

Perineal Endometriosis

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Endometriosis is an enigmatic disease of menstruating females affecting the reproductive life. We report herein a case of perineal endometriosis in a 33-year-old woman. The patient presented two rounded, thickened plaques bilaterally on the perineum for about four years. The plaque on the right side was located mainly at the episiotomy scar. Histologic examination showed confirmative feature of glandular structures embedded in the cellular stroma and predecidual changes of late secretory phase of endometrium.

Radical operation of total hysterectomy with bilateral oophorectomy and partial excision of perineal lesions were performed. There was complete resolution with no relapse by the 6-month follow-up.

• This is of a very rare occurrence and three cases have been reported in the Korean literatures up to date. (*Ann Dermatol* 6:(2) 196-199, 1994)

Key Words: Endometriosis, Perineum

Endometriosis(E) is the condition in which tissue like that of the uterine mucosa occurs outside the uterus as a solitary lesion during the reproductive life^{1,2}. It most often affects the pelvis but may involve other areas, including the skin. Endometriosis occurring on the perineum, perineal endometriosis (PE) is quite rare^{1,3}, being reported in 3 of 138 cases with E(2.2%) in the Korean literatures up to date⁴⁻¹¹.

We describe herein a case of PE which occurred bilaterally on the perineum and was diagnosed by histologic examination.

REPORT OF A CASE

A 33-year-old housewife presented to our De-

partment of Dermatology had been referred from the Department of Gynecology for the evaluation of bilateral plaques occurring on the perineum. The plaques were developed insidiously about 4 years ago and have progressively worsened in spite of various treatment. The patient frequently suffered from dyspareunia and felt on occasion dull pain and discomfort with tenderness to the touch in the lesional areas. During the menstrual period, the lesions worsened with painful swelling and spotty bleeding.

Menarche had occurred at age 15, with regular 28- to 30-day cycles. The patient had a history of 2 normal vaginal deliveries with right mediolateral episiotomies, 8 and 9 years ago, and subsequent 3 abortions of unknown time(G5P2A3). Her family history was non-contributory. The routine laboratory studies were negative or within normal limits. The hormonal values of estrogen, luteinizing hormone and follicle stimulating hormone were 153.3 pg/ml(N: 61-440), 6.7IU/ml(N: 4-20) and 18.3IU/ml(N: 4-20) respectively. The general physical examination disclosed no abnormalities other than the perineal lesions.

The perineum had two rounded, dark-brown

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Fig. 1. The thickened plaques with irregular surfaces are located bilaterally on the perineum (right : 6 to 7 o'clock direction, left : 5 o'clock direction).

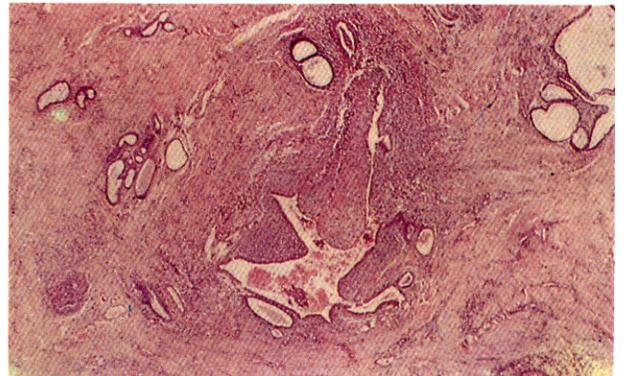


Fig. 2. Numerous glandular structures are embedded in the cellular stroma, showing architecture of endometrium (H & E stain, × 40).

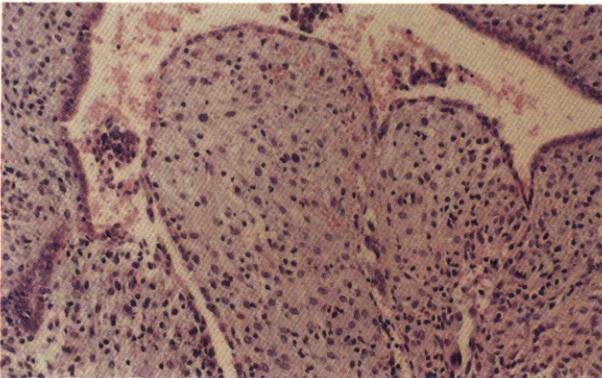


Fig. 3. Predecidual changes are seen in the stroma (H & E stain, × 200).



Fig. 4. The lesions are not improved with a 4-month hormonal therapy.



Fig. 5. The lesions show remarkable resolution like a scar one month after radical operation.

colored, thickened plaques, each measuring approximately 3.0x3.0cm(6 to 7 o'clock direction) on the right and 2.0x1.7cm(5 o'clock direction) on the left side, with rubbery consistency and perilesional hyperpigmentation. Their surfaces were irregularly studded with 2 to 3mm sized cobblestones(Fig. 1). The lesion on the right side was located mainly at the episiotomy scar.

