Endometriosis of the Eyelid, an Extraordinary Extra-abdominal Location Highlighting the Spectrum of Disease

Kevin G. Sharghi, MD,* Nicholas A. Ramey, MD,† Patrick S. Rush, DO,*‡ and Douglas J. Grider, MD‡

Abstract: Cutaneous endometriosis is an uncommon dermatologic disorder predominantly seen in young women. Most commonly, it presents within a region of a previous surgical scar, often in relation to a gynecologic procedure on the abdomen or in close proximity to the umbilicus. The typical clinical presentation is that of papules or nodules with monthly cyclical pain and size variation. Histologically, the lesions are composed of endometrial stroma and glands. The pathophysiology is not well understood but is believed to be due to metastasis or seeding of endometrial cells. When this uncommon disorder occurs outside of the most typical clinical setting, it may cause some diagnostic difficulty. In this report, we present the first known case of cutaneous endometriosis on the eyelid.

Key Words: endometriosis, eyelid, CD10, estrogen receptor, progesterone receptor

(Int J Dermatopathol 2019;41:593–595)

INTRODUCTION

Cutaneous endometriosis is a unique entity in which endometrial stroma and glands are located within the cutaneous and subcutaneous tissue. Of women with endometriosis, only 0.5%–1% have cutaneous disease. It presents as red, brown, or violaceous papules and nodules that can vary in size or be painful in a cyclical manner, often corresponding to a patient’s menstrual cycle, and can even bleed. These cyclical symptoms are unique and associated with cutaneous endometriosis.¹

Scars at gynecologic surgery sites are most frequently involved and is believed to be due to seeding of cells during the procedure. The umbilicus is another site, even without a history of surgery, and is believed to occur from migration of endometrial cells.²,³ Here, we describe a case of cutaneous endometriosis that presented as a solitary telangiectatic macule on the right upper eyelid.

CASE REPORT

A 41-year-old woman presented to clinic with complaints of a growth on her right upper eyelid. It had been present for 6 months, and the lesion itself was asymptomatic. She did endorse xerophthalmia and ocular pruritus treated with artificial tears. Her eyesight was not affected. The remainder of her medical history was not contributory at the time of evaluation.

On physical examination, a 1 × 2-mm nonblanching macule of telangiectasias was present on the right upper eyelid (Fig. 1). A biopsy was obtained and sent to dermatopathology, which revealed a nodule of endometrial glands and stroma, which demonstrated strong expression of CD10, estrogen receptor, and progesterone receptor, by immunohistochemistry (Figs. 2 and 3).

The diagnosis of cutaneous endometriosis was made. After discussion with the patient, she endorsed having irregular and painful menstrual cycles. She was advised to follow-up with gynecology.

![FIGURE 1. The end of a cotton tip applicator pointing to the telangiectatic macule on the right upper eyelid.](image-url)
DISCUSSION

To our knowledge, this is the first known reported case of eyelid involvement with cutaneous endometriosis. The patient did not endorse any pain or variations of size in a cyclical fashion—contributing more to its extraordinary presentation. This case serves to highlight the spectrum of disease in cutaneous endometriosis and provides a reminder to the biologic ability of these lesions to occur in anatomical locations other than the umbilicus.

The innermost lining of the uterus is composed of endometrial stromal cells and glands to form the endometrium. This glandular and vascular lining functions to promote implantation of a blastocyst during the initial phases of pregnancy and forms the placenta. This layer is also shed during menstruation. In the more typical setting of endometriosis, ectopic endometrial cells are found outside of the uterus, within the abdominal and pelvic cavity due to retrograde menstruation. Patients commonly endorse pelvic pain, dysmenorrhea, and infertility.4

The pathobiology of cutaneous and subcutaneous involvement of endometriosis is unfortunately less understood. Lopez-Soto et al published their retrospective study of patients with a histopathologic diagnosis of cutaneous endometriosis in 2017. With 20 years’ worth of data, their group documented 33 patients with the diagnosis of cutaneous endometriosis: 84.84% within scars and 15.15% outside of scars.2 It is believed that seeding of endometrial cells during gynecologic procedures leads to ectopic tissue implantation and development within surgical sites, while umbilical involvement, as well as at other cutaneous sites, is likely due to metastatic endometrial cells.1

Differentiating between a classical presentation of cutaneous endometriosis and other diagnoses should be straightforward, as the cyclical growth and pain are specific to cutaneous endometriosis. Within the clinical differential diagnosis of lesions occurring on the abdomen is Sister Mary Joseph’s nodule, which can be a painful or tender nodule at the umbilicus and is a cutaneous manifestation of abdominal or pelvic malignancy.5

Histopathological findings are pathognomonic: endometrial glands, tall columnar epithelial cells set within a field of monotonous basophilic spindled endometrial stromal cells, and loose concentric fibromyxoid tissue. Metaplastic changes may be seen. In keeping with the tissue origins, erythrocytes and hemosiderin deposits are often present. Immunohistochemical staining demonstrates glandular and stromal cells to be positive for estrogen and progesterone receptors with expression of CD10 within endometrial stromal cells.6–8

Excision of cutaneous endometriosis should be considered, with or without preoperative hormone therapy to reduce size. The incidence of recurrence is not known. Purvis and Tyring9 documented some benefit with both danazol and leuprolide, but the patients later went on to have excisions. Nevertheless, patients should be referred to their primary care physician or gynecologist for evaluation of pelvic disease.

FIGURE 2. Hematoxylin and eosin stain of right upper eyelid macule demonstrating endometrial glands and stroma as well as erythrocyte extravasation.

FIGURE 3. A, CD10 stain highlighting the endometrial stroma. B, Estrogen receptor showing strong diffuse nuclear positivity. C, Progesterone receptor showing strong nuclear positivity within the endometrial stroma and glands.
REFERENCES