Original Research Article

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A Clinicopathological Study of Endometriosis with Special Reference to Endometriosis in Very Rare Locations

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Abstract

Endometriosis is a chronic debilitating disease that occurs in 6-10% of the female population and about 35-50% of women with pain, infertility or both and characterized by endometrial tissue outside the uterine cavity. Endometriosis can be divided into pelvic and extrapelvic sites. The most common sites affected are ovaries, however it can occur in rare sites like gastrointestinal tract, pulmonary structures, diaphragm, umbilicus, urinary system, laparotomy and episiotomy scars, breasts, perineum, extremities and even in central nervous system. The clinicopathological spectrum of endometriosis with emphasis on occurrence at unusual sites were studied with total of 30 cases from different sites diagnosed as endometriosis by FNAC (4 cases) and histopathology in all cases over a period of 17 years in a tertiary care hospital. The most common site located is ovary in 23 cases (76.6%) followed by abdominal wall in four cases (13.3%). The uncommon extrapelvic locations , include each in vulva (3.33%), perianal (3.33%), and bladder (3.33%). Clinical presentation in different location of endometriosis varies. Malignant transformation is very rare and may occur in less than 1% of women, the most common site being ovary. Regarding pathogenesis, several theories are suggested. Follow up varied from 1-2 years. The typical presentation and local findings allows to make a correct diagnosis in clinically suspicious endometriosis. Proper caution must be taken during lower abdomen surgical procedures. Complication like bleeding from cyst, bowel obstruction in intestinal, reduced fertility etc, may be circumvented, if early diagnosis and treatment is instituted.

Keywords: Endometriosis; Chocolate Cyst; Scar Endometriosis; Extra Pelvic Endometriosis.

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Introduction

Endometriosis is a benign progressive disorder of the female genital tract, principally characterized by endometrial tissue consisting of glands and/or stroma found outside the uterine cavity. It is an enigmatic disease because neither the etiology, nor the natural history nor the mechanisms of the associated pelvic pain and/or infertility are completely understood. Malignant transformation is rare and may occur in less than 1% of women, the most common site being ovary [1,2].

In general, endometriosis can be divided into pelvic and extrapelvic sites. The most common sites affected are ovaries, uterine ligaments, recto and vesicovaginal septae, pelvic peritoneum, cervix, labia and vagina. Very occasionally it is found in extrapelvic sites such as gastrointestinal tract, pulmonary structures, diaphragm, umbilicus, urinary system, laparotomy and episiotomy scars, breasts, perineum, extremities and even in central nervous system [3,4]. Typically endometriosis causes pain, dysmenorrhoea, dyspareunia and infertility, although 20-25% of patients are asymptomatic [5].

Endometriosis is a chronic debilitating disease that occurs in 6-10% of the general female population and about 35-50% of women with pain, infertility or both. About 25-50% of infertile women have endometriosis and 30-50% of women with endometriosis are infertile. Recent data indicate that the incidence of endometriosis has not increased in the last 30-35 years and remains at 2.37-2.49/1000/y which equals to approximate prevalence of 6-8% [5]. Several classifications exist. The Acosta classification emphasizes the presence and volume of endometriosis. The revised American Fertility Society (AFS) classification takes into account the extent of the disease and the amount and severity of adhesions [2].

The present study was undertaken to analyse clinicopathological spectrum of endometriosis with an emphasis on occurrence at unusual sites and also on prevention and treatment.

Methodology

The records of endometriosis from different sites, were retrieved from the department of pathology, MR Medical college, over a period of 17 years from January 2001 to December 2017. A total of 30 cases from different sites diagnosed as endometriosis by histopathological examination, on slides stained with haematoxylin and eosin were reviewed in all the cases. Special stains and IHC were done where ever necessary. Follow up information was available in 18 cases over a period of 1-2 years. The criteria for diagnosing endometriosis was presence of endometrial glands and/or stroma, in old and fresh haemorrhage and haemosiderin-laden macrophages.

Results

Observations

The present study included 30 cases of endometriosis in age group of 24 to 45 years, with a mean age of 29.8 years. The most common clinical feature is cyclical pain followed by local swelling at the site.

The operative procedures performed included total abdominal hystrectomy with salpingoophrectomy of 19 cases and oophrectomy of affected ovary of 4 cases. Excision biopsy was done in six cases and bladder biopsy in one case Table 1.

