Int J Clin Exp Med 2014;7(2):446-450
www.ijcem.com /ISSN:1940-5901/IJCEM1312037

Case Report

Recurrent squamous cell carcinoma arising in a neglected pilonidal sinus: report of a case and literature review

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Received December 28, 2013; Accepted January 22, 2014; Epub February 15, 2014; Published February 28, 2014

Abstract: Squamous cell carcinoma (SCC) is a rare complication observed mainly in chronic, recurrent and untreated primary pilonidal disease. It is associated with poor prognosis, and its recurrence rate after surgery is high. Here we present a patient diagnosed with SCC arising from a neglected pilonidal sinus. A 44-year-old male, who is a heavy truck driver, applied to the hospital with chronic discharge from sacrococcygeal region. He had symptoms of the disease with multiple recurrent abscesses and spontaneous drainage for 10 years. The patient underwent cyst excision and the defect was left open for secondary healing. One year later, the patient was admitted to the hospital with a mass of 3 x 3 cm in the same area. Re-excision and a sigmoid colostomy were performed. The patient died after two years. This case study illustrates that since SCC is a fatal complication of recurrent and long-standing pilonidal sinus, the proper treatment of this disease should be carried out as soon as the diagnosis is established. To this end, we believe, that all pilonidal sinus lesions should be sent for pathologic examination.

Keywords: Pilonidal sinus, squamous cell carcinoma, neglect

Introduction

Pilonidal disease (PD) is a very common, well recognized, benign disease which occurs with the greatest frequency in men. PD occurs with a frequency of 0-5% in the general population and is often complicated by infection [1-3]. It is usually located in the sacrococcygeal area, but is also found at, other sites such as in the umbilicus, the axilla, on the sole of the foot, the penis, the clitoris and in the anal canal [4].

Malignant degeneration is a rare complication observed mainly in chronic, recurrent and untreated primary pilonidal disease and is associated with poor prognosis compared with regular non-melanoma skin cancer. The incidence of malignant degeneration occurs in approximately 0.1% of patients with recurrent pilonidal disease [5, 6]. Fewer than 70 cases of malignancy arising in a pilonidal sinus have been published in the literature since 1900 [5, 7]. The first case was reported by Wolff in 1900 [8]. The histological type of tumour that arises from a pilonidal sinus is often SCC (90%) [9, 10]. Additionally, basal cell carcinomas, adenocarcinomas and verrucous carcinomas (an uncommon type of SCC) have also been reported. Wide excision with tumour-free margins is the treatment of choice. Radiotherapy and chemotherapy are of limited value [9, 11, 12]. Our aim was to present SCC arising in pilonidal sinus disease in diabetes mellitus (DM) patient and discusses the present literature.

Case report

A 44-year-old male heavy truck driver applied to the hospital with chronic discharge from sacrococcygeal region. The patient was diagnosed with pilonidal sinus. He had symptoms of the disease with multiple recurrent abscesses and spontaneous drainage for 10 years. He had a history of diabetes mellitus for 12 years. The patient underwent cyst excision and the defect was left open for secondary healing. One year later the patient was admitted to the hospital with a mass of 3 x 3 cm in the same area. A

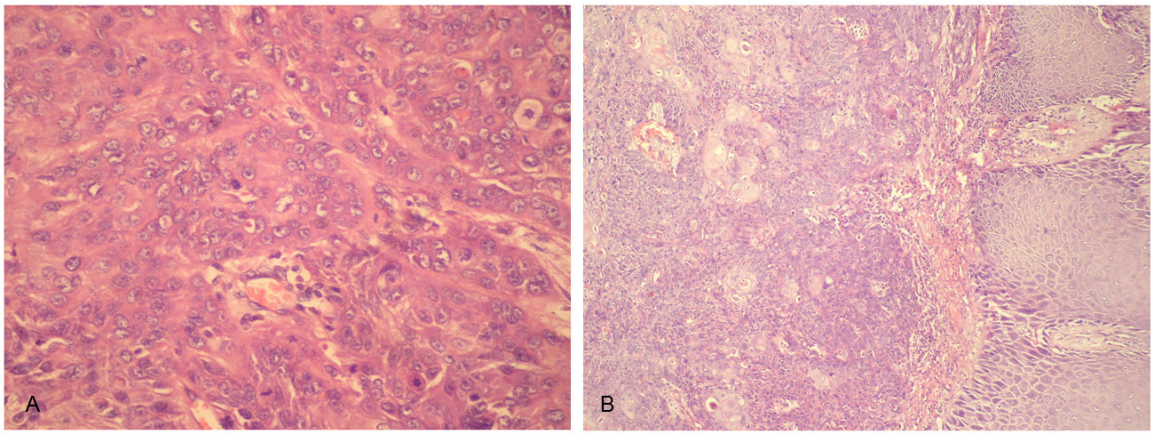

Figure 1. The histological appearance of the wound. A: The normal squamous epithelium and adjacent area with squamous carcinoma can be visualized (HE X 100); B: The atypical mitotic figures can be seen (HE X 400).

