Cutaneous Endometriosis of the Umbilicus
--- Report of a Case and Review of the Literature ---

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A 46 year-old woman presented with a flesh-colored umbilical tumor which was rhythmically painful and waxed and waned with menstruation. A biopsy of the lesion revealed ectopic endometrial glands surrounded by endometrial stroma and scattered hemosiderin-laden macrophages and was diagnosed as endometriosis. Although cutaneous endometriosis of the umbilicus is a rare occurrence, it is sometimes of great importance in the differential diagnosis of malignancy of the umbilicus.

Key words: Endometriosis, Umbilicus, Skin neoplasms.

Introduction

Endometriosis is defined as the presence of ectopic endometrial tissue outside the endometrium and myometrium. The disease has been described in almost every organ of the body, as well as in males. ¹ An umbilical tumor may be diagnosed on the bases of its clinical characteristics as a hernia, malignant melanoma, or metastatic neoplasm. We present a relatively rare case of cutaneous endometriosis of the umbilicus which occurred spontaneously without association of operation.

Case history

A 46 year-old woman, gravida 3 and para 3, presented with an umbilical nodule for one year duration. She had suffered from lower abdominal pain for 2 years. Rhythmic pain of the lesion was worse in the premenstrual period. A little blood-tinged fluid discharged from the lesion occasionally.

She did not have serious illness or surgery. Menarche occurred at the age of 14 years, and the menstruation was quite regular. She did not take any oral contraceptives and denied history of dysmenorrhea. Grossly, the tumor was approximately 1.2 cm in diameter arising from the umbilicus ( Fig. 1 ) and was completely excised under 1 % Xylocaine local anesthesia. Histopathological examination showed a piece of fibrous tissue, which contained many ectopic endometrial glands surrounded by

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endometrial stroma (Fig. 2). Some of them contained blood. Scattered hemosiderin-laden macrophages were also present (Fig. 3). The diagnosis of cutaneous endometriosis of the umbilicus was made. No recurrence happened later.

Discussion

Since Villar first described cutaneous endometriosis of the umbilicus in 1886, more than 200 cases have been reported. The incidence is estimated to be 0.5 to 1% of all women with endometrial ectopia. It occurs in women between 30 and 40 years of age, as with all other types of endometriosis. Although cutaneous endometriosis of the umbilicus can occur after gynecological surgery, it is generally spontaneous. The other locations of spontaneously occurring cutaneous endometriosis are inguinal area and perianal regions.

It is usually firm to hard and ranges from 0.5 to 6 cm in diameter. The color is often blue, dark blue or black but occasionally pink or brown, depending upon the depth of the lesion in the skin and age and amount of hemorrhage. The spontaneously occurring umbilical lesions are always precisely associated with the umbilical dimple. Sometimes secretion or some bleeding may occur through the cutaneous lesions, concomitant with menses, which used to be called the "menstruating tumor".

The endometriosis may cause pain, swelling, itching or tenderness, mainly in the premenstrual period, as this ectopic endometrial tissue responds cyclically but not consistently to the ovarian hormones. The disclosure, on examination, of a tender, indurated umbilicus containing cystic structures which exude old blood on exposure, is pathognomonic. Symptoms of pelvic endometriosis are usually absent and pelvic involvement is confirmed in only one-half of the patients at exploratory operation.

The possibility of endometriosis should be considered in the differential diagnosis of masses that located in any area of the female body. The ultrasonographic examination is useful in determining whether a cutaneous mass is cystic, solid, or mixed. The fine needle aspiration cytology was reported to be performed to rule out a neoplasm, which may be benign or malignant.

The differential diagnosis of a lesion of the umbilicus including hernia, primary and metastatic neoplasms, embryological rest masses and various granulomata, all these must be excluded before diagnosis of cutaneous endometriosis of the umbilicus is confirmed. However, massive size, rapid growth, and extensive ulceration give rise to a clinical aspect suspicious of malignancy. In case of continual recurrence, the possibility of malignancy should be kept in mind.

The cut surface is typically gray-white, with or without focal areas of recent or old hemorrhage. Nodular lesions that are poorly circumscribed occurred in the dermis and less often in the subcutaneous tissue. The major pathologic changes are glandular element and stroma resembling those of the endometrium, and peripheral reactive fibrosis to these ectopic structures. Disintegration of the epithelium and presence of the erythrocytes in the
Fig. 1. The flesh-colored tumor arises from the umbilicus.

