Malignant Struma Ovarii

F2Neeranat Simasing
Harinroat Gladcharoen, MD
Ratchaneewan Kwanchareon, MD

Endocrine division, Internal Medicine Department
Faculty of Medicine Vajira Hospital
Navamindradhiraj University

Malignant Struma Ovarii

- In 1895, Malignant struma ovarii(MSO) was first reported
- Struma ovarii is a form of ovarian teratoma
  - 0.5 – 1% of ovarian teratoma
  - Comprised ≥ 50% thyroid tissue
  - Mostly on one side, ≤ 5% on both sides
- Malignant struma ovarii is very rare (5-10% of struma ovarii)
    - 5-23% metastasis

Front. Oncol. 11:645156

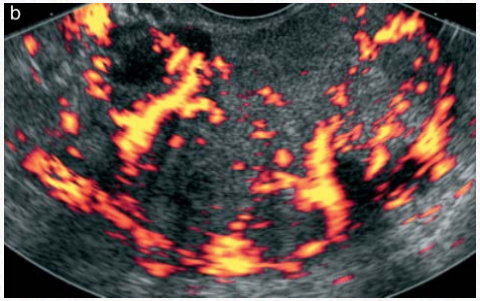
Clinical Feature

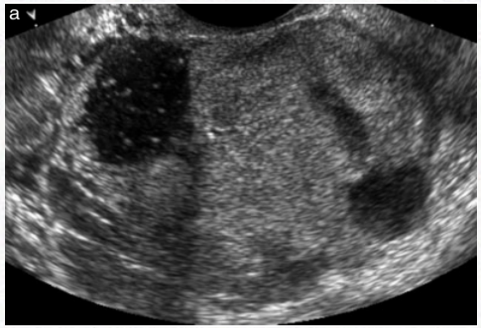
- Average age at diagnosis: 40-50 years (12-78 years)
- Clinical presentation
  - Asymptomatic (40%)
  - Pelvic mass, abdominal distension, abdominal pain
  - Ascites(rarely contained malignant cell)(17-33%)
  - Thyrotoxicosis(5-8%)
- Tumor Characteristics
  - Size 8.65 ± 4.38 cm
  - 8.27% bilateral ovaries

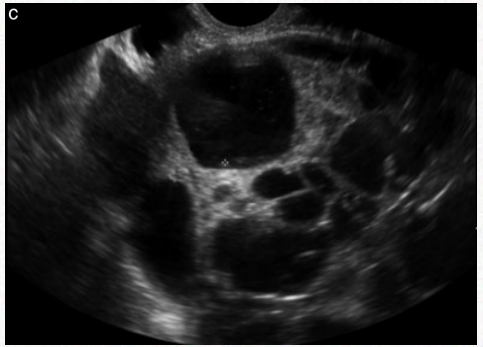
Li et al. BMC Cancer (2021) 21:383,Front. Oncol. 11:645156, Leuștean et al. Thyroid Research (2022) 15:14

Pelvic Ultrasound

- Solid tumor with irregular internal echogenicity and cystic spaces containing anechogenic cyst fluid
- Cystic tumor with ‘Struma pearl’
- Multilocular solid/ cystic tumor
- Occasionally with ascites
- Color doppler: variable well-vascularized solid component







Ultrasound Obstet Gynecol 2008; 32: 210–219

CT and MRI

- CT findings in struma ovarii
  - Non-contrast: high attenuation area and calcification
- MR findings in struma ovarii
  - The punctuate foci of high intensity on T1-weighted images (solid component)
  - Areas of very low intensity on T2- weighted images (solid component)
  - Cystic components show a variety of signal intensities on MR images

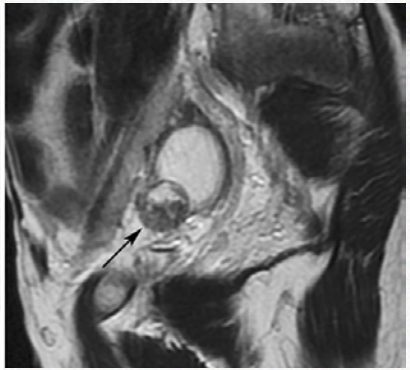


Fig. 2. 61-Year old woman with struma ovarii of the right ovary. Sagittal T2-weighted image shows a solid and cystic lesion; the solid component appears as a mass. The solid component shows heterogeneous signal intensities from high to intermediate intensity on T2-weighted image (*arrow*).

