

Squamous carcinoma in pilonidalis sinus: case report and review of literature

A. DELVECCHIO¹, R. LAFORGIA¹, M.G. SEDERINO¹, M. MINAFRA¹,
G. CARBOTTA², G. BALDUCCI², G. FABIANO¹, S. FEDELE¹, N. PALASCIANO¹

SUMMARY: Squamous carcinoma in pilonidalis sinus: case report and review of literature.

A. DELVECCHIO, R. LAFORGIA, M.G. SEDERINO, M. MINAFRA,
G. CARBOTTA, G. BALDUCCI, G. FABIANO, S. FEDELE, N. PALASCIANO

Aim. We report a case of squamous carcinoma arising from a pilonidal sinus.

Case report. Patient of 83 years old, that after 30 years had a recurrence of pilonidal sinus revealed by a sacral abscess studied with pelvic CT scan and MRI. After clinical investigation, traditional open surgical technique was performed and pathologic studies revealed a squamous carcinoma. The patient performed a new CT scan with persistence of disease, and a second surgical look with mass excision until the presacral fascia and V-Y flap was performed. Histological exami-

nation was found to be positive for squamous carcinoma on the margin and the patient underwent adjuvant radiotherapy cycles with a close follow-up with evidence of free disease survival. He died after 5 years for old age.

Discussion. The incidence of carcinoma arising from a pilonidalis sinus is about 0.1% and the most important risk factor is represented by a chronic abscess from 20-30 years. In literature there are about 100 cases. Gold standard treatment is surgery with complete excision of the presacral fascia, while radiotherapy decrease the risk of recurrence.

Conclusion. All cases reported in literature are submitted as case report. Sacro-coccyx fistula should be treated early because chronic inflammation can determine neoplastic degeneration. Histological examination should be performed routinely. Gold standard is surgery with wide excision. There is no evidence about the gold standard for the reconstructive time.

KEY WORDS: Squamous cell carcinoma - Sinus pilonidalis - Sacrococcyx fistula - VY flap.

Introduction

Sacro-coccyx fistula (SCF) or sacro-coccyx cysts or sinus pilonidalis is one of the most common benign affections, described in 1847 by Anderson and whose name derives from Hodges in 1889, defined as a chronic inflammatory process located in the sacrococcygeal region. Etiology is not exactly known: in recent years, it has overtaken the idea of an acquired origin, related to an invagination of loose hair into the skin of natal cleft that results in foreign body reaction with inflammation, abscess and fistula formation. There is also the idea of a

congenital origin, although in 20-50% of cases there are not hairs in the cystic cavity (1).

The incidence is 0-5% in the general population with the greatest frequency in man, working age population, between 15 and 40 years, with poor personal hygiene and with attitude for sitting posture (2, 3).

An untreated FSC can evolve in multiple fistulae formation, recurrent abscesses, cellulite, osteomyelitis of the sacrum and coccyx.

Several surgical treatments have been reported in literature, including simple incision and drainage, phenol application, total sinus excision with primary or secondary intention method closure, with marsupialization or with flap closure (4, 5).

Malignant degeneration of an SCF is a rare complication, with an incidence of 0.1% and occurs in untreated cases or recurrences. Therefore it is more common in the elderly population with a previous

¹ Department of Emergency and Transplantation of Organs, University of Bari, Bari, Italy

² General Surgery Unit "V. Bonomo", "Azienda Consorziale Ospedaliero-Universitaria Policlinico", Bari, Italy

Corresponding author: Salvatore Fedele, e-mail: salvatorefedele.md@gmail.com

clinical history of SCF. In 88% of cases histological examination revealed a squamous carcinoma. Less than 100 cases have been published in literature, the first one was reported by Wolff in 1900 (6).

Case report

A 83-years-old man, with a previous history of drainage of sacro-coccyx abscess 30 years earlier, was admitted to our General Surgical Unit for a consultation because of recurrence of sacro-coccyx abscess. His clinical history reported also Hepatitis C-related hepatocellular carcinoma, IPB, megaloblastic anemia.