The most common site located is ovary in 23 cases (76.66%) followed by abdominal wall in four cases (13.33%). The uncommon location each in vulva (3.33%), perianal (3.33%), and bladder (3.33%) of extra-pelvic endometriosis is seen. Clinical presentation in different location of endometriosis is depicted in Table 2 and 3.

In ovarian endometriosis, patients clinically presented with intermittent spasmodic pain abdomen. On USG, unilateral ovary was affected with cystic loci of fluid collection and minimum vascularity was noted. Diagnostic laproscopy was performed in three cases, which showed haemorrhagic foci, and with powdered burnt areas. All patients were fertile having three to four children. FNAC was not carried out in ovarian lesions. The gross appearance of cut section of endometriotic cyst looks like "chocolate cyst " Figure 1. The lesions vary in size (4-8cms) in relation to phases of menstrual cycle. Microscopy revealed, typical lesions composed of endometrial glands, and stroma with fresh and old (hemosiderin containing) hemorrhagic foci (Figure 2).

Table 1: Operative procedures

Operative Surgery	Number of cases
Total abdominal hysterectomy + Salpingoophrectomy	19
Oophrectomy	04
Excision biopsy	06
Bladder biopsy	01
Total	30

Table 2: Site distribution

Age(years)	Site	Number of cases	%
24-45	Ovary	23	76.66
22-30	Abdominal wall	04	13.33
25	Vulva	01	3.33
28	Perianal	01	3.33
28	Urinary Bladder	01	3.33
29.8 (mean age)	Total	30	99.98

Table 3: Clinical presentation

Ovary	Cyclical pain abdomen
Abdomen wall	Pain at the site, nodular swelling
Vulva	Pain, swelling and discharge
Perianal	Recurrent swelling and cyclical pain at the site
Urinary bladder	Lower abdominal pain, hematuria and white discharge

In all four patients of abdominal wall lesions (Figure 3), presented with nodular swelling of abdomen, with previous history of surgery, with gap between 10-12 years. FNAC in 30 year female patient presented with per abdomen swelling since 3 years and pain at the site during menstrual cycle. Fine needle aspiration cytology (FNAC) (Figure 4) diagnosed as scar endometriosis and was confirmed by histopathology study. Thirty two years old patient clinically diagnosed as umbilical granuloma, was 2 inches away from scar tissue measuring 4x3cms. The cytology findings and histology was consistent with endometriosis. Another patient of 30 years old, clinically diagnosed as fibrolipoma, was histologically diagnosed as endometriosis. The fourth case, clinically diagnosed as mass per abdomen, with cut open tissue of 2-3cms size, shows fibrofatty tissue with gray white and haemorrhagic areas and histopathologically diagnosed as scar endometriosis (Figure 5).

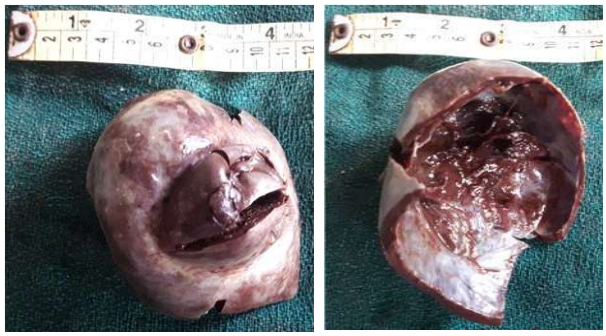

Fig. 1: Ovarian endometriosis: Gross image of ovary & cut-section showing haemorrhage at the centre of cyst(chocolate color)