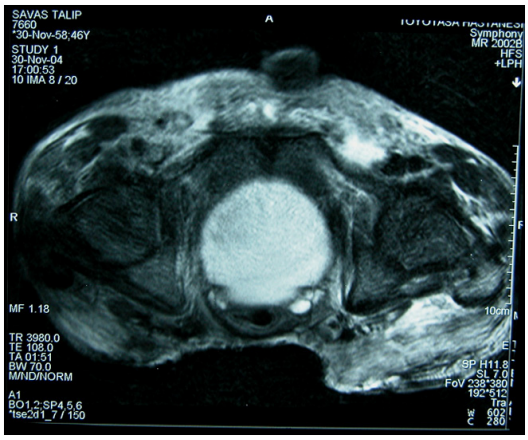

Figure 2. Pelvic axial fat-suppressed T2-weighted imagins: Subcutaneous tissue, presacral fascia, muscles and coccyx are resected (Bold white arrows) and bilateral inguinal lymphadenopathy (thin white arrows).

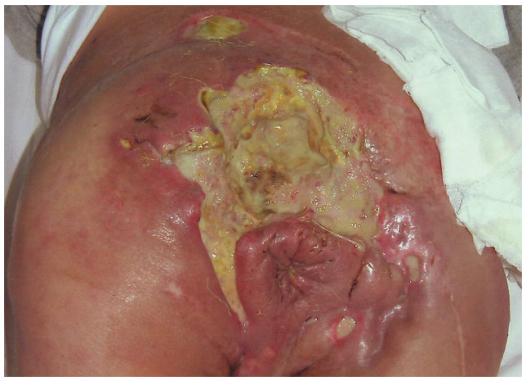

Figure 3. The terminal stage of the tumor involving adjacent structures like anal canal.

conglomerated inguinal lymphadenomegaly (LAM) was noticed in physical examination. The lesion was widely excised. Histopathological examination revealed SCC (low grade differentiation) on top of chronic inflammation (**Figure 1**). Thorax computed tomography (CT) did not show any signs of metastases. Abdominal CT and pelvic magnetic resonance imaging (MRI) examination showed inflammation and tumoral infiltrate in the gluteal region (**Figure 2**). The mesorectal plane was infiltrated with tumour and pathological lymph nodes were noted perirectally. Additionally, conglomerated LAM's were found in both inguinal regions. The uncontrolled DM and the tumoral growth required repeated excisions. An excisional biopsy results showed a tumour of squamous cell origin with low grade differentiation upon histological examination. There was no vascular and perineural invasion. The tumour invaded the deep muscular level and continued through the surgical margins. The patient received radiotherapy as adjuvant treatment. Two years later, the tumor had metastasized to involve adjacent pelvic structures including gluteal muscles and anal canal with a diameter of 15 cm (**Figure 3**). No distant metastases were detected under vigorous investigation. Re-excision was performed. To protect the wound, which was circumferentially very close to anus, a sigmoid colostomy was performed. Both appropriate antibiotic treatment against anaerobic bacteria and hyperbaric O_2 treatment were instituted. The patient died two years after treatment.

Discussion

Pilonidal sinuses are found in the 3 to 5 cm sacrococcygeal region superior to the anus.

They are believed to arise from the frictional impaction of hair shafts which penetrate into the dermis, producing a foreign body reaction with subsequent chronic inflammation [1-3, 7, 10]. Squamous-cell carcinoma arising in a pilonidal sinus is mostly seen in males (90%), with a mean age of 50 years. The average duration of antecedent pilonidal disease is 25 years, and it is associated with obesity and is frequently referred to as "Jeep Disease" [7, 9, 11, 12]. Our patient was a 44-year-old white male truck driver and had diabetes mellitus for 12 years. He first had symptoms of pilonidal disease 10 years prior to this submission.

The disease may exist as an asymptomatic sinus, as an abscess or it may include cellulites or a chronically draining sinus/fistula [9]. Symptomatic pilonidal sinus abscesses are drained and symptomatic pilonidal sinuses are excised. Although pilonidal sinuses are common, malignant degeneration arising from them is rare [5-7]. The process of malignant degeneration is similar in pilonidal squamous-cell carcinomas and other chronically inflamed wounds, such as burns, osteomyelitis, scars, skin ulcers and fistulas [11-13]. Malignant degeneration occurs in approximately 0.1% of patients with chronic untreated or recurrent pilonidal disease [10]. A well-differentiated squamous cell carcinoma is the main histological type, but basal cell carcinoma, mixed squamous and basal cell carcinoma and adenocarcinoma (sweat gland type) and unspecified tumour have also been reported [13].

