Basic Data

- Birthday: 1951-07-02
- Gender : Female
- Admission date : 2004-06-28

Chief Complaint

 A protruding mass over RLQ abdomen for many years.

Present Illness & Past History

- Pseudomyxoma peritonei s/p laparotomy and debulking of adhesion at Chung Shin Municiple hospital on 2003-08-26.
- CEA: 11.93, CA199: 102.7 at our OPD for cancer follow up on 2004-06-17.
- DM for 1~2 years under regular control.
- HTN for 1~2 years under regular control.

Review of systems & PE

- Review of systems: no specific findings
- Physical examination :
 - Abdomen : 4x4 fixed palpable mass

Lab Data

採樣時間	2004-06-17
CEA (< 4.6 ng/ml)	11.93
CA199 (< 37 U/ml)	102.70

Lab Data

檢驗日期	2004-06-28	
WBC	3600/uL	
RBC	3.17 (10 ⁶ /uL)	
Hb	10.6 g/dL	
Hct	33%	
MCV	104.1 fL	

Image Findings-CT pre-contrast

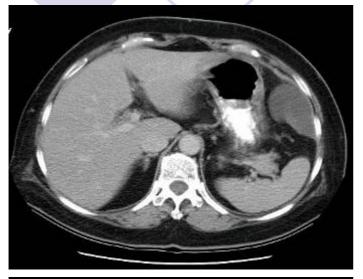








Image Findings-CT post-contrast



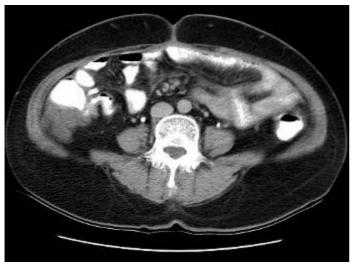






Image Findings-Conclusion

- Multiple non-enhanced intraperitoneal masses (HU: 22~25)
- Bowel wall thickening and folds swelling
- Impression : Pseudomyxoma peritonei

Image Findings-D/D

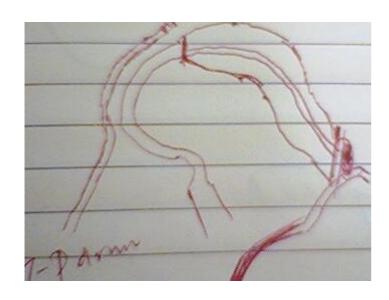
- Ascites.
- Sclerosing peritonitis.
- Intraperitoneal abscess.
- Retroperitoneal lymphadenopathy.



Recurrent pseudomyxoma peritonei

Operation Findings

- 2004-07-02
 - R't hemicolectomy
 - Hartmann's procedure
 - Debulking of tumor





Pathological Findings-Grossly

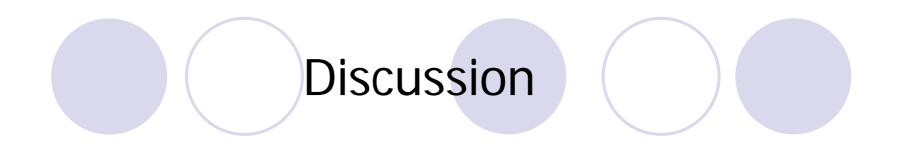
- R't hemicolectomy
 - Marked fibrous adhesion and mucinous tumor involving on the serosa and the omentum are noted.
- Hartmann's procedure
 - The outer surface of the colon also shows marked mucinous tumor coating and fibrous adhesion.
- Debulking
 - Mucinous substance admixed with adipose tissue
- All the tumor is limited to serosa and subserosal areas.
- Impression : Pseudomyxoma peritonei

Pathological Findings-Microscopically

- Large mucin pools
- Only a few mucin pools have an epithelial lining
- Some mucin-producing cells are arranged in glands, nests, or ribbons.
- Benign in appearance
- No invasive components
- Frozen diagnosis : Pseudomyxoma Peritonei



Recurrent Pseudomyxoma Peritonei



Pseudomyxoma Peritonei

Introduction

 Pseudomyxoma Peritonei is a slowly progressive disease that produces extensive mucus accumulation within the abdomen and pelvis.

Symptoms & Signs

- Gradual increase in abdominal girth.
- The increase of intraperitoneal pressure prevents the patient from eating normally.
- Increase in body weight.

