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A case report and review of the literature

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Perineal scar endometriosis involving the anal sphincter. A case report and review of the literature

BACKGROUND: *Perineal endometriosis is the presence of endometrial tissue in the perineal region. Early diagnosis and treatment is important due to anal sphincter involvement in almost half of the patients. Endoanal ultrasonography is a reliable technique in the assessment of perineal endometriosis with anal sphincter involvement. This report describes the presentation, clinical investigation, and surgical treatment of a perineal endometriosis case*
CASE REPORT: *32-year-old female patient presented with cyclic pain and swelling of an old episiotomy scar. Three-dimensional endoanal ultrasonography showed a lesion with involvement of the external anal sphincter muscles, and it was completely excised and primary sphincteroplasty was performed for the external anal sphincter defect.The final pathology result was reported as endometriosis. Postoperative periods were uneventful and anal incontinence was not observed.*
CONCLUSION: *Perineal endometriosis is a rare disease and may involve the anal sphincter muscles. Incomplete excision to protect the sphincters is associated with high recurrence, while extensive excision can cause anal sphincter damage that may cause anal incontinence. Endoanal ultrasonography may be necessary in surgical planning. Primary sphincteroplasty with excision may be necessary in cases of perineal endometriosis with external anal sphincter muscle involvement.*

KEY WORDS: Case Report, External Anal Sphincter, Endoanal Ultrasound, Sphincteroplasty Perineal Endometriosis

Introduction

Endometriosis is the ectopic growth of the endometrial tissue outside of the uterus. It is seen in about 15% of women of childbearing age, and while it is most commonly seen in the pelvic region, it is very rare in the perineal [1-4]. Despite the development of diagnostic and therapeutic methods, the etiology of the disease is still unknown. The retrograde menstruation theory may provide an explanation for the disease in the pelvic region, while the transplantation theory may be an explanation for the development of perineal scar endometriosis [5]. It is important to evaluate the relationship of the disease with the anal sphincters. Endoanal ultrasound is a reliable technique for detecting anal sphincter [6] In this article, we present a case report of perineal endometriosis and a review of the literature.

Case Report

Chief Complaints
Swelling, pain, and pruritus of the perineum correlated with the menstrual cycle.

History of Present Illness
A 32-year-old woman (G3P3) presented with cyclic pain, swelling, and pruritus of the perineum starting 1 year after her first delivery 8 years ago. Her complaints began to increase after the other two births. After not being diagnosed in the local hospitals, she applied to our clinic.

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History of Past Illness
The patient had three spontaneous vaginal births and an episiotomy at the first delivery. No history of perineal trauma, perineal surgery, or laparotomy. No history of endoscopic imaging.

Personal and Family History
Personal and family history were unremarkable.

Physical Examination
Anogenital examination revealed blue swelling of the episiotomy scar upon inspection (Fig. 1). A 3×2 cm rigid mass was, 2 cm far away the anal verge, palpated on the episiotomy scar. Although anal tonus and squeezing were considered normal in digital rectal examination, the mass was suspected to be related to the external anal sphincter.

Laboratory Examinations
Complete blood count and biochemistry values of the patient were as follows: white blood cell count, 4800/μL; hemoglobin, 11.3 g/dL, thrombocyte count, 268×10^3/μL. The Ca 125 level was in the normal range.

Imaging Studies
Pelvic MRI revealed a mass lesion in the right perineal region whose relationship to the anal sphincters could not be clearly assessed. A hypoechoic irregular mass of 3×2 cm ibetween the episiotomy scar and external anal sphincter was detected in 3D endoanal ultrasonography (Fig. 2).

Preoperative Diagnosis
Perineal endometriosis involving the anal sphincter.

