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Two Identical Cases of Extreme Fistulation of Buttocks Originating from a Mesorectal Cyst with Keratinizing Squamous Epithelium: A Novel Clinical Entity

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Abstract

Background: Fistulation in the buttock area are mostly of cryptogladular origin, associated with pilonidal sinus or caused by an underlying disease, e.g. Crohn's disease. Several other rare conditions may cause such Fistulation. This paper describes two identical cases that did not fit with any of these differential diagnoses.

Patients: Two patients are described that suffered from persistent extensive Fistulation in the perianal and buttock region. Details on the history or the condition, comorbidity, treatments, imaging, pathology and laboratory findings, both previous and during final treatment, were retrieved from the notes.

Results: Both patients had a retrorectal cyst lined with keratinizing stratified squamous epithelium and an extensive mesh of fistula tracts on the buttock with the same epithelial lining. Both were cured by excision of the cyst and epithelialised tracks.

Discussion: The condition of these two patients is compared with the diseases know to cause fistular disease and the various types of retro rectal cysts.

Conclusion: condition described appears to be a separate clinical entity. The cyst and fistulation appear to require complete excision of the structures lined with stratified squamous epithelium for definitive treatment.

Introduction

Suppurative lesions of the perianal region are a common problem in surgical practice. Hydradenitis, pilonidal sinus, perianal fistulae of crypto glandular origin and Crohn's disease are the most common causes.

Retro rectal lesions such as Mesorectal cysts, dermoid cysts, enteric or rectal duplication cysts, cystic teratoma and tailgut cysts may also cause perianal inflammation and fistulas. We have treated two patients with very extensive suppurative lesions of the buttocks caused by fistulas and a Mesorectal cyst, lined by keratinizing squamous epithelium without skin annexes. To our knowledge such lesions have never been described in literature before.

Case Report

Patient A

A Caucasian male born in 1944, known with heart failure and chronic obstructive pulmonary disease , a history of alcohol abuse (until 2005) and of carcinoma of the tongue, had his first fistulectomy for a perianal fistula in 1984. There was no history of previous surgery or trauma in the perineal region. Since 2003, recurrent perianal abscesses and fistulas were treated surgically, without success. In 2006 he was referred to our clinic with extensive fistulas of both buttocks and the perianal and inguinal areas.

Repeated cultures showed mixed intestinal flora. All cultures, cytology and histology were always negative for fungi, actinomyces and mycobacteria. CT with intravenous contrast and MRI (with gadolinium based contrast) scan showed extensive inflammatory lesions involving the subcutaneous fat and skin of the buttocks, perineum and scrotum but no involvement of the sphincters. A Mesorectal cyst was present close to the dorsolateral wall of the distal rectum (Figure 1). Flexible sigmoidoscopy revealed some sigmoid diverticulosis but no signs of diverticulitis.

A diverting colostomy was fashioned. Excision of all lesions was

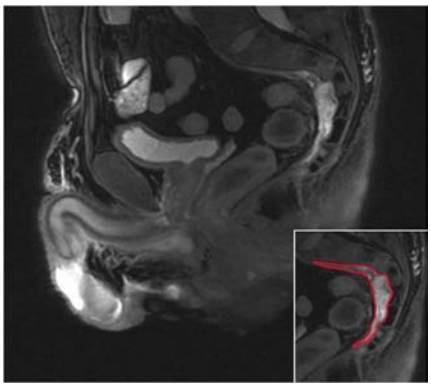

Figure 1: MRI image (T2 haste after gadolineum based contrast) : Mesorectal cyst (patient B).

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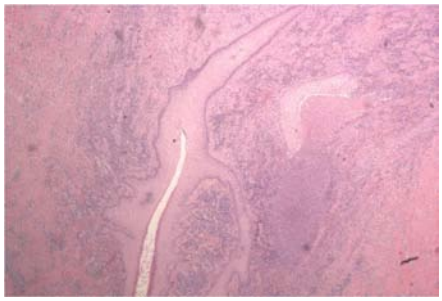

Figure 2: Keratinizing stratified squamous epithelium (patient B).

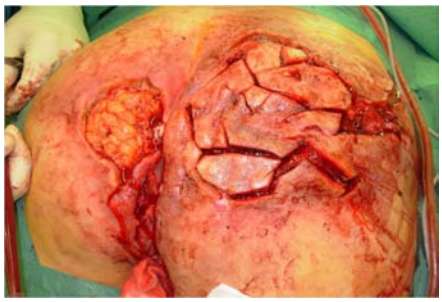

Figure 3: Extensive network of fistulas (patient B).

performed: in two operations the fine mazed network of fistulas on the buttocks, scrotum and inguinal skin and the Mesorectal cyst were excised radically, including resection of a large area of skin and subcutaneous tissue (Figure 3). Split skin grafts were used to cover the resulting defects. After complete wound healing, the colostomy was reversed and the patient was fully continent. The patient remained free of fistulae during 4 years of follow-up and was discharged from our care.

