

Left Adnexal Struma Ovarii with Papillary Thyroid Carcinoma: A Case Report

A A George, R L Hutton, J S Saenger, E P Fillman

Citation

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Abstract

Struma ovarii are rarely encountered ovarian teratomas composed entirely or predominantly of thyroid tissue. Malignant transformation of these tumors is quite uncommon with less than 5% of them demonstrating evidence of malignancy. The presence of thyroid carcinoma in struma ovarii is even rarer. This article reports a case of struma ovarii with papillary thyroid carcinoma (PTC) in a 56 year old female, diagnosed in our department and confirmed by both intra- and extra-departmental consultation. Due to the low incidence of this tumor, we believe this case is noteworthy and will help in their diagnosis and management in the future.

DISCLOSURE

The above authors are employees of the US Federal Government and the United States Army. The opinions or assertions contained herein are the private views of the authors and are not to be construed as official or reflecting the views of San Antonio Military Medical Center, Brooke Army Medical Center, the Department of Defense, or United States government.

INTRODUCTION

Teratomas account for 15%-20% of ovarian tumors¹. Ovarian teratomas that contain thyroid tissue as their primary cellular component are known as struma ovarii. They are quite rare tumors, with a reported incidence being 3%-5% of ovarian teratomas and roughly 1% of all ovarian neoplasms entirely^{2,3}. The development of malignant changes in struma ovarii has an even lower incidence, occurring in about 5% of the tumors⁴. With such a low frequency, malignant struma ovarii are interesting both from a clinical and histopathological standpoint; however, because of their scarcity the diagnosis and management are rather unclearly defined, resulting in the need for additional case reports to more consistently characterize the tumors histologically and provide additional data for clinicians regarding clinical presentation, management and associated patient outcomes.

CASE

A 56 year old G2P2002 female with two term vaginal

deliveries presented to the Emergency Room with dull aching pain in the left lower quadrant, associated with intermittent 9/10 sharp pain in the same region. She denied nausea, vomiting, diarrhea or constipation. She also denied night sweats, dysuria or urinary symptoms, and admitted to normal appetite with no unintentional weight loss or gain. She reported having the same type of pain about 3 months prior to presentation that resolved with over-the-counter ibuprofen. She attempted the same self-medication with little relief this time. Her past medical history is significant for asthma which is well-controlled, and essential hypertension. Last menstrual period was 2 months prior. She reported a history of regular menstrual cycles with 5-7 days of menses up until 12 months prior at which time her cycles started to space out to roughly every 1-2 months. She denied a history of abnormal Paps or sexually transmitted diseases, and denied being sexually active. Surgical history was limited to a tonsillectomy. Family history was unremarkable. She denied alcohol, tobacco or recreational drug use. Current medications include Flonase, Advair, Singulair, and a daily multivitamin.

Physical examination:

Vital signs: Blood pressure 153/93, heart rate 97, temperature 98°F, O₂ 100% on room air.

The abdomen was soft and mildly distended with a palpable pelvic mass extending up to the level of the umbilicus, with no rebound tenderness or guarding. Pelvic exam further demonstrated the mass lesion with minimal tenderness to

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palpation. Rectal exam was unremarkable with negative stool guaiac.

The remainder of the physical exam was unremarkable.

Labs and radiographic imaging:

CBC and electrolyte panel were unremarkable. Urine HCG was negative. Urinalysis showed moderate leukocyte esterase and 12 WBC/HPF. CA 19-9 and CEA were both elevated at 128 U/mL (normal 0-35 U/mL) and 3.5ng/mL (normal 0-3.4ng/mL) respectively. CA-125 and AFP were within normal limits at 25.7 U/mL and 6.78 ng/mL respectively. Thyroid function tests were not drawn.

Pelvic ultrasound showed a complex left adnexal mass (11.1 x 9.3 x 8.4 cm) with no sonographic evidence of ovarian torsion, and a 1.8 x 1.7 x 1.7 cm uterine fibroid. The right ovary was not visualized.

Hospital course:

The patient was initially evaluated by Obstetrics & Gynecology and discharged with Ciprofloxacin and adequate pain control, and referred for CT scan at a later date with outpatient follow up. She returned two days later with worsening pain and a CT scan was performed which showed a large (15.1 x 14.2 x 8.1 cm) heterogeneously enhancing, mixed solid and cystic pelvic mass with free fluid concerning for malignant ascites (Image 1).

Image 1

Mixed solid and cystic pelvic mass with free fluid.