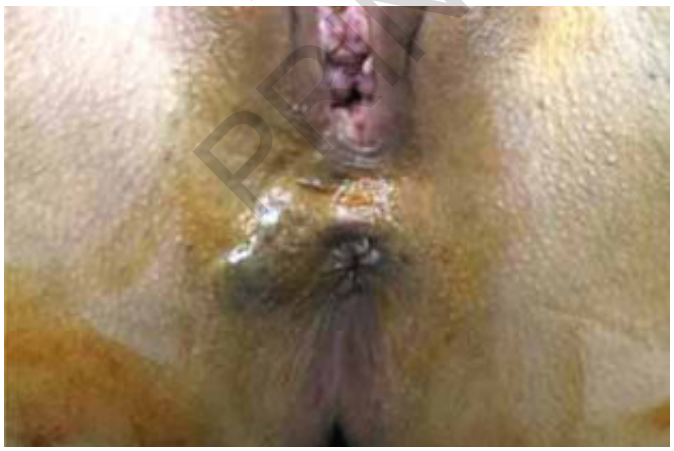

Fig. 1: A case of perineal endometriosis on the right mediolateral episiotomy scar.

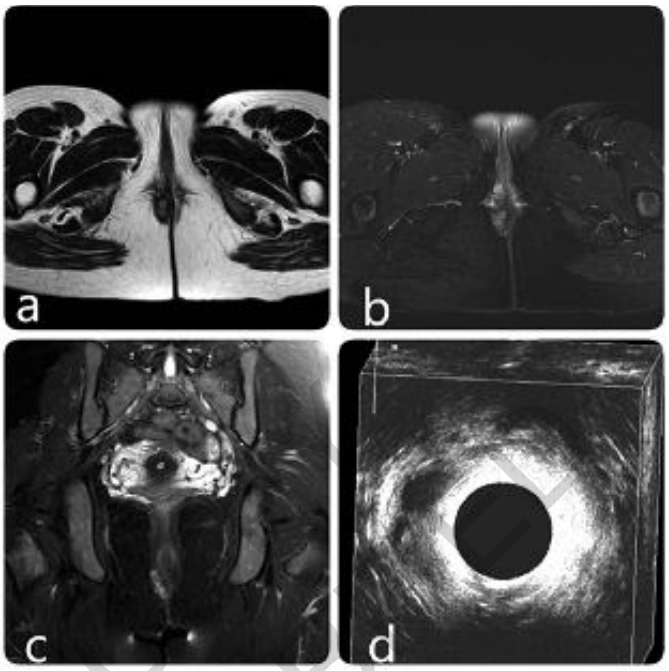

Fig. 2: A) Hypointense heterogeneous, well-circumscribed nodular lesion on the right lateral perineal midline on axial T1-weighted MRI; B) Increased enhancement in the center of the lesion on T1-weighted MRI with axial fat suppression; C)Contrast-enhanced T1-weighted MRI with coronal fat suppression showed contrast enhancement in the lesion; D) Hypoechoic nodular lesion involving the external anal sphincter on 3-dimensional endoanal ultrasonographic examination.

Treatment
The lesion was totally removed under general anesthesia with the affected sphincter muscles. External anal sphincter muscle damage that developed after excision was repaired by primary sphincteroplasty with 3/0 vicryl (Fig. 3).

Final Diagnosis
Pathology results were reported as endometriosis externa (Fig. 4).

Outcome and Follow-Up
The patient has been followed for two years since the surgery, has no complaints, and is fully continent.

Discussion

Perineal endometriosis is a rare disease that occurs on an episiotomy scar or postpartum tearing scar. The incidence is low, which can be explained by the bacteria present in the perineal wound causing infection or local necrosis. This does not provide a suitable environment for the survival of transplanted endometrial tissues, and the reduced estrogen level after delivery complicates the growth of transplanted endometrial cells [7,8].