Pathologic examination showed fistulae and a retrorectal cyst, all covered with keratinizing squamous epithelium without skin adnexes (Figure 2). No other components of bowel wall, such as smooth muscle fiber, were present in the cyst's wall. No hairs were present in the fistulae. Staining for microorganisms was negative.

Patient B

A Caucasian male born in 1958, known with an aortic valve stenosis, first presented with a perianal abscesses in1991. There was no history of previous surgery or trauma in the perineal region. The initial treatment, with antibiotics only, failed. In 1998, he was referred to our department for recurrent abscesses of the scrotal skin and fistula openings on the iliac crest on both sides. The buttocks and anus were severely scarred and deformed with multiple fistulae.

Fistulography and CT scan showed an extensive network of sinuses in the skin and subcutaneous fat. Subsequent MRI with gadolinium based contrast showed multiple fistulae and sinuses but no connection with the anale canal. A thin-walled cyst was lying dorsolateral to the rectum. Colonoscopy revealed a normal and intact mucosa. All cultures were negative for fungi, actinomyces and mycobacteria. Repeated surgical drainage and broad-spectrum antibiotics did not resolve the problem. A loop ileostomy was created and a radical excision of all fistula tracks and the Mesorectal cyst. During surgery part of the coccyx was excised for access and an

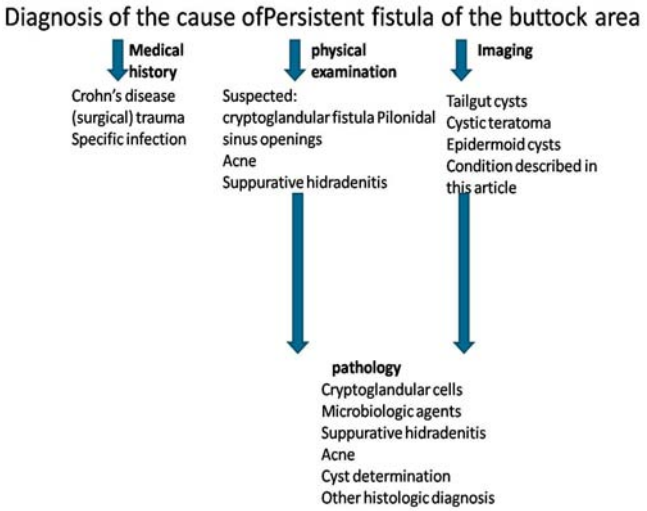

Figure 4: Algorithm for the diagnosis of persistent fistula in the buttock area.

iatrogenic lesion of the rectum was caused by resection of the cystic wall that was adherent to the dorsal aspect of the rectum. The rectal wall was sutured. Split skin grafts were used to cover the skin defects and the ileostomy was reversed after complete healing of the skin. At 5 years follow up the patient remains free of fistulae and was discharged from our care. The patient turned out to be fully continent.

Histological examination showed that both the fistulas and the retrorectal cyst were covered with keratinizing squamous epithelium without skin adnexes and hairs. No other components of bowel wall, such as smooth muscle fiber, were present in the cyst's wall. No microorganisms were found in Gram, Ziehl-Neelsen and Periodic Acid Schiff (PAS) stains

Discussion

The present case reports describe two patients, unique in their extensive suppuration in the perianal region due to a Mesorectal cyst and multiple fistulas, all lined with keratinizing squamous epithelium. This clinical entity that not has been described before. Both patients had a very long history of recurrent suppurative lesions that did not heal after surgical drainage and antibiotic treatment. Ultimately, radical excision of the Mesorectal cyst and the fistula tracts led to complete recovery.

Differential diagnosis: fistulae/sinus

Suppurative lesions of the perianal region are common. Perianal fistulas are of cryptoglandular origin, may be associated with Crohn's disease or caused by trauma or specific infections such as tuberculosis. These fistula tracts are either lined by crypto glandular cells, mucosa or granulation tissue and may sometimes involve the Mesorectal fat [1].

Crohn's disease was considered, but no granulomas were seen in samples taken by repetitive flexible endoscopy. The lesions of these patients did not communicate with the anus or bowel. An etiology of crypto glandular or Crohn's Fistulation can be rejected in these two cases.

In neither of the patients there was any history of previous surgery

or trauma in the area. Imaging techniques and histologic findings did not indicate any scarring that could not be associated directly with the fistulae and the associated surgery during the previous attempts to treat the condition.

Perianal tuberculosis was rejected because mycobacteria were never found in specific cultures and on histology.

