Ovarian remnant syndrome

Philippe R. Koninckx

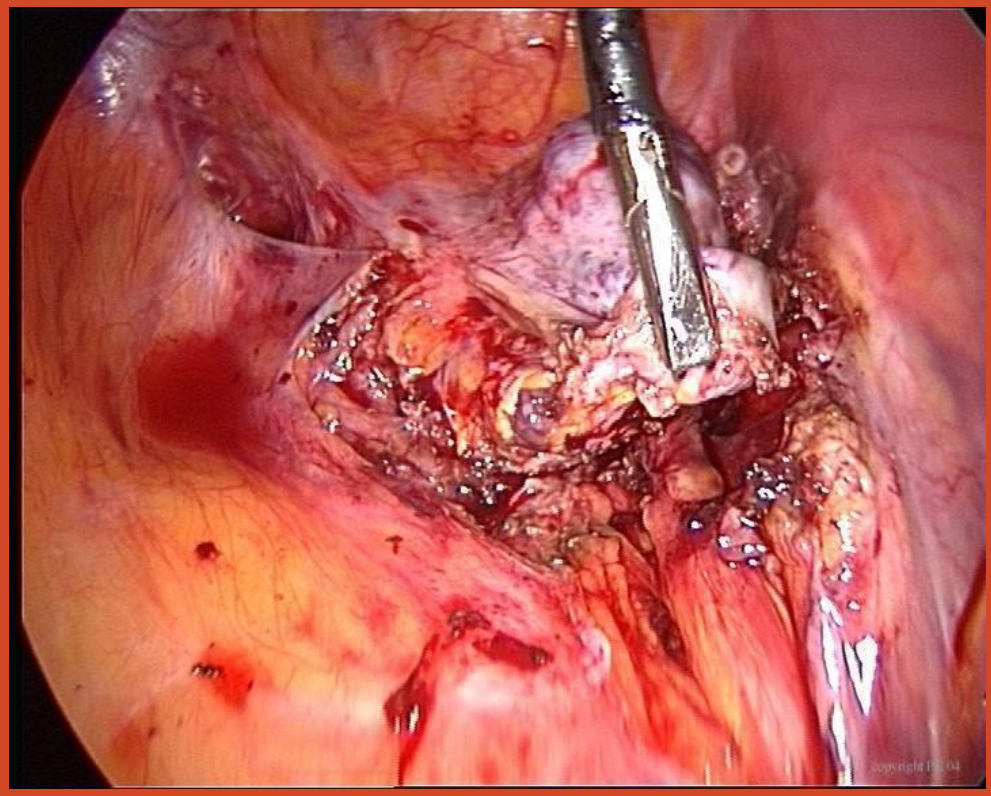
Prof em KU leuven Belgium, Univ Oxford UK, Univ Sacro Cuore, Italy, Hon Prof Moscow Univ ; Honorary Consultant UK

Director Research, Latiffa Hospital Dubai

Consultant Univ Sacro Cuore, Gemelli, Roma

Gruppo Italo Belga, Leuven –Rome, Belgiumly.

