EXTRAPELVIC ENDOMETRIOSIS

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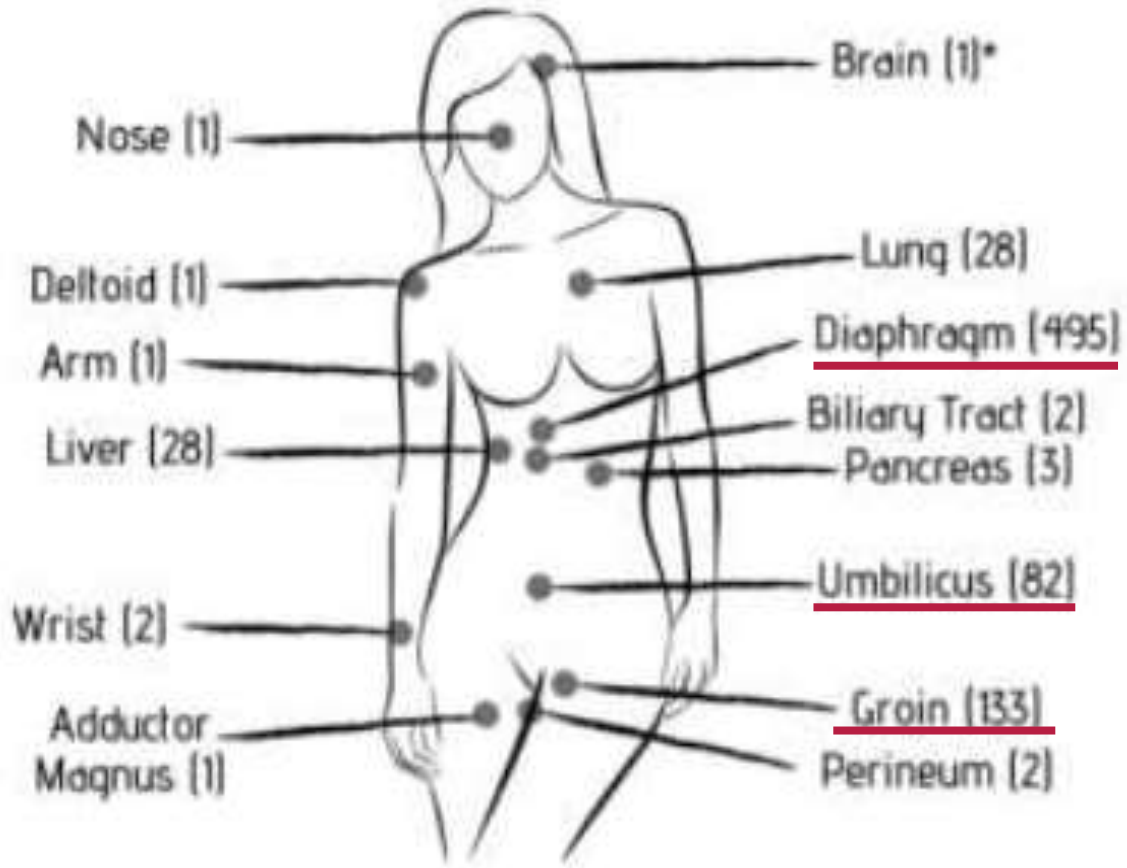
DEFINITION

Extra-pelvic endometriosis is a rare type of endometriosis, which occurs in a distant site from gynecological organs.

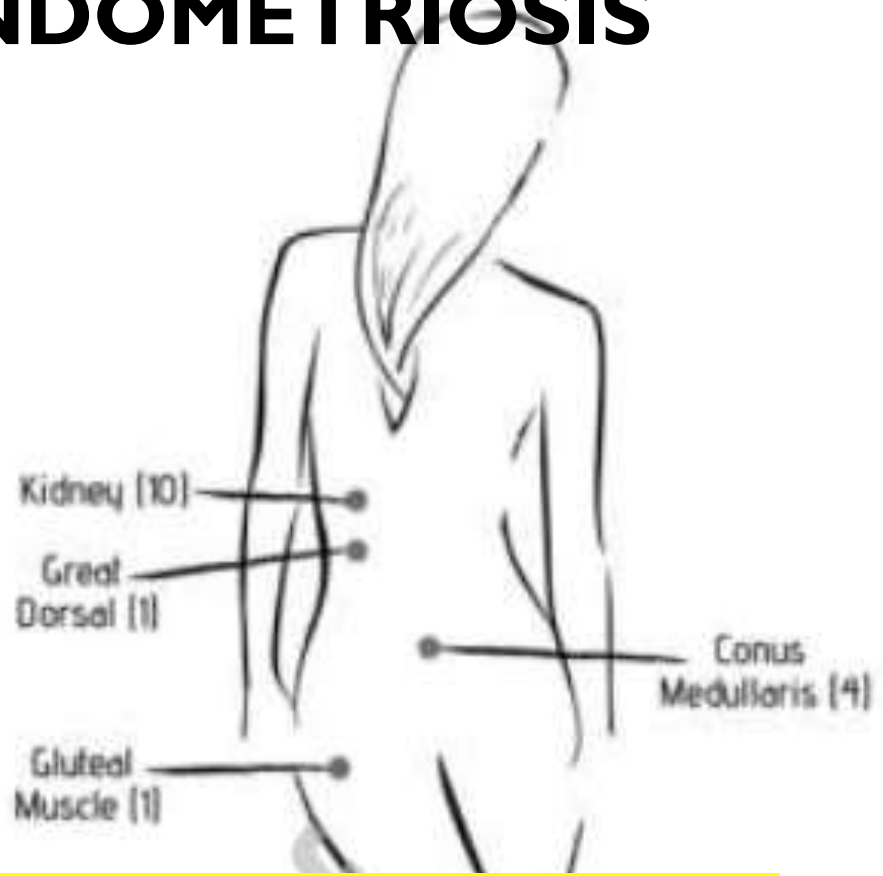
In a recent systematic review, most of the reported cases with extra-pelvic endometriosis (84%) were treated by non-gynecologic clinicians.

Andres MP, Arcoverde FVL, Souza CCC, Fernandes LFC, Abrao MS, Kho RM. Extrapelvic endometriosis: a systematic review. JMinim Invasive Gynecol. 2020;27(2):373-389

SITES OF EXTRAPELVIC ENDOMETRIOSIS



*In parentheses is the N of patients from each region studied



SYSTEMATIC REVIEW OVER 20 YEARS

Andres MP, Arcoverde FVL, Souza CCC, Fernandes LFC, Abrao MS, Kho RM. Extrapelvic endometriosis: a systematic review. JMinim Invasive Gynecol. 2020;27(2):373-389

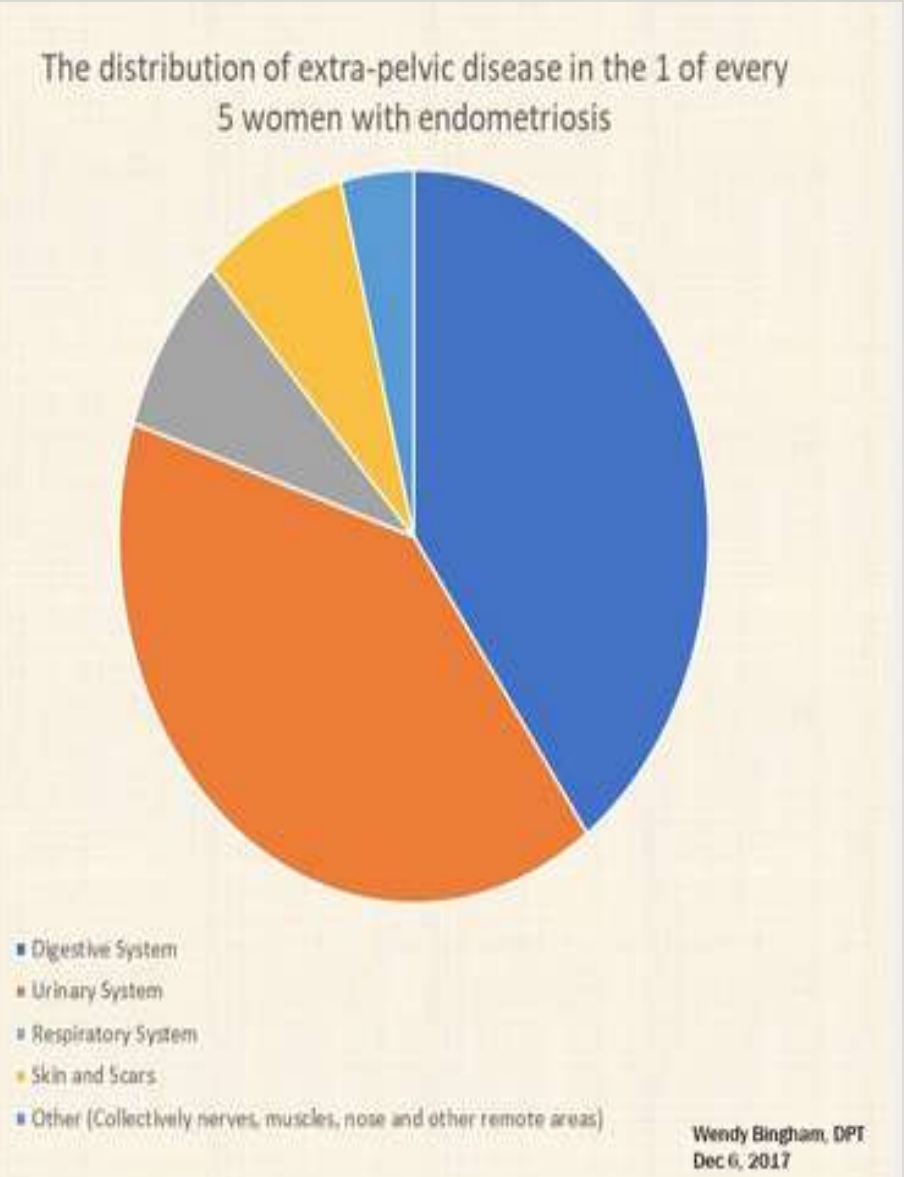
Endometriosis has been found in all 11 systems of the body. But most disease is found in these 4 systems:

Digestive
Urinary
Respiratory
Skin and Scar
Other*

Other* includes all 'rarer' locations of disease collectively. (ex: nerve, blood vessel, muscles, bones, nose, tear ducts...)

The pie chart shows the probabilities of a female-gendered person with extra-pelvic endometriosis – and likelihood of disease within each systems.

40% probability each of Digestive or Urinary
8% probability each of Respiratory or Skin/Scar
4% probability of 'Other'



NOT SO RARE!

People used to say that endometriosis had been found in every part of the body except the spleen. What was so different about the spleen that it was not affected by endometriosis?

It turns out that there is nothing different about the spleen! Endometriosis has been found on the spleen as well. The first case of endometriosis on the spleen was reported in 2022.

ABDOMINAL WALL ENDOMETRIOSIS (AWE)

The presence of ectopic endometrial tissues superficial to the peritoneum of the abdominal wall (0.04 – 5.5%).