Histological classification of mucinous lesions

- DPAM-disseminated peritoneal adenomucinosis
- PMCA-peritoneal mucinous carcinomatosis
- Hybrid tumor

From: Mod pathol 2001;14:664-671 Abstract quote

Pathogenesis

MUC2-expressing goblet cell

From: Am J Pathol 2002 Aug; 161(2):551-64 Abstract quote

Tumor Markers

- For diagnosis and follow-up
- CEA (75%) & CA199 (58%)
- CA199 was shown to be a more useful tumor marker than CEA for follow-up.

From : Ann Surg Oncol. 2002 Dec;9(10):961-7. Abstract quote



- Multiple low-attenuation masses within the peritoneal cavity
- Scalloping of the liver margin
- Calcifications



Figure 26.7. Pseudomyxoma Peritonei. A CT scan of a 65-year-old male with a ruptured appendiceal mucocele demonstrates copious ascites (a) with prominent septations (closed arrows) and mass effect displacing bowel loops (b). Punctate calcifications (open arrow) are also present on peritoneal surfaces.

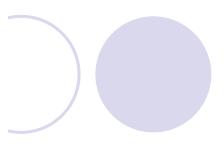




Fig. 20.40 CT reveals the liver scalloping typical of pseudomyxoma.

http://www.surgicaloncology.com/atct.ntm

Treatment (

- Watch and wait
 - OMonitor the situation closely
- Surgery
- Systemic chemotherapy
 - The evidence for the use of systemic chemotherapy in the management of pseudomyxoma peritonei has yet to be established



- Debulking
 - Traditional surgical approach
 - OHigh recurrent rate
- Cytoreductive surgery plus peri-operative chemotherapy(HIPEC)
 - Aggressive removal or destruction of all visible tumors
 - After cytoreduction, chemotherapy is administered directly into the peritoneal cavity.

Post-operative Mortality & Morbidity

- Mortality rates of complete cytoreduction: 3~5%
 - Main complication: lung infections and heart failure
 - Other complications : clots in main leg vein
- Significant morbidity: 30%
 - 20% of patients require further surgery
 - 20% of patients need a stoma
 - Leakage of anastomosis

Prognosis (

- Prognostic factor for survival
 - Completeness of cytoreduction (P<0.0001)</p>
 - OHistopathological charater (P<0.001)
 - Extent of previous surgical interventions (P=0.001)

From: Eur J Surg Oncol 2001 Apr;27(3):239-43 Abstract quote

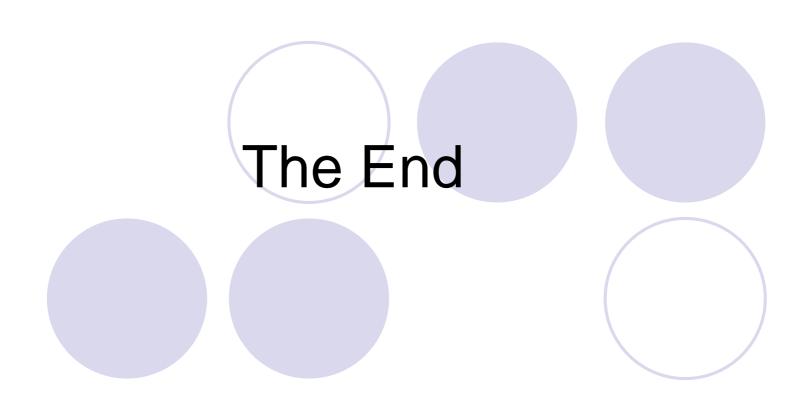
Prognosis

Survival rates	5 years	10 years
DPAM	75%	68%
PMCA-I/D	50%	21%
PMCA	14%	3%

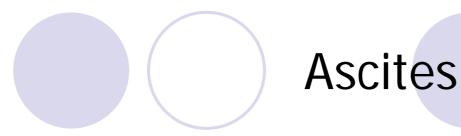
• From: Cancer 2001 Jul 1;92(1):85-91 Abstract quote

