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A rare case of squamous cell carcinoma arising from chronic sacrococcygeal pilonidal disease

Pilonidal disease is a chronic disorder which is observed in boys and men between ages of 15 and 40 years old in Caucasian race. Malignant degeneration and carcinoma arising from chronic sacrococcygeal pilonidal disease are observed extremely rare and are described in the group of Marjolin cancer. Carcinoma can be seen on a place with long-term inflammation process as it was in the case described in the patient's article. First time squamous cell carcinoma arising from chronic sacrococcygeal pilonidal disease was described in 1900 by Wolff. The incidence of carcinoma arising in pilonidal disease is less than 0,1% and in the whole medical journals less than 100 patients are reported. This article presents the case of a patient with diagnosed squamous cell carcinoma arising from chronic sacrococcygeal pilonidal disease. In this article was presented the clinical features, pathogenesis, treatment options and prognosis of this rare disease.

KEY WORDS: Pilonidal disease, Skin carcinoma, Squamous cell carcinoma, Surgery

Introduction

Pilonidal disease is a chronic disorder which is observed in boys and men between ages of 15 and 40 years old in Caucasian race [1-4]. Squamous cell carcinoma is an extremely rare complication observed in chronic and recurrent pilonidal disease. Performing the correct diagnosis before surgery and effective treatment makes the results of treatment of the disease should be good. This article presents the case of a patient with diagnosed squamous cell carcinoma arising from chronic sacrococcygeal pilonidal disease. In this article was presented the clinical features, pathogenesis, treatment options and prognosis of this rare disease.

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Case Report

A 58-year-old man, Caucasian race, was admitted to the Department of Surgical Oncology because of diagnosed squamous cell carcinoma arising from chronic sacrococcygeal pilonidal disease. In an interview with the patient, reported that he had been treated for 30 years because of the pilonidal disease, and in this period there was inflammation very often. The patient reports that he had about 15 to 20 incisions and abscesses evacuation in this area in the past. In the last 6 months, he observed rapid tumor growth and pain in this area. After reporting to the outpatient surgical oncology, he had a tumor specimen removed that showed highly differentiated squamous cell carcinoma in the sample.

The patient had no any other symptoms, there was no history of weight loss and loss of appetite. He was not treated for chronically diseases and there was no history of carcinoma in patient family. Blood test and other routine hematological examinations and biochemical tests were within normal limits.

On physical examination, the skin tumor had a diameter of about 7 centimeters. Exophytic tumor, under which there was a large amount of purulent content.

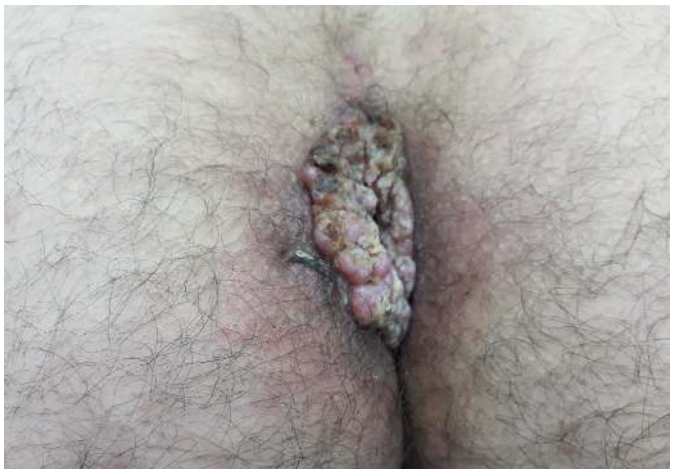

Fig. 1: Squamous cell carcinoma arising from chronic sacrococcygeal pilonidal disease.

