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Is Histological Examination Necessary when Excising a Pilonidal Cyst?

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Study Design A
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Statistical Analysis C
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Conflict of interest: None declared

Patient: Male, 77
Final Diagnosis: Pilonidal cyst
Symptoms: Severe pain • bleeding mass
Medication: —
Clinical Procedure: Local radiation therapy • neoadjuvant chemotherapy • surgical resection
Specialty: Surgery

Objective: Rare disease
Background: Pilonidal disease is a common inflammatory condition mostly affecting young males. Malignant degeneration of a pilonidal cyst is rare, with incidence estimated at 0.1%. The most common type is squamous cell carcinoma and the treatment of choice remains en block resection of the lesion.
Case Report: We present the case of a patient with locally advanced squamous cell carcinoma arising in a pilonidal cyst, due to misdiagnosis of the disease during his first treatment.
Conclusions: Detailed histological examination of all excised pilonidal cyst lesions is essential and any histological suspicion should prompt a wider excision.

MeSH Keywords: Carcinoma, Squamous Cell • Pilonidal Sinus • Sacrococcygeal Region

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Background

Pilonidal disease is a chronic inflammatory condition mostly affecting white males ages 15–40 years. It is thought to be an acquired infection of natal cleft hair follicles, which become distended and obstructed and rupture into the subcutaneous tissues to form an abscess [1]. According to the literature, black and Asian people are rarely affected by the disease [2].

Untreated pilonidal disease may result in many complications, including multiple draining sinuses, recurrent abscess formation, local cellulites, and, less frequently, osteomyelitis of the sacrum or coccyx [3].

Despite the high incidence of the disease, malignant degeneration of a pilonidal cyst is rare. The incidence is estimated at 0.1% and is triggered by a chronic inflammatory process, occurring mostly in neglected primary lesions; therefore, it is most commonly seen in older people with longstanding disease [4]. In 88% of the cases, the histological type is squamous cell carcinoma [4,5].

The purpose of this paper is to present the case of a patient with locally advanced squamous cell carcinoma arising in a pilonidal cyst, as a consequence of misdiagnosis of the disease, and to review the literature.

Case Report

A 77-year-old white male patient was admitted in our clinic complaining of severe pain in the sacrococcygeal area due to recurrent pilonidal disease. In his medical history, he mentioned 2 previous operations for a pilonidal cyst existing more than 10 years. The first operation was performed in March 2013, and the second was 1 year later, due to recurrence of the disease. No histological confirmation of the excised specimen was done in either of the previous interventions.

Until the time of admission, the patient was being treated for a second recurrence of pilonidal disease. Due to severe pain, he received large doses of analgesics, including opioids and NSAIDs, with no result.

On clinical examination, he presented with a 5×5 cm ulcerated, friable, bleeding mass in the sacrococcygeal area (Figure 1). Left inguinal lymphadenopathy was also noticed.

Laboratory tests on admission showed mild anemia with hemoglobin of 11.7 g/dl and hematocrit of 36.8% and mild leukocytosis with WCC of 11.500 K/ μ L, 80% segmented.



Figure 1. Squamous cell carcinoma in sacrococcygeal area.



Figure 2. Pelvic MRI shows the mass, which reaches the elevators ani muscles.

On a pelvic MRI performed 1 month before, a 3.3×2.7×4 cm inflammatory mass was demonstrated, extending beyond the last coccygeal vertebra between the major gluteal muscles, reaching the junction of the elevators ani muscles (Figure 2).

An abdominal CT scan was performed, which revealed that the mass had destroyed the coccyx and invaded the elevators ani. It also extended to and affected the mesorectal fat, but the posterior wall of the rectum was intact. In addition, it demonstrated involvement of the left inguinal lymph nodes. No distant metastasis was observed (Figures 3 and 4).

We decided to seek histological confirmation of the disease. Biopsies were taken from the lesion and the prominent lymph nodes from the left inguinal area. The results from the histological examination showed a well-differentiated squamous cell carcinoma of the sacrococcygeal area and metastatic infiltration of the left inguinal lymph nodes.



Figure 3. Abdominal CT scan shows the mass invading the coccyx and the elevators ani muscles.



Figure 4. Abdominal CT scan shows the mass extending to the mesorectal fat without involving the posterior wall of the rectum.

Due to the patient's severe general condition, it was decided to first treat him with local radiation therapy and chemotherapy.

Discussion

Pilonidal disease is generally a benign condition and malignancy is extremely rare, with an incidence estimated at 0.1%. Reviewing the literature, the low frequency of malignant degeneration of a pilonidal cyst is confirmed, with fewer than 100 cases reported [6].

The first squamous cell carcinoma arising in a pilonidal sinus was described in 1900 by Wolff in a 21-year-old woman [7]. Ever since, there has been an increased concern of the possibility of malignant degeneration of a pilonidal cyst complicated with recurrent episodes of inflammation [3].