A review describing 445 cases of AWE revealed that 57%, 11%, and 13% of cases were associated with a prior cesarean section, hysterectomy, and other surgery, respectively.

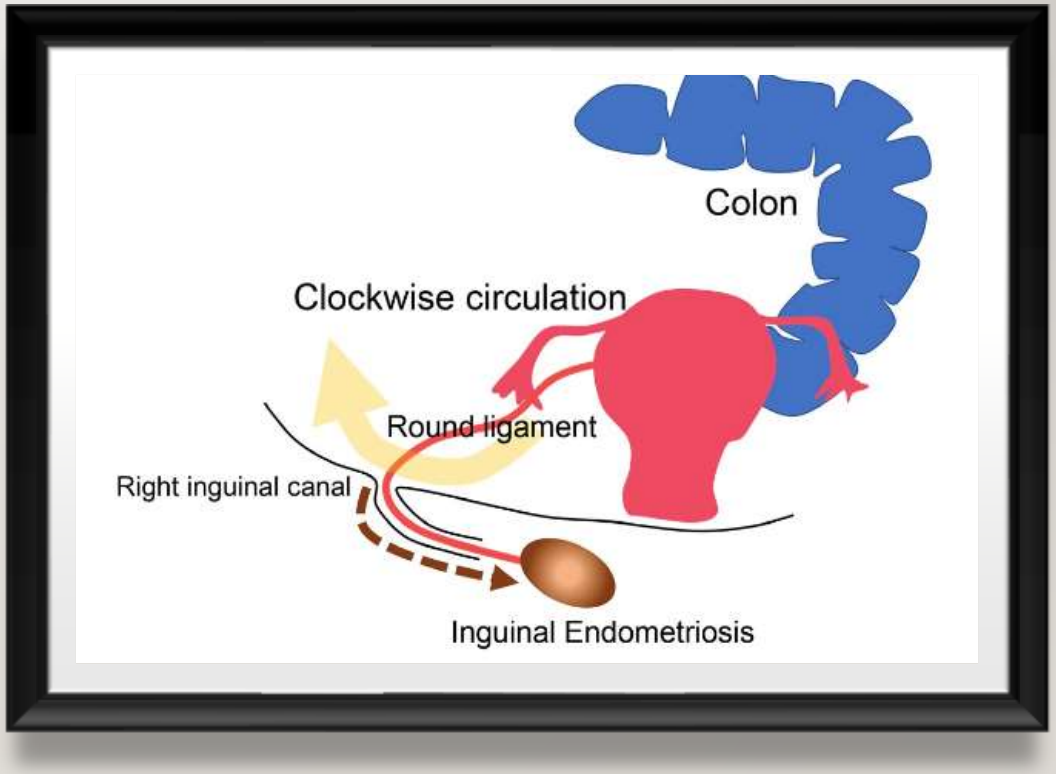
Twenty percent of cases were spontaneous with no surgical history and were preferentially found at the umbilicus or in the groin.

AWE is defined to include scar endometriosis, umbilical endometriosis, and inguinal endometriosis.

Horton JD, DeZee KJ, Ahnfeldt EP, Wagner M. Abdominal wall endometriosis: a surgeon's perspective and review of 445 cases. Am J Surg. 2008;196(2):207-212

INGUINAL ENDOMETRIOSIS

- 0.3 to 0.6 % incidence
- First case reported in 1896
- Pain and groin lump – mimics inguinal hernia or hydrocele of Canal of Nuck
- 94% right sided



CASE 1 – ISOLATED INGUINAL ENDOMETRIOSIS

- 30 year old female
- G1P1 (LSCS)
- c/o pain in lower abdomen, PMS and delayed period
- Clinical examination - NAD
- UPT – negative
- USG pelvis – 3 cm clear left ovarian cyst
- Put on OCP – improved.

5 MONTHS LATER

- **Presented with severe dysmenorrhea**
- **Continuing PMS**
- **LMP -18/05/2011**
- **USG pelvis – 4.5 x 3.9 cm cyst in left adnexa with 2.9 cm solid irregular marginated lesion, left ovary not separately visualised. Right ovary normal.**
- **CA 125 = 35.6**

MRI


GE MEDICAL SYSTEMS
SIGNA EXCITE EXCITEHR
ARS
SARAL DIAGNOSTICS
NITIKA DUGGAL
F31Y
07/1364
May 26 2011
11:14:57 AM
Mag = 1.50
FL:
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WW: 2470 WL: 1077


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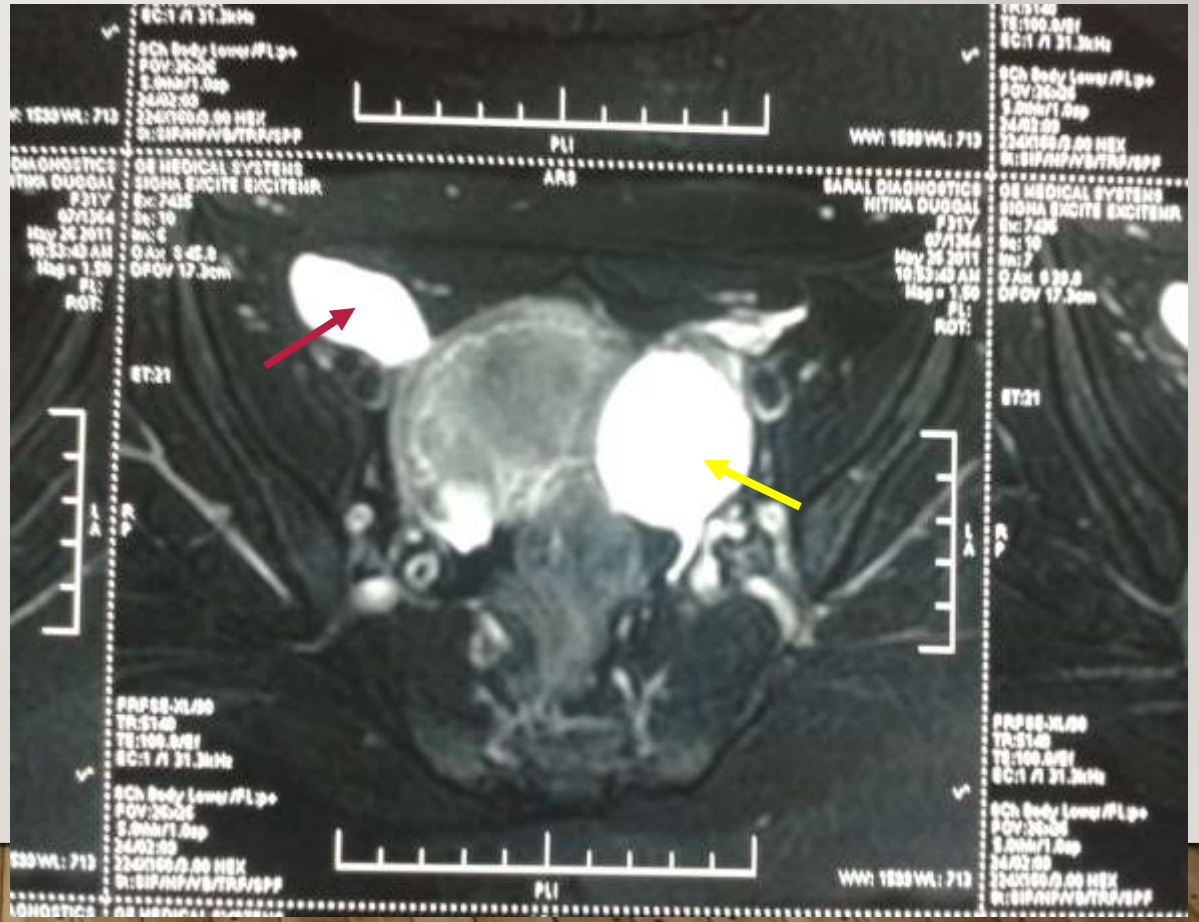
MRI



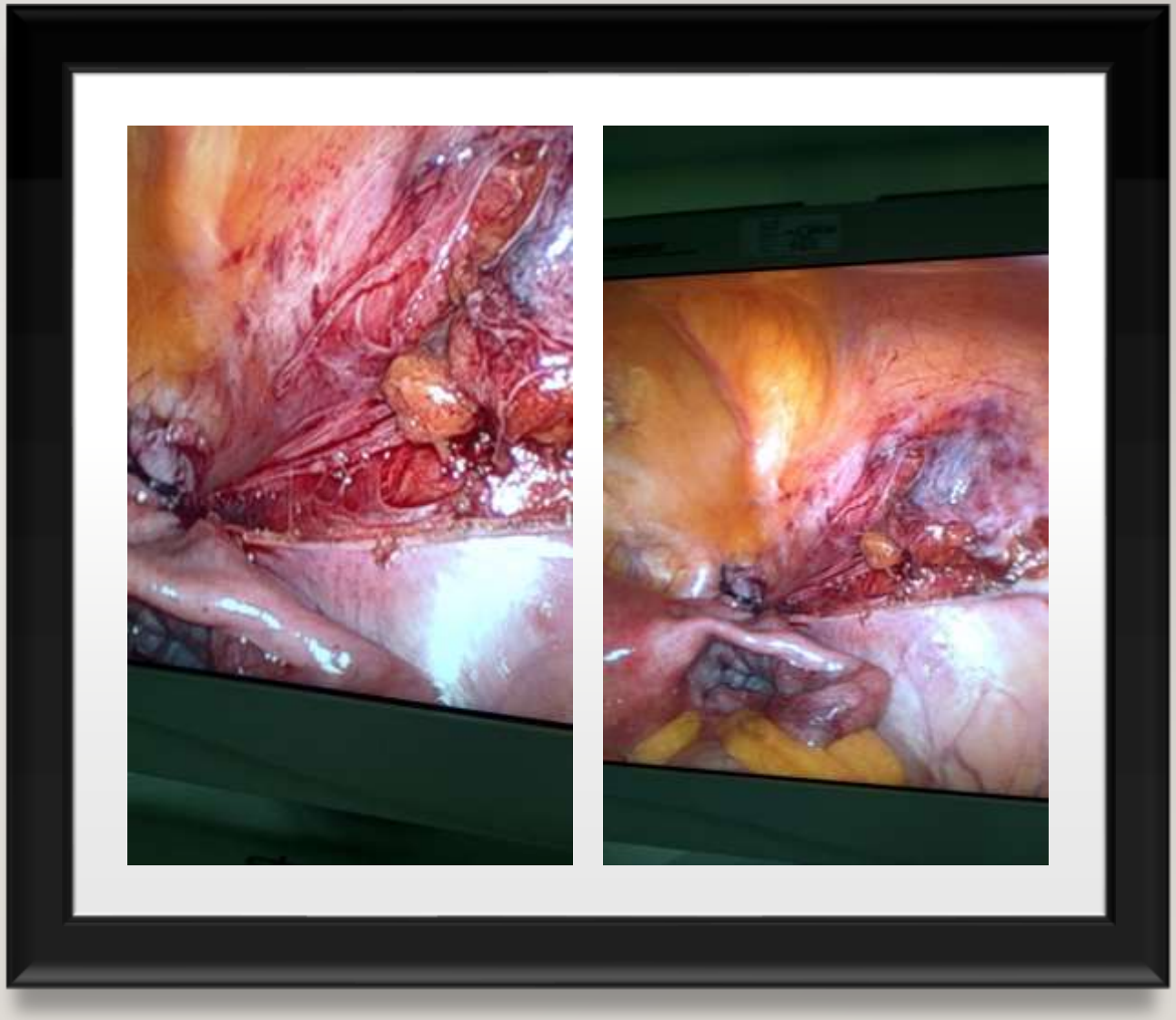

Normal pelvis and adnexae

INGUINAL CANAL EXPLORATION

INGUINAL CANAL EXPLORED WITH RESECTION OF ROUND LIGAMENT AND DISSECTION OF HERNIAL SAC.