The biopsy specimen from the lesion on the right side showed confirmative feature of numerous glandular structures embedded in the cellular stroma and edema in some areas. The glandular lumina were lined with a single row of columnar or cuboidal cells(Fig. 2). The stroma demonstrated some predecidual changes which consisted of plump cells with abundant cytoplasm and small

nuclei, consistent with features of late secretory phase of endometrium (Fig. 3). Disintegration of the glandular epithelium and bleeding were also noted. Accordingly, a few phases were apparent in a specimen. Immunohistochemical stains for S-100 protein, cytokeratin and carcinoembryonic antigen revealed negative reaction in glandular epithelium and stroma cells. The study of estrogen-progesterone receptors using the paraffin block failed.

Treatment was instituted with hormonal therapy with oral danazol 800mg per day for 2 months and buserelin(Suprecur)[®], gonadotropin-releasing hormone agonist, 0.9mg per day nasally for 2 months. Post-treatment values of estrogen, luteinizing hormone and follicle stimulating hormone were 84.1 pg/ml, 1.0IU/ml and 3.0IU/ml, resulting in a considerable decline in comparison with values prior to treatment. The perineal lesions and subjective symptoms, however, did not improve appreciably(Fig. 4). Therefore, the radical operation of total hysterectomy with bilateral oophorectomy and partial excision with cauterization of perineal lesions was carried out. There was no evidence of endometriotic foci in any other sites of the pelvis in the operation field. One month after the operation, the lesions showed remarkable resolution with the flattened features like a scar(Fig. 5). The patient was placed on hormone replacement therapy of estrogen(Premarin)[®], 0.625mg daily, because of severe estrogen-deprived symptoms, but this remedy was rapidly discontinued as it stimulated the redevelopment of the lesions. There was complete resolution of the lesions with no more relapse at the 6-month follow-up.

DISCUSSION

E occurs chiefly in reproductive aged women, usually between the ages of 25-45 years, affecting 1-2% of women in the United States^{12,14}. It is rare before menarche and after menopause, since endometrial tissue depends upon stimulation of estrogen for growth and propagation¹³. The lesion appears varied as a firm or hard tumor, mass or thickened nodule with cobblestones or a shotty feel¹³. The clinical symptoms of severity are also varied. The severity of symptoms such as dysmenorrhea and dyspareunia does not correlate with the extent of the disease¹³. The preferred sites are the ovaries, uterosacral ligaments, pelvic peritoneum,

rectovaginal septum, cervix, vagina and large bowel¹⁴. The skin may be rarely involved in the umbilicus, inguinal areas, lower abdominal area, labia and perineum¹. PE is particularly rare¹³. In a review of 82 cases with cutaneous E reported by Steck and Helwig¹, PE was observed in only two and, multiple lesions in two. In Korea, only 4 cases of PE have been reported including the present case^{4,11}.

The mechanism by which the development of E occurs is not yet clarified. Among several theories proposed to explain it, two major theories have been summarized^{1,13}: 1) the direct implantation theory is that the endometrial tissue is implanted directly through surgery, or by the lymph or blood stream; 2) the celomic metaplasia theory is that the embryonal multipotential cells in any tissue with a celomic epithelium are stimulated, under the proper stimuli, to differentiate into endometrial tissue. The former theory has been favorably recognized, but no single theory explains all cases. In a Steck and Helwig's review¹, 56 cases(68%) arose in the surgical scar. Recently, autoimmune factors may be implicated as seen by the higher incidence of E in infertile women(15-25%)^{12,13}. Besides, the lower level of estrogen receptor in decidual tissue seems to take part in the etiopathogenesis^{16,17}. It was assumed most likely that the present case occurred as a result of implantation of endometrial tissue into the episiotomy wound or laceration associated with parturition.

The leading histologic features are those of endometrium. The endometrial stromas surround the glandular structures lined with a single row of columnar or culoidal epithelium and have cyclic changes seen normally in uterine mucosa. More than one phase is apparent in a single specimen^{13,18}. Malignant transformation occurs very rarely. Immunohistochemically, expression of vimentin is present on the surface and glandular epithelial cells involving decidualization. The level of estrogen-progesterone receptors in the lesional tissue seems to be lower than those in the corresponding normal endometrium¹⁶, and the endometriotic foci show no cyclic changes in the localization of estrogen receptors^{13,17}. In the present case, an inconclusive result was obtained in the immunohistochemical studies of estrogen-progesterone receptors.

In the differential diagnosis, vulvar lichen simplex

chronicus or neurodermatitis, hypertrophic scar, kraurosis vulvae and benign tumor such as apocrine hidrocystoma should be considered. PE is easily differentiated by the characteristic histologic examination¹⁸.

Effective therapy is mainly hormonal suppression of the production of estrogen by the ovaries or surgical ablation of endometriotic lesion¹⁴. Danazol therapy results in symptomatic and objective improvement in 90% of cases, but its major side effects due to hyperandrogenic state may be problematic¹⁴. Gonadotropin-releasing hormone agonists such as nafarelin, buserelin and so on are at times necessary and effective¹⁴. However, these approaches often provide only temporary relief and recurrence is common as long as the ovaries remain^{13,14}. The radical therapy is total ablation of the uterus, tubes and ovaries, when pregnancy is not a consideration^{13,14}. In the present case, the hormonal therapy with danazol and buserelin was done for about 4 months, with no good clinical result. Consequently, radical operation and partial excision with cauterization of perineal lesions were carried out, with complete resolution at the 6-month follow-up.

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