Disclosures: Shareholder Endosat NV

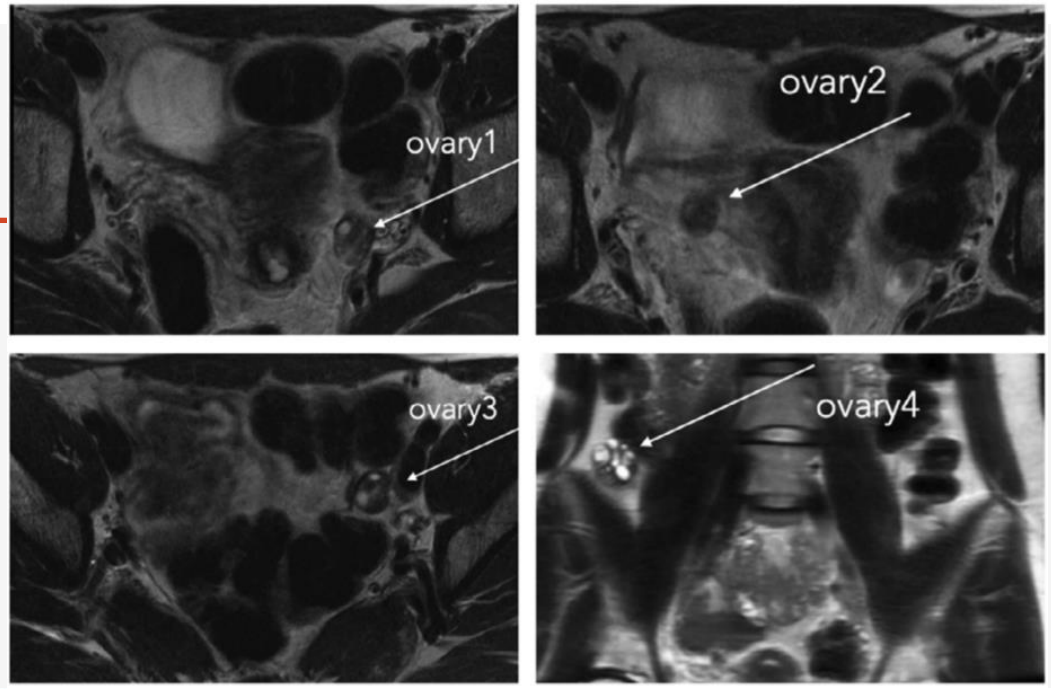


Definitions of Ovarian remnant

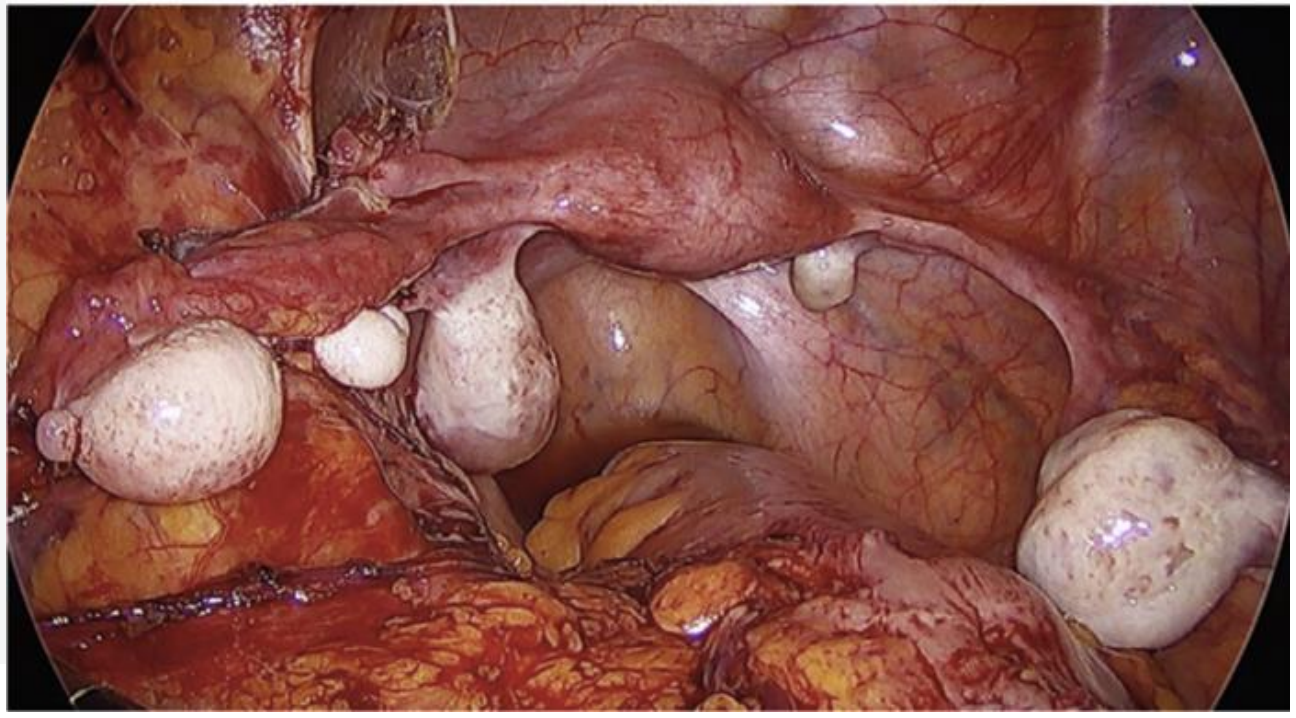
- (Functional) ovarian tissue after ovariectomy
  - Ectopic / accessory /supernumerary n=127
    - Incidence 1/30,000- 7/90,000
    - up to 26% to 75% other anomalies müllerian anomalies, renal agenesis, duplicated ureters, bladder diverticulum, accessory adrenal glands, and a lobulated liver
    - **Supernumerary ovary**: :a disruption of the normal gonadocyte migration from the yolk sac endoderm through the dorsal mesentery before reaching thegenital ridge during embryogenesis
    - **Accessory ovary** : additional ovaries that are found along the genital ridge within any of the gynecologic ligaments.
  - Incomplete ovariectomy
- Fibrosis

Accessory ovary

- Functional ovarian tissue after ovariectomy



- MOVILLA P, OHLIGER M, WOOD S. Prepare for the Unexpected: Accessory Ovaries with Abernethy Malformation. J Minim Invasive Gynecol 2019;26:977-82

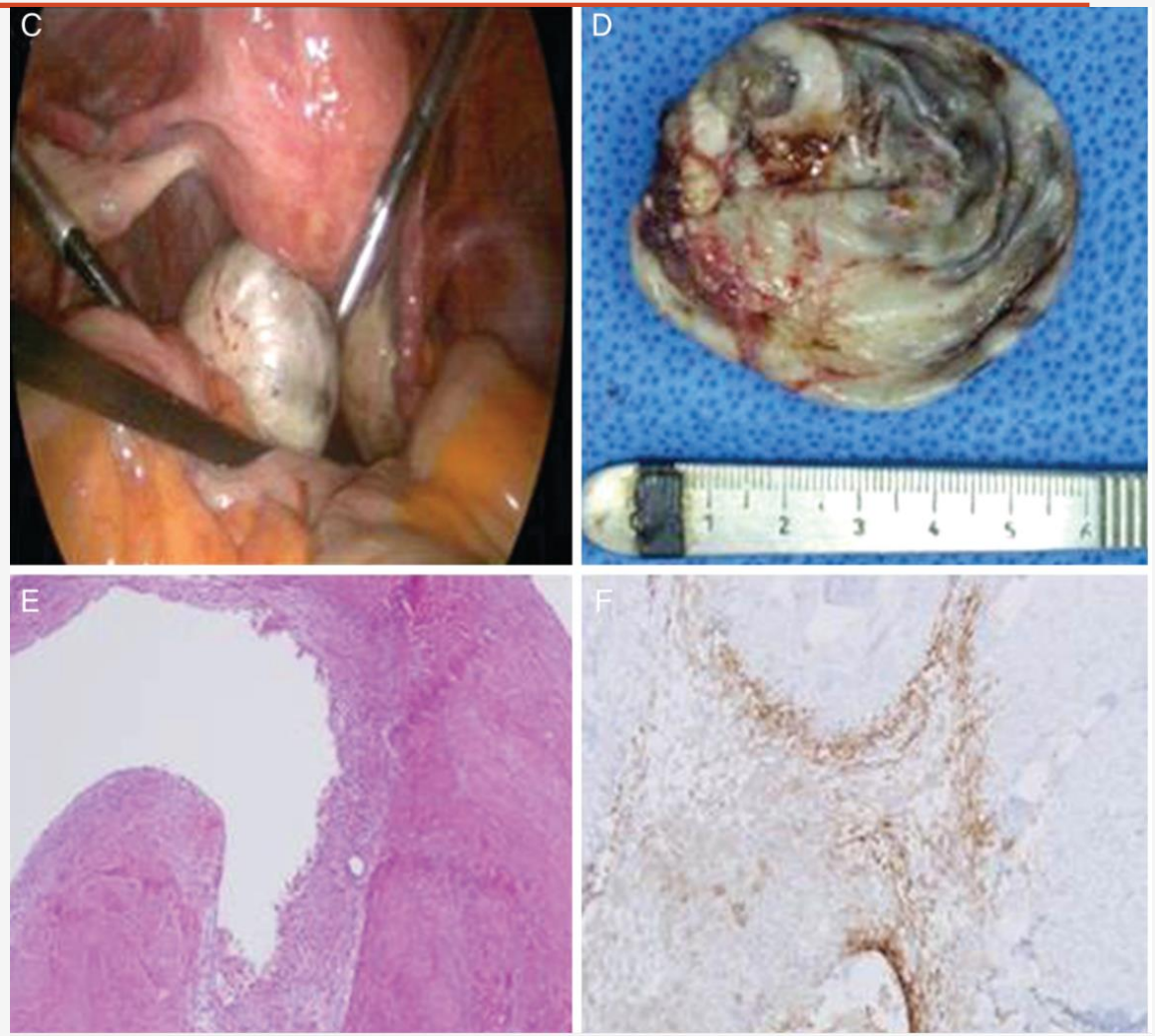


Supernumerary ovary

- Lim CK, Kim HJ, Pack JS, et al. Supernumerary ovary on recto-sigmoid colon with associated endometriosis. Obstet Gynecol Sci 2018;61:702-06.