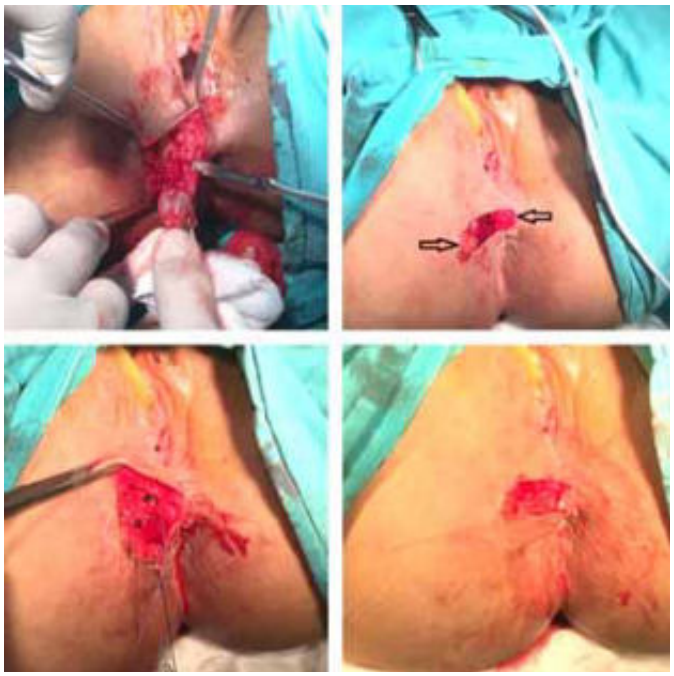

Fig. 3: Intraoperative findings. After the complete removal of the lesion (A) the muscle defect that developed on the external sphincter (B, arrows) was repaired with primary sphincteroplasty (C) and can be seen here after the repair (D).

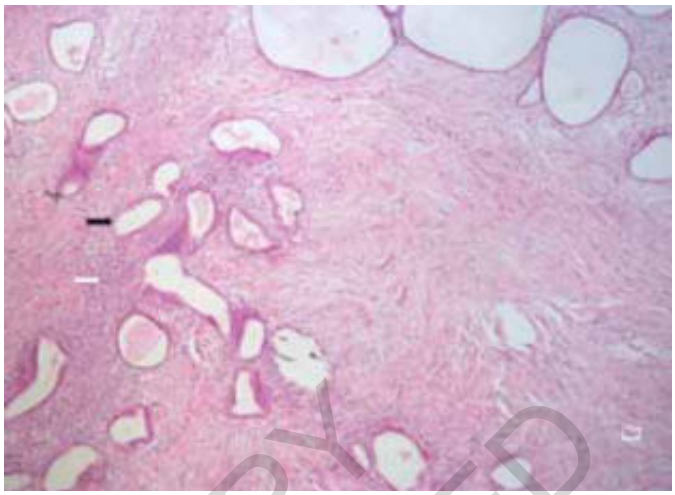

Fig. 4: Histological analysis. Endometriosis externa focus consisting of endometrial stroma (white arrow) and endometrial glands (black arrow) in subcutaneous soft tissue (H&E, 400×).

Perineal endometriosis occurs as a normal brown or blue-black cyst, papule, or nodule near the surgical scar. Most patients have complaints of cyclic pain and swelling in the perineal region during the menstrual cycle [7]. Cyclic bleeding from the perineal mass may be observed in some patients [9].

Early diagnosis and treatment of perineal endometriosis is important to prevent progressive involvement of the surrounding tissue, especially the anal sphincters, and to reduce the risk of postoperative fecal incontinence. For clinical diagnosis, a classical triad of cyclic pain, perineal mass, and history of rupture due to episiotomy or vaginal delivery is sufficient [5]. It may be useful to perform a physical examination during the menstrual cycle when the symptoms are most pronounced. Differential diagnosis includes suture granuloma, abscess, hematomas, lipomas, sebaceous cysts, and desmoid or malignant tumors [10]. The disease itself may undergo malignant transformation, albeit rarely [11-13]. Although fine-needle aspiration biopsy is recommended by some authors for definitive diagnosis, others are opposed to the idea, saying that this may cause new endometriotic implants according to the iatrogenic implant theory [11,14-16].

Pelvic endometriosis may also be present in patients with perineal endometriosis. Pelvic examination and imaging may be necessary when there is clinical suspicion [5].