The patient underwent pelvic CT scan and MRI with evidence of an oval mass (about 20 cm), apparently with a fluid level, with a 4 cm thickness, extended from S3 to coccyx, without apparent infiltration of the bone structures. Clinical examination reported a 15x7cm partially ulcerated inflammatory sacro-coccyx cyst, with multiple fistulas; the puncture revealed blood serum liquid (Figure 1).

The surgical team chose a traditional open surgical technique with secondary intention method closure: elliptical incision, removal of the front wall of the cyst, debridement, curettage, toilette and diathermoagulation of the posterior wall, iododoform gauze packing. Cefazoline intravenous antibiotics were administered and the patient was discharged in 1st POD. Re-packing wound treatments were performed every 48 days.

Histological examination was found to be positive for moderately differentiated ulcerated squamous carcinoma, infiltrating soft tissues with tumor-free resection margin.

Therefore the patient underwent a complete pre-operative evaluation: laboratory blood tests; physical examination which did not highlight the presence of inguinal lymphadenopathy; Total Body CT scan with evidence of unequal solid tissue, diameter of about 5 x 4 cm, related to the coccyx area, with intact bone tissues and without signs of remote metastasis; endoscopy, which was negative for neoplastic infiltration. The second surgical look was performed in conjunction with plastic surgeons: lozenge surgical incision with extensive excision of the tumor including the presacral fascia (Figure 2); plastic with bilateral VY flap, double suction drainage, layer suture, then primary intention method closure (Figure 3).

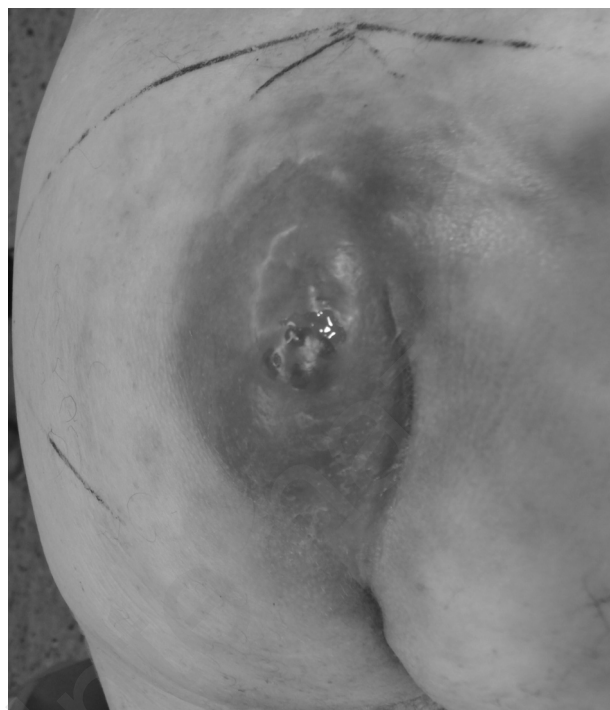


Figure 1 - Clinical examination.

Histological examination evidenced a moderately differentiated squamous carcinoma that insists on the deep margin. The patient underwent oncological evaluation and started adjuvant radiotherapy cycles with a strict follow-up. He was disease-free at 5 years and unfortunately he died of another disease.

Discussion

In literature, about 100 cases have been published, the first one in 1900 by Wolff; in 1962, 15 cases were reported, followed in 1980 by 34 cases; therefore in 2001, 44 cases were published and in 2014 about 70 cases (7-10).

In all case reports, the surgical treatment and postoperative management are different, so there are no standardized therapeutic and surgical strategies.

The incidence of malignant degeneration occurs in 0.1%, and in males in 80-90% with a mean age of 50 years (5). The most important factor in the incidence of malignant degeneration is the untreated disease duration (about 20-30 years) or with recurrent pilonidal disease.