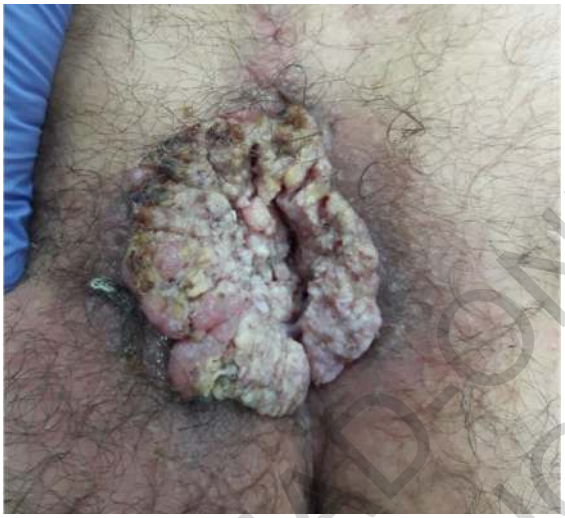

Fig. 2: A large amount of purulent content was present under the tumor.

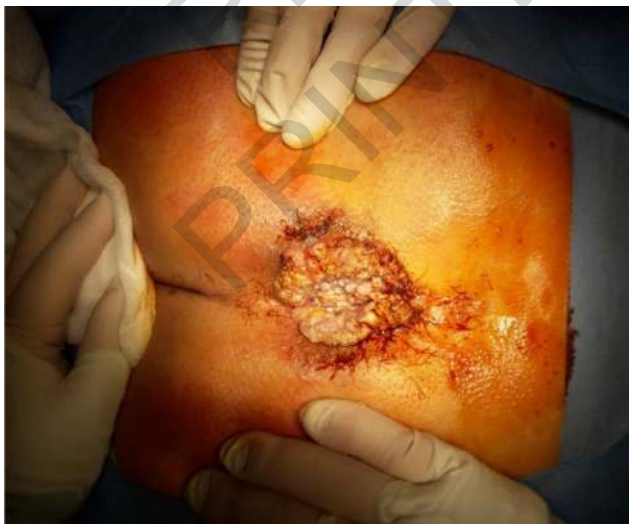

Fig. 3: Operating field. Visible squamous cell carcinoma arising from chronic sacrococcygeal pilonidal disease.

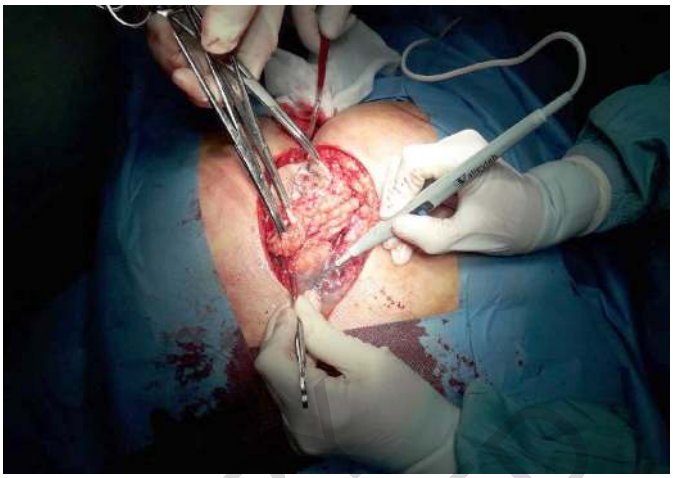

Fig. 4: Operating field. Visible squamous cell carcinoma cutting.

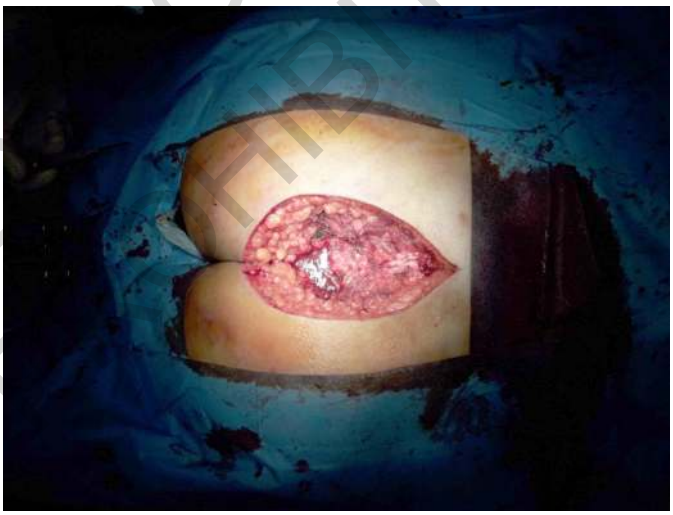

Fig. 5: Operating field. State after cutting squamous cell carcinoma.