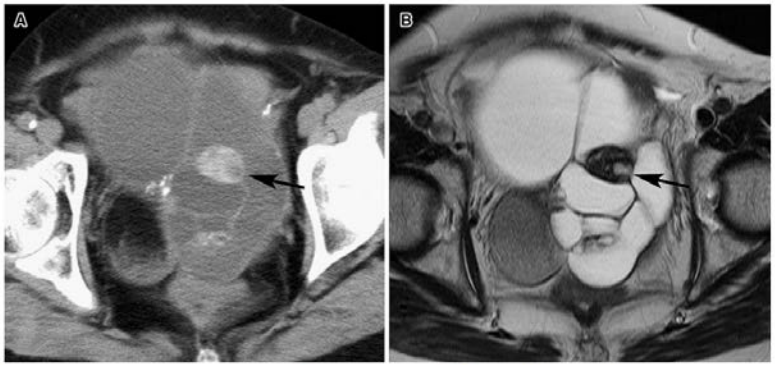


Fig. 5. 78-Year old woman with struma ovarii of the right ovary. **A** Axial T2 weighted image shows a multicystic lesion with a solid component like a mass, which shows low intensity (*arrow*). **B** The solid component shows high attenuation on non-contrast CT image (*arrow*).

Diagnosis

- Most case are diagnosis after surgical remove ovarian cyst or mass
- Histopathological examination
  - Typical papillary thyroid carcinoma: 50%
  - Follicular thyroid carcinoma: 26%
  - Follicular variant of papillary thyroid carcinoma: 18%
  - Papillary and follicular mixed thyroid carcinoma: 3%
  - Anaplastic thyroid carcinoma: 1%
  - Medullary carcinoma: 1%
- CA 125 positive: 51%
  - Sensitivity 71-82%, specificity 75-94%