Fig. 2. The ectopic endometrial glands are surrounded by endometrial stroma.
Fig. 3. The hemosiderin-laden macrophages are present.

lumina resemble menstruation. Macrophages with ingested hemosiderin are frequently observed. Every stage of the endometrial cycle is exhibited by the ectopic glands, and more than one stage is often apparent in a single specimen. However, no good correlation exists between the histologic appearance of the lumina in the cutaneous lesion and the menstrual stage, possibly associated with adequacy of the local blood supply or various response of endometrial tissue to the normal hormone fluctuations of the menstrual cycle. The synchrony between the eutopic and ectopic endometrial glands varies from 44 to 80%. The decidualized endometriosis with a pseudomalignant appearance due to myxoid change and the vacuolated cells present in the myxoid areas which can assume a signet-ring appearance, similar to the cells characteristic of mucin-producing adenocarcinoma, can be misinterpreted as metastatic malignancy by the dermatologists not familiar with general pathology.

There are two major theories of the cause and pathogenesis of cutaneous endometriosis, the transportation theory and the metaplasia theory. The transportation theory proposes that the endometrial tissue in some way is transported to an extraterine location. Scott et al demonstrated lymphatic channel between the pelvis and umbilicus, that blood, dye, and radioactive materials can migrate by retrograde flow. The endometrial cells may be existent and transported in the lymphatic system because pelvic lymph-nodes containing
endometrium-like glands or stroma, or both has been proven in a necropsy study. So, it may explain cases of spontaneous endometriosis in the umbilicus. The other supposed mechanisms of transportation possibly associated with cutaneous endometriosis of the umbilicus were suggested that spread by implantation in a hernia sac and embolization of endometrial cells via blood vessels. Scar endometriosis, which often appears after gynecological operations or cesarean sections, may be explained by iatrogenic mechanical transportation due to secondary surgical maneuvers. The metaplasia theory suggests that endometriosis may develop from the pleuripotent cells of the celomic mesothelium under some stimulation. An unusual case of cutaneous endometriosis in a patient affected by a granulosa cell tumor was reported and endometrial metaplasia may be promoted by high estrogen levels was suggested. So, it is reasonable that 'Endometrial cells themselves stimulate imitative metaplasia or cellular replication when they are transported by any means to a susceptible tissue that has a high degree of primitive pleuripotentiality' described by Steck and Helwig. Immunological process also plays a part in the development and progression of endometriosis. Women with endometriosis have endometrial cells that are somewhat less sensitive to the normal body defense mechanisms and, as a result, implant, and grow more easily in ectopic sites. Furthermore, the accompanying altered immune response acts inappropriately with the production of increased tissue growth factors such as fibronectin stimulating a fibrotic reaction that protects the endometriotic implants. No single theory can explain the spontaneous occurrence of cutaneous endometriosis of the umbilicus. The development is probably a multifactorial event.

Medical therapy for cutaneous endometriosis using anabolic hormone manipulation with Danazol or Nafarelin acetate is inappropriate because of the risk of side effects of the medication. Simple excision of a solitary lesion of cutaneous endometriosis of the umbilicus, sparing the umbilicus when possible, is sufficient. Although malignant transformation of endometriosis is very rare, follow up in cases of cutaneous endometriosis is advisable and a curettage should be performed to examine the possibility of malignant changes whenever an endometrioid carcinoma is discovered.

Reference


中華皮誌 十三卷二期 八十四年六月
摘要

臍部皮膚子宮內膜異位
--- 病例報告暨文獻回顧 ---

黃柏翰 蔡仁雨 陳明君 曾興隆

長庚紀念醫院 皮膚科

一位 46 歲女性於臍部出現一疼痛與月經週期相關之膿色腫瘤，病灶切片可見異位之子宮內膜腺為子宮內膜基質包被，並有含黏膜質之吞噬細胞，而診斷為子宮內膜異位。雖然臍部皮膚子宮內膜異位並不常見，但是在鑑別臍部惡性腫瘤時，仍應考慮之。
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