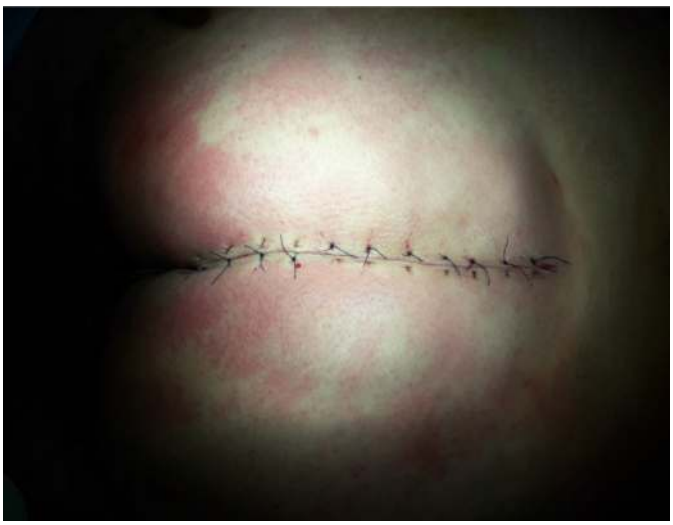

Fig. 6: Operating field. Wound after tumor removal.

(Figs. 1, 2). Due to the exuding of purulent content from under the tumor, swabs for bacteriological examination were collected. The following bacteria were found in the bacteriological test: Escherichia coli pathogen, Prevotella oralis pathogen and Corynebacterium spp. Before the surgery, targeted antibiotic therapy was used. Digital rectal examination showed no tumor infiltration on the rectum. There were not enlarged in the both inguinal regions lymph nodes in palpable examination. Also ultrasound examination of the both inguinal lymph nodes did not show pathologically enlarged lymph nodes. CT scan of the abdominal and pelvic cavity did not show tumor cell infiltration on bone structures or on the rectum or anus. In the CT examination local lymph nodes were not enlarged and pathologically changed, in the remaining organs no abnormalities were found.
The patient was qualified for surgery. The tumor was removed and sent for routine histopathological examination (Figs. 3, 5). The duration of surgery was 45 minutes. Pathological examination showed highly differentiated squamous cell carcinoma. The tumor was removed entirely with a margin of 7 millimeters and with sacral fascia. Patient after surgery felt good and did not complain of pain. The postoperative period was uncomplicated and the patient left the ward in the third day after operation. The patient is in the care of outpatient surgical oncology and was qualified for radiotherapy.

Discussion

Pilonidal disease is a chronic disorder which is observed in boys and men between ages of 15 and 40 years old in Caucasian race [1,4]. The most frequent complications of pilonidal disease are abscess, cellulitis and fistula forming [2-4]. Malignant degeneration and carcinoma arising from chronic sacrococcygeal pilonidal disease are observed extremely rare and are described in the group of Marjolin cancer. Carcinoma can be seen on a place with long-term inflammation process as it was in the case described in the patient's article. First time squamous cell carcinoma arising from chronic sacrococcygeal pilonidal disease was described in 1900 by Wolff [5]. The incidence of carcinoma arising in pilonidal disease is less than 0.1% and in the whole medical journals less than 100 patients are reported [4]. Squamous cell carcinoma arising from chronic sacrococcygeal pilonidal disease are observes more often in males, the average age is 52 years and in higher rate in immunosuppression patients [6].