Table 1. Site and diseases in 18 cases with tumors and/or endometriosis originating in a supernumerary ovary

Case	Authors	Reported Year	Age	Site	Diseases
1	Kriss [11]	1947	49	Omentum	Cystic teratoma
2	Wharton [1]	1959	37	Pelvic	Granulosa cell carcinoma
3	Wharton [1]	1959	39	Side of the right ovary	Serous cystadenoma
4	Hogan et al. [12]	1967	21	Omentum	Cystic teratoma
5	Printz et al. [5]	1973	23	Left retroperitoneum	Mucinous cystadenocarcinoma
6	Huhn [13]	1975	36	Left retroperitoneum	Mucinous cystadenoma
7	Roth and Ehrlich [14]	1977	48	Omentum	Cystic teratoma
8	Cruikshank and van Drie [15]	1982	36	Left retroperitoneum	Adenocarcinoma
9	Mercer et al. [16]	1987	34	Omentum	Cystic teratoma
10	Barik et al. [18]	1991	45	Omentum	Fibroma, Meig's syndrome
11	Barik et al. [18]	1991	28	On pregnant uterus	Endometrioma
12	Besser and Posey [19]	1992	30	Left retroperitoneum	Mucinous adenocarcinoma
13	Badawy et al. [7]	1995	32	Retrouterine	Serous adenocarcinoma
14	Irving and Clement [3]	2011	21	Omentum	Cystic teratoma
15	Kamiyama et al. [20]	2013	47	Left retroperitoneum	Serous adenocarcinoma
16	El-Gohary et al. [17]	2015	20	Left retroperitoneum	Endometrioma
17	Ogishima et al. [8]	2016	40	Rectovaginal pouch	Cystic endoterioma and fibroma
18	Present case	2016	42	Anterior sigmoid colon	Cystic endometrioma



Supernumerary ovary

- OGISHIMA D, SAKAGUCHI A, KODAMA H, et al. Cystic Endometrioma with Coexisting Fibroma Originating in a Supernumerary Ovary in the Rectovaginal Pouch. Case Rep Obstet Gynecol 2017;2017:7239018

FIGURE 1: The rectovaginal mass on pelvic MRI (T2-weighted, sagittal line). The mass with multilocular cyst (indicated by C) and solid (indicated by S) components and a maximal diameter of 9 cm locates in the rectovaginal pouch. The uterus is indicated with an

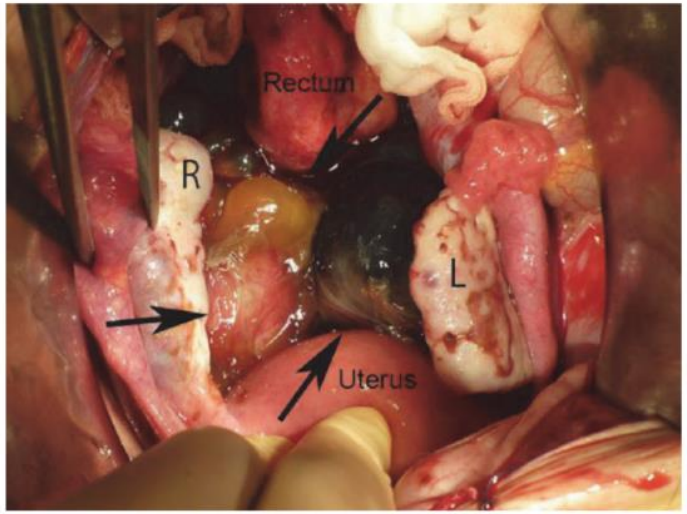
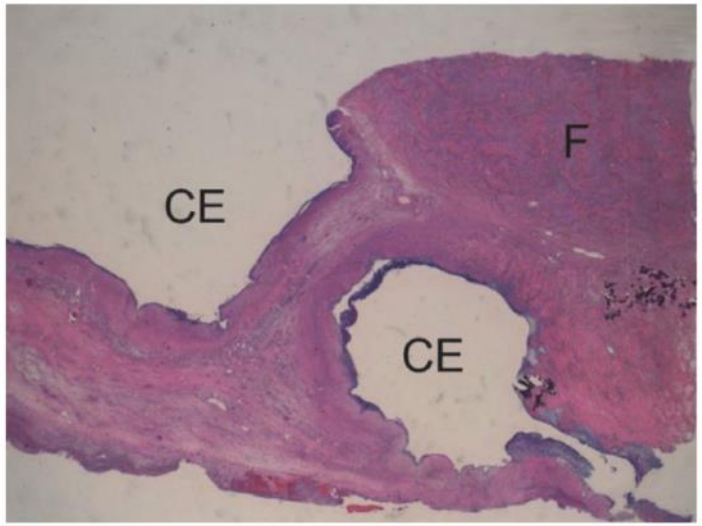
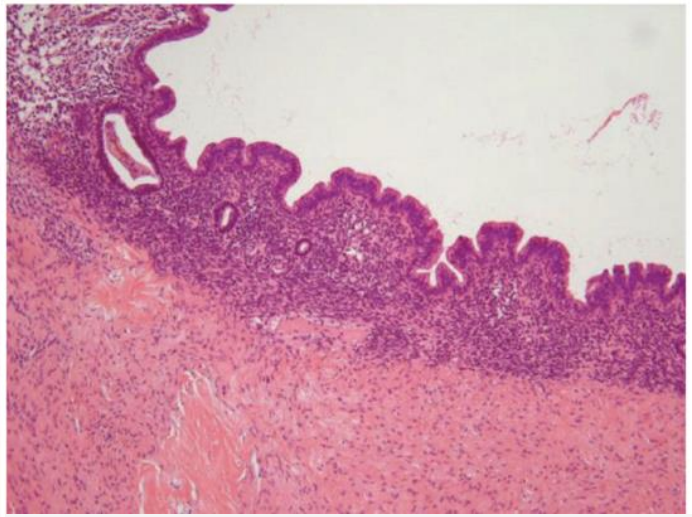


