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clinical practice

Primary abdominal wall endometriosis: presentation of rarely seen two cases

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SUMMARY: **Primary abdominal wall endometriosis: presentation of rarely seen two cases.**

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Abdominal wall endometriosis is a pathology which usually develops after preceding surgeries on the surgical incision line and shows clinical manifestation especially during menstrual cycle. However, primary abdominal wall endometriosis is seen very rarely and it is a condition developing without a previous history of surgery. In this paper, we aimed to provide a contribution to the theories of pathogenesis of the disease by presenting two cases of primary abdominal wall endometriosis in two patients without previous history of surgery.

KEY WORDS: Endometriosis - Abdominal wall - Endometrioma.

Introduction

Endometriosis is defined as the presence of an ectopic endometrial gland and connective tissue outside the uterine cavity and myometrium (1). While the frequency of endometriosis varies in the literature, disease prevalence is considered to be between 7% and 10% among women of reproductive age (2). Reported patients of abdominal wall endometriosis comprise of cases occurring usually in 20-40 years of age women 3 months-10 years after a previous a caesarean section delivery (3). But, contrary to incisional endometrioma and scar endometrioma, it is known that primary abdominal wall endometriosis is defined as presence of an ectopic endometrial tissue on the parietal peritoneum irrespective of a previous surgery. Therefore, while endometriosis in areas of incision is a well-described lesion, endometriosis in areas not including incision is a very rare entity with unclear pathophysiology. In this paper, we aimed to make a contribution to the theories of pathogenesis of the disease by presenting two cases of primary abdominal wall endometriosis in two patients without a previous history of surgery.

Case 1

A 48-year-old female patient presented to outpatient clinic with complaint of a palpable, painful, solid and draining mass in her umbilicus. The patient described an increase in her complaints of pain for last three years especially more specifically during menstrual cycles. It was learned that the patient was treated with diagnosis of umbilical sinus and local antibiotic therapy was administered in different healthcare institutions. The patient had a history of two normal vaginal deliveries and two diagnostic curettage due to the uterine wall thickness. The patient had no previous abdominal surgery, abdominal incision and coexisting disease. At the physical examination, there was a solid well-circumscribed mass in size of 2 cm with nodular millimetric thickening in places and appearing to be localized to the subcutaneous tissue (Figure 1). At laboratory investigations of the patient, it was determined that hemoglogin value was low, sedimentation and C-reac-

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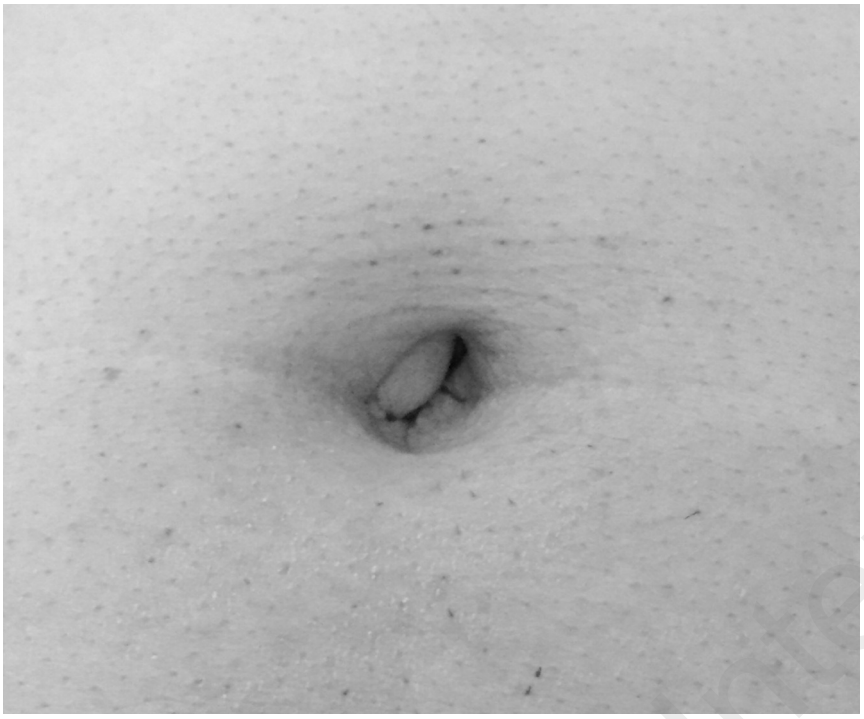