From a clinical point of view pilonidal carcinoma looks like ulcerated mass with strong pain and often bleeding during examination [1,4]. Physical examination and rectal examination is obligatory. During the palpation examination, it is necessary to check the mobility of the tumor, infiltration of the bone structure and determine whether there is any infiltration on the anal sphincters. In the digital rectal examination, the doctor examine whether there is an infiltration on the rectum. Any patient with a bleeding, exophytic tumor who is suspected of having a cancerous process should have a sample for histopathological examination before surgery. Squamous cell carcinoma is the most frequent carcinoma arising from chronic sacrococcygeal pilonidal (90% cases) but basal cell carcinoma, sweat gland adenocarcinoma and verrucous carcinoma are also observed [1,7,8]. Usually a chronic inflammatory process lasts for months or even years, as in the case of the patient described, therefore it is necessary to take swabs for bacteriological examination to treat the patient with a targeted antibiotic before surgical treatment [9].
In the pre-operative diagnosis, the patient needs X-ray of the chest and CT of the abdominal cavity and pelvis. In the CT examination of the abdominal cavity and pelvis, the depth of tumor infiltration is assessed for the surrounding structures - in particular, it involves infiltration into bone structures [4]. Most cases of the pilonidal carcinoma are found deep inside the subcutaneous tissue, but up to 8% have invided the bone [10]. In the CT examination, regional lymph nodes, iliac and para-aortic lymph nodes are also evaluated. Ultrasound examination may also be helpful, as it precisely assesses the morphology of inguinal lymph nodes. In the case of the described patient, no metastatic lymph nodes were observed in imaging studies, and in the palpation examination the inguinal lymph nodes were not enlarged. MRI examination is a good tool to show inflammation and malignant infiltration in the gluteal region on the second admission. Positron emission tomography is a tool, which can be use to find metastases [10,11].
Surgical treatment is the primary method of treatment carcinoma arising from chronic sacrococcygeal pilonidal disease. Wide surgical excision with strict oncologic techniques of en bloc resection and minimizing violation of the tumor margins and the lesion is indicated [12]. A wide margin of skin, subcutaneous tissue, presacral fascia and gluteal muscle should be excised [13]. The sacrum and coccyx should be removed in the case of infiltration neoplasm cells into bone structures [13]. Prophylactic inguinal lymphadenectomy is not recommended [14]. Radical excision of the superficial squamous cell carcinoma has good results [7]. After surgical treatment of invasive tumors which infiltrate sacrum and/or coccyx and perforate the sacral fascia, there are often recurrences of the carcinoma process [4]. Local recurrence rates reach 50% and are observed usually 9-16 month after surgery [4]. Adjuvant radiotherapy with proper oncological surgery can decrease local recurrence to 30% [15]. Re-excision of local recurrence is necessary and may prolong survival [15]. The 5-year survival rate of patients with squamous cell carcinoma arising from chronic sacrococcygeal pilonidal disease is between 55-61% [12,13]. The role of adjuvant chemotherapy is unclear, but some authors believe that adjuvant chemotherapy may be effective in connection with radical surgery and radiotherapy for high-risk lesions [4,7].

Patients suffer from squamous cell carcinoma arising from chronic sacrococcygeal pilonidal disease should be treated in high-specialized oncological centers where different therapies can be also used.

Riassunto

La malattia pilonidale è una malattia cronica che si osserva in ragazzi e uomini di età compresa tra 15 e 40 anni nella razza caucasica. La degenerazione maligna e il carcinoma derivanti da malattia pilonidale sacrococcigea cronica sono osservazioni estremamente rare e sono descritte nel gruppo del cancro di Marjolin. Il carcinoma può essere la conseguenza di un processo infiammatorio di lunga durata, come nel caso descritto nella presente osservazione.

Un carcinoma a cellule squamose derivante dalla malattia pilonidale sacrococcigea cronica fu descritto per la prima volta nel 1900 da Wolff. L'incidenza di questa degenerazione neoplastica della malattia pilonidale è inferiore allo 0,1% e in tutte le casistiche tale evento è riportato in meno di 100 pazienti. Questo articolo presenta il caso di un paziente con carcinoma a cellule squamose diagnosticato quale evoluzione di malattia pilonidale sacrococcigea cronica. Vengono presentate le caratteristiche cliniche, la patogenesi, le opzioni di trattamento e la prognosi di questa malattia rara.

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