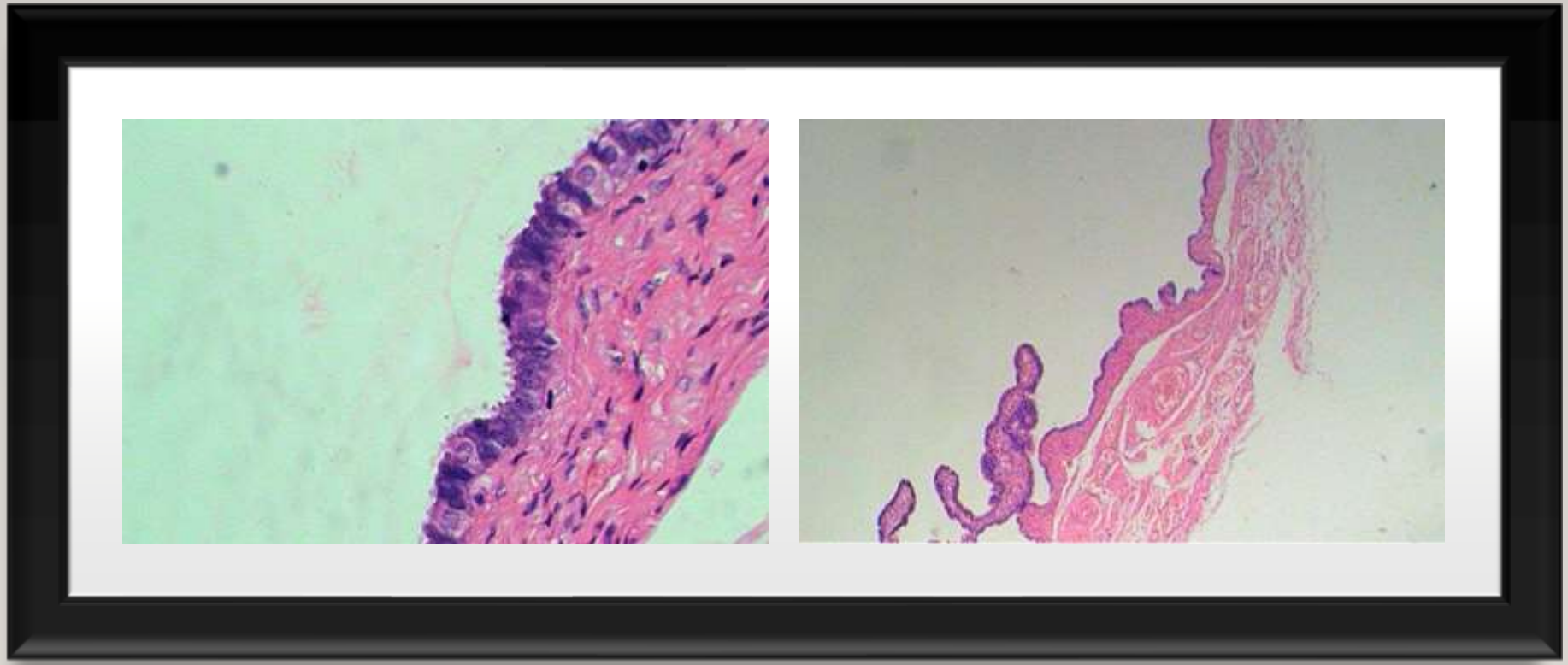
LAPAROSCOPY AFTER HERNIA REPAIR

- As a radical surgical treatment, an en bloc resection of tumor and the round ligament has been reported.
- The theory is to suppress recurrence by removing the round ligament, which is considered to be the transmission route of endometriosis.



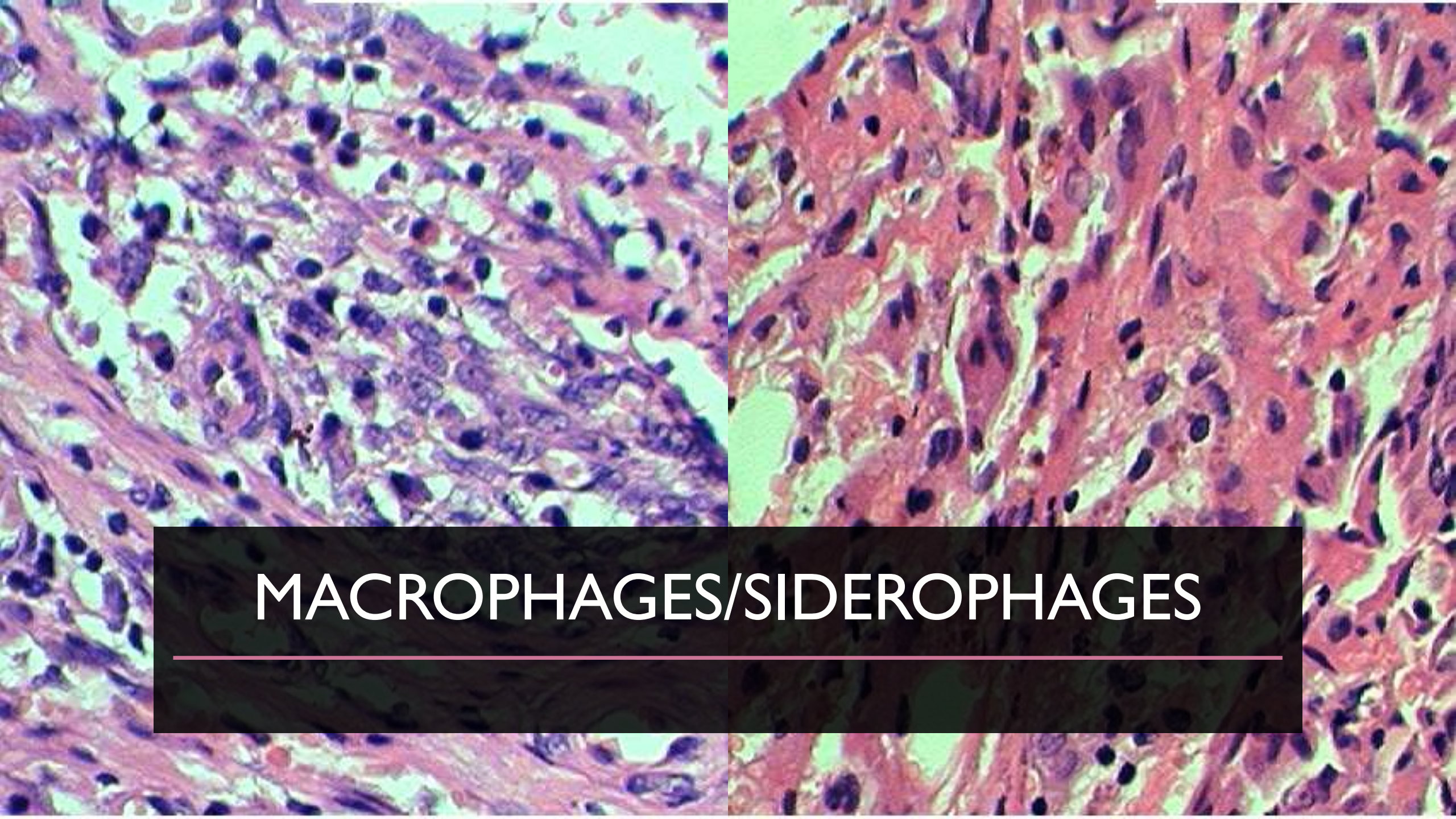
FINAL WOUNDS

- Many patients that have been surgically treated by general surgeons for an inguinal hernia or Nuck's canal hydrocele are pathologically diagnosed with inguinal endometriosis after surgery.