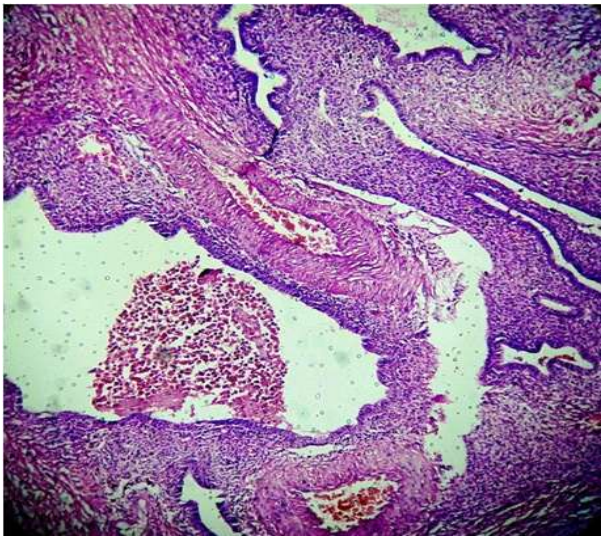

Fig. 2: (10x) Ovarian endometriosis: Cystically dilated endometrial glands with intraluminal hemorrhages amidst fibrocollageneous tissue. (H&E, LP)

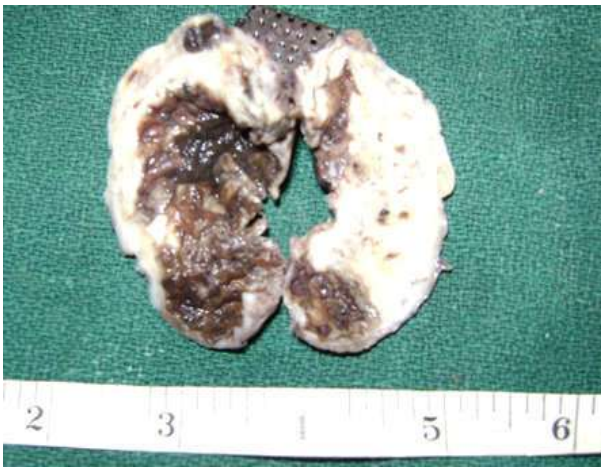

Fig. 3: Scar endometriosis gross: Skin covered tissue measuring 3x2x1 cms with haemorrhagic area at the centre

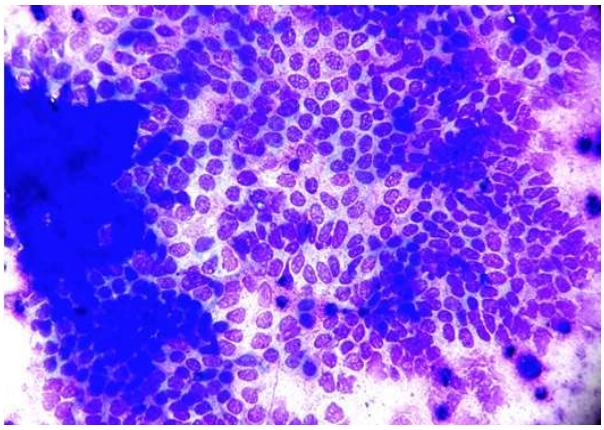

Fig. 4: (40x): Scar endometriosis: Smear with two population of cells, predominant glandular cells with anisonucleosis, hyperchromasia and small but conspcious nucleoli. (Giemsa, HP)

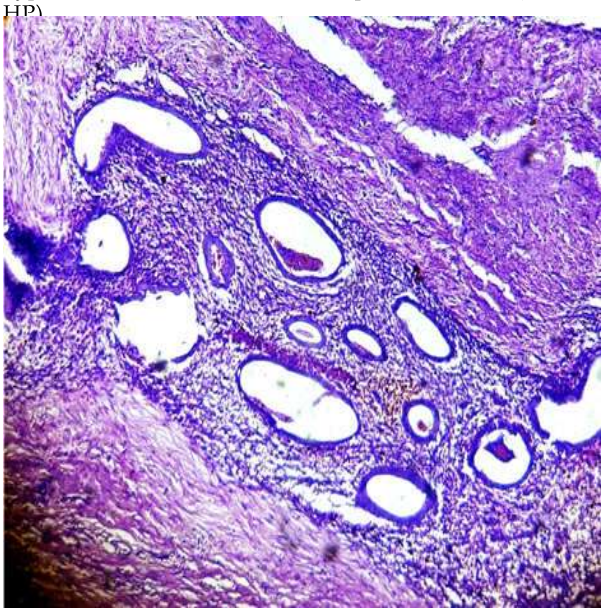

Fig. 5: (40x) Scar endometriosis: Fibrocollagenous tissue with varaible sized endometrial glands with luminal secretion surrounded by endometrial stroma and foci of haemorrhage. (H&E, HP)

A twenty eight year female patient presented with perianal swelling measuring 4x3cms (Figure 6), since 6 months, 2 inches away from episiotomy scar following vaginal delivery. She was treated with antibiotics which was misdiagnosed as abscess. Ultrasonography of perineum was suggestive of haemangioma in left pararectal space. FNAC diagnosis was benign epithelial tumor/skin adnexal tumor (Figure 7). The cut section of excised perianal nodule appeared grayish white mass with gray to brown colored small cyst, which histologically diagnosed as perianal endometriosis. (Figure 8).