In 88% of cases, the tumor is a locally invasive squamous carcinoma with bone metastases (such as coccyx and sacrum in 8% of cases), rectal and anal



Figure 2 - Lozenge surgical incision.



Figure 3 - Bilateral VY flap.

canal involvement, inguinal lymph node metastases (14%) and absence of distant metastasis (11).

In literature, the preoperative evaluation comprehends the physical inguinal lymph node examination, CT Scan, pelvic MRI and proctosigmoi-

doscopy (11, 12).

Our clinical case is in agreement with the literature for patient's history of 30 years of sacro-coccygeal abscess history and recurrence (8, 9), the histological type and pre-operative staging.

The surgical procedure of choice is considered a complete resection of the mass, including the presacral fascia, gluteal muscle resection, resection of bone metastases (the sacrum) and also total mesorectal excision if infiltrated (13-16).

The closure method is different between open, semi-closed (marsupialisation) and closed with direct suture or plastic suture and is the same for neoplastic and benign disease. In literature there is not a gold standard treatment for plastic procedure, because each technique presents indications, contraindications, complications and none is prevailing.

The best method that should be selected may present fewer complications, shorter hospital stay, low recurrence rate, successful esthetic results and earlier return to daily activities.

The treatment for acute pilonidal abscess is characterized by incision, drainage and antibiotic endovenous therapy, treatment in chronic phase is excision with primary closure but the recurrences should be treated by excision with secondary healing or flap closure (17).

Flaps are very often performed in cases of recurrence and after wide excision and their superiority has been reported in many papers (15).

Recently, fasciocutaneous V-Y advanced flap (VYF) and Limberg Transposition Flap (LTF) are preferred surgical procedure for closure step for primary healing and in cases of recurrent pilonidal sinus disease (PSD).

Bahadir et al. compare VYF with LTF and reported that there were no significant differences in term of complications, operating time, hospital stay, recurrence rate between the two techniques.

Other studies favored LTF because it allows a reduction in operating time, hospitalization and earlier return to daily activities, but VYF is favored in

large substance leakage and lower seroma incidence. The recurrence rate of the V-Y flap is 0-11% (18, 19).

The post-operative management is also controversial: adjuvant radiotherapy (RT) is recommended with a 30-44% recurrence rate since 1980. Chemotherapy has not a well-defined role, but it is normally associated to RT in high-risk patients. Prophylactic lymphadenectomy is not recommended. Patients should undergo a strict follow-up program every 3 months for the first two years and then every 6 months for 3 years and after each year. Five years survival rate is 55-61% if there are free margins and recurrence rate in 44-50% (20, 21).

Conclusions

Sacro-coccyxal fistula should be treated early because chronic inflammation can determine neoplastic degeneration. Histological examination should be performed routinely. The gold standard for the treatment of squamous carcinoma arising out of pilonidal sinus is wide excision with presacral fascia with free margins, but there is no uniform therapeutic strategy for reconstruction. Our surgical approach is considered in literature as the best one for this clinical presentation. More attention should be paid to recurrent Sacro-coccyxal fistula and patients' clinical history.

Consent section

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

References

1. Parpoudi SN, Kyziridis DS, Patrivas DCh, Makrantonakis AN, Iosifidis P, Mantzoros IG, Tsalis KC. Is histological examination necessary when excising a pilonidal cyst? *Am J Case Rep.* 2015 Mar 21;16:164-8.
2. Eryılmaz R, Bilecik T, Okan I, Ozkan OV, Coşkun A, Sahin M. Recurrent squamous cell carcinoma arising in a neglected pilonidal sinus: report of a case and literature review. *Int J Clin Exp Med.* 2014 Feb 15;7(2):446-50.
3. Nunes LF, Castro Neto AK, Vasconcelos RA, et al. Carcinomatous degeneration of pilonidal cyst with sacrum destruction and invasion of the rectum. *An Bras Dermatol.* 2013;88:59-62.
4. Yamashita Y, Nagae H, Hashimoto I. Ambulatory Surgery for Pilonidal Sinus: Tract Excision and Open Treatment Followed by At-Home Irrigation. *J Med Invest.* 2016;63(3-4):216-8.
5. Gul VO, Destek S, Ozer S, Etkin E, Ahioglu S, Ince M, Cimin V, Sen D, Erbil Y. Minimal-ly Invasive Surgical Approach to Complicated Recurrent Pilonidal Sinus. *Case Rep Surg.* 2015;2015:759316.
6. Wolff H. Carcinom auf dem Boden des Dermoids. *Arch Klin Chir.* 1900;62:731-8.
7. Velitchkov N, Vezdarova M, Losanoff J, Kjossev K, Katrov E. A fatal case of carcinoma arising from a pilonidal sinus tract.

