Sacro-coccygeal 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 SCF 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 congenital origin, although in 20-50% of cases there are not hairs in the cystic cavity (1).
Squamous carcinoma in pilonidal sinus: case report and review of literature

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 sacred-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 15x7 cm 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, toilettte and diathermoagulation of the posterior wall, iododiform 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 preoperative 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).

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...
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 proctosigmoidoscopy (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.
Squamous carcinoma in pilonidal sinus: case report and review of literature

The surgical procedure of choice is considered a complete resection of the mass, including the presacral fascia, glutal 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 pilonidalis 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

A. Delvecchio et al.