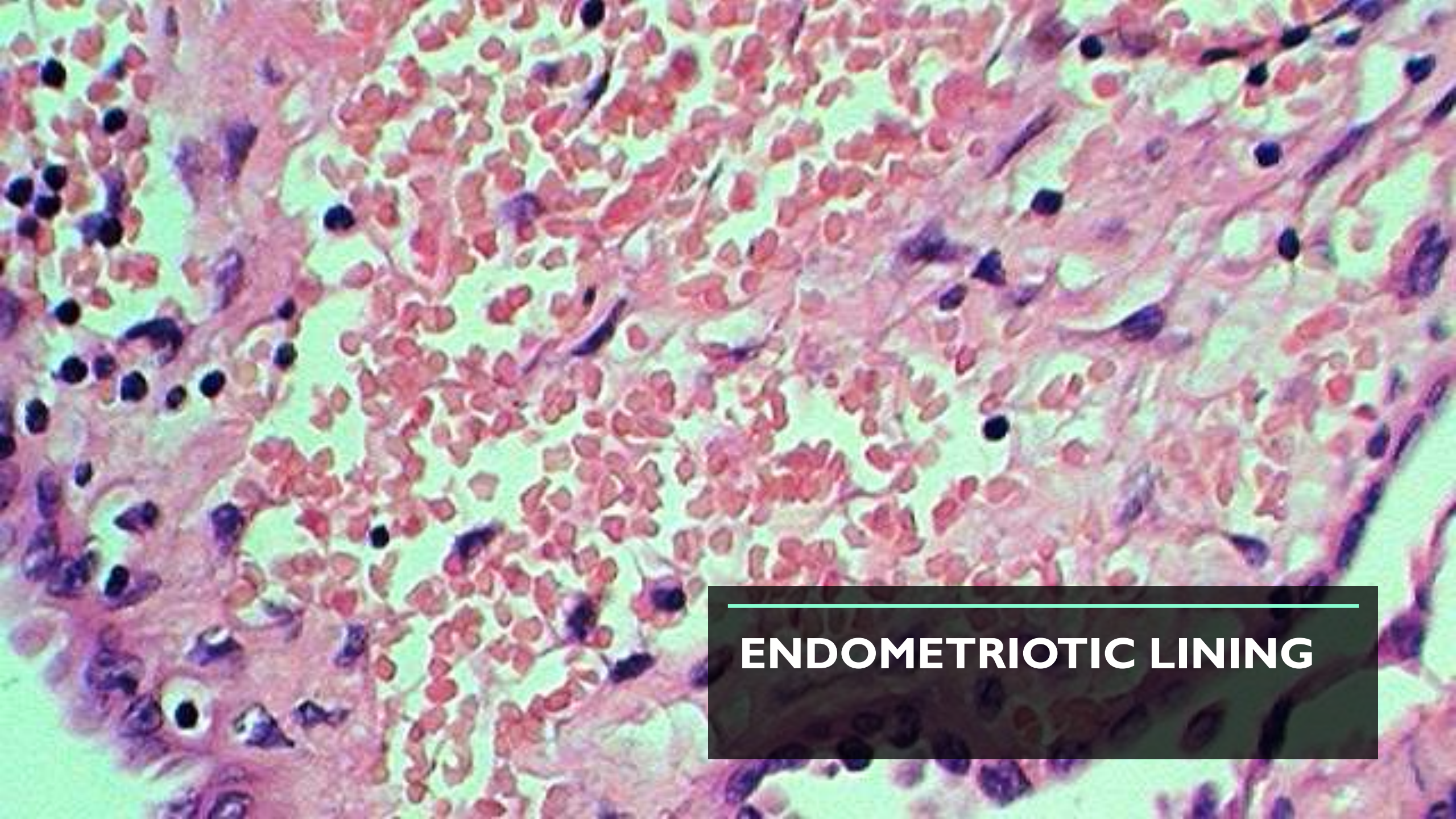


PAROVARIAN CYST – FIBROMUSCULAR WALL WITH BLAND CUBOIDAL LINING EPITHELIUM AND PORTION OF FOLIAR PAPILLAE – FIMBRIAL CYST.

HISTOPATHOLOGY – CONGESTED CONNECTIVE TISSUE (HERNIAL SAC CONTENTS)



MACROPHAGES/SIDEROPHAGES


ENDOMETRIOTIC LINING

POSTOPERATIVE RECURRENCE

There have been a few reports describing postoperative recurrence rates ranging from 0%to 16.6%.

A subcutaneous tumor was formed at the site distal to the resection site from the inguinal ring, which suggests that a part of the lesion was newly transplanted during the initial surgery or that a small lesion remained at the distal site of the excised lesion and the round ligament.

Arakawa T, Hirata T, Koga K, et al. Clinical aspects and management of inguinal endometriosis: a case series of 20 patients. J Obstet Gynaecol Res. 2019;45(10):2029-2036.

HORMONAL THERAPY

Although there have been few reports on the effects of hormone therapy, the administration of dienogest (2 mg/d) was associated with improved groin pain in 6 of 7 patients.

Dienogest can be an option for patients who do not want primary surgery or reoperation after recurrence.

Further study is required, because there are limited data on the effects of hormonal therapy on inguinal endometriosis.

Accordingly, surgical treatment is recommended as the first choice, because the postoperative recurrence rate is not high and symptoms improve after surgery.

SCAR ENDOMETRIOSIS

Most common of AWE – iatrogenic form of endometriosis predominantly occurs at the cesarean scar, followed by uterine surgery scar, and at the laparoscopic port site.

Dissemination of endometrial tissue during a cesarean section or uterine surgery is biologically plausible because of the opportunity to inoculate the abdominal wall with endometrial cells from a hysterotomy.

Abdominal mass (96%) and pain during menstruation (87%) are the most common presenting symptoms.

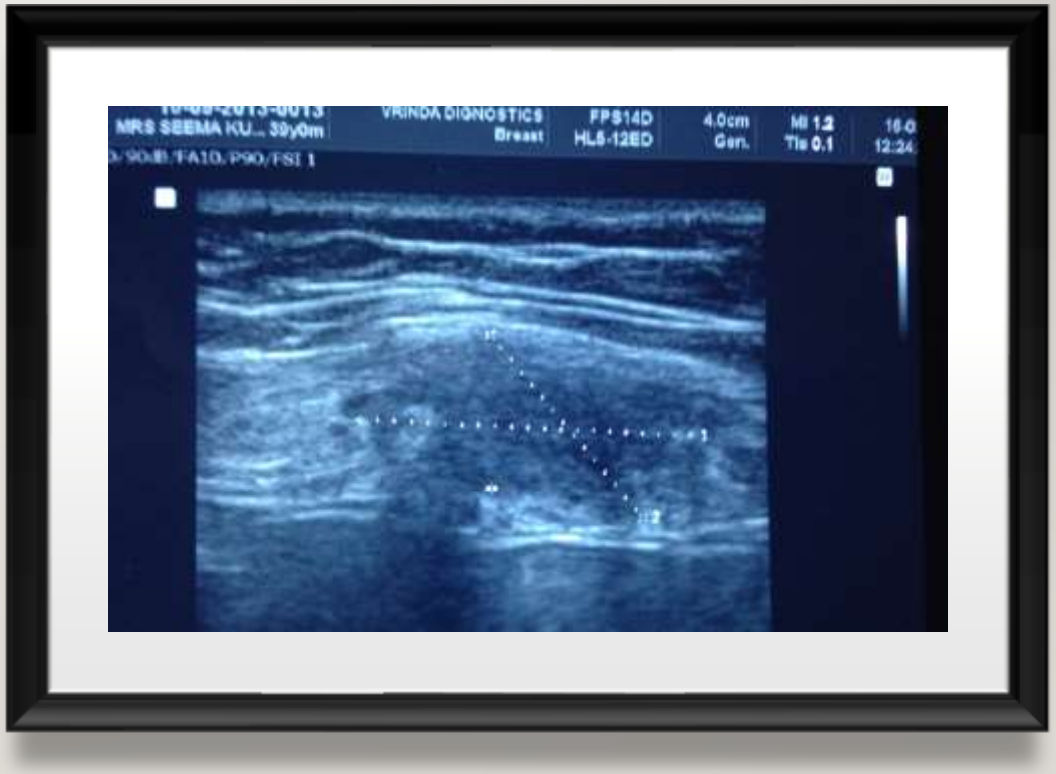
CASE 2 - HISTORY

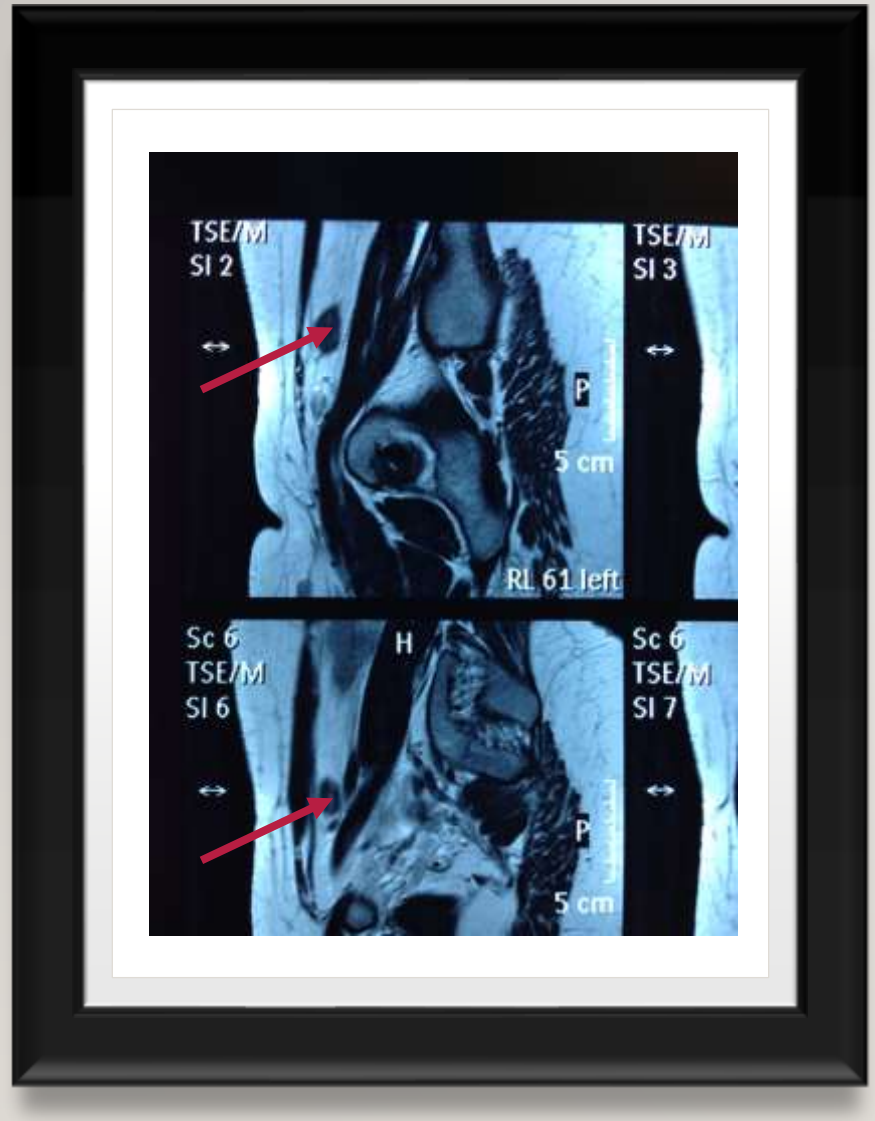
- **39 year old female**
- **G1P1 (LSCS 3 years ago)**
- **c/o cyclical pain just below umbilicus**
- **Clinical examination – 3 x 2 cm firm lump in parietal wall just below umbilicus.**

DIAGNOSIS - ULTRASOUND

Small irregular hypoechoic solid nodule in rectus abdominis muscle in anterior abdominal wall in infra-umbilical region + multiple small uterine fibroids

FNAC can be done if malignant transformation suspected.





MRI

Small nodule in rectus abdominis muscle in infraumbilical region + small intramural fibroids

Helps clarify depth of lesion along with muscle and fascial involvement.

Presence of blood components within lesion.

TREATMENT

To date, there has been no report demonstrating that hormonal therapy is effective for AWE.

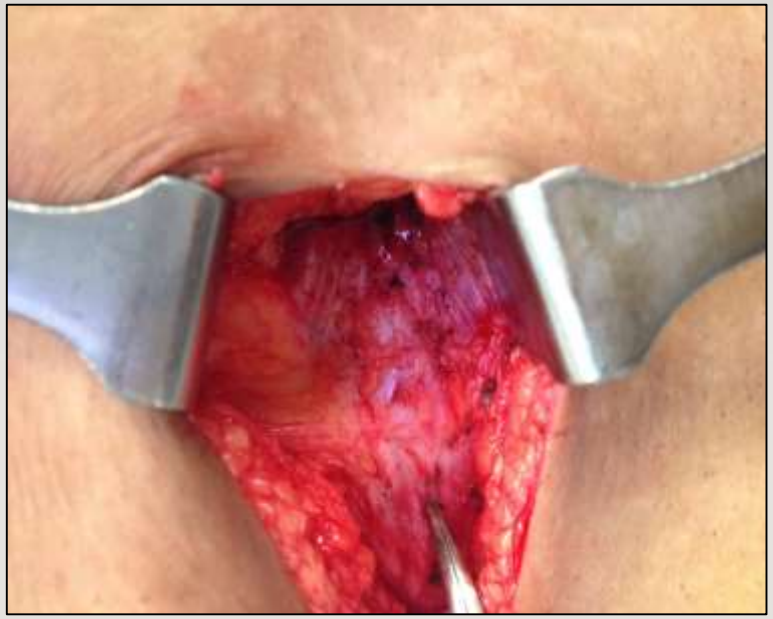
Oral contraceptives (OC), progestin, or gonadotropin-releasing hormone (GnRH) agonists may be effective in improving symptoms and can be an option for patients who do not want surgery.