Figure 1 - Outer appearance of subcutaneous umbilical mass of the patients.

tive protein levels were normal. Ultrasonography of superficial soft tissue mass revealed a well-circumscribed mass in the umbilicus, localized to the subcutaneous tissue, without no attachment with intra-abdominal organs and structures. A computed tomography scan of the pelvis showed multiple uterine myomas with the biggest one in size of 3 cm, but pelvic endometriosis was not observed. The mass was excised with a pre-diagnosis of umbilical endometriosis and intact surgical margins under local anesthesia. Pathological evaluation of the mass was reported to be consistent with endometriosis (Figure 2). At the postoperative first month physical exam and ultrasonographic assessment of the patient, no recurrence was observed.

Case 2

A 29-year-old female patient presented to outpatient clinic with complaints of left lower-quadrant pain exacerbating especially during menstrual cycles and a palpable mass for last six months. The patient had no history of previous surgery. At the physical examination, there was a palpable well-circumscribed mass approximately in size of 4 cm with a rubbery consistency in left inguinal region in the neighborhood of the rectus muscle. Ultrasonography of superficial soft tissue mass revealed a well-circumscribed mass approximately in sizes of 4x3 cm in the right lower-quadrant of the abdomen on external oblique fascia without no attachment with subcutaneous tissue and muscle tissue planes. The patient underwent surgery for histopathological sampling and a well-circumscribed mass was determined on the left external oblique muscle fascia densely adherent to muscular fascia and the lesion was excised with intact surgical margins. The fascial defect was repaired primarily. The patient was discharged on the postoperative 1st day. Histopathological examination of the specimen was found to be consistent with endometriosis. At the postoperative twelfth month physical exam and ultrasonographic assessment of the patient, no recurrence was observed.

Discussion and conclusion

Although abdominal wall endometriosis is a rarely seen condition in clinical practice, its exact incidence was reported to be between 0.03% and 1% (4). Among all of these case presentations, primary abdominal wall endometriosis without a previous history of surgery could be identified in only 20% of