Twenty-eight year female with chief complaints of pain abdomen, increased frequency of micturation, discoloration of urine and history of white discharge, underwent vaginoscopy, which revealed mass measuring 3x2cm and biopsy shows florid granulation tissue. Cystoscopy revealed sessile mass in urinary bladder and biopsy tissue shows endometrial tissue with intervening muscle fibers. (Figure 9)

A 25 year old lady, presented with swelling, dragging pain at genital region and brownish discharge. USG finding suggested vulval cyst measuring 3x2cms. FNAC diagnosed as endometriosis (Figure 10). Histology revealed fibrocollagenous tissue with endometrial glands surrounded by endometrial stroma. Few cystically dilated glands also seen with luminal secretion in them. Focal areas of haemorrhage confirmed as vulval endometriosis. (Figure 11).

Three cases of ovarian endometriosis were clinically suspected and laproscopically diagnosed as endometriosis in this study. FNAC was performed in only 4 cases out of 30 cases. In 3 cases, FNAC revealed cytological diagnosis of endometriosis. In one case FNAC diagnosis was skin adnexal tumor where as histologically it was diagnosed as perianal endometriosis [3].

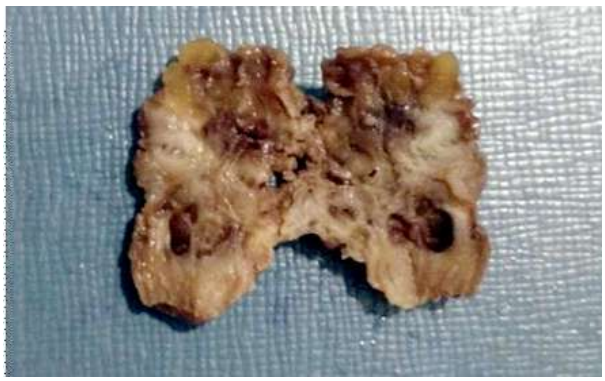

Fig. 6: Perianal endometriosis gross: Cut section of excised perianal nodule appeared grayish white mass with gray to brown colored small cyst measuring 4x3cms

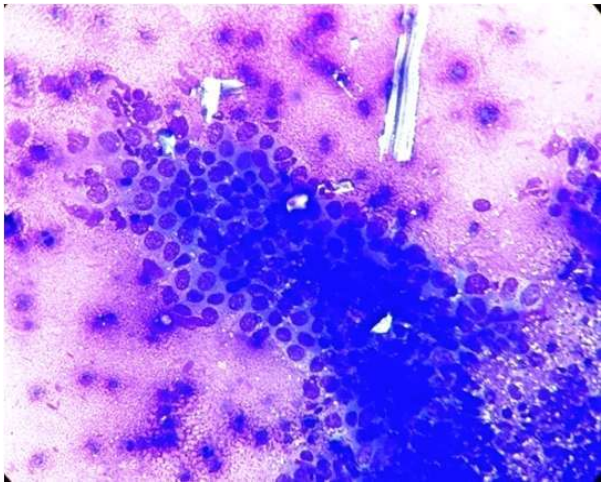

Fig. 7: (40x) Perianal endometriosis: Loose cluster and sheets of epithelial cells, anisonucleosis old haemorrhage in the background. (Giemsa, LP)