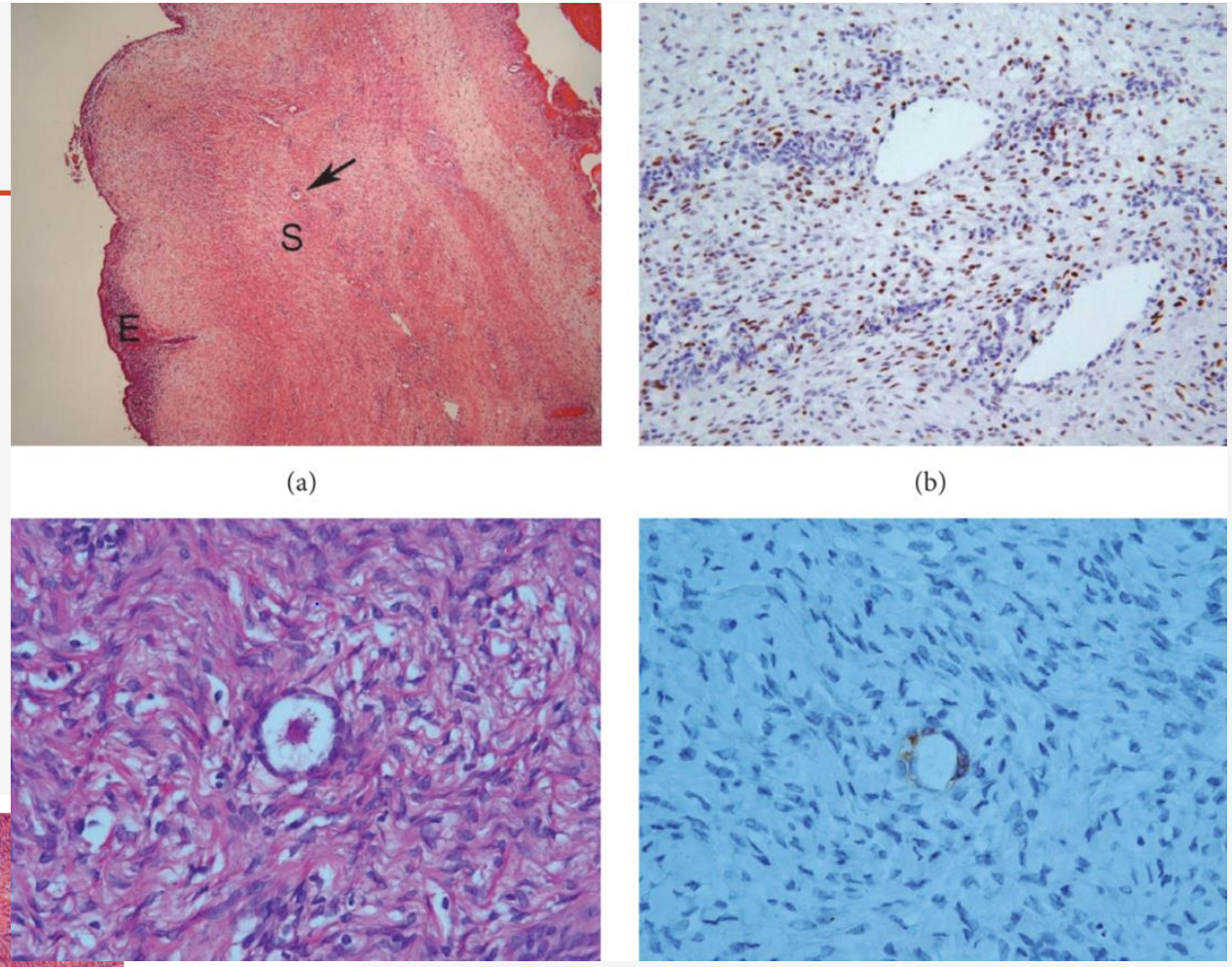
FIGURE 2: View of the rectovaginal pouch during the operation. The cystic and solid mass, indicated by arrows, is present in the



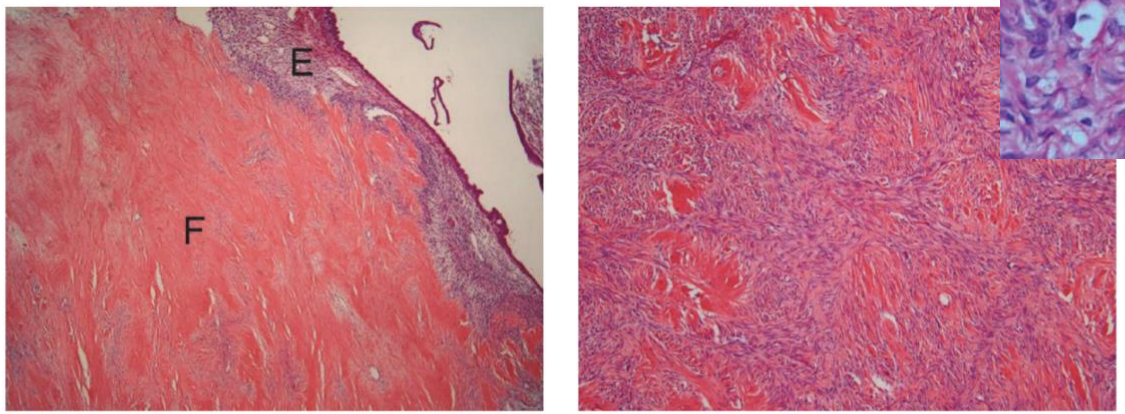


Supernumerary ovary

- OGISHIMA D, SAKAGUCHI A, KODAMA H, et al. Cystic Endometrioma with Coexisting Fibroma Originating in a Supernumerary Ovary in the Rectovaginal Pouch. Case Rep Obstet Gynecol 2017;2017:7239018



(a) (b)



(a) (b)

FIGURE 7: Histological appearance of the fibroma. (a) The fibroma (indicated by F) is present adjacent to endometrial tissue (indicated by E) of cystic endometriosis. HE stain. (b) High-power of the fibroma. Note the increase of the admixture of fibroblasts and collagen fibers. HE