Ultrasonography and MRI can provide valuable information in the evaluation and differential diagnosis of the lesion [17,18]. Computed tomography is not very useful in the diagnosis of endometriosis [19]. Because it is a rare disease, sonographic findings have been reported infrequently. In some publications, the lesion has been described as an irregular hypoechoic mass, and in some cases, it was reported that it can be seen as a heterogeneous mass with cystic anechoic and hyperechoic areas [7,18,20]. Although perineal ultrasonography may be helpful in diagnosis, it is insufficient to show the relationship of the lesion with the anal sphincters. Endoanal ultrasonography is an effective imaging modality to show the relationship of the lesion with the anal sphincters [7,17].

Optimal treatment of the disease appears to be total excision [5]. However, it is important to determine the relationship between the lesion and anal sphincters. It is recommended that a lesion not associated with the sphincters be removed with a margin of 0.5-1 cm. Incomplete excision to protect the sphincters is associated with high recurrence, while extensive excision can cause anal sphincter damage that may cause anal incontinence. In a study of 36 patients, all 7 patients who underwent incomplete excision due to external anal sphincter involvement developed early recurrence within 6 months. At the end of the study, wide excision of ectopic endometrial tissue with a healthy surgical margin was recommended, even if primary sphincteroplasty was required for anal sphincter involvement [5]. However, if the lesion is massive or anal sphincter involvement is high, hormonal treatment is recommended to minimize the sphincter damage by reducing the size of the perineal mass before surgery. In the same study, a patient who developed recurrence and underwent hysterectomy with bilateral salpingo-oophorectomy reported that the mass disappeared after 2 years. It was reported in another study that bilateral salpingo-oophorectomy with hysterectomy could be performed after discussion with

patients over 40 years of age who have masses too large for excision and a risk of anal incontinence [5,7]. In patients close to menopause, limited or incomplete resection with subsequent hormone therapy can be discussed with the patient to reduce the risk of incontinence caused by sphincter resection [7].

Conclusion

Perineal endometriosis is a rare disease. Although extensive surgical excision is the recommended treatment method, it is important to evaluate its relationship with the anal sphincters. This relationship can be clearly assessed by endoanal ultrasonography. Primary sphincteroplasty is recommended in cases of sphincter damage. High incidence of recurrence may occur after incomplete excisions to avoid sphincter damage.

Riassunto

L'endometriosi perineale è la presenza di tessuto endometriale nella regione perineale. La diagnosi e il trattamento precoci sono importanti a causa del coinvolgimento dello sfintere anale in quasi la metà dei pazienti. L'ecografia endoanale è una tecnica affidabile nella valutazione dell'endometriosi perineale con coinvolgimento dello sfintere anale. Questo rapporto descrive la presentazione, l'indagine clinica e il trattamento chirurgico di un caso di endometriosi perineale
Esso riguarda il caso di una paziente di 32 anni che presentava dolore ciclico e gonfiore di una vecchia cicatrice da episiotomia. L'ecografia endoanale tridimensionale ha mostrato una lesione con il coinvolgimento dei muscoli dello sfintere anale esterno. La lesione è stata asportata completamente ed è stata eseguita la sfinteroplastica primaria di riparazione del difetto dello sfintere anale esterno. Il referto anatomo-patologico definitivo è stato di endometriosi. Il periodo postoperatorio è decorso senza incidenti e non si osservava incontinenza anale.
Conclusione: l'endometriosi perineale è una malattia rara e può coinvolgere i muscoli dello sfintere anale. L'escissione incompleta per proteggere gli sfinteri è associata a un'elevata recidiva, mentre un'escissione estesa può causare danni allo sfintere anale che possono causare incontinenza anale. L'ecografia endoanale può essere necessaria nella pianificazione chirurgica. La sfinteroplastica primaria con escissione può essere necessaria nei casi di endometriosi perineale con coinvolgimento del muscolo sfintere anale esterno.

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