Thoracic Endometriosis
Case Presentation

43F, P0, presented on 10/19/15 w/ SOB

PMHx: HTN, endometriosis, uterine fibroids and ovarian cysts, PE (9/2015)

PSHx: UHR (childhood), resection of abdominal wall endometrioma (2005)

Meds: warfarin, norethindrone

NKDA

No toxic habits
Presented with acute onset dyspnea x 5 days.

LMP 5/1/15

HGB 9.6; CA 125 87.4; CEA & CA 19-9 WNL

Large right pleural effusion

Thoracentesis in ED, cloudy, bloody fluid. Negative for malignant cells.

PT refused surgery

Discharged with PleurX catheter
A Rough Year…

Returned with SOB

Right PTX with small effusion on CXR

4 day course uneventful

Chest tube removed after resolution of PTX and effusion

Discharged with plan for VATS biopsy, pleurodesis
A Rough Year…

(Started norethindrone on 7/1)

Underwent VATS biopsy and pleurodesis

Gross: Adhesions, no obvious lesions

Path: pleura with fibrosis, hemorrhage, chronic inflammation, hemosiderin laden macrophages

Postoperative course uneventful

Discharged POD3
A Rough Year…

Sept 8

Returned with SOB

Loculated pleural effusions

IR guided drainage. Same bloody, cloudy fluid

Bedside pleurodesis

Discharged HD 6
A Rough Year…

Presented with SOB, chest pain.

CTA demonstrated b/l PE w/ loculated right effusions

Enoxaparin to warfarin
A Rough Year…

Oct 21

Presented with SOB for 2 days.

LMP 2 days prior w/ heavy bleeding

BP 117/57   HR 100   RR 36   T 97F   SpO2 75%

HGB 3.4   INR 10.2

Bedside Sono w/ free fluid in abdomen

CXR w/ right pleural effusion
A Rough Year…  

Oct 21

Transferred to SICU

BiPAP

Resuscitated, normalization of INR

Chest tube placed

Discussion about GYN surgery - pt refused.
HD 5

Chest tube w/ sudden increase in bloody output, 2.3 liters

HGB 9.8 to 7.8 over 4 hours despite 4 units

Tachycardic to 102. No change in abdominal exam

Taken to OR for VATS exploration

Several dark purple lesions noted on pleura and diaphragm.

PATH: endometrial stroma, fibrosis. No epithelial component, ER and PR negative.
Postoperative Course

Uneventful

Transferred to floor

Restarted on coumadin

Initiated on Danazol

No recurrence to date
Questions?
Thoracic Endometriosis
Outline

Endometriosis

Thoracic Endometriosis - terminology
Endometriosis

Endometrial glands and stroma at extrauterine sites

Common, benign, chronic, estrogen-dependent

Sx: pelvic pain, dysmenorrhea, dyspareunia, infertility

RF: nulliparity, early menarche/late menopause, short menstrual cycles, prolonged menses

Sites: usually pelvic, but can be anywhere!

Most common extra-pelvic: thoracic

Others - abdomen, skin, brain, eyes
Thoracic Endometriosis

Terminology:

Thoracic Endometriosis - endometrial tissue (hormone-receptor positive stroma and glands) identified on histology from chest tube aspirate, thoracotomy, bronchoscopy

“Probable” Thoracic Endometriosis - suggestive but not definitively diagnostic

Thoracic Endometriosis Syndrome - no histologic confirmation but thoracic manifestations in association with menstruation
Thoracic Endometriosis

Meigs Syndrome - Triad benign ovarian tumor (fibroma), pleural effusion, and ascites, WITH disappears after removal of pelvic tumor

Joe Vincent Meigs
Josiah V. Meigs

MEIGS’ EXPERIMENTAL RAILWAY
JOSEPH V. MEIGS, INVENTOR AND ENTREPRENEUR SUCCESSFULLY TESTED HERE A STEAM POWERED ELEVATED MONORAIL TRAIN INTENDED FOR RAPID TRANSIT USE IN BOSTON 1886
Pathogenesis

Several theories for thoracic endometriosis

- Autotransplantation
- Metastasis
- Coelomic Metaplasia
- Altered Immunity
Autotransplantation

Sampson’s Theory - retrograde movement of endometrium during menstruation through fallopian tube and into peritoneum.