Supernumerary ovary: complete since 2000

- GUPTA R, VERMA S, BANSAL K, JAIN V, SENGAR M, MOHTA A. Mature Teratoma in a Supernumerary Ovary in a Child: Report of the First Case. J Pediatr Adolesc Gynecol 2016;29:e5-7.
- NOMELINI RS, OLIVEIRA LJ, JAMMAL MP, ADAD SJ, MURTA EF. Serous papillary cystadenocarcinoma in supernumerary ovary. J Obstet Gynaecol 2013;33:324.
- BAE HS, RYU MJ, KIM IS, KIM SH, SONG JY. Cancer of the supernumerary ovary in Mayer-Rokitansty-Kuster-Hauser Syndrome: A case report. Oncol Lett 2013;5:598-600.
- ABEDALTHAGAFI M, JACKSON PG, OZDEMIRLI M. Primary retroperitoneal mucinous cystadenoma. Saudi Med J 2009;30:146-9.
- ZHIGANG Z, WENLU S. An intrarenal supernumerary ovary concurrent with a completely duplicated pelvis and ureter. International urogynecology journal and pelvic floor dysfunction 2007;18:1243-6.
- IMIR G, ARICI S, CETIN M, KIVANC F. Supernumerary ovary on sigmoid colon resembling an endometriotic lesion. J Obstet Gynaecol Res 2006;32:613-4.
- HARTIGAN K, PECHA B, RAO G. Intrarenal supernumerary ovary excised with partial nephrectomy. Urology 2006;67:424.e11-24.e12.
- SONNTAG B, LELLE RJ, STEINHARD J, BRINKMANN OA, HUNGERMANN D, KIESEL L. Retroperitoneal mucinous adenocarcinoma occuring during pregnancy in a supernumerary ovary. J Obstet Gynaecol 2005;25:515-6.
- REDMAN R, WILKINSON EJ, MASSOLL NA. Uterine-like mass with features of an extrauterine adenomyoma presenting 22 years after total abdominal hysterectomy-bilateral salpingo-oophorectomy: a case report and review of the literature. Arch Pathol Lab Med 2005;129:1041-3.
- LITOS MG, FURARA S, CHIN K. Supernumerary ovary: A case report and literature review. J Obstet Gynaecol 2003;23:325-7.
- VENDELAND LL, SHEHADEH L. Incidental finding of an accessory ovary in a 16-year-old at laparoscopy. A case report. J Reprod Med 2000;45:435-8.

Ectopic / accessory /supernumerary ovary

- Assumed to be embryological and functional

- But quid when not functional
  - OMORI M, KONDO T, FUKUSHIMA J, et al. Extraovarian Fibroma With Minor Sex Cord Elements: A Case Report and Literature Review. Int J Surg Pathol 2017;25:472-76.

  - This patient 66 year postmenopausal
  - 17 cases 22-78 year;
  - ½ oestrogen secretion + risk of adenocarcinoma

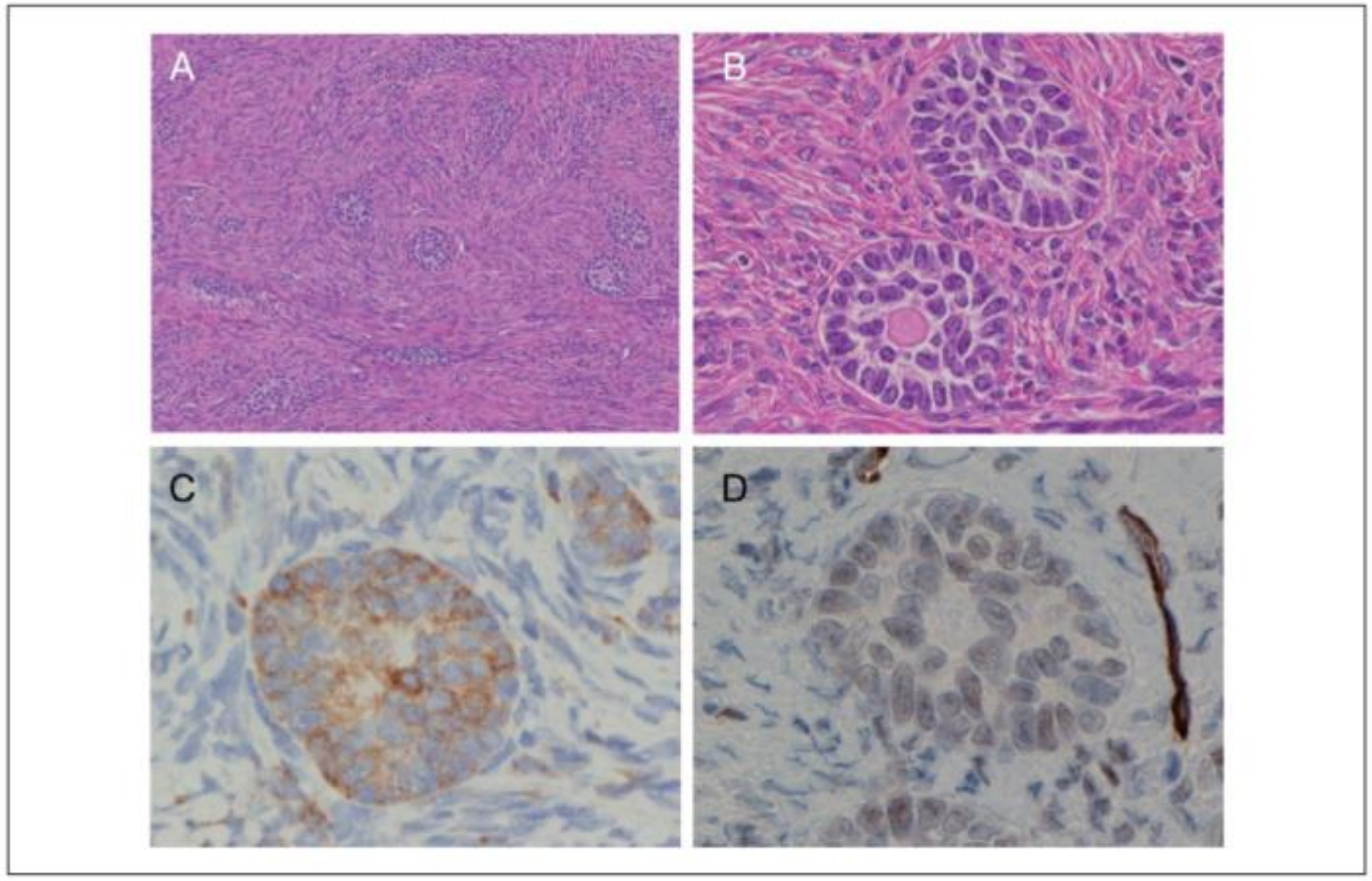
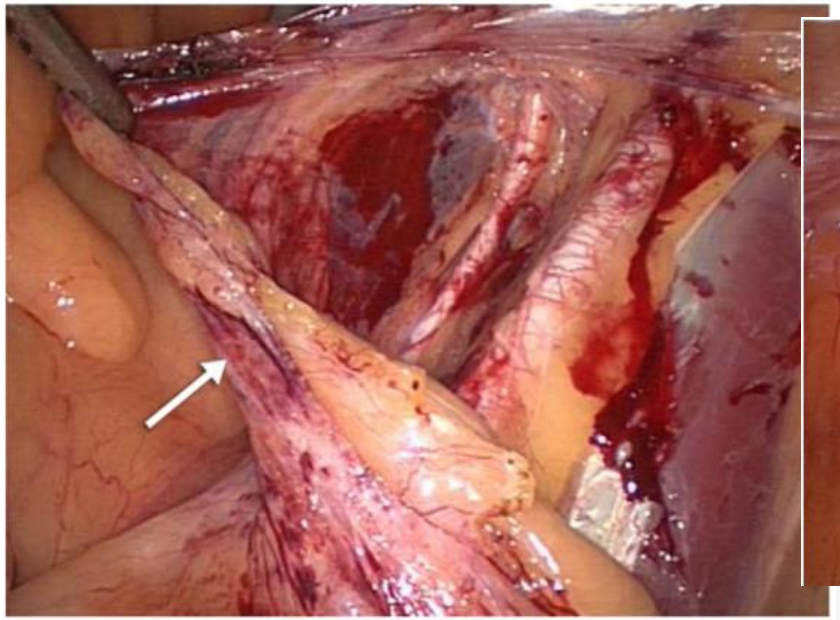
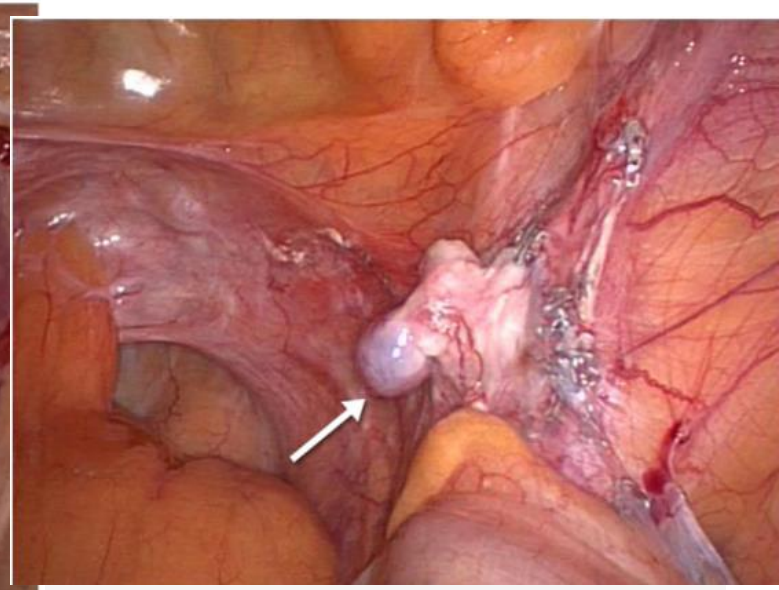