However, the symptoms are likely to recur after hormonal therapy is discontinued.

Given the low postoperative recurrence rate and the low invasiveness of surgery, surgical treatment can be recommended as the first choice.

Rindos NB, Mansuria S. Diagnosis and management of abdominal wall endometriosis: a systematic review and clinical recommenda- tions. Obstet Gynecol Surv. 2017;72(2):116-122.

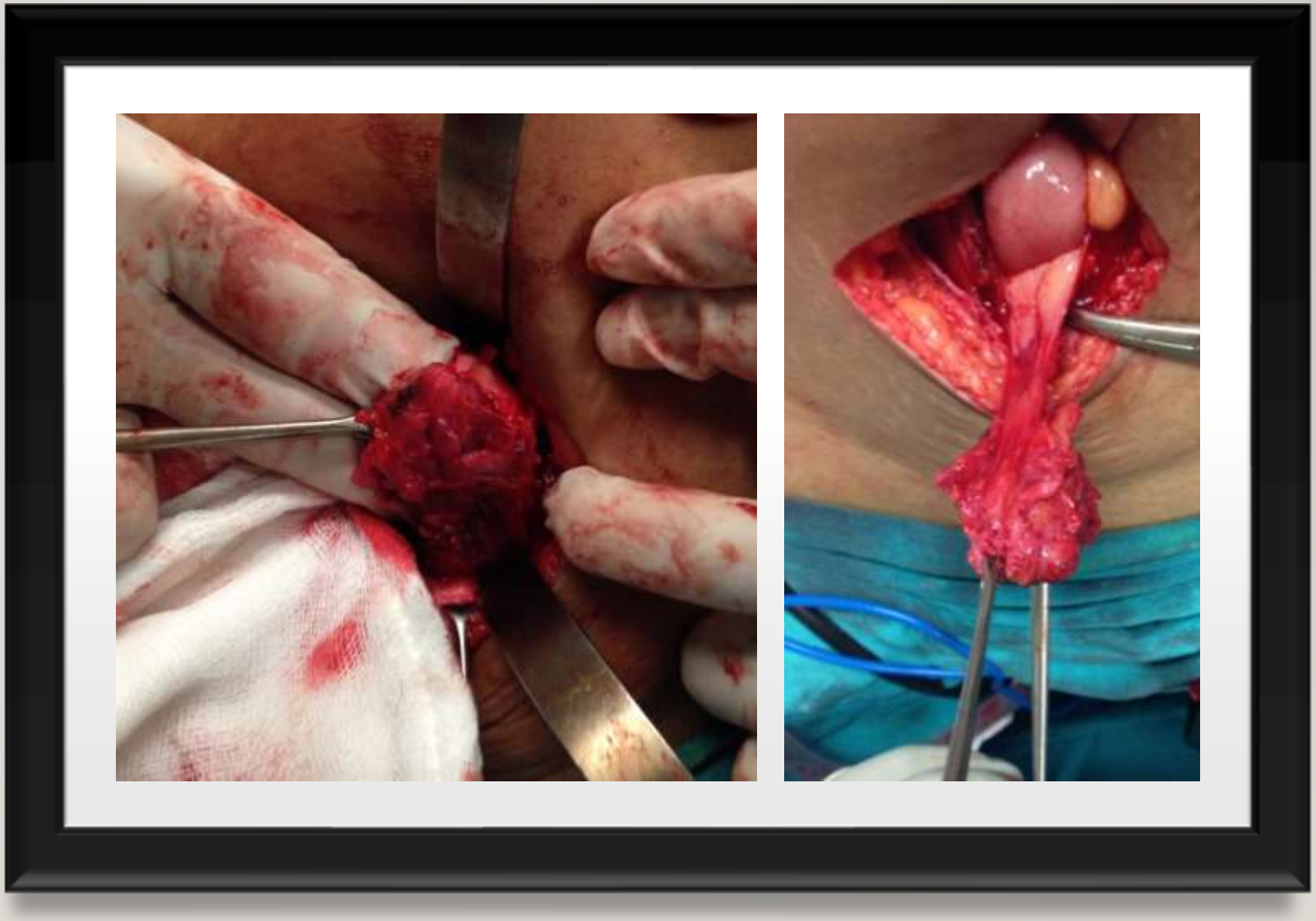
WIDE LOCAL EXCISION OF LESION



- **Surgical excision with 1-cm margins on all sides of endometriotic lesion should be optimal.**
- **Mesh repair of fascia may be required if fascial defect is significant.**

Bektas H, Bilsel Y, Sari YS, et al. Abdominal wall endometrioma; a 10-year experience and brief review of thel iterature. J Surg Res. 2010;164(1):e77-e81.

WIDE LOCAL EXCISION



PEARLS OF WISDOM

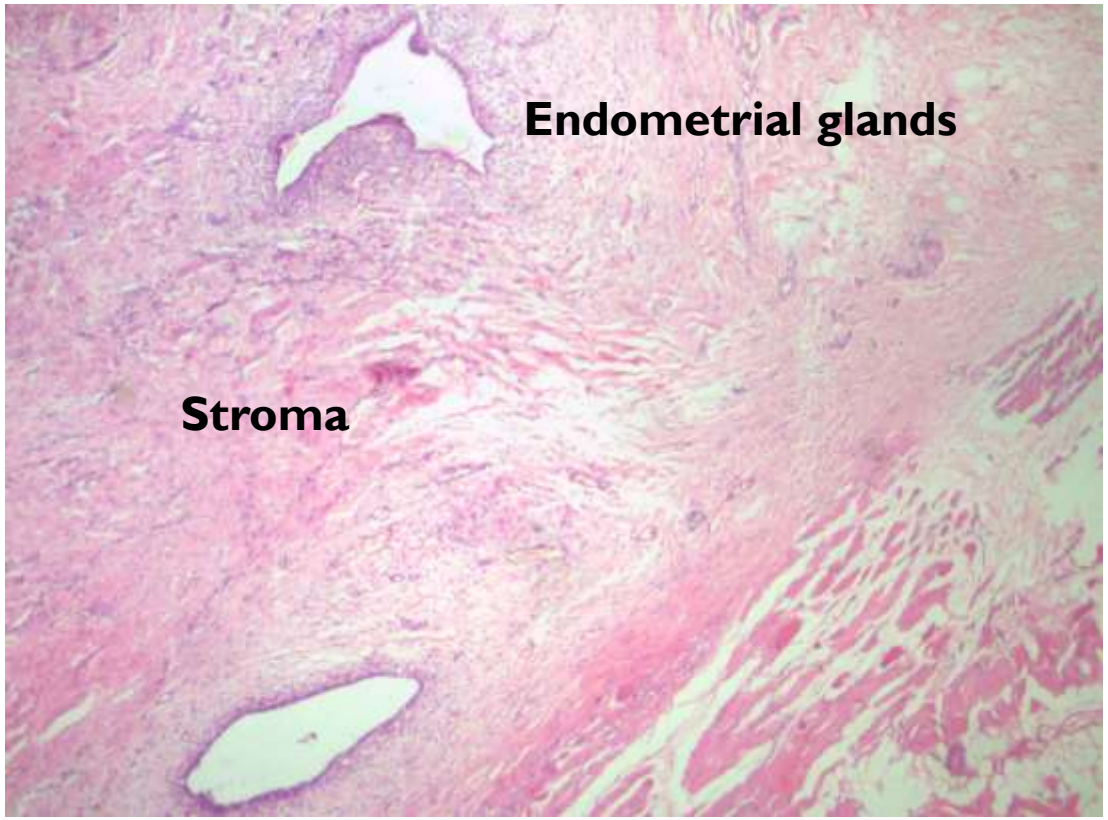
The postoperative recurrence rate of AWE is 4.5%-11.2%, lower than that of ovarian endometriosis.

Malignant transformation of scar endometriosis is reported to have a very poor prognosis.

The average time between initial surgery and diagnosis of endometriotic malignant transformation was 19.3 years (n=48), which suggested a slow evolution of the disease.

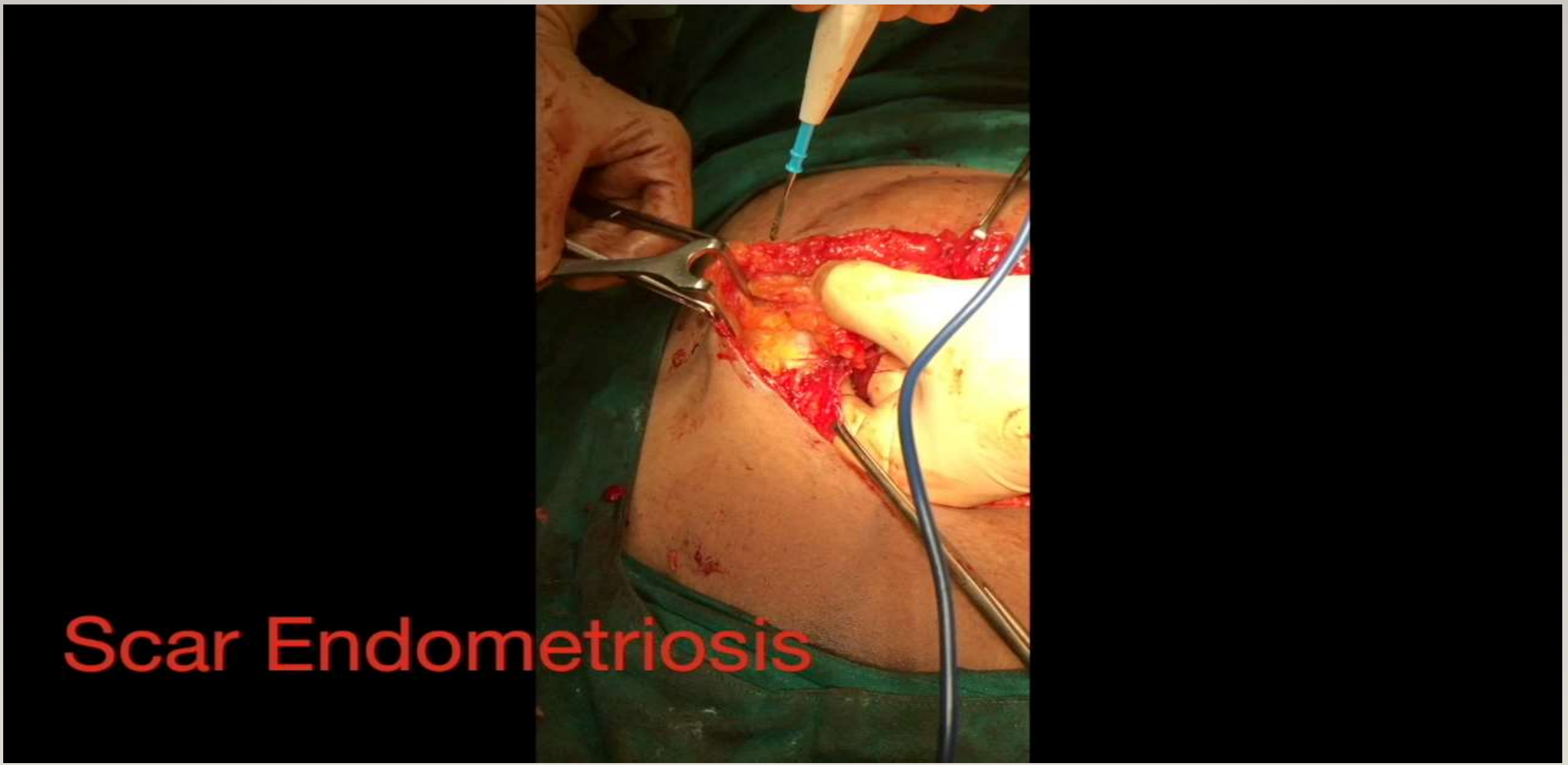
Mihailovici A, Rottenstreich M, Kovel S, Wassermann I, Smorgick N, Vaknin Z. Endometriosis-associated malignant transformation in abdominal surgical scar. Medicine. 2017;96(49):1-5.