Histological finding

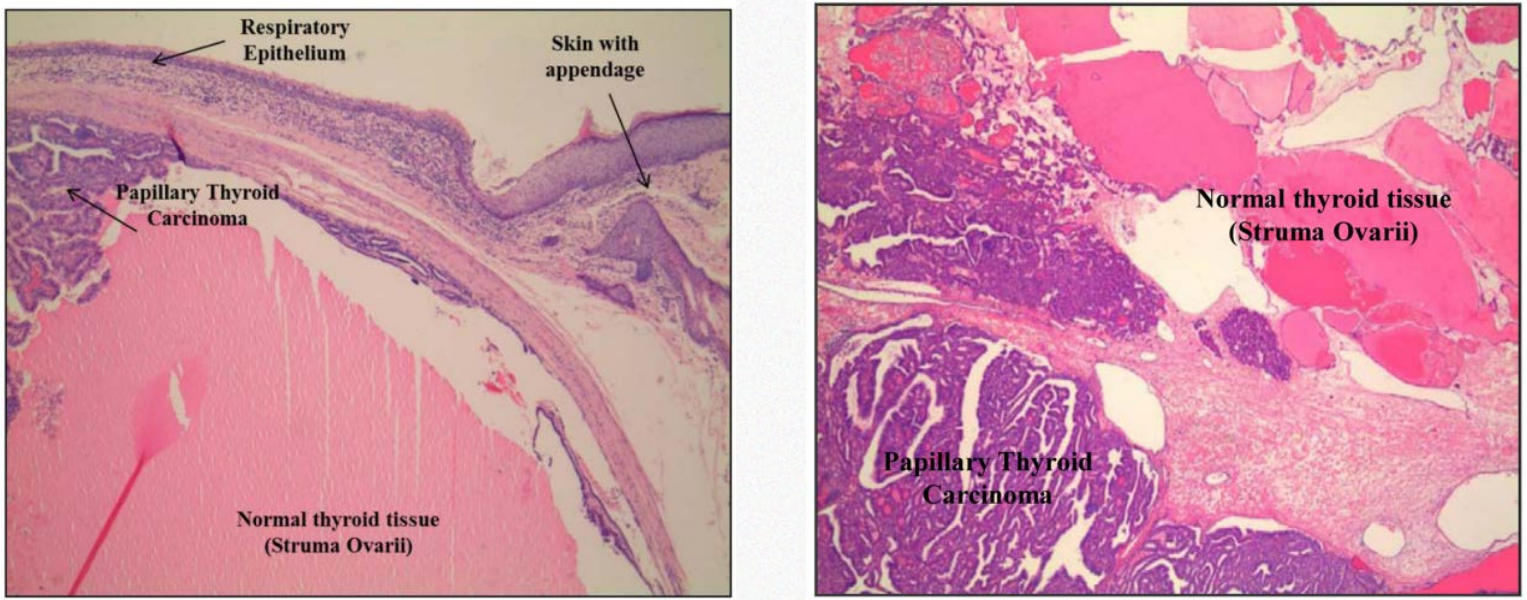


Figure 1. Histology slides at 40× magnification. Top slide shows classical type, PTC arising from struma ovarii. Bottom slide shows PTC arising from struma ovarii and benign cystic teratoma, featuring respiratory epithelium and skin with hair follicles.

J Clin Endocrinol Metab, December 2013, 98(12):4599 – 4605

Coexistence of Malignant Strumarii and Cervical PTC

Table 1. Differences in Clinical Characteristics, Dissemination Patterns, and Prognosis Between Synchronous Tumors and Advanced Metastatic Disease

Characteristics	Synchronous Tumors	Metastasis From Malignant Struma Ovarii	Metastasis From Primary Thyroid Tumor
Dissemination pattern	Negative for disseminated metastasis	Pelvic/abdominal nodes, ascites, liver metastasis	Cervical/mediastinal nodes, lung and bone metastasis
PET-CT ^{131}I whole body scan			
Stimulated Tg	Low	Elevated	Elevated
Ovarian tumor	Unilateral	Unilateral	Bilateral
Teratomatous features (eg, thyroid tissue, dermoid cyst)	Present	Present	Absent
Prognosis	Favorable	Adverse	Adverse

- Routes of metastasis
  - Regional lymphatic to pelvic and paraaortic lymph nodes
  - Direct spread to the omentum and peritoneal cavity
  - Hematological dissemination to the bone, lung, liver, brain

J Clin Endocrinol Metab, December 2013, 98(12):4599 – 4605

Immunohistochemistry

- Thyroid tissue
  - Thyroglobulin
  - TTF-1
- Malignant thyroid tissue
  - HBME-1(Hector Battifora mesothelial(cell) 1)
  - Galectin-3
  - Cytokeratin -19(CK-19)
  - CD-56

Li et al. BMC Cancer (2021) 21:383,Nature Clinical practice EndoMet, August 2008, 4(8):469–72

Tumor Profiling

- BRAF (v-raf murine sarcoma viral oncogene homolog B1)
  - Common in papillary thyroid carcinoma
  - 2/ 3 of malignant struma ovarii with papillary feature
- RAS(codon61: HRAS,NRAS), RET/ PTC
  - Follicular variant papillary thyroid carcinoma
- TERT: no report of malignant struma ovarii

Endocrine. 2021;71(1): 216-224, Nature Clinical practice EndoMet, August 2008, 4(8):469–72

Treatment

- The management of malignant struma ovarii is still controversial
- Pelvic surgery
  - Conservative, Unilateral oophorectomy
  - Complete staging
- Adjuvant treatment
  - Total thyroidectomy with radioactive iodine ablation
  - Thyroxine
- Follow up
  - Thyroglobulin(Tg)
  - Whole body I-131 scan(WBS)

J Clin Endocrinol Metab, December 2013, 98(12):4599 – 4605,Nature Clinical practice EndoMet, August 2008, 4(8):469–72, Front. Oncol. 11:645156