Prognosis

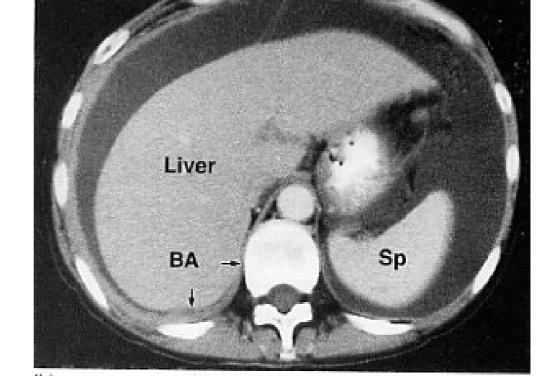
- From: Mod Pathol 2001;14:164-171 Abstract quote
- Unsuccessful second-look surgery is often related to an inaccurate initial histologic classification of appendiceal mucinous tumor.
- Assessment of tumor histology can predict the outcome if a uniform surgical treatment is used in patients







- Low density fluid accumulation
- Common sites :
 - Douglas pouch
 - Morrison's pouch
 - Paracolic gutter
- Retroperitoneal free







Sclerosing Peritonitis

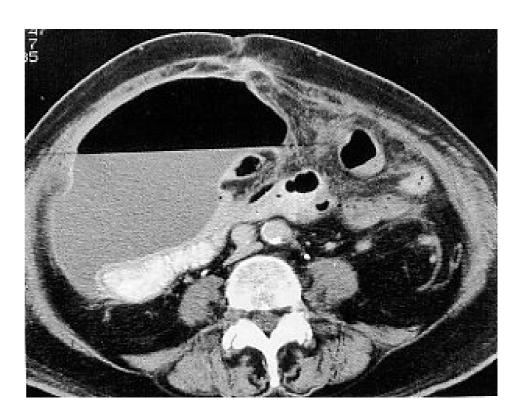
- Adhesion
- Calcifications



Intraperitoneal Abscess

- Air-fluid level
- Thin enhanced wall

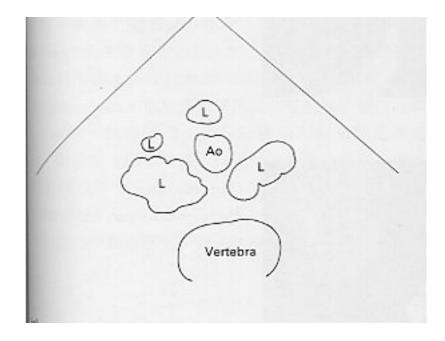
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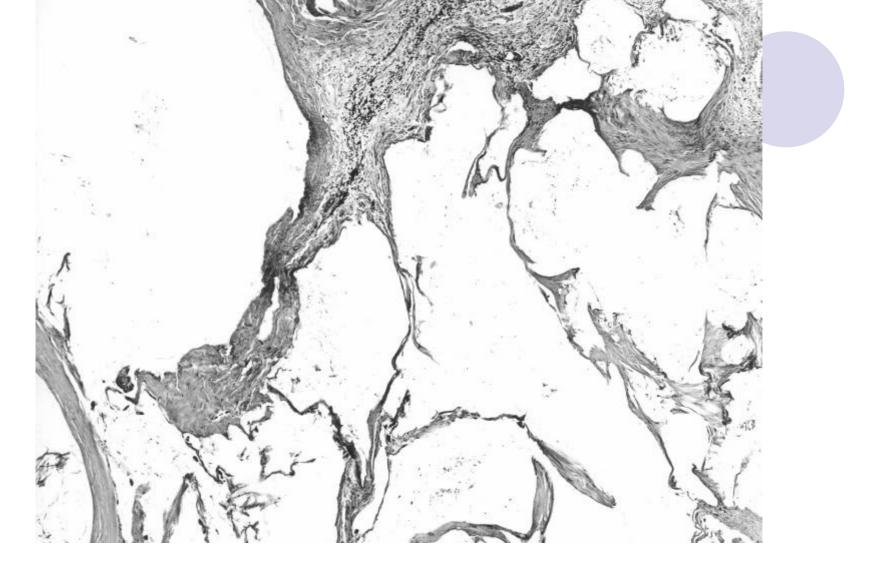


Retroperitoneal Lymphadenopathy



Over lymph node chain





Strips of mucinous epithelium with minimal cytologic atypia are found associated with pools of mucin and fibrous tissue. (H&E). Ronnett BM, Schmookler BM, Sugarbaker PH, Kurman RJ: Pseudomyxoma peritonei: New concepts in diagnosis, origin, nomeclature, and relationship to mucinous borderline (low malignant potential) tumors of the ovary. In: Fechner RE, Rosen PP (Eds): Anatomic Pathology 1997 ASCP Press, Chicago, IL, 1997.