She underwent an exploratory laparotomy, total abdominal hysterectomy/bilateral salpingo-oophorectomy (TAH/BSO) with uneventful post-operative course. The patient is

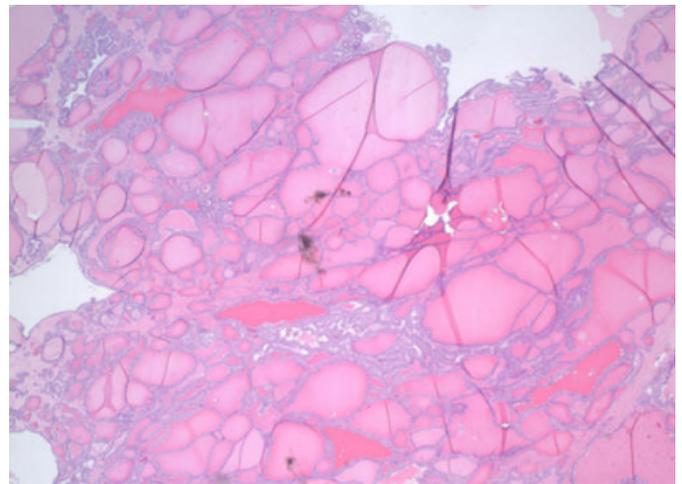
currently almost 1 year post-surgery and is thus far without evidence of recurrence or metastases.

Histopathologic examination:

On gross examination the mass was a 388 gram specimen, measuring 14.5 x 11.5 x 8.5 cm with a smooth surface and a multi-cystic red-brown to yellow hemorrhagic cut surface. An intra-operative consultation was obtained and a diagnosis of cystic teratoma was rendered based on limited sampling, with final classification deferred to permanent sections. Figure 1 shows an H&E section of the mass demonstrating thyroid tissue consistent with a struma ovarii.

Figure 1

4 x magnification showing mature teratoma composed predominantly of thyroid tissue (struma ovarii), with increased cellularity in the center, Hematoxylin & Eosin (H&E).



Figures 2 & 3 show features of papillary thyroid carcinoma, to include cellular elongation, nuclear crowding, and optically clear or grooved nuclei. Figure 4 demonstrates an epidermal inclusion cyst-like area of keratinization within the teratoma, surrounded by unremarkable ovarian stroma.

Figure 2

20 x magnification showing nuclear crowding and elongation and occasional nuclear grooving, which are features of papillary thyroid carcinoma, H&E.

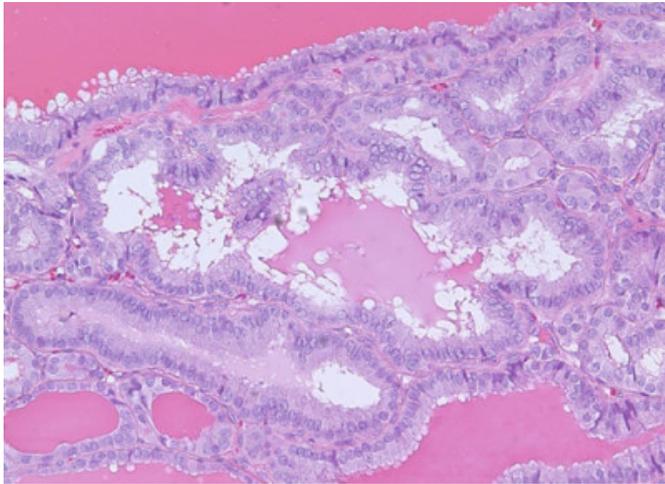


Figure 4

2x magnification showing Epidermal Inclusion Cyst-like area within the teratoma with unremarkable ovarian stroma, H&E.

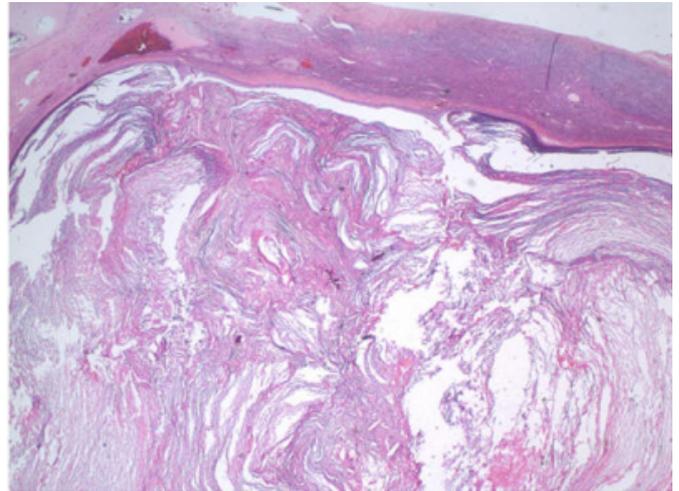
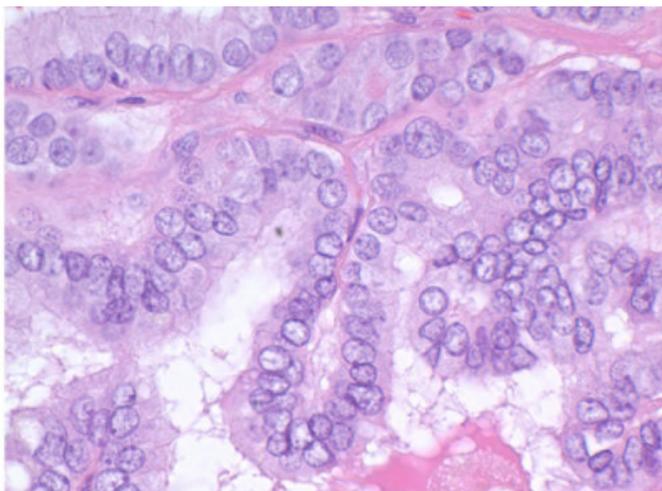


Figure 3

60 x magnification showing cleared, crowded grooved nuclei which are diagnostic of papillary thyroid carcinoma, H&E.