Figure 2. Histological findings of the tumor located in the left broad ligament show 2 types of small cell nests surrounded by abundant fibrous tissue. One nest is an aggregate resembling the Call-Exner bodies of granulosa cell tumors containing eosinophilic amorphous material in the center, and the other nest is a sheet-like aggregate of undifferentiated sex cord–type cells (A, hematoxylin and eosin [H&E], ×10). Granulosa cell–type elements display oval to round nuclei, eosinophilic cytoplasm, small nucleoli and occasional longitudinal nuclear grooves (B, H&E, ×40) and are immunopositive for inhibin (C, ×40) and WT1 (D, ×40).

Tissue remaining after incomplete ovariectomy

- KHO RM, ABRAO MS. Ovarian remnant syndrome: etiology, diagnosis, treatment and impact of endometriosis. Curr Opin Obstet Gynecol 2012;24:210-4.
- Associated with Endometriosis & difficult surgery
- Reimplantation of ov tissue
  - Wall
  - pelvis





Kho. Ovarian remnant syndrome. Fertil Steril 2007.

Table 1. ORS studies since 2006

Study	*N* (number of ORS patients)	Indication for primary oophorectomy	Pathology	Initial presentation	ORS surgery
Mahdavi *et al.*, 2007 [3]	3	Endometriosis – 1; pain – 1	Ovarian serous cystadenoma – 3	Pain – 2; mass – 1	Laparoscopy – 3
Donnez *et al.*, 2007 [5]	1	Endometriosis	Ovarian adenocarcinoma	Pain	Laparoscopy
Kho *et al.*, 2007 [4]	20	Endometriosis – 8; mass – 6	18 – benign; 2 – malignant (mucinous cystadenocarcinoma, endometrioid adenocarcinoma)	Pain – 18; mass – 2	Laparotomy – 1; laparoscopy – 14; robotics – 5
Chao, 2008 [6]	1	Endometriosis	Benign ectopic ovarian tissue, endometriosis	Pain	Laparotomy
Fat *et al.*, 2009 [7]	7	Ovarian cyst – 6; unspecified – 1	Benign – 7	Pain – 7	Laparotomy – 3; laparoscopy – 4
Arden and Lee, 2011 [8▪]	30	Pain or endometriosis – 16; ovarian cysts – 7	All benign	Pain – 29; mass – 1	Laparotomy – 1; laparoscopy – 29

Tissue remaining in infundibulo pelvic ligament after ovariectomy

- FENNIMORE IA, SIMON NL, BILLS G, DRYFHOUT VL, SCHNIEDERJAN AM. Extension of ovarian tissue into the infundibulopelvic ligament beyond visual margins. Gynecol Oncol 2009;114:61-3.
  - In 14 %
  - From 0,2-1,4 (3,5)cm
  - Ovarian remnant