- Ulster Med J. 2001 May;70(1):61-3.
8. Pilipshen SJ, Gray G, Goldsmith E, Dineen P. Carcinoma arising in pilonidal sinuses. *Ann Surg.* 1981 Apr;193(4):506-12.
 9. Boukalik WF, Salwan FA. Squamous cell carcinoma arising in a pilonidal sinus: case report. *Ann Surg.* 1962 Jul;156:157-60.
 10. Hall A, Lee JG. Squamous-cell carcinoma complicating a pilonidal sinus. *Cancer.* 1956 Jul-Aug;9(4):760-2.
 11. Malek MM, Emmanuel 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;50:1475-77.
 12. Tirone A, Gaggelli I, Francioli N, et al. Degenerazione maligna di una cisti pilonidale. Caso clinico. *Ann Ital Chir.* 2009;80:407-9.
 13. Bahadır Oz, Alper Akcan, Ertan Emek, Muhammed Akyuz, Erdogan Sozuer, Hızır Akyıldız, Husnu Aydın. A comparison of surgical outcome of fasciocutaneous VeY advancement flap and Limberg transposition flap for recurrent sacrococcygeal pilonidal sinus disease. *Turkey Asian Journal of Surgery.* 2015;20:1e6.
 14. Kulaylat MN, Gong M, Doerr RJ. Multimodality treatment of squamous cell carcinoma complicating pilonidal disease. *Am Surg.* 1996;62:922-9.
 15. Arslan S, Karadeniz E, Ozturk G, Aydinli B, Bayraktutan MC, Atamanalp SS. Modified Primary Closure Method for the Treatment of Pilonidal Sinus. *Eurasian J Med.* 2016 Jun;48(2):84-9. doi: 10.5152/eurasianjmed.2015.0059.
 16. Lorant T, Ribbe I, Mahteme H, Gustafsson UM, Graf W. Sinus excision and primary closure versus lay-ing open in pilonidal disease: a prospective randomized trial. *Dis Colon Rectum.* 2011 Mar;54(3):300-5. doi: 10.1007/DCR.0b013e31820246bf.
 17. Lee PJ, Raniga S, Biyani DK, Watson AJ, Faragher IG, Frizelle FA. Sacrococcygeal pilonidal disease. *Colorectal Dis.* 2008;10:639e650.
 18. Altintoprak F, Dikicier E, Arslan Y, Ozkececi T, Akbulut G, Dilek ON. Comparison of the Limberg flap with the V-Y flap technique in the treatment of pilonidal disease. *J Korean Surg Soc.* 2013;85:63e67.
 19. Milito G, Cortese F, Casciani CU. Rhomboid flap procedure for pilonidal sinus: results from 67 cases. *Int J Colorectal Dis.* 1998;13:113e117.
 20. Gil A, Amondarain JA, Aribé X. Squamous-cell carcinoma on pilonidal disease. *Kirurgia.* 2006;5:1-4.
 21. De Martino C, Martino A, Cuccuru A, et al. Epitelioma a cellule squamose e malattia del seno pilonidale. Presentazione di un caso clinico e revisione della letteratura. *Ann Ital Chir.* 2011;82:511-4.