

Extrapelvic Endometriosis: A Study of 17 Cases

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ABSTRACT

Endometriosis is defined as the presence of tissue, histologically similar to endometrium outside the uterine cavity. Endometriosis at unusual sites is a diagnostic and therapeutic challenge for the clinician.

Materials and methods: This is a study of 17 cases presented in last 15 years at our institute with extrapelvic location and unusual symptomatology.

Observation and discussion: Extrapelvic endometriosis can affect unusual sites, like abdominal scars, episiotomy scars, umbilicus, bladder, GI tract and on rare occasions may be found at undersurface of diaphragm. The clinical presentation in each case is different and often misleading. Diagnosis can be reached by high degree of clinical suspicion. Histopathology of tissue is most reliable for diagnosis. CT and MRI offer higher diagnostic accuracy compared to ultrasonography. Complete excision of the tissue is ideal treatment however medical methods, like progesterone/danazol, can be tried with varied success in difficult cases.

Conclusion: High degree of clinical suspicion along with energetic treatment of extrapelvic endometriosis is the goal of this uncommon but challenging pathology.

Keywords: Extrapelvic endometriosis, Scar endometriosis, Endometriosis externa, Endometrioma.

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INTRODUCTION

Endometriosis is characterized by the presence of functional endometrial tissue consisting of glands and/or stroma located

outside the uterus. It is a painful chronic disease occurring in 5 to 15% of menstruating women.¹

Endometriosis seldom occurs as a solitary lesion but when it does, it can still exhibit the total gambit of destructive activity. It is also a disease that seldom affects extragenital organs without pelvic involvement. While endometriosis is a diagnostic and therapeutic dilemma, its undetectable masquerading presentation poses a bigger challenge.

Study of 17 such rare and unusual cases of endometriosis is presented to offer the diagnostic criteria and to impress upon that how it can be unraveled with ease and confidence.

MATERIALS AND METHODS

In last 15 years, 17 cases of endometriosis externa presented at our institution with very unusual symptomatology. No presurgical invasive method for diagnosis in these cases was used. Surgical excision and biopsy study was the mainstay of diagnosis. Careful interpretation and correlation of surgical and pathological findings in the context of patient's clinical presentation helped us to arrive at the diagnosis.

DISCUSSION

Presentation of 17 such rare cases is summarized in Table 1.

The most common location of endometriosis is within the pelvis. However, extrapelvic endometriosis is a fairly uncommon disorder and difficult to diagnose. The various sites for extrapelvic endometriosis are urinary system, bowel, omentum, lymph nodes, lungs, pleura, umbilicus, hernia sacs, episiotomy and nasal mucosa, and on rare occasions may be found on the undersurface of the diaphragm. It is a common disease of unknown etiology. Many theories, like Sampson's retrograde menstruation theory, Minh's theory—metaplastic

Table 1: Observation

Sr. no.	Site of endometriosis	No. of cases	Presenting complain
01	Abdominal scar	2	Poor healing of abdominal scar
		2	Scar tenderness which aggravates during periods
		2	Nodular cystic swelling which gets tender at time of menses
		1	Keloid-like presentation
		Total = 7	
02	Episiotomy scar	3	Thickened and tender scar, dyspareunia. Excision and biopsy
03	Umbilicus	1	Bluish cystic swelling which increases in size and tenderness during menses
		2	Disrupted wound with brownish discharge at the time of menses
		Total = 3	
04	Bladder	2	Frank hematuria, menstrual strangury During menstruation, diagnosis of hematuria confirmed by passing catheter, sent for HPE which confirmed endometrial tissue. Cystoscopic fulguration and danazol therapy
05	Rectum	1	Bleeding piles, melena. Tenesmus related to menstruation. Piles removed surgically and HPE confirmed endometriosis. Danazol therapy.
06	Omentum	1	Acute abdomen and lump. Laparotomy done to find the cause. There was torsion of omental mass which was removed and sent for HPE. Endometriosis confirmed. Danazol was given.

transformation of pluripotential peritoneal mesothelium, migration of cells through the lymphatic system or via hematogenous spread, endometrial nodules formation from metaplasia of mullerian remnants or iatrogenic displacement of the deciduas during a cesarean section. The 'neurological hypothesis' is a new concept in the pathogenesis of endometriosis: The lesions seem to infiltrate the large bowel wall along the nerves, at a distance from the primary lesion. However, other factors, immunological, genetic and familial, could be involved in the pathogenesis of this disease. The iatrogenic route explains the presence of endometriosis at episiotomy and laparotomy scars.²

Scar endometriosis most commonly occurs after operation on the uterus and tubes. Incidence of scar endometriosis following hysterotomy is 1.08 to 2%, whereas after cesarean section, it is 0.03 to 0.4%. The reason for higher incidence after hysterotomy has been given as the early deciduas has more pluripotential capabilities and can result in cellular replication producing endometriosis.

Endometriosis usually becomes apparent in the reproductive years, when the lesions are stimulated by the ovarian hormones. Forty percent of the women present symptoms in a cyclic manner which are usually related with menses.³ In our patients, symptoms relapsed irregularly and were not related with menses.

The gross clinical and pathological picture changes as lesions age. The endometrioma often becomes totally buried in scar tissue. Such lesions encased in dense scar tissue are still perfused cyclically with hormones which stimulate the hemorrhagic response at menstruation. The vascular perfusion sometimes becomes inadequate because of scar tissue, leading to a vascular necrosis. Biopsy of suspected areas should provide a definitive histological diagnosis. To be conclusive, the presence of both endometrial glands and stroma should be present.⁴

Various diagnostic methods have been described in the literature. Till recently, the use of ultrasonography (USG) have hardly been reported in detail and anecdotal reports have described it as nonspecific, can give a varied picture of hypoechoic mass with scattered internal echoes. USG and color Doppler when combined with clinical data may substantially contribute to the preoperative diagnosis. FNAC has been reported to be accurate in some cases, but not in all cases.⁵ MRI is more helpful than CT in detecting the planes between muscles and abdominal subcutaneous tissue. Nowadays, CA-125 and other immunochemistry with cytokeratin (CK) of different molecular weight (CK 7 and CK 20) are claimed to be helpful for the diagnosis of endometriosis externa.⁶

Treatment of choice is wide excision of the lesion and medical management, if required. Only medical treatment with the use of progestogens, oral contraceptive pills and danazol is not effective and gives only partial relief in symptoms. Recently, there has been report of use of gonadotrophin agonist but only with the prompt improvement in symptoms with no change in the lesion size. These patients need to be followed up because

of the chances of recurrence, which require re-excision. In cases of continual recurrence possibility of malignancy should be kept in mind.⁷ To prevent the occurrence of scar endometriosis, it has been suggested that at the end of surgery, especially on uterus and tubes, the abdominal wall wound should be cleaned thoroughly and irrigated vigorously with high jet solution before closure.⁸

Despite more than 80 years of intense interest in the condition, it is probably true to say that we are no closer even today to arrive at a consensus to either pelvic pain or infertility, or the optimum form of the management.

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