    in 6,5-18%

    > vaginal ovx

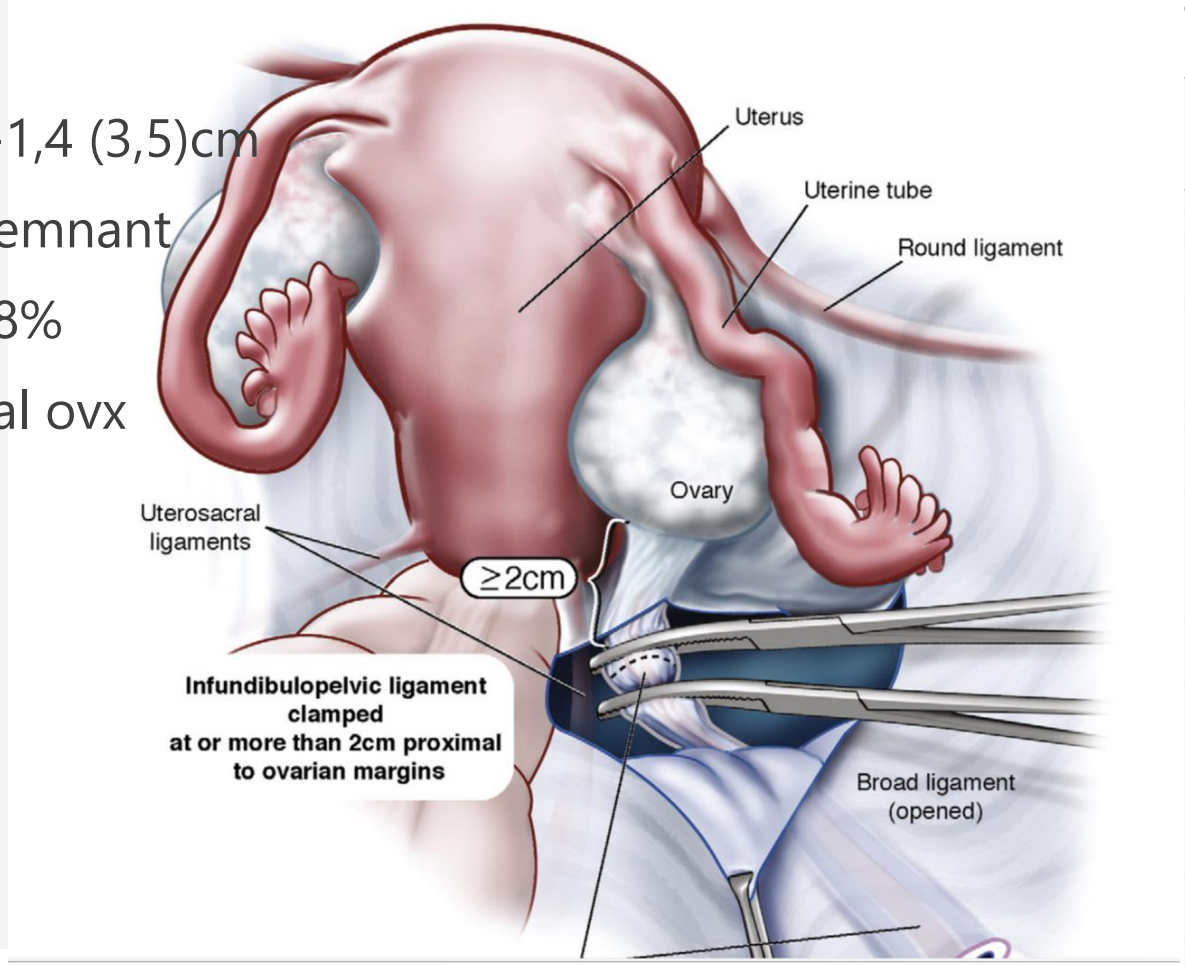


Table 1
Infundibulopelvic (IP) ligament measures and extension of the ovarian tissue.

Menopausal status	Ovary[a]	Operative measure of IP (cm)	Fresh specimen measure of IP (cm)	Extension of ovarian tissue into IP (cm)	Final pathology (ovaries)
Premenopausal					
#1	R IP	4.0	3.0	0.2	Normal tissue
	L IP	4.5	3.0	0.0	Normal tissue
#2	R IP	3.5	2.5	0.0	Endometriosis
	L IP	2.5	2.0	1.4	Normal tissue
Postmenopausal					
#1	R IP	4.5	3.5	0.8	Normal tissue
	L IP	3.5	2.5	0.0	Normal tissue
#2	R IP	4.0	2.5	0.0	Papillary serous carcinoma
	L IP	4.0	3.0	0.2	Papillary serous carcinoma
#3	R IP	5.0	4.0	0.0	Normal tissue
	L IP	4.0	3.0	0.2	Normal tissue
#4	R IP	3.3	2.5	0.0	Metastatic adenocarcinoma of endometrium
	L IP	3.0	2.0	0.8	Metastatic adenocarcinoma of endometrium
#5	R IP	5.0	2.0	0.0	Normal tissue
	L IP	3.5	1.6	0.2	Normal tissue
#6	R IP	3.5	2.0	0.4	Fibroma
	L IP	3.5	1.5	0.0	Normal tissue

Diagnosis of Ovarian remnant

- Most diagnoses
  - BSO + cystic mass at ovarian location
  - BSO + estrogen secretion
- Unclear
  - Diagnosis of stroma by pathology ??
  - Presence of follicles ??

Diagnosis of ORS

- VILOS GA, MARKS-ADAMS JL, VILOS AG, ORAIF A, ABU-RAFEA B, CASPER RF. Medical treatment of ureteral obstruction associated with ovarian remnants and/or endometriosis: report of three cases and review of the literature. J Minim Invasive Gynecol 2015;22:462-8.
- Endometriosis & difficult BSO
- Endometriosis + ureteral obstruction + unclear source of estrogens
  - Pragmatic therapy by GNRH + add-back
  - Even not an attempt to find the source of estrogens