Pilonidal sinus is common cause of sinus and fistulae in the perianal region. Pilonidal sinus can be asymptomatic. When it is symptomatic it may present as an abscess, or as a chronic or recurrent sinus. The sinus has a small opening in the midline of the natal cleft, lined by stratified squamous epithelium [2]. Additional sinuses are often present, with multiple lateral openings. These cysts are lined with chronic granulation tissue. It may contain hair shafts, epithelial debris and young granulation tissue. The sinus itself is lined by chronic granulation tissue containing neutrophils, lymphocytes and plasma cells. Giant cells are a frequent finding in reaction to hair shafts. The pathogenesis remains unclear. There are theories suggesting a congenital origin, whereas others suggest an acquired origin such as an invading loose hair [3]. The histological picture is definitely different from the two patients described here.

Suppurative hidradenitis is a chronic recurrent suppurative disease of the skin of the axillar, groin and anal regions. It is manifested by abscesses, sinuses, fistula's and scarring. Although there is no consensus about the pathogenesis, hidradenitis suppurative seems to originate from the hair follicle and the usual microorganism involved is S.Aureus [4]. Smoking is a major risk factor. Patients can develop a subcutaneous network of abscesses and fistulae in these areas in which micro organisms are present. Mesorectal involvement is not seen in suppurative hidradenitis. The same applies to acne variants [5]. The Mesorectal cysts, the microorganisms involved and the histologic picture of these two patients therefore do not comply with suppurative hidradenitis.

Fistulae may also originate from pilomatrixomas [6]; calcifying epithelioma of Malherbe which, however, occurs in the head and neck area in the majority of cases. This diagnosis was considered because of its aseptic origin and because 'ghost cells' were seen on histology. But, pilomatrixomas are calcified nodular lesions, originating from the outer sheath cell of the hair follicle [7]. Almost always these are solitary lesions. Calcification wasn't found on histology of the cases presented here. This diagnosis could therefore be rejected in these cases.

Differential diagnosis and embryologic considerations; mesorectal cysts

Benign cystic processes such as sacrococcygal pilonidal sinus and developmental cysts, such as dermoid cysts, epidermoid cysts, enteric or rectal duplication cysts, cystic teratoma and tailgut cysts can occur in the mesorectum [8-11]. The origin of the mentioned developmental cysts remains a matter of debate, because of the complex embryological development of the perianal and the sacrococcygal area where all three germ layers come together. Theories suggest that retro rectal cysts have their origin in remnants of the tailgut or the neurenteric canal [9]. Others suggest that dermoid and epidermoid cysts are not developmental cyst but inclusional cysts, originating from implanted dermal cells after trauma.

Tailgut cysts contain various types of epithelium [9] as do cystic teratomas.

Enteric duplication cysts are lined by epithelial lining specific for hollow organs, and usually contain deeper wall layers like muscularis [10,11].

Dermoid cysts are characterized by squamous epithelium and dermal adnexal structures.

Epidermoid cysts are lined by keratinized sqaumous epithelium.

Because the Mesorectal cysts of both patients were covered with keratinized squamous epithelium without skin adnexes, all described primary Mesorectal cysts could be excluded except for epidermoid cyst. Single fistulas originating from epidermoid cysts are reported [7] but an extensive maze like network of sinuses and fistulas like in these two patients, has never been described.

The origin of the Mesorectal cyst in the present two cases is unknown. It may be an epidermoid cyst originating from the anoderm or the transitional zone. Infection, subsequent fistula formation with secondary epithelialization of the fistulas may have caused the present clinical picture. An extensive network of fistulas could develop during many years since no causal therapy excising the congenital cyst was performed.

Because these patients had been treated for a wrong diagnosis they presented after years of symptoms and unsuccessful treatment. We consider these two identical clinical pictures to be a clinical syndrome that was never described before: cases of extensive and extremely inter digitating fistulation of buttocks in connection with a mesorectal cyst, all covered with keratinized stratified squamous epithelium.

Development of new fistulas and abscesses was halted abruptly by radical excision of all ectopic keratinizing squamous epithelium in fistulae and the Mesorectal cyst. Therefore an (aggressive) debridement of the mesorectum and cutaneous and subcutaneous parts of the buttocks, perianal and scrotal region was performed. Fecal diversion was used to prevent infection of the wounds and of the covering spit skin grafts. No recurrences were seen during long term follow-up.

An algorithm for the diagnosis of conditions that may lead to persistent fistula in the buttock area is presented in Figure 4.

Conclusion

The differential diagnosis of atypical perianal fistulation should be extended with the syndrome reported here. Besides all previously described diagnoses this syndrome should be considered, if retro rectal cyst and lining with keratinizing stratified squamous epithelium are present, especially in cases of therapy resistant Perianal Fistulation.

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