HISTOPATHOLOGY



CASE 3 – SCAR ENDOMETRIOSIS IN CS SCAR

- 30 year old lady with a scar endometrioma on the prev CS scar.
- Before excision could be planned, she conceived again – scar endometriosis excision was done during her repeat Caeserean.
- The nodule, which was no longer palpable, was marked pre – operatively using ultrasound.


Scar Endometriosis

UMBILICAL ENDOMETRIOSIS

Umbilical endometriosis includes secondary umbilical endometriosis, which is thought to develop iatrogenically at the port site after laparoscopic surgery, and primary umbilical endometriosis, which has no operation history and occurs spontaneously.

Unlike scar endometriosis, approximately 70% of patients with umbilical endometriosis did not have a history of prior surgery, suggesting a mechanism other than direct contact implantation.

The most likely cause of primary umbilical endometriosis is hypothesized to occur by migrating through blood or lymphatic vessels.

Hirata T, Koga K, Kitade M, et al. A national survey of umbilical endometriosis in Japan. J Minim Invasive Gynecol. 2020;27(1):80-87.

CASE 4 - HISTORY

- 40 year old female
- Para 2 (NVD, LCB 11 years back)
- c/o bluish discoloration of umbilicus
- Cyclical bleeding with pain in the nodule
- MH – Regular cycles
- H/o Diagnostic laparoscopy for infertility in 1997.
- H/o Appendectomy in 2003

CASE 4 – CLINICAL EXAMINATION

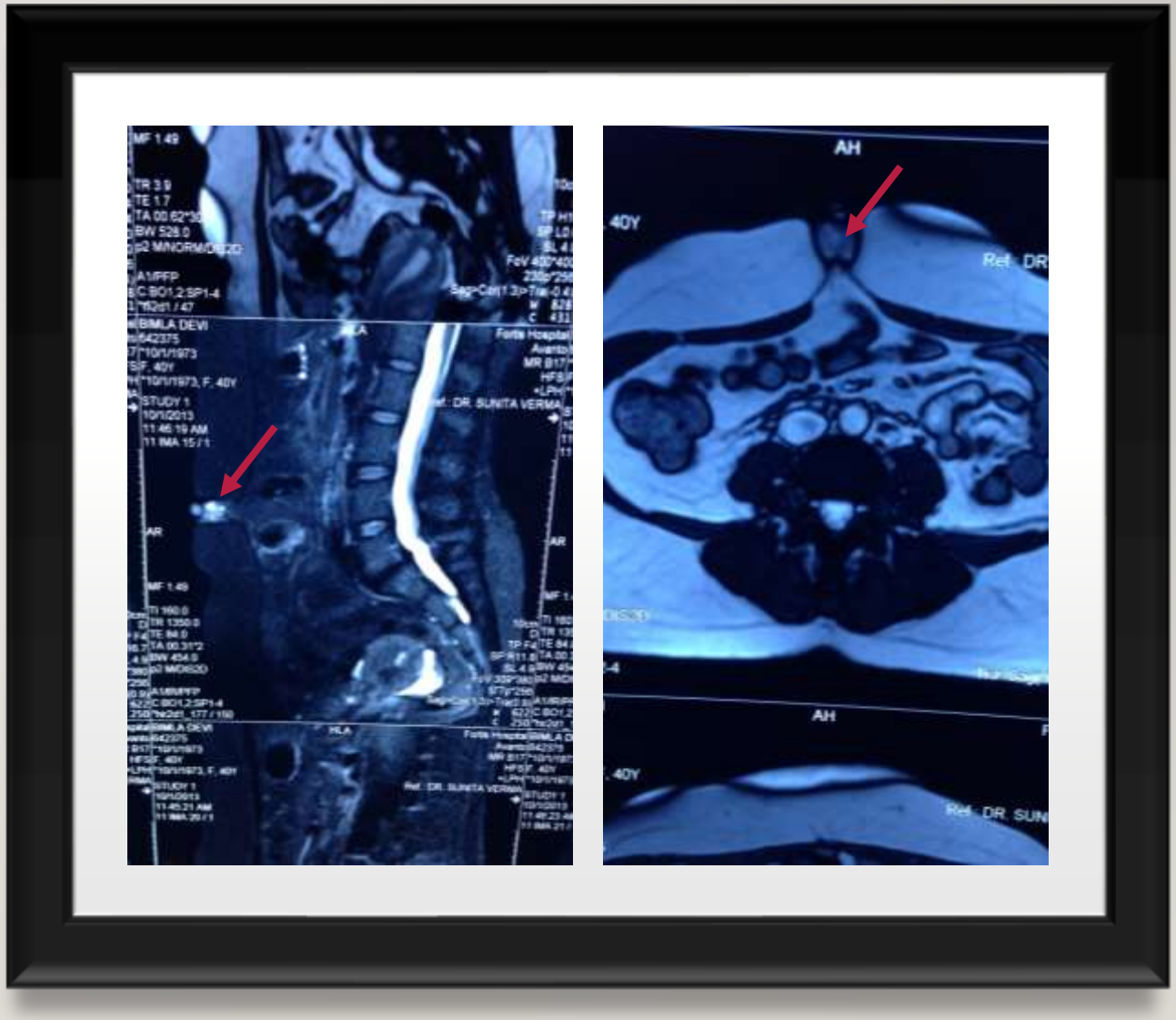
Bluish mass in umbilicus – 2X2 cm

SPOT DIAGNOSIS

MRI

USG – Septated cyst in right ovary, 44 x 23 mm, rest NAD

MRI – Small endometriotic cyst in right ovary with endometriotic deposit in umbilicus



LAPAROSCOPY WITH WIDE LOCAL EXCISION

Entry through Palmer's point

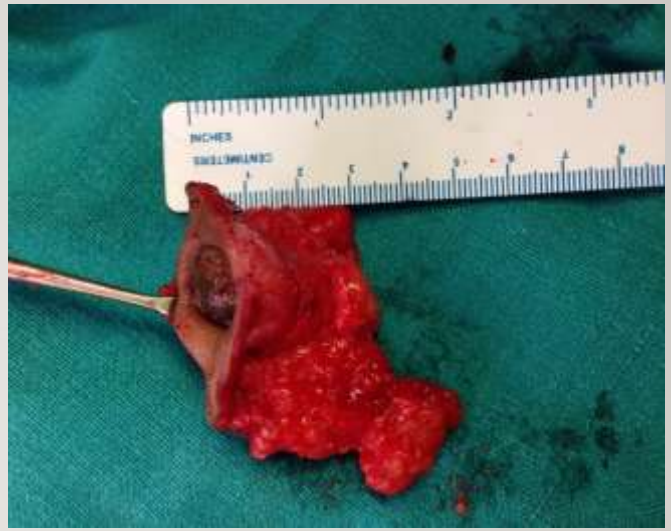

Entry through Palmar's point

WIDE LOCAL EXCISION

Surgical treatment should involve a wide local resection to avoid recurrence and malignant transformation.

The recurrence rate after local resection of umbilical endometriosis is approximately 10%, which is considerably lower than the recurrence rate of ovarian endometriosis.

In particular, no postoperative recurrence was reported in cases of radical surgery resected to the peritoneum.

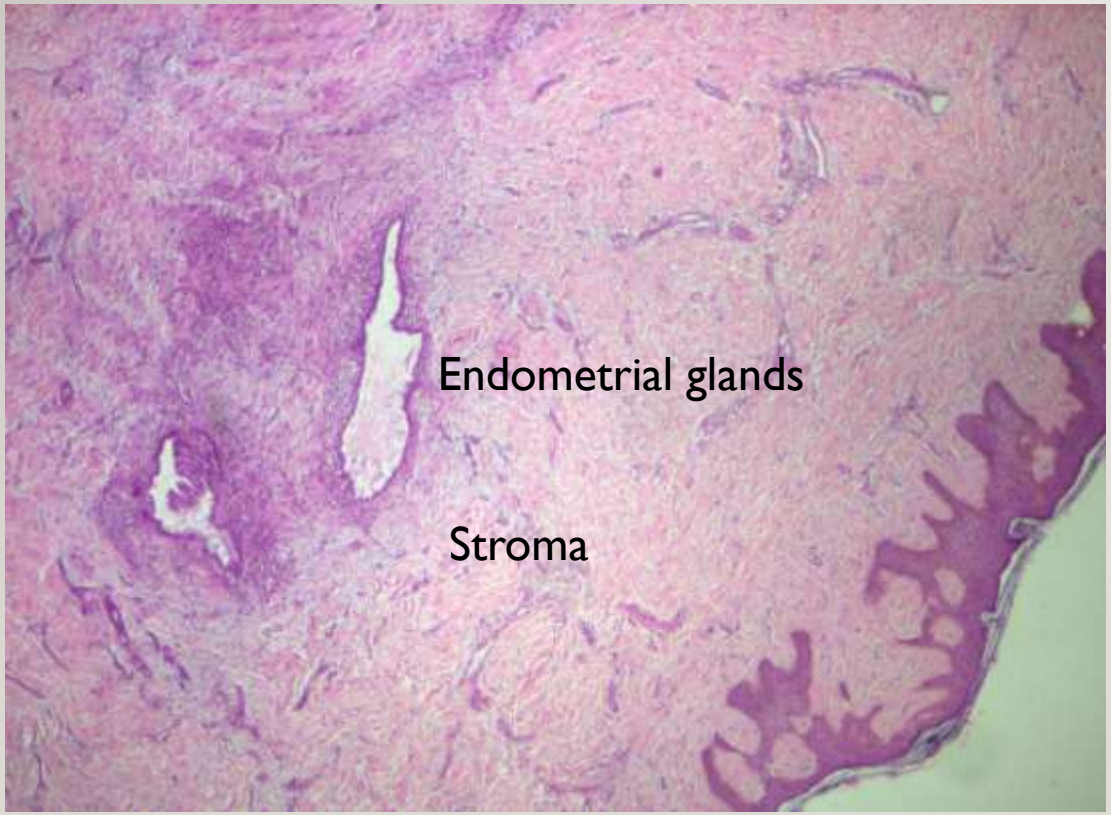


UMBILICAL RECONSTRUCTION

There have been very few reports on hormone therapy for umbilical endometriosis.