Histologically the mass was determined to be a struma ovarii with papillary thyroid carcinoma, featuring classical, follicular, and tall-cell patterns. Capsular or vascular invasion was not identified within the papillary component; neither was extra-ovarian spread.

DISCUSSION

Struma ovarii are usually found incidentally on routine gynecologic exams or typically present due to abdominal pain or pelvic mass. They are usually diagnosed post-operatively, due both to their rarity and the lack of specific clinical presentation. They tend to present around the 5th decade of life, and are typically unilateral^{5,6}. Like the thyroid gland, strumal tissue can develop carcinomas and can also be hormonally active; however, they rarely produce hormonal levels that are elevated enough to cause symptoms^{5,7}. Several types of thyroid carcinoma may arise in struma ovarii, with papillary thyroid carcinoma being the most common type (~51%, 53 out of 102 struma ovarii), according to a recent summary of the literature on thyroid malignancies of struma ovarii reported between 1924-2008⁸. There are currently no standardized diagnostic criteria for thyroid carcinoma arising in a struma ovarii other than those in use for primary thyroid carcinoma³. Our patient's struma ovarii contained papillary thyroid carcinoma which histologically included a papillary architecture with crowded, round or oval-shaped nuclei that are optically cleared and enlarged with occasional nuclear grooves³ (Figures 2 & 3).

Due to the rarity of these tumors, the prognosis of papillary thyroid carcinoma of struma ovarii is not entirely certain;

however, a handful of reports demonstrate a relatively good prognosis with a 5- and 25- year survival rate of 92% and 79% respectively, which is comparable to that of papillary thyroid carcinoma arising in the thyroid gland^{3,8}. The treatment of these tumors is usually surgical, involving complete resection with staging, but the exact management (TAH/BSO vs. oophorectomy vs. strumectomy or other procedures) has yet to be determined. Post-operative management appears to be even more unclear as some authors recommend they be treated like their thyroid equivalent (with subsequent thyroidectomy and 131I therapy)⁵ while others seem to suggest that these adjuncts be utilized in cases with residual disease after surgery, recurrence, and/or metastases¹.

Our patient underwent complete resection with staging, however did not undergo subsequent thyroidectomy or receive adjuvant 131I therapy. The patient is currently almost 1 year post-procedure and appears to be without evidence of metastasis, tumor recurrence, or return of symptoms at this time. This time-frame however may be too short to determine if the current management has been successful or if recurrence is imminent, as the average time to detection of recurrence has been found to be four years¹. Makani et al.'s review of 39 cases through 2004 found recurrence in 15% and metastasis in 23%⁶.

It will be interesting to note if recurrence does occur as patients with papillary thyroid carcinoma of struma ovarii have been found to have a mean survival of 8 years and a median survival of 3.5 years, with death occurring anywhere from 2 weeks – 21.5 years after treatment³. If this patient's tumor was definitively treated, close follow up is still warranted as recurrence has been found to be more likely in tumors > 10cm⁸.

Unfortunately pre-operative thyroid function tests were not performed to determine if the tumor was hormonally active; however, surveillance with serum thyroid studies will still need to be a component of future follow up exams. Therefore obtaining serum thyroid studies at initial evaluation is an important factor to consider in a patient whom this entity is suspected.

Our case clearly demonstrates an instance of papillary thyroid carcinoma arising in a struma ovarii. The clinical presentation and histologic features observed fit the typical patterns of the tumor as described throughout the literature. The management is consistent with standard treatment historically, while supplemental treatment modalities were not administered. The patient's outcome thus far has been successful but cannot be definitively determined at this time. Undoubtedly, additional research on this particular tumor type is needed to further define it histologically; however, due to its infrequent occurrence more extensive research is difficult. Therefore it is our hope that this article will contribute to the limited available data regarding these tumors and will help with both their clinical and histologic diagnosis, as well as their management.

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Author Information

Alan A. George, DO

From the Department of Pathology and Area Laboratory Services, San Antonio Military Medical Center
San Antonio, Texas, USA
alan.a.george@us.army.mil

Robert; L. Hutton, MD

From the Department of Pathology and Area Laboratory Services, San Antonio Military Medical Center
San Antonio, Texas, USA

Jeffrey S. Saenger, MD

From the Department of Pathology and Area Laboratory Services, San Antonio Military Medical Center
San Antonio, Texas, USA

Eric P. Fillman, MD

From the Department of Pathology and Area Laboratory Services, San Antonio Military Medical Center
San Antonio, Texas, USA