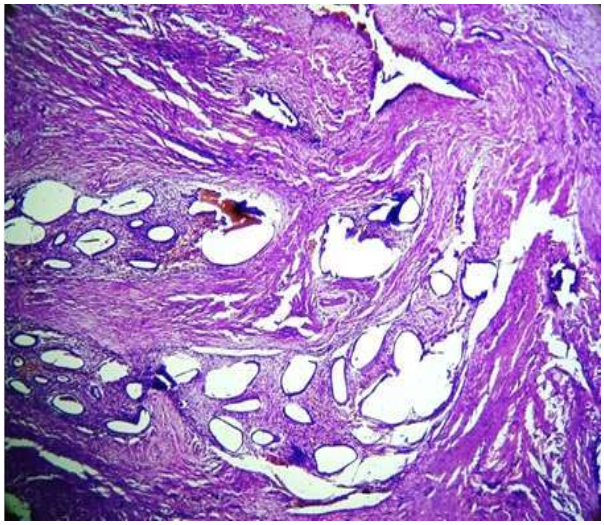

Fig. 8: (10x) Perianal endometriosis: Fibrocollageneous tissue and muscle tissue, showing variable cystically dilated endometrial glands & stroma. (H&E, LP)

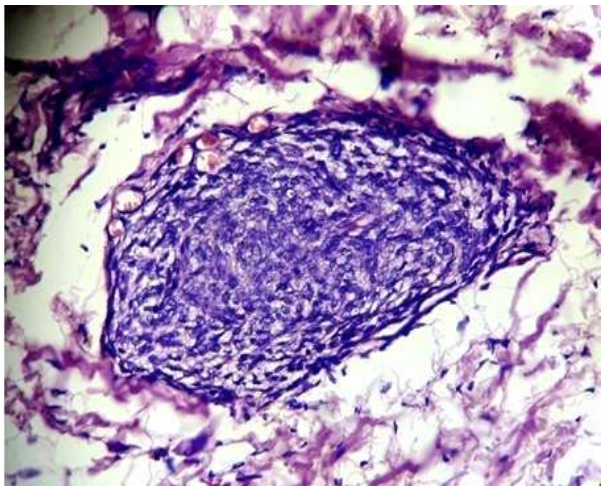

Fig. 9: (40x) Bladder endometriosis: Island of endometrial stroma in the lamina propria. (H&E, HP)

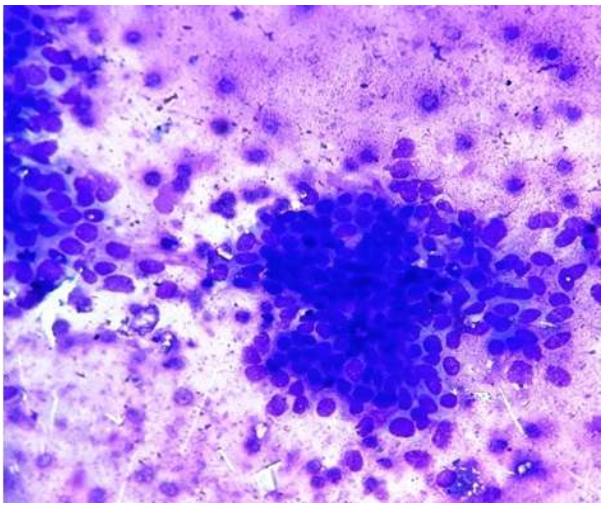

Fig. 10: (40x) Vulval endometriosis: Cluster of epithelial cells with, round to oval nuclei with scant cytoplasm in proteinaceous fluid background. (Giemsa, LP).

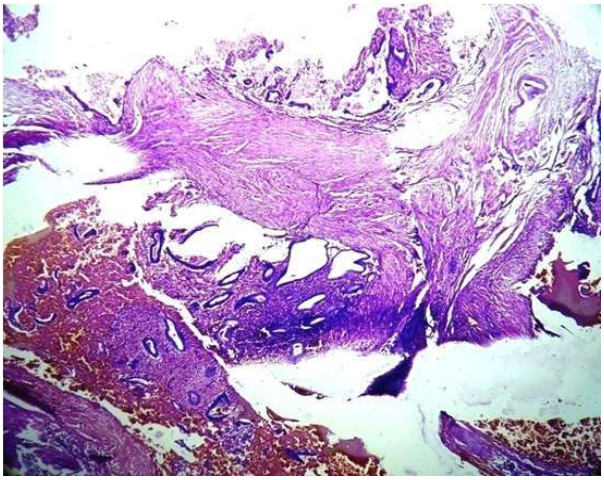

Fig. 11: (10x) Vulval endometriosis: Fibrocollageneous tissue, areas of fresh & old haemorrhage & many endometrial glands with stroma (H&E, LP)

Discussion

Endometriosis was first mentioned by Von Rokitansky in 1860, but the first detailed description was introduced by Sampson in 1921. Endometriosis is defined as the implantation of uterine mucosa at ectopic sites. It is one of the debilitating progressive diseases in women of reproductive age. Occurrence of endometriosis may involve any part of body [5].

