

Malignancy in pilonidal disease: Uncommon occurrence

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ABSTRACT

Development of malignancy in pilonidal sinus is a rare complication. The disease occurs most frequently in men. About 69 cases have been reported so far in literature. The lesion is mostly a well-differentiated squamous cell carcinoma. Best treatment is local wide excisions along with a short period of local wound care. After healthy granulation tissue, definitive closure of the defect is done with rotational flaps or skin grafts. Fine needle aspiration cytology is mandatory for palpable inguinal nodes. The prognosis is very poor if inguinal node has metastatic deposits. Radiation therapy may be used as palliative therapy for local bone or soft tissue recurrences. Recently, both topical and systemic chemotherapy have been used without promising results.

Key words: Pilonidal disease, squamous cell carcinoma, wide excision

INTRODUCTION

Pilonidal disease is a common condition. The most common complications of pilonidal disease are cellulitis, abscess or chronic infection, and fistula formation. Malignancy, as complication, is very rare and these changes can occur if the disease is long duration. More than 69 cases have been reported in literature.^[1] All occurred in chronic pilonidal disease, with a mean duration of 23 years.^[2] Estimated incidence is about 0.1%. Radical surgical excision with tumor-free margin is gold standard.^[3] Wide excision with free margin produces disease-free 5-years survival rate in only 55% of patients. The recurrence rate is quite high (50%). Regional (inguinal) node metastases (14%) are unusual at presentation and is associated with a poor prognosis and survival rate is usually not more than 2 years. Radiation therapy is not curative and may be used as palliative therapy for locally recurrent disease or residual disease in the soft tissues and sacrum. We report such a case of squamous

cell carcinoma arising in a chronic pilonidal sinus, due to its rarity.

CASE REPORT

A 70-year-old was admitted with a complaint of an enlarging and bleeding ulcer of the sacrococcygeal region. Since the age of 49, he had got pilonidal abscesses drained four times. During the 6 months before admission, the pilonidal ulcer produced persistent pain and bleeding. Local examination revealed an ulcer of 7.5 × 5 cm over the sacrococcygeal area. The ulcer was associated with multiple sinus tracts. The genito-rectal and sigmoidoscopic examinations were normal. There was no inguinal adenopathy.

Laboratory data included a leukocyte count of 16,000 with a shift to the left. Lumbosacral spine films revealed no osteomyelitis or destruction of bone. A biopsy specimen taken at the margin of the ulcer demonstrated squamous cell carcinoma [Figures 1 and 2]. The wound cultured *Pseudomonas aeruginosa* and *Staphylococcus aureus*. Wide excision with rotation flap was planned but patient went home due to his wife's death and lost the follow-up.

DISCUSSION

Pilonidal disease is a common disease. A malignant change rarely occurs and happens only when the disease is of

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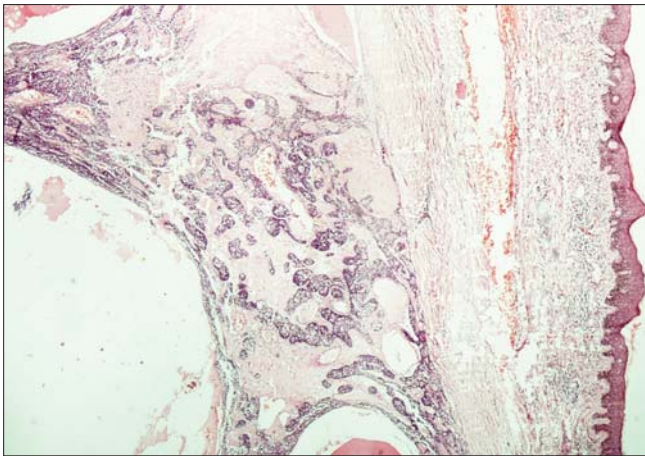


Figure 1: Hyperplastic stratified squamous epithelium

long duration. Predisposing factors are low immunity and infection with human papillomavirus.^[1]

The first case of squamous cell carcinoma arising in pilonidal sinus was published in 1900.^[4] Around 69 cases of pilonidal squamous cell carcinoma have been reported in the literature^[1] and mostly males are affected (80% of the reported cases) as in our case. The mean age at diagnosis was 50 years, and the average duration of existence of pilonidal disease was 23 years as in our case.^[5] It is believed that predisposed area becomes prone to malignant changes due to abnormal changes in repair mechanisms of chronic inflammation. Chronic inflammatory process usually release free oxygen radicals which may result in malignant changes. Stimulation of inflammatory cells (neutrophils, macrophages, and eosinophils) results in release of free oxygen radicals. Free oxygen radicals change the behavior of specific clones of cells and thus acts as dynamic force in creation of malignancy by assisting clonal growth.^[6]

Pilonidal carcinoma is diagnosed by clinical examination as a growing, ulcerated mass with indurated edges, which may be painful and often bleeds (as in our case).^[4] The incidence of carcinoma arising in pilonidal inflammation is estimated to be 0.1%.^[3]