Hormones and ORS

- 50% have ‘normal estrogens’
- Not useful for diagnosis

- However HRT estrogens are often not measurable in Radio-immuno-assay

“ovarian mass” concept

In rats

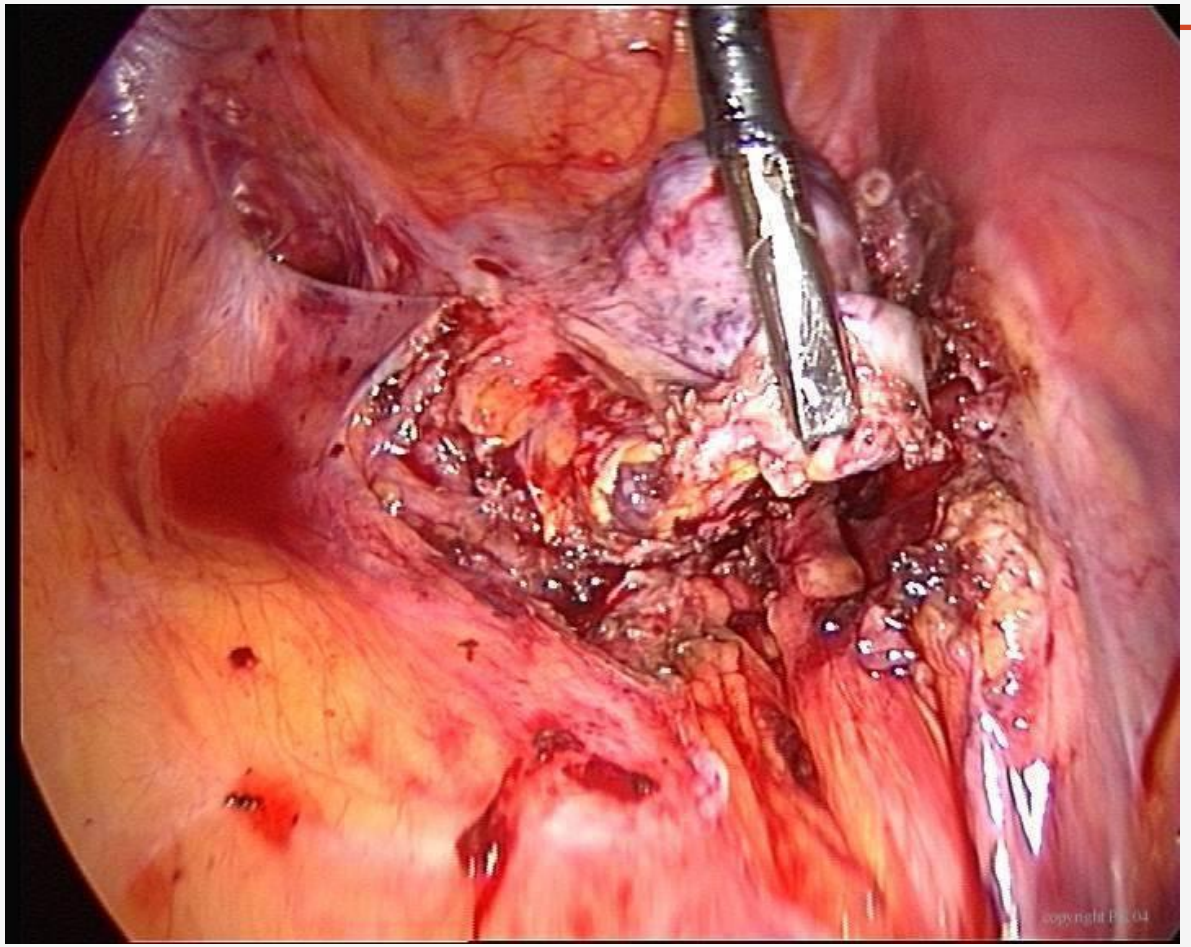
- + extra ovary -> remains inactive ovary
- Removal of 1 ovary -> becomes active

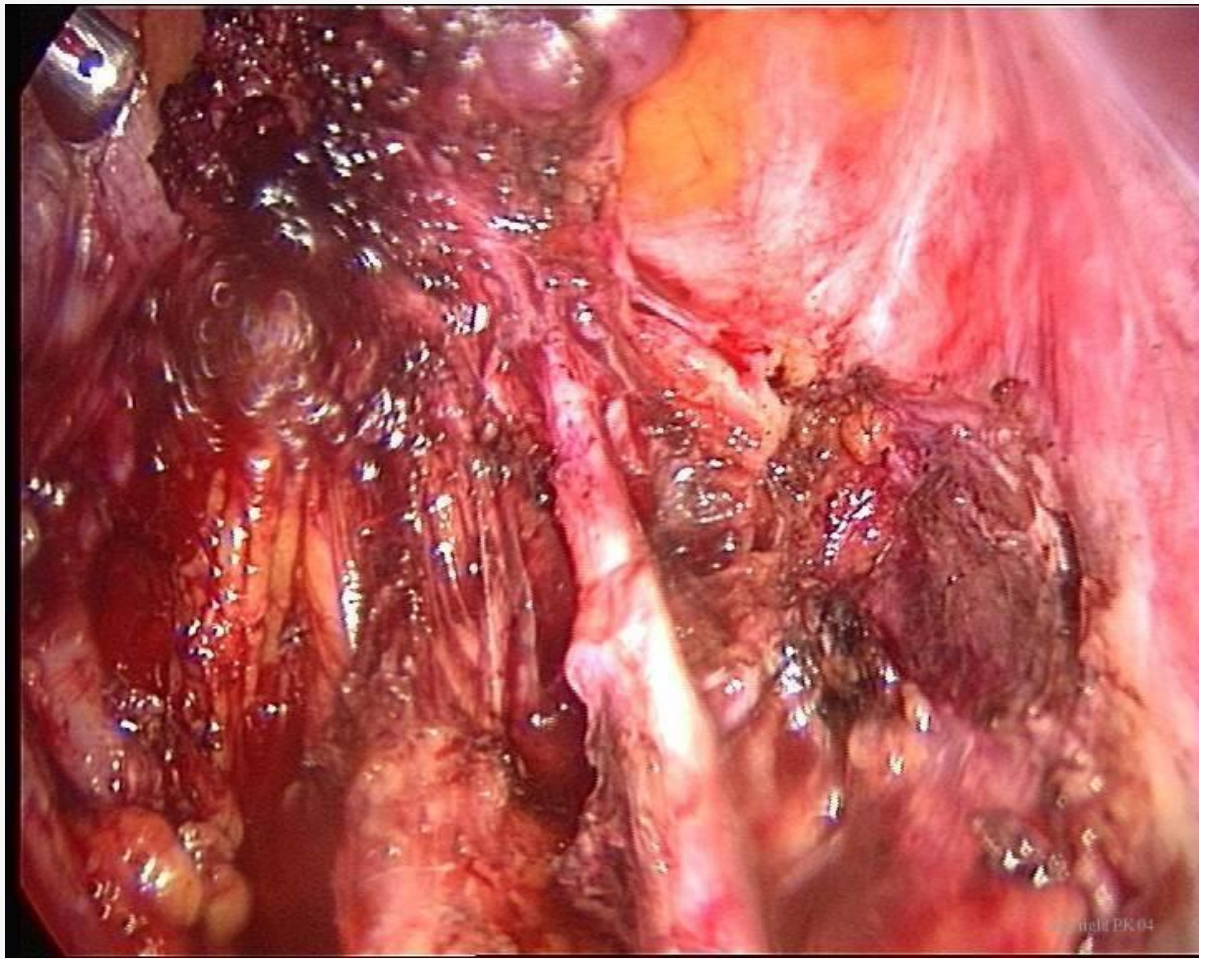
Translated to humans

- Ovarian “remnants” risk to become active after ovariectomy

-

Fibrotic … Ovarian remnant surrounding the ureter





Conclusions: ovarian remnant

- 1/20,000 surnumerary ovaries
- Ovarian remnant
  - Ovarian stromal remnant in 7-18% after ovariectomy
  - More after endometriosis surgery -> more difficult planes of cleavage & more ovarian tissue
- Ovarian remnant 50/50 hormonal active/inactive
- Symptoms : mainly pain
- If only stromal component and inactive : unclear diagnosis and clinically fibrosis
- Always difficult surgery since ureter circularly involved
- Unclear why pain and why around ureter
- MRI + US + surgical observation

Rare cases

At laparoscopy : Inspection : slow and in detail

you only recognise what you know

be a clinician