation. With the above clinical data, pilonidal cyst was diagnosed. Surgical procedure was exeresis and marsupialization with margins of ~1 cm from the pilonidal cyst. Histopathological examination of the surgical specimen reported pilonidal cyst.

Eight days later, the patient presented for a follow-up with discharge of serous and abundant material form at the surgical site. The physician’s indication was to change the dressing at the site three times a day. Within 15 days after surgery, the patient continued with discharge of seropurulent material without adequate wound healing. An increase in body temperature of 38°C was added to his symptoms along with other tumors of ~2 to 3 cm around the surgical wound.

The patient presented to the Emergency Department of Hospital Juárez of México due to continuation with seropurulent secretions from the surgical wound, pain, increased body temperature of 38°C, and weight loss of ~5 kg in 1 month, with no other clinical data.

Physical examination revealed a conscious, calm patient with regularly hydrated mucosa. There were no cardiopulmonary conditions and abdomen demonstrated no hepatosplenomegaly. In the left inguinal region an enlarged, painless lymph node of ~2.5 cm was palpated and in the surgical wound in the sacral region of ~10 × 8 cm with irregular edges. The fascia of the sacrum was observed with necrosis and abundant discharge of hemopurulent material. The rest of the physical examination continued without any pathological data.
Physicians from the General Surgery Service assessed the patient and found abundant discharge of hemopurulent material and tissue necrosis of the surgical wound. Therefore, they decided to treat him surgically with an excision of necrotic tissue and four lesions (~2 to 3 cm in diameter) were found on the edge of the surgical wound. Resection of ~15 × 10 cm of necrotic tissue was done around the wound until reaching apparently healthy tissue. The wound was opened in order to accomplish healing of the second intervention (Figure 2). Histopathology of the resected tissue sent to the Pathology Department reported moderately differentiated squamous cell carcinoma (Figures 3 and 4). The patient progressed adequately with surgical wound healing. Thoraco-abdominal simple and contrast computed tomography (CT) was carried out to assess metastases reported pulmonary mediastinal, liver and inguinal metastases of the left side (Figures 5 and 6).

With the histopathological findings and the CT, the patient was examined in conjunction with the medical oncology service. The surgical wound was observed to be clean, with a small amount of drainage of serous material, presacral fascia was exposed and lower wound borders with
proximity to the perianal region. It was decided to perform a colostomy and to begin radiation treatment. Treatment resulted in an appropriate response, decreasing the size of the lesion in the sacral region. Despite this, the patient had a torpid evolution and died 8 months after treatment.

**DISCUSSION**

Malignant degeneration occurs in ~0.1% of patients with chronic pilonidal disease that goes untreated or relapses during its course. It is believed to be caused by the release of oxygen free radicals by activated cells that induce neoplastic transformation induced by genetic alterations. In addition, the normal mechanism of DNA repair is damaged with chronic inflammation and predisposes to malignant degeneration. According to Davis et al., the first case was described by Wolff in 1900 and, to date, 62 cases have been reported in the literature. Our case has now been added to other published cases. It was confirmed by examination and biopsy of chronic ulcers with friable and necrotic margins, although it can also be found in the histological examination of pilonidal sinus material.

Lesions are usually large and often measure >5 cm in diameter. In most cases, malignant lesions are deep and invade the subcutaneous tissue. Bone tissue is affected in 8% of the cases. Rarely is the diagnosis established after fine needle aspiration of a metastatic inguinal node. In order to exclude extension to the rectum, a colonoscopy may be needed. CT and/or magnetic resonance (MR) have indications to show local extension and to exclude intra-abdominal metastases, as well as extension to iliac and para-aortic nodes. The treatment of choice should be surgical, with en bloc removal, which can present enormous difficulties because the tumor is able to extend through the fistulas into wide areas of the sacrocccygeal and perineal regions. The excision should include, at least, the presacral fascia, but often portions of the sacrum, coccyx and rectum. Closure of the defect can be done with free grafts or LLL-plasty or by using the gluteal or hamstring gracilis. In some cases, use of these is impossible. Therefore, healing should be done by secondary intention, the resulting defect or use of more complex techniques such as myocutaneous plasty of the rectus abdominis or omentum plasty followed by skin grafting. In the case reported it was not possible to cover the defect. Local recurrence after surgery is common (44%) and recurrent disease occurs early. The combination of radical surgery with pre- or postoperative radiotherapy appears to reduce the rate of recurrence (30%). In our case, colostomy was performed because the lesions extended...
almost to the perianal region, warranting treatment with radiotherapy.

Data are lacking for the preferred sequence of surgery and radiotherapy, although preoperative radiotherapy is preferred because it avoids the risk of graft viability and plasty. The results of combined chemo- and radiotherapy are similar to those observed in anal squamous carcinoma. Inguinal lymph node metastases are a sign of poor prognosis and are associated with median survival time of only 7 months. Our patient, who had lymphatic metastases, survived for 8 months.

Controversy exists as to which is the appropriate treatment for these tumors and, according to some authors, regional lymphadenectomy is indicated. Chemoradiotherapy or radiotherapy are considered unproven benefit options. Other authors suggest that for the treatment of anal squamous cancer cells, associated inguinal lymphadenectomy is recommended. However, although chemo- and radiation therapy are the primary treatment in locally advanced anal cancer, its application in pilonidal sinus cancer is not sufficiently proven. In all cases of preoperative diagnosis according to whatever way the patient is intervened, it is convenient to include inguinal lymphadenectomy. Lymphatic mapping with sentinel node biopsy to identify metastases does not seem practical.

The biological behavior of carcinoma of the pilonidal sinus is much more aggressive than squamous carcinoma of other sites. Therefore, knowledge of the possibility of malignant degeneration in pilonidal disease is crucial for early recognition and treatment and for subsequent improved prognosis. Adequate information about the locoregional extent of the disease should lead to the appropriate planning approach and the surgical procedure.

In conclusion, diagnosis of this reported case was established based on examination and biopsy of friable and necrotic tissue. Without a doubt, pilonidal sinus that is removed can be a histological finding. Lesions are usually large and often >5 cm in diameter and with >10 years of evolution. In the vast majority of cases, malignant lesions are deeply invasive in the subcutaneous tissue. Bone tissue is affected in 8% of cases. CT and MR are indicated to demonstrate the extent of and to exclude intra-abdominal metastases and infiltration to iliac or para-aortic lymph nodes and, above all, the inguinal lymph nodes affected early. Treatment with radiotherapy and colostomy, in our case, was a treatment option due to the lesion being very close to the perianal region.

REFERENCES