Several papers have reported that hormonal treatment is inadequate for treating umbilical endometriosis, while other authors have reported that OC, dienogest and GnRH agonist were effective in improving symptoms.

HISTOPATHOLOGY



To date, four cases of malignant transformation have been reported.

Obata K, Ikoma N, Oomura G, Inoue Y. Clear cell adenocarcinoma arising from umbilical endometriosis. J Obstet Gynaecol Res.2013;39(1):455-461.
Koguchi-Yoshioka H, Oshima H, Manago E, et al. Sister MaryJoseph's nodule: malignant transformation of umbilical endometriosis. J Dermatol. 2016;43(12):1449-1450.

TAKE HOME MESSAGE

For abdominal extrapelvic endometriosis, surgical removal is the preferred treatment when possible, to relieve symptoms. Hormone treatment may also be an option when surgery is not possible or acceptable.	⊕◯◯◯

BLADDER ENDOMETRIOSIS

Bladder endometriosis is defined by the presence of endometriosis in the detrusor muscle, including partial or full thickness infiltration, and/or the bladder epithelium.

Urinary tract endometriosis is present in approximately 0.3–12% of individuals with endometriosis.

BE is the most common manifestation, accounting for 70–85% of urinary tract endometriosis classified as primary, where it occurs spontaneously, or secondary, following iatrogenic lesion deposition, or bladder injury in pelvic surgery such as caesarean section or hysterectomy

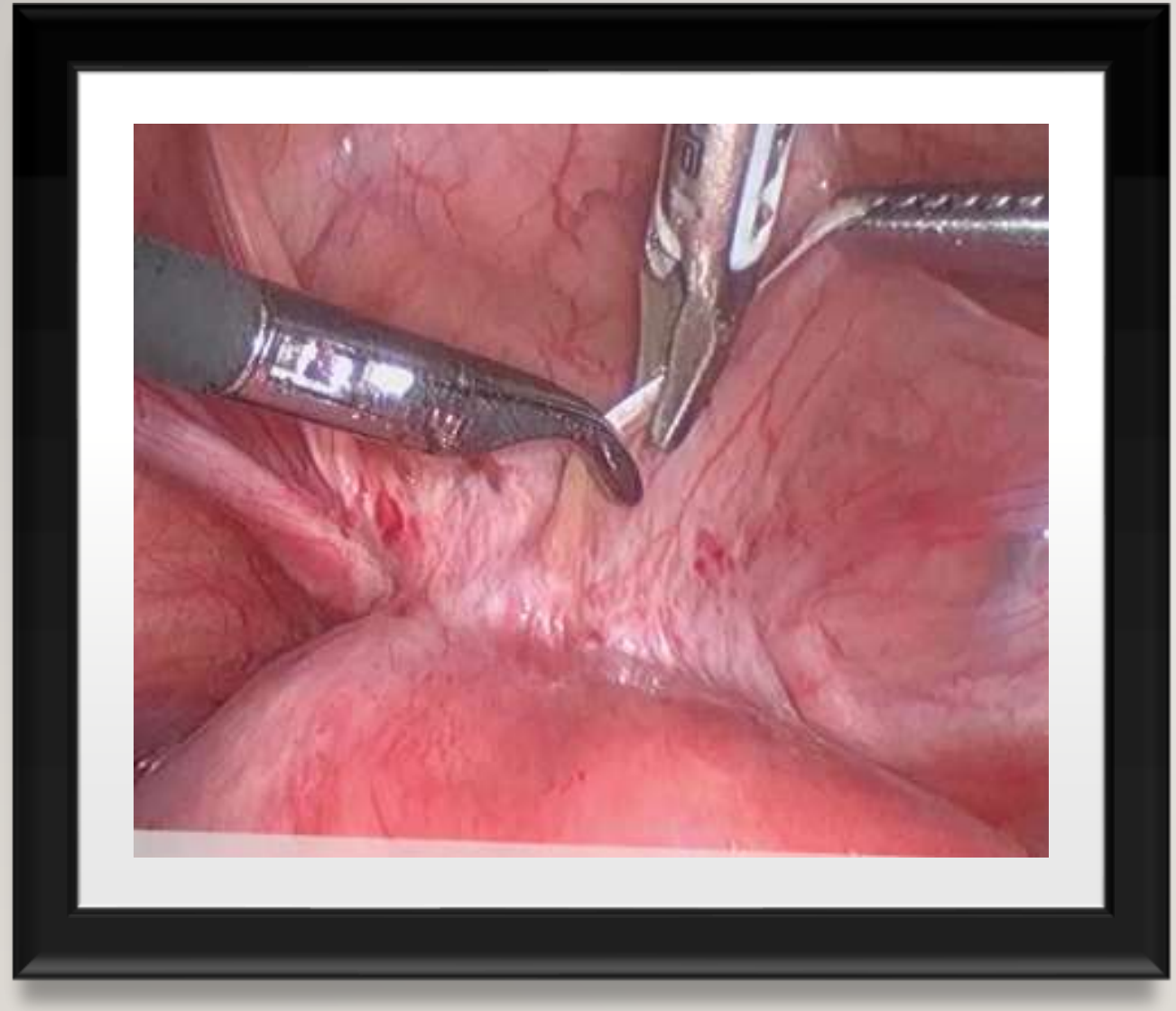
CASE 5 - BLADDER ENDOMETRIOSIS

- 35 year old patient, Para 2 (both LSCS).
- c/o severe dysmenorrhea, LUTS (more during menses).
- O/E – Uterus bulky, mobility restricted.
- P/R – NAD
- USG – small mass in urinary bladder, adenomyotic changes in uterus
- CA 125 - WNL

MRI

Heterogeneous mass in the posterior bladder wall (arrow) protruding into the bladder lumen and containing hyperintense spots.

A thin hyperintense fat plan (white open arrow) separates the mass from the uterus.



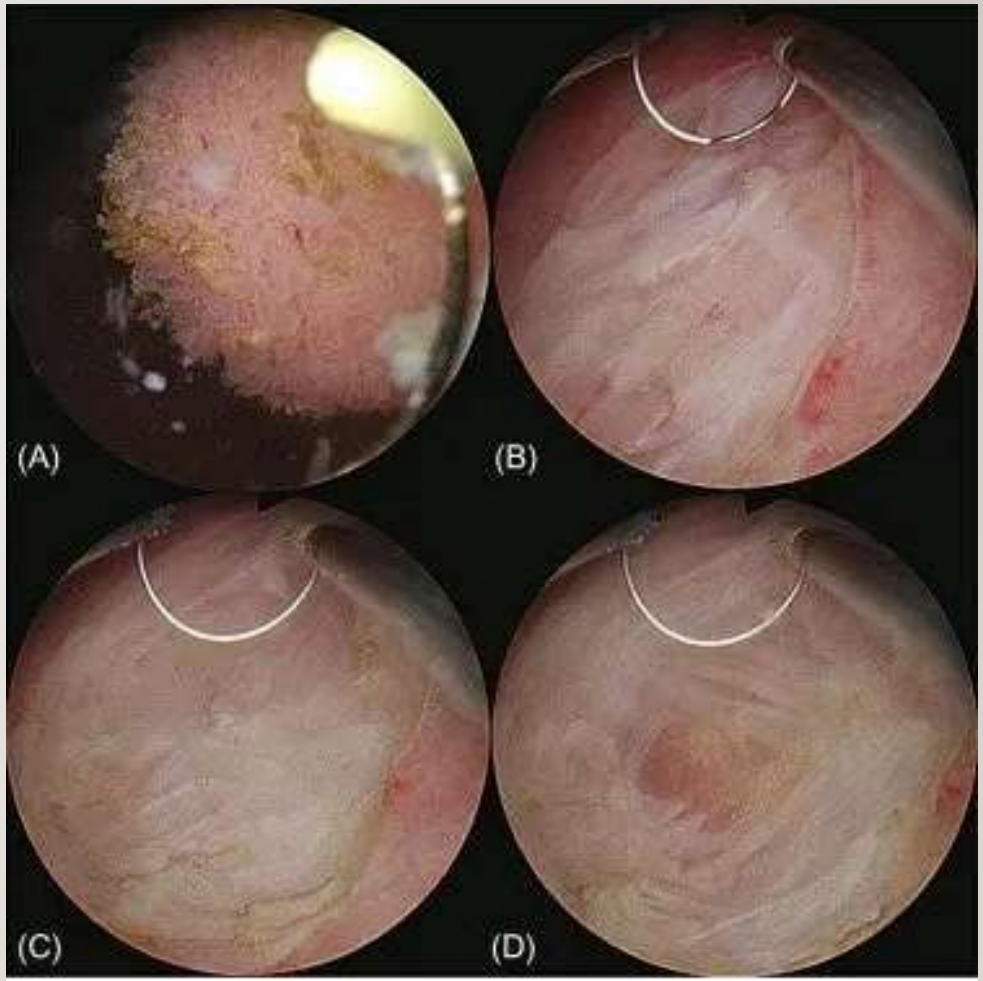
LAPAROSCOPY

Adhesiolysis with LNG – IUD insertion done (due to adenomyosis).

No evidence of endometriosis.