A biopsy and examination of a chronic ulcer displaying friable and necrotic margins is one method to diagnose malignant degeneration. Alternatively, a histological examination of an excised pilonidal sinus may be used. If malignant lesions are found, they are generally large tumors, (usually more than 5 cm in diameter) [10]. Most of the malignant lesions are found deep within the subcutaneous tissue and up to 8 percent have invaded the bone [9]. Fine-needle aspiration of a metastatic inguinal lymph node does not usually detect tumours arising from chronic and untreated pilodinal sinuses [14]. Although tumors arising from pilodinal sinuses can be locally extensive, it is rare to find distant metastases. Since malignant lesions are often deeply invasive, it is important to determine the extent of invasion and presence of metastases during the preoperative workup by examining both the lesion and the inguinal region. Positron emission tomography is a good tool to find metastases [7, 9]. In order to ascertain that the tumors have not spread to the rectum, an endoscopy should be performed. Metastasis to the intra-abdominal region, such as the iliac and para-aortic lymph nodes, can be determined using computerized tomography or magnetic resonance imaging in addition to physical examination of the inguinal lymph nodes [15, 16]. Although, there was not any metastatic disease in our case at onset, abdominal CT and pelvic MRI examination showed inflammation and tumoral infiltrate in the gluteal region on the second admission. The mesorectal plane was infiltrated with a tumour and pathological lymph nodes were noted perirectally. Additionally, conglomerated LAM's were found in both inguinal regions.

Among malign degenerations, chronic inflammation and recurrent infections are common. A tumour is usually associated with long–standing pilonidal inflammation the mean duration of pilonidal sinus disease that leads to tumours is over 20 years [11-14]. We observed this phenomenon in our patient. Similar to other chronic ulcerative and scarifying cutaneous disorders, it is believed that the development of tumours due to pilonidal sinus is caused by the release of free oxygen radicals by activated inflammatory cells, inducing genetic damage and neoplastic transformation. Additionally, the normal repair DNA mechanism is impaired in chronic inflammation and predisposes patients to malignancy [16].

The first case was reported by Wollf in 1900 and an additionally 68 such cases have been reported in the world literature [12-17]. Frequently pilonidal disease is present two or more decades before development of carcinoma. The tumour grows locally before metastasizing to inguinal lymph nodes. A pilonidal carcinoma often has a distinctive appearance, and diagnosis can frequently be made on inspection. A central ulceration is often present, with a friable, indurate, erythematous and fungating margins. Continuity with a cyst is usually easily demonstrated [18].

The surgical treatment options range from observation, incision and drainage, limited excision, unroofing, adypo-lypo facial flap, z plasty, v-y plasty and aggressive excision. After exci-

sion, the wound is closed primarily or left open to heal by secondary intention. Lesions are usually large tumours and frequently more than 5 cm in diameter [18, 19]. Recurrent disease, delayed wound healing and a chronic open wound occur in 5-20% of the cases [19]. Our patient underwent cyst excision and the defect was left open for secondary healing.

The treatment of choice remains en-bloc resection, which may present enormous difficulties, because the tumour is able to extend along fistula tracts in a wide area of the sacrococcygeal and perineal regions. This means resection has to include at least the presacral fascia, subcutaneous tissue, muscle, but often also portions of the sacrum, and coccyx and rectum [5-7, 12, 14, 19]. Currently, surgery appears to be the only way to achieve a cure. In the past literature, most of the patients received excision as the primary mode of therapy (80%). In our patient two years later after surgery, patient had extensive metastasis to adjacent pelvic structures including gluteal muscles and anal canal with a diameter of 15 cm. No distant metastases were detected under vigorous investigation.

Closure of the ensuing defect may be accomplished with split thickness skin grafts or vascularized flaps, including gluteal and graceless rotation flaps, free latissimus dorsi myocutaneus flap, local flaps, gluteal advancement flaps [5-7]. To fill up the enormous defect a rectus abdominals myocutaneous flap with a pedicle containing the epigastric vessels and an omentoplasty can be used. Transposition of the rectus abdominis myocutaneous flap based on the inferior epigastric vascular pedicle is a very useful, but rarely used, reconstruction technique for closure of large defects in the inguinal, perineal, and sacrococcygeal area after extended oncological resections [5-7, 9, 12, 14, 19].

Even with surgery, there is still a very low survival rate (55%) and the very high recurrence rate (50%) in patients who have pilonidal carcinoma [11]. One year later after surgery our patient was admitted to the hospital with a mass of 3 x 3 cm in the same area that surgery had previously been performed. The patient died two years later.

It is important to note that it is frequently difficult to ascertain the margins of the tumour since there is often pseudocarcinamataous hyperplasia of the squamous epithelium, as is seen in the association with any severe inflammatory reaction. It is, therefore, important to adequately mark the margins of the surgical resection with India ink as well as to make certain of the margins of resection clinically [20].

Malignancies of this location often have a more aggressive clinical course than SCC of other skin locations. Therefore, the proper treatment of the disease should be carried out as soon as the diagnosis is established. Careful inspection of the pilonidal sinus disease in all chronic and long-standing inflammatory processes is important and should be evaluated for malignant transformation We believe that all pilonidal sinus lesions should be sent for pathologic examination. A second important aspect of this case that deserves mention is that even with detailed histologic examination of the specimen there is no absolute certainty regarding the presence of carcinoma.

Disclosure of conflict of interest

The Authors have nothing to disclose.

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