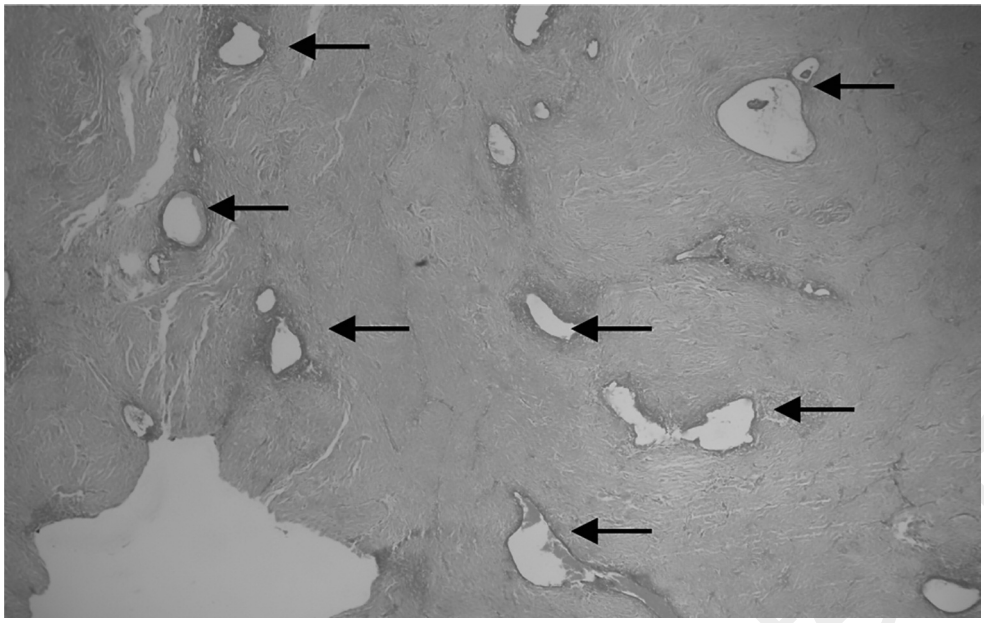

Figure 2 - Many foci comprising of endometrial gland and stroma located within the fibrous stroma are seen in the section (black arrows) (Hematoxylin Eosin 4x).

all patients (5). Patients often consult a physician with complaint of cyclic pain consistent with menstrual cycles. Since it is rarely seen and its symptoms show clinical variation, even in presence of a strong pre-clinical suspicion, diagnosis of primary abdominal wall endometriosis can be overlooked by an experienced surgeon. In differential diagnosis, suture granuloma, abscess, presence of benign or malignant tumor and abdominal wall hernias may mimic this disease (6, 7).

Although it is very difficult to make a definite diagnosis of primary abdominal wall endometriosis with imaging methods, they are used efficiently to determine the extent of disease and for planning of surgery especially in recurrent large masses (6). Diagnosis can be made with percutaneous fine-needle aspiration biopsy performed from suspected lesion (3), but it is still controversial due to presence of risk for ectopic endometrial tissue scattering along needle tract. Demonstration of ectopic endometrial tissue histologically following removal of the mass by ensuring intact surgical margins is the most definitive diagnosis and treatment method.

Although underlying pathophysiology of endometriosis is not known considerably, a great deal of progresses were made in understanding the development of the disease. Accepted theory on the pathogenesis of endometriosis as development of Mullerian residues and implantation due to retrograde menstruation remains incapable of describing the cases of extragenital endometriosis. Occurrence of cases of distant organ localization like primary abdominal wall endometriosis can be explained when it is supported by theories of genetic predisposition, dysfunctional immune response, and the coelomic metaplasia, the lymphatic or vascular metastasis (9). However, experimental demonstration of both of coelomic metaplasia and development of Mullerian tissues in menstrual debris showed that these theories took part together and complementary to each other (theory of induction) (10). Additionally, a composite theory indicating a direct invasion and implantation of endometrial tissues through lymphatic or vascular metastasis was also described in development of cases of extragenital endometriosis (6, 11). In the presence of these theories, extra uterine adhesion, invasion and angiogenesis of ectopic endometrial tissue, responses of macrophages and Natural Killer (NK) cells to ectopic tissue, local concentration of hormone, changes in the cellular and humoral immune system play an important role in occurrence of endometriosis (12). Additionally, observation of endometriosis more frequently in first-degree relatives of affected women, demonstration of familial predisposition in various studies (2) and association of endometriosis with monozygotic twins reaching up to 87% (1) highlight that this disease can be a genetically inherited disorder due to

underlying immune system disorder (13).

However, since cases of extragenital endometriosis are rarely observed, valid and efficient studies can not be performed in identification of phenotypic subgroups of this disease and follow-up of the role of these subgroups on disease prognosis and treatment response (14). Besides, until today, a molecular and genetic test efficient in diagnosis of endometriosis, guiding in disease prognosis, determinative in treatment efficiency could not be developed (15).

In conclusion, while endometriosis in the previous incision scar areas is a well-described condition, we think that presentation of this case with primary abdominal wall endometriosis can provide a contribution to the theories of development of the disease. More comprehensive and reliable information will be obtained related to diagnosis, pathogenesis and clinical course of the disease with phenotypic, molecular and genetic studies which will be performed about rarely seen primary abdominal wall endometriosis. Prospective, multicenter and systematic studies which will be performed in the near future will be guiding about this disease.

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