TREATMENT GUIDELINES

- **Surgical management offers potentially definitive treatment. Excisional surgery via bladder shave or partial cystectomy offers good improvement in symptoms with relatively low rates of serious complications and recurrence.**

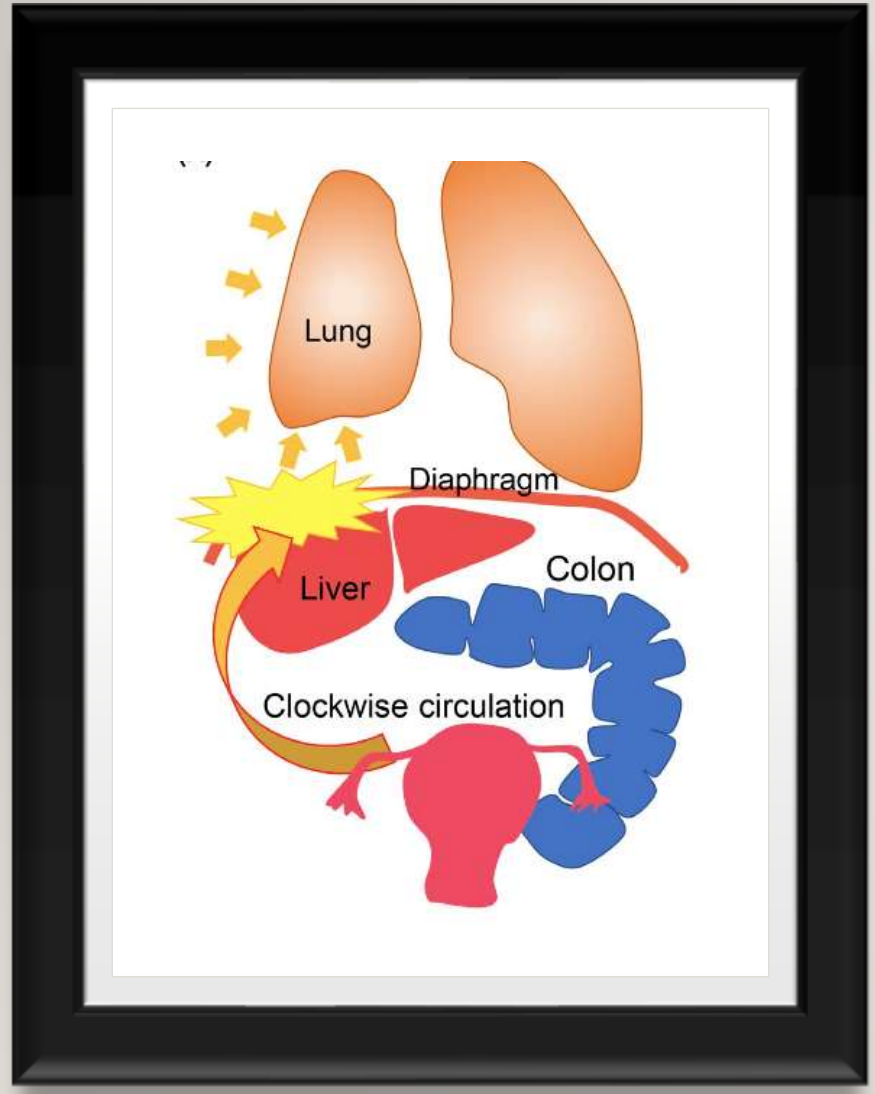


THORACIC ENDOMETRIOSIS

Symptoms have a catamenial (cyclical) pattern, occurring between 24h before and 72h after the onset of menses, and typically recurring.

Catamenial pneumothorax, hemothorax and lung nodules.

Clinicians should be aware of symptoms of extrapelvic endometriosis, such as cyclical shoulder pain, cyclical spontaneous pneumothorax, cyclical cough, or nodules which enlarge during menses.	GPP



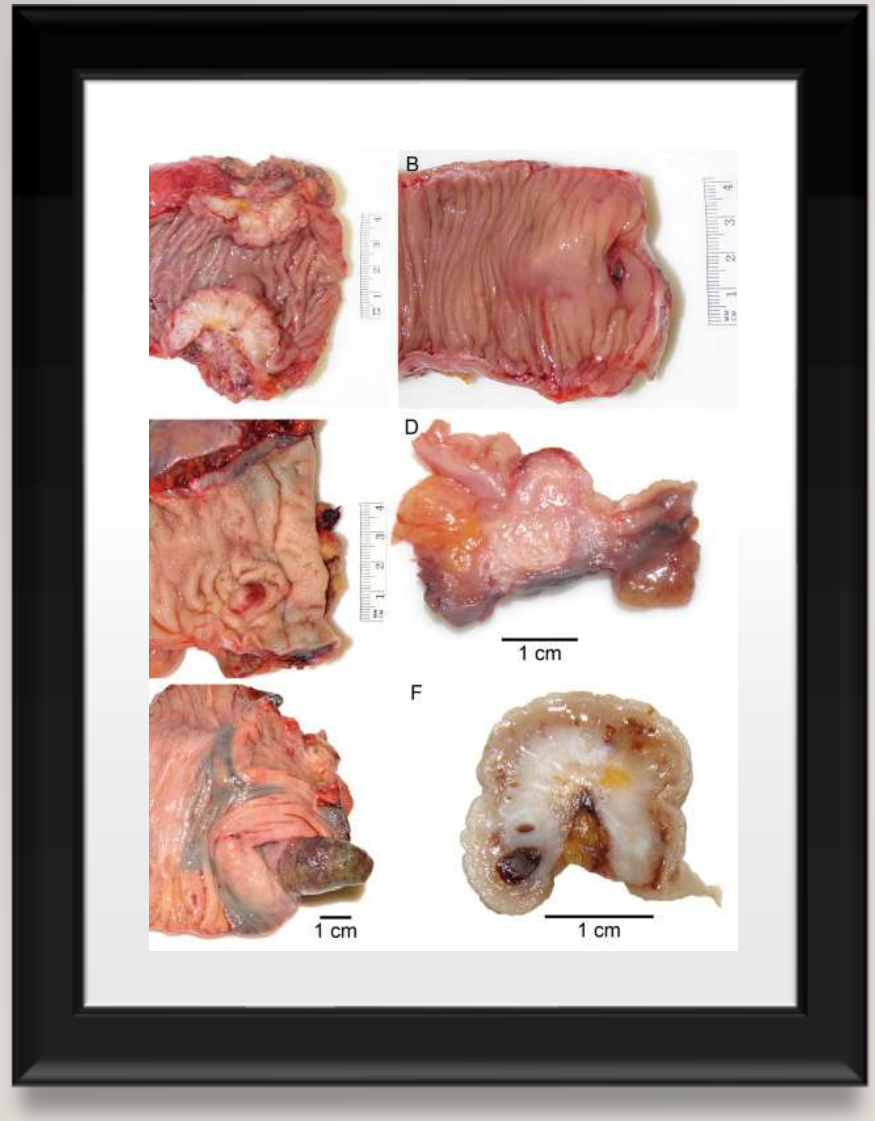
90% - RIGHT SIDE

Surgical treatment (VATS).

High rate of recurrence – combined surgical and hormonal therapy (GnRH agonists and antagonist, OCP, dienogest).

Multidisciplinary approach.

For thoracic endometriosis, hormone treatment can be offered. If surgery is indicated, it should be performed in a multidisciplinary manner involving a thoracic surgeon and/or other relevant specialists.	⊕○○○



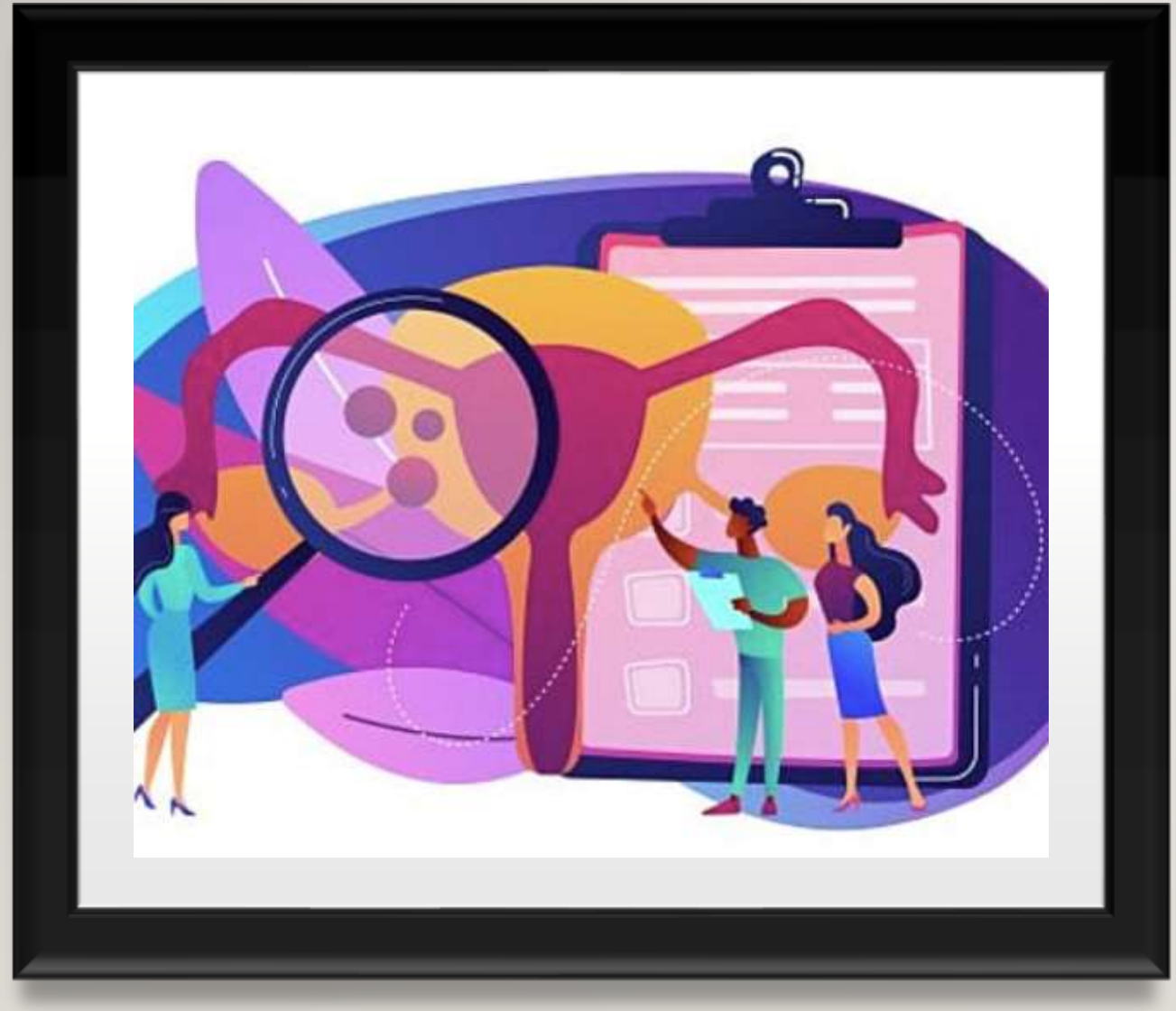
VISCERAL ENDOMETRIOSIS

35 year old patient, para 1 (NVD) operated for colonic mass by GI surgeon. Colonic resection done. No reported evidence of endometriosis on laparoscopy or imaging.

Referred to us because histopathology revealed endometriosis.

She was put on GnRH for 3 months followed by dienogest and is following up with us.

She had a repeat surgery for ventral hernia – no evidence of endometriosis seen.



THE EVERYWHERE DISEASE

A mimic, a thief, a constant guest,
It tests endurance, robs of rest.
No boundary sacred, no border tight,
It spreads its pain in endless night.