In 1899, Russell was the first person to describe external endometriosis, skin and soft tissue endometriosis makes upto 3.5%, with most common cause following surgical operated scar. Scar endometriosis incidence is between 0.03% to 0.4% of women undergoing cesarean section [2,6]. Incidence of rare endometriosis site involving genitourinary tract is between 0.01 % and 1.2 % [5,6,7]. Rarely endometriosis is presented before menarche, and its frequency tends to decline after menopause.

Regarding pathogenesis, several theories are suggested viz. Transplantation theory (Halban's, Sampson's retrograde transport, and direct wound implantation), Metaplasia theory, Induction theory, Steroid hypothesis, Immunologic theory, and Genetic theory [5,8,9].

Obstruction of menstrual outflow (e.g, mullerian anomalies), exposure to diethylstilbestrol in utero, prolonged exposure to endogenous estrogen (e.g., because of early menarche, late menopause, or obesity), short menstrual cycles, low birth weight, and exposure to endocrine-disrupting chemicals are the various risk factors for endometriosis [10].

In the present study, ovarian endometriosis was most common (23 cases) and the second most common in the abdominal wall (4 cases) as reported by others [3]. In this study we found that abdominal wall endometriosis located at the site of previous surgical scar from cesarean section or hysterectomy was common among soft tissue endometriosis. Because of the increasing number of cesarean sections and laparoscopies being performed, there has been rise of scar endometriosis in the present era. In Rao, et al. study, all 4 cases were abdominal post-operative scar endometriosis and Santosh, et al study shows, 2 abdominal and 1 episiotomy scar endometriosis, as seen in present study following previous surgical scar [11,12].

Any female patient with symptoms and swelling in the peritoneal wall referred to surgeons, should be suspected to be endometriosis. Scar endometriosis should not be misinterpreted for recurrence of any other primary tumor. The common differential diagnosis are suture granuloma, infective granuloma, rectus hematoma, melanoma, anal canal cancer, desmoid tumor, and incisional hernia [7,12,14]. Treatment of choice for scar endometriosis and also for recurrent lesions is local wide excision, with at least 1 cm of margin [7,11].

Perianal endometriosis is commonly diagnosed as anal abscess, fistula, thrombosed haemorrhoid, dermoid cyst, sebaceous cyst or malignancy. Anal endosonography shows characteristic findings, which will rule out differentials and reach a correct diagnosis. Early diagnosis and treatment of perianal endometriosis is important, because progressive encroachment of anal sphincter may cause fecal incontinence, thus reduces adverse risk of post-operative outcome [13,14,15].

Care must be taken in the management of occult cancer in patients with extensive endometriosis of ovary. They have increased frequency of developing ovarian cancer. Endometriosis are at increased risk of developing autoimmune diseases, non-Hodgkin lymphoma, and melanoma. Endometriosis is considered as a precancerous lesion for ovarian cancer and is risk factor for the development of invasive epithelial ovarian cancer, such as low grade serous, endometrioid, and clear-cell ovarian cancer [10,16].

FNAC method is diagnostic in majority of cases, cost effective, easy, least time consuming procedure and in providing medical hormonal treatment to affected patient,thus sparing surgical treatment. In the present study, FNAC was done in only 4 cases, as 23 cases were ovarian endometriosis and only 7 cases were at other sites. Among the 4 cases, 2 were diagnosed by FNAC in the scar tissue, one in vulval. In a case with perianal nodule, FNAC was done, it could not be diagnosed by cytology.

Conclusion

The typical presentation and local findings allow us to make the correct diagnosis in clinically suspicious endometriosis. Proper caution must be taken during lower abdomen surgical procedures to avoid transplantation of endometrial tissue to the abdominal wall. Complication of endometriosis like bleeding from cyst, bowel obstruction in intestinal, reduced fertility etc, may be circumvented, if early diagnosis and treatment is instituted.

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