The etiologic factors of the development of a squamous cell carcinoma in the sacrococcygeal area appear to be the same as with all chronically inflamed wounds such as burns, scars, ulcers, and fistulas, which may undergo malignant transformation [8]. In 1828, Jean Nicolas Marjolin was the first to describe the occurrence of malignant transformation in unstable scars and chronic ulcers [9,10]. The mechanism that leads to malignant degeneration seems to be related to a compromise of DNA-repairing mechanisms due to chronic inflammation, which causes the release of free oxygen radicals by activated inflammatory cells [11]. The development of carcinoma follows a certain sequence of events in which there is a transition from squamous cell to squamous hyperplasia to carcinoma *in situ* to invasive carcinoma. This seems to be a long latent period of symptomatic pilonidal disease during which early lesions go unrecognized and untreated [5].

Therefore, the most important factor in the incidence of malignant degeneration of a pilonidal sinus is the duration of the disease. Longstanding untreated or recurrent pilonidal disease appears to have a higher incidence of development of squamous cell carcinoma. The average age at presentation appears to be 49 years, with an average duration of 23 years [12]. Seventy percent of patients with malignant degeneration of a pilonidal cyst have been symptomatic for at least 10 years [11].

During the Second World War, in a series of 86 333 pilonidal cysts treated in the U.S. Armed Forces, no malignancy was reported [13]. Apparently, early treatment in this young population prevented malignant degeneration and supports the theory of chronic transformation [8].

On clinical examination, pilonidal carcinoma usually appears as a painful, ulcerated, bleeding mass. According to the patient's medical history, it complicates a longstanding pilonidal disease with recurrent episodes of abscess formation [6].

Carcinomas arising from pilonidal cysts grow slowly, but have a tendency towards local invasion and metastasis [14,15]. Eight percent of the patients appear with bone involvement on diagnosis [16]. Inguinal lymphadenopathy at the time of diagnosis is related to a poor prognosis [17].

Preoperative evaluation should be designed to identify both the extent of local invasion of the disease and the presence of distant metastasis. An accurate clinical examination of the patient's sacrococcygeal area, including inguinal areas for lymph node involvement, is essential. Preoperative lower endoscopy (sigmoidoscopy or colonoscopy) is used to exclude extension of the disease into the rectum, as well as CT scan of the abdomen and MRI of the pelvis to evaluate the local extent of the disease, distant metastasis, and iliac or para-aortic lymph node involvement [11]. If the degree of local inflammation is

severe, the patient may preoperatively require a short period of wound care and intravenous antibiotics [18].

The treatment of choice remains en block resection of the lesion, including the presacral fascia, with tumor-free margins. Surgery appears to provide the only means of cure. In patients with local extent to adjacent structures, a wider excision is necessary, including coccygeal excision, decortication or resection of the sacrum, and total mesorectal excision [4]. Frequently, it is difficult to determine the margins of the tumor due to severe inflammatory reaction in the area. Therefore, it is crucial to clearly mark the margins of resection before surgery [8]. Defect reconstruction can be accomplished either with skin grafts or local flaps (gluteal rotation or gluteal advancement flaps) or even by using free muscular or musculocutaneous flaps [6]. If the patient's clinical status does not allow major interventions, the defect can be left open to close by secondary intention [4].

Postoperative radiation therapy and/or chemotherapy are recommended³. In a series of patients, Gill et al. reported a reduction in local recurrence rate from 44% to 30% when radiation therapy was initiated postoperatively [17].

Almeida-Goncalves presented a series of 7 patients diagnosed with advanced disease and treated with liquid nitrogen cryotherapy. Cryosurgery accomplishes deep and inclusive neoplastic sterilization, destroying the sacral fascia but not the bone, and without inducing metastasization. Local eradication was achieved in all cases (100% complete response). This demonstrates the value of cryosurgery as a promising intervention for such advanced cancers³.

Postoperative follow-up includes regular check-ups every 3 months for the first 2 years, every 6 months for the next 3 years, and once every year after that [6]. It includes clinical examination of the sacrococcygeal and inguinal area, ultrasound (if there is any sign of inguinal lymphadenopathy), and CT of the abdomen to rule-out local recurrence or distant metastasis [18].

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The 5-year disease-free survival rate after wide excision is 55–61%. The local recurrence rate is 44% and it often occurs the first year after surgery. Metastasis appears in 14% of patients and is usually fatal. Prophylactic inguinal lymph node dissection is not recommended because there is not enough evidence to support an impact on patient survival [6,8,19]. Postoperative radiotherapy has been shown to decrease local recurrence to 30%. The role of chemotherapy is unclear and it has been suggested it be used in combination with resection and radiotherapy for high-risk patients [16].

In our department, we consider that all excised pilonidal cyst lesions should be sent for detailed histological examination. Differential diagnosis should be made between squamous cell carcinoma and pseudocarcinomatous hyperplasia of squamous epithelium, which is related to the chronic inflammatory process [5]. Because cellular atypia and increased mitotic rate is found in pseudocarcinomatous hyperplasia as well, careful study of a wide tissue sampling is required to establish final diagnosis. Any histological suspicion of carcinoma should prompt a wider excision [20].

Conclusions

Pilonidal disease should be treated early in its history and all excised specimens should be sent for histological examination, especially in older patients (>40 years old) with longstanding disease. When dealing with a macroscopically suspicious lesion or when delayed healing is observed, the possibility of malignancy should be considered and new lesion biopsies should be performed [12].

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Competing interests

The authors declare that they have no competing interests.

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