Occasionally, the lesion is discovered as an incidental finding in an otherwise routine specimen. Multiple biopsies of the margin of the ulcer provide the histological diagnosis. Continuity with the cyst or sinus is easily demonstrated. Carcinomas arising in association with pilonidal disease are typically differentiated epidermoid carcinomas that may have focal keratinization. The tumors are of the same histological type that are found in association with fistulas-in-ano or sinuses of chronic osteomyelitis. There is no single, well-established chemical or physical carcinogen concerned in the development of carcinoma except for chronic inflammation, often with bacterial infection. As there is spread of the tumor beyond the resection via

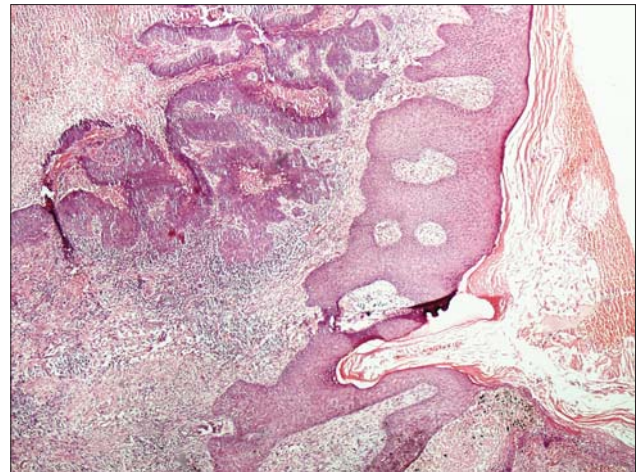


Figure 2: Well-circumscribed tumor cells (basophilic) underneath normal looking skin

vessels or perineural spaces, operating surgeon is always in dilemma about the extent of excision and worries that lesion may not be left behind. So, if on initial biopsy there is cellular atypia and an increased mitotic rate then a wider excision of ulcerated pilonidal sinus should be done.^[7]

As inguinal lymph node involvement is not frequent, prophylactic inguinal node dissections have not been recommended. The incidence of inguinal node enlargement was about 14-22%. Enlargement can be due infection or due to metastasis. FNAC of the lymph nodes shall be done if they are clinically palpable. Reported incidence of inguinal node metastasis is about 14% and that of recurrence is about 34-50%.^[8] The survival rate of patients with proven inguinal node metastasis is poor.

de Bree, E has reported 30% incidence of mortality, 39% of recurrence in total number of 59 patients (47 males and 12 females) who were treated mainly with surgery. The local recurrence rate was lower when radiotherapy was added to surgical treatment alone (30% vs. 44%).^[9]

Radiotherapy and/or chemotherapy may be added to the treatment regimen. When radiotherapy is added to surgery alone, recurrence rates decrease from 44% to 30%. Re-excision of local recurrence resulted in some long-term survivals.^[2] Prophylactic chemotherapy, cytoxan and methotrexate with regional BCG injections can be used for a completely excised lesion. Even topical 5-FU, systemic Bleomycin, and high-dose radiotherapy to the lesion (9500r Co60) can be used for recurrent disease.

CONCLUSIONS

Carcinoma arising in pilonidal disease is extremely rare and it is seen in patients with chronic disease. Early surgical treatment of pilonidal disease is best prevention.

Surgical team work is required for proper management of carcinoma arising on pilonidal disease and in cases of lymph metastasis, inguinal dissection is mandatory.

REFERENCES

1. Malek MM, Emanuel PO, Divino CM. Malignant degeneration of pilonidal disease in an immunosuppressed patient: Report of a case and review of the literature. *Dis Colon Rectum* 2007;50:1475-7.
2. Chatzis I, Noussio G, Katsourakis A, Chatzitheoklitos E. Squamous cell carcinoma related to long standing pilonidal-disease. *Eur J Dermatol* 2009;19:409.
3. Martino A, Martino CD, Pisapia A, Maharajan G, Evangelista M. Squamous-cell carcinoma arising in a pilonidal sinus: Case report and review of the literature. *BMC Geriatr* 2011;11 (suppl):A28.
4. Kovacevic P, Visnjic M, Vukadinovic M, Kovacevic T, Mihajlovic D. Carcinoma arising in pilonidal disease: Report of two cases. *Med Biol* 2007;14:133-7.
5. Cilingir M, Eroqlu S, Karacaoqlan N, Uysal A. Squamous carcinoma arising from chronic pilonidal disease. *Plast Reconstr Surg* 2002;110:1196-8.
6. Gur E, Neligan PC, Shafir R, Reznick R, Cohen M, Shpitzer T. Squamous cell carcinoma in perineal inflammatory disease. *Ann Plast Surg* 1997;38:653.
7. Velitchklor N, Vezdarova M, Lossanoff J, Kjossev K, Ketrov EA. Fatal case of carcinoma arising from a pilonidal sinus tract. *Ulster Med J* 2001;70:61-3.
8. Abboud B, Inqea H. Recurrent squamous-cell carcinoma arising in sacrococcygeal pilonidal sinus tract: Report of a case and review of the literature. *Dis Colon Rectum* 1999;42:525-8.
9. de Bree E, Zoetmulder FA. Treatment of malignancy arising in pilonidal disease. *Ann Surg Oncol* 2001;8:60.

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