Treatment

Surgical management

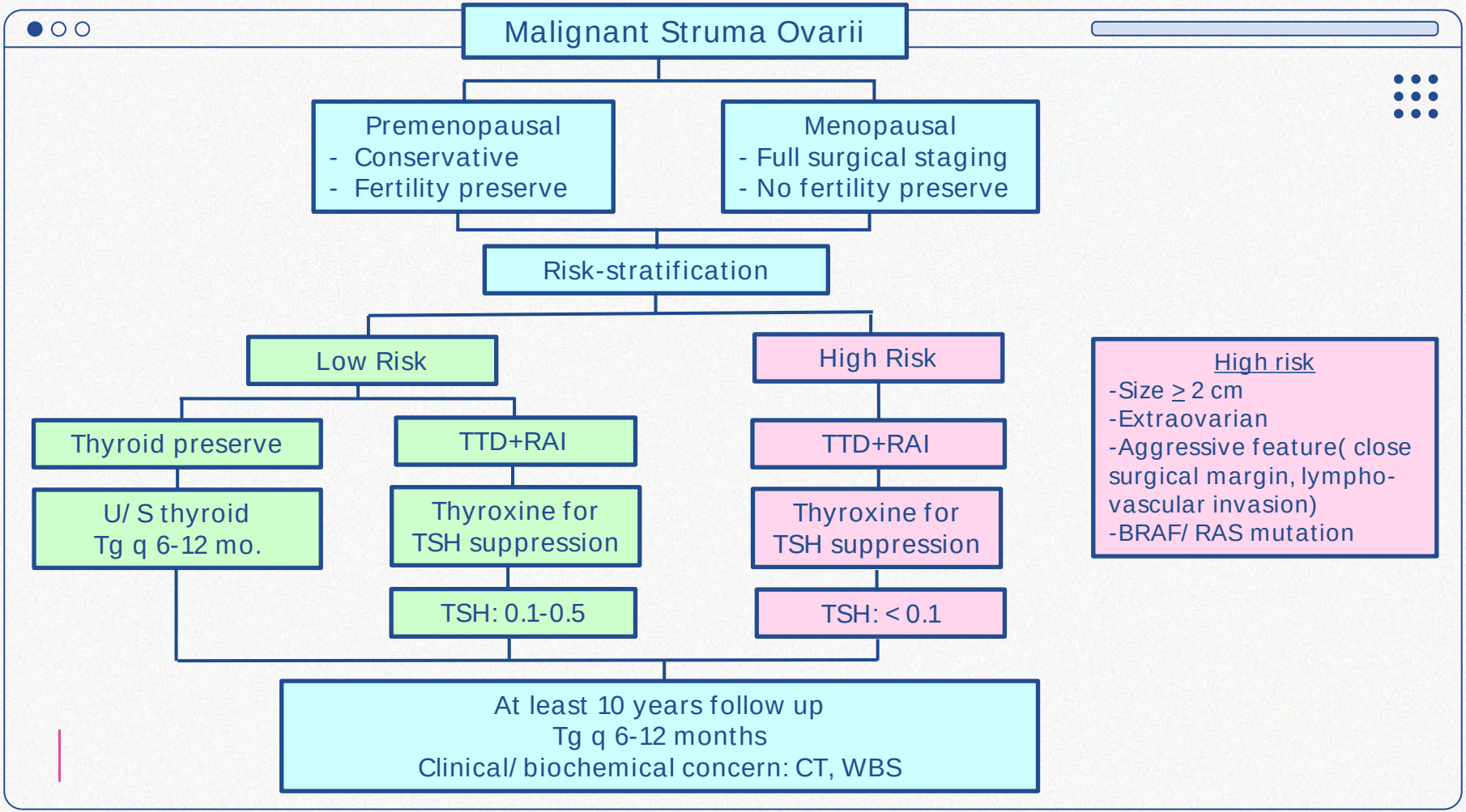
- Unilateral oophorectomy or simple tumor resection
  - Desired to retain fertility
  - After successful fertility, regular follow-up
- Conservative surgery
  - Total abdominal hysterectomy with bilateral salpingo-oophorectomy
- Complete surgical staging
  - TAH with BSO, include cytological abdominal washing, pelvic and para-aortic lymph node sampling, and omentectomy

Treatment

Adjuvant treatment

- Total thyroidectomy and eventually undergo I-131 ablation
  - High risk
    - Tumor ≥ 2 (>1) cm
    - Extra-ovarian extension
    - Aggressive histological feature: close surgical margin, lympho-vascular invasion
    - BRAF, RAS mutation
- Thyroxine(reduce TSH secretion)
  - TSH goal
    - Low risk: 0.1-0.5 mIU/l
    - High risk: < 0.1 mIU/l