Migration through diaphragmatic defects

Defects are uncommon

Thoracic endometriosis predominantly right-sided

Ectopic tissue differs from eutopic tissue on path
Metastasis

Microemboli through venous or lymphatic system

explains brain and eye foci

still the right sided problem
Coelomic Metaplasia

Peritoneal and Thoracic cavities contain undifferentiated mesenchymal cells

Triggered into developing into endometrial tissue

Promoted by hormone exposure
Altered Immunity

Inability to recognize presence of ectopic endometrial tissue

Reduced NK cell activity leading to decreased cytotoxicity to autologous endometrium
Pathophysiology

Endometrial implants undergo proliferative luteal phase followed by decidualization

In the chest, cause hemothorax/pneumothorax or hemoptysis during menses

Physiologic theory - (non-endometriosis catamenial PTX)

  vasoconstriction and bronchospasm caused by prostaglandin F2 induced alveolar rupture

Another Theory - retrograde air during menses pass through diaphragmatic defects...
Epidemiology

Incidence unknown - reported at <1% of women undergoing pelvic surgery for suspected or known pelvic endometriosis

in women with primary spontaneous pneumothorax - 3-6%

Recurrent pneumothorax - 6-20%

Catamenial pneumothorax - 65-89%

Women of reproductive age, peak 30-34

not all have pelvic disease! (16%)
Risk Factors

Besides risk factors for pelvic endometriosis infertility (OR 4.21) and a history of uterine surgery or scraping (OR 2.85) strongest predictors of catamenial / endometriosis related pneumothorax

Genetic predisposition for pelvic endometriosis

No association reported for thoracic endometriosis
Gross Pathology

implants are raised, up to a few centimeters in size, red/purple, gray, black white
Histology

two major components - stroma and glands

also with fibrous tissue, blood, and cysts

implants stain positive for estrogen or progesterone receptors

frequently observe stroma without HR+ (Probable thoracic endometriosis)
Clinical Presentation

Onset within 72 hours of menstruation of chest pain and dyspnea

Right sided (>80%)

Intermittent (months to years)

Most common presentation is catamenial pneumothorax (70%)

Hemothorax in <15%, self-limiting

Hemoptysis in <14%

PTX without temporal relation to menses (10%)
Diagnosis

Often a clinical diagnosis, delayed for several episodes

CXR and CT chest

Bronchoscopy

pleural fluid, gross findings, histopathology

CA 125 and CA19-9 may be elevated, not sensitive nor specific

Differential - Lymphangioleiomyomatosis, Birt-Hogg-Dubé syndrome, Langerhans cell histiocytosis, lymphocytic interstitial pneumonia
Treatment

Hemo/Pneumothorax

Initial therapy - Tube thoracostomy

Diagnosis - Thoracoscopy, biopsy/resection

Prevention of recurrence - pleurodesis

+/- hormone therapy 6-12 months
Treatment

Hemoptysis

Locate and control bleeding

Bronchoscopy - biopsy, Nd:YAG laser

Distal endobronchial lesion - wedge resection

Massive hemoptysis - lobectomy, pneumonectomy

+/- hormone therapy 6-12 months
Hormone Therapy

Difficult to remove all endometrial tissue from the chest surgically...

Hormone therapy similar to pelvic disease

Reduces recurrence

Significant side effects
Hormone Therapy

Gonadotropin-Releasing Hormone Analogs (Lupron)

Suppress ovarian hormone production -> inhibit growth of endometrial tissue

Side effects - menopause, osteoporosis

Oral contraceptives + danazol
Refractory Disease

Options include

Switching hormonal therapy

Pleurectomy, repeated pleurodesis

Hysterectomy + BSO
Summary

Thoracic endometriosis most commonly presents as catamenial pneumothorax

Evaluate as close to onset of menses and symptoms - CT and VATS

Clinical diagnosis - histopathology can be difficult to prove

Include hormone suppression