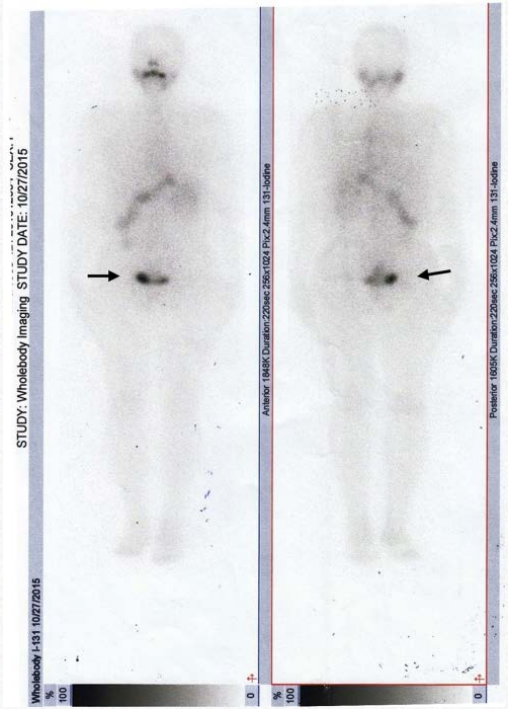
Archiev of Gynecology abd Obstetric 303, 863-870(2021), Nature Clinical practice EndoMet, August 2008, 4(8):469–72, Li et al. BMC Cancer (2021) 21:383



Adapt from J Clin Endocrinol Metab, 2013, 98(12):4599 – 4605,Nature Clinical practice EndoMet, 2008, 4(8):469–72, Archives of Gynecology and Obstetrics(2021)303:863-70.

Follow Up

- Not well established strategy
- Prophylactic total thyroidectomy and RAI therapy
  - Tg, Tg antibody q 6-12 months
  - Pelvic MRI q 1-2 years
  - Other imaging(WBS, ultrasound, CT)
    - Locoregional or distance recurrence
    - Increase Tg, Tg antibody
- Preserve thyroid
  - Neck ultrasound
  - Thyroglobulin level
- Follow up at least 10 (20) years



Leuștean et al. Thyroid Research (2022) 15:14, Front. Oncol. 11:645156,Li et al. BMC Cancer (2021) 21:383

Prognosis

- Median recurrence duration: 14 years (95%CI , 9.5-18.5)
- The 5-year and 10-year RFS rate were 27.1 and 35.2%
- Mean OS rate: 37 years(95%CI , 33.4-40.5)
- The 5-year, 10-year and 20-year OF rate were 95.3, 88.7 and 88.7%

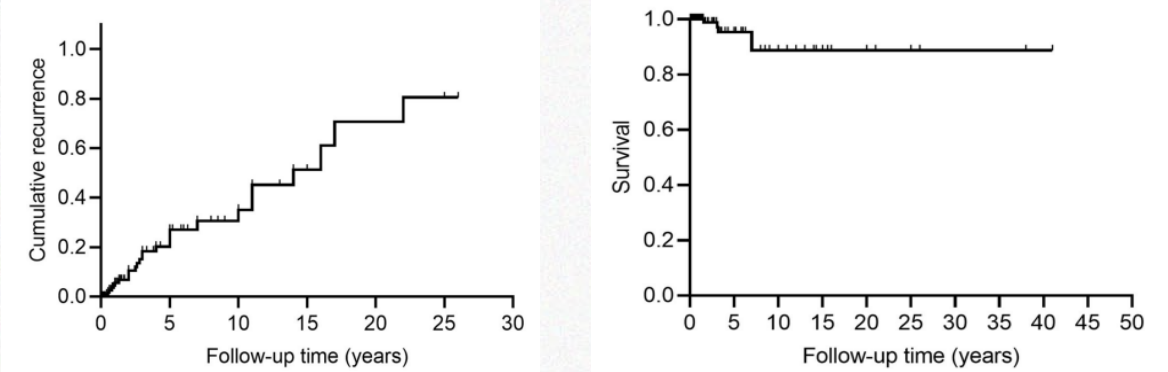


Fig. 4 Survival curves in patients with MSO confined to ovary. **a** Overall survival of all patients enrolled in this study. **b** The cumulative recurrent rate was 27.1% at 5-year and 35.2% at 10-year in our study ($n = 124$)

Tumor Recurrence

- Recurrence rate after initial surgical treatment: 35%
- Median time to recurrence: 4 years
  - All recurrence: no additional surgical or radioiodine treatment after pelvic surgery
- Site of recurrence: adjacent pelvic structure, contralateral ovary, lymph node, lung, bone, brain, liver
- Treatment for recurrent disease: radioactive iodine, external beam radiation

Nature Clinical practice EndoMet, August 2008, 4(8):469–72

Thank You