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Malignant struma ovarii found in a teratoma of an undescended ovary: A case report

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Abstract
Teratomas are the most common neoplasm of the ovary but transformation of a teratoma is very rare. Furthermore, transformation to papillary thyroid carcinoma is even more uncommon. Mullerian anomalies and undescended/maldescended ovaries are also rare congenital conditions that have a low estimated prevalence. We present an extremely rare case of a 33-year-old female presenting with a struma ovarii with malignant transformation to papillary thyroid carcinoma within a teratoma arising from an undescended ovary and fallopian tube with a concomitant Mullerian anomaly. While there are several cases of similar pathologies reported separately, there has not been a similar case to our patient. Undescended ovary and fallopian tube, unicornate uterus, stuma ovarii, and malignant transformation of a struma ovarii and mature teratoma are all extremely rare conditions that have not been reported to co-occur within a single patient in the current literature.

Keywords: Ovarian teratoma, struma ovarii, Mullerian anomalies, unicornate uterus, undescended/maldescended ovary

Introduction
Teratomas are the most common neoplasm of the ovary, accounting for 15-20% of ovarian tumors [1]. Among teratomas, there are several subtypes, including the rarer struma ovarii, accounting for less than 5% of teratomas [1-3]. Malignant transformation of a teratoma is very rare, with a rate of less than 2%. When a malignant transformation occurs, the most common transformation is to squamous cell carcinoma, although among struma ovarii there can also be transformation to papillary thyroid carcinoma [1-5]. We present an extremely rare case of a 33-year old woman presenting with a struma ovarii with malignant transformation to papillary thyroid carcinoma within a teratoma arising from an undescended ovary and fallopian tube with a concomitant Mullerian abnormality.

Case Report
Patient is a 33-year-old female with obstetric history of two cesarean sections and one miscarriage. Medical history includes congenital solitary right kidney and recurrent urinary tract infections since her childhood. She originally presented to our institution with one day of persistent gross hematuria with dysuria and lower abdominal pain described as a constant aching and cramping with no radiation. Patient had recently completed a seven-day course of Augmentin for previous urinary tract infection. She had no associated symptoms such as fever, nausea and vomiting, and shortness of breath. She has no travel history or recent sick contacts. Urine pregnancy test at the time of admission was negative. During her second cesarean seven months prior, a unicornate uterus and presumed absence of the left fallopian tube and ovary were noted. A tubal ligation after her second cesarean is her only surgical history.

She originally presented to the emergency room with vitals notable for hypertension and tachycardia. Complete blood count revealed a white blood cell count of 17.3 (x10(3)/uL) with a neutrophilic predominance. Urinalysis showed red, turbid urine with 3+ protein, 3+ blood, 3+ leukocyte esterase, and 2+ bacteria. A computed tomography (CT) scan of the abdomen and pelvis without intravenous (IV) contrast revealed an absent left kidney, malrotation of the gastrointestinal tract, and a 6.9cm x 5.1cm calcified fatty dense lesion in the left iliac fossa. The mass was interpreted as potentially being a dermoid cyst. After treatment with ceftriaxone, the patient became stable and was discharged home with a referral to surgical oncology.

At an outpatient visit, it was determined her mass was most likely a teratoma, and surgical intervention via diagnostic laparoscopy and robotic assisted resection of the left adnexa was planned.

On the day of surgery, a diagnostic laparoscopy revealed the left mass to be a left ovary and fallopian tube which was malpositioned in the left colic gutter (as seen in figure.1). The left round ligament was inserted into the inferior aspect of left ovary, and the left fallopian tube and ovary were separate from the uterus. Grossly, the left ovary appeared to be enlarged to 6cm with cystic and solid mass components. Appearance was consistent with a mature teratoma and was excised robotically with intraoperative consent of the patient's spouse. After excision, fat and hair were grossly visible within the sample. Cystoscopy during this operation was unremarkable, revealing no obvious sources of hematuria. The excised sample was sent for pathological examination, and no complications arose during or immediately after the surgery.

Pathology examination revealed a 6.5 cm x 4.0 cm x 3.0 cm focally disrupted ovarian mass. The majority of the teratoma consisted of tan, friable material admixed with hair and cyst containing serosanguinous fluid. Other components included a 0.3 cm x 0.2 cm plaque-like gray discoloration, a 2.5 cm x 2.0 cm x 1.5 cm red-purple cyst containing minimal dark fluid, and a 2.8 cm x 2.2 cm focally calcified fatty nodule. Within the ovary, a 1mm focus of papillary carcinoma was found arising from the struma ovarii component of the excised mature teratoma, completely encapsulated by the ovary and approximately 2mm away from the nearest ovarian surface. The excised fallopian tube was admixed with fibro-membranous tissue, hair, and tan friable debris with no evidence of malignancy. The samples were negative for endometriosis. Histology is shown in figures 2-4 which on Figure 4 in 400x magnification you can see nuclear chromatin clearing, nuclear overlapping, and occasional nuclear grooves in the tumor cell population, all of which can be seen in the context of papillary thyroid carcinoma (PTC).

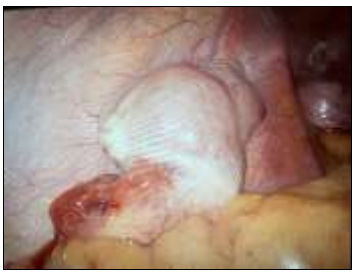

Fig 1: Left ovary and fallopian tube in left Paracolic gutter.

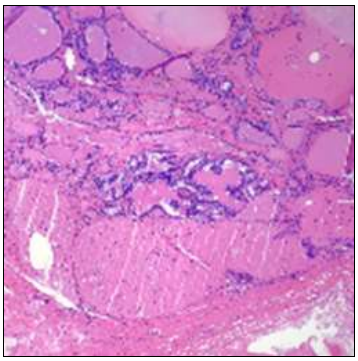

Fig 2: PTC in struma stained by hematoxylin and eosin, 100x

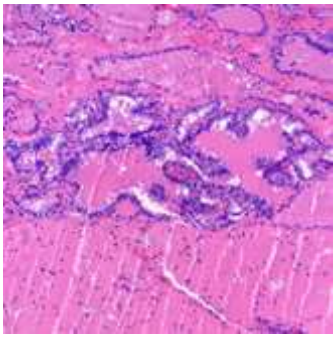

Fig 3: PTC in struma stained by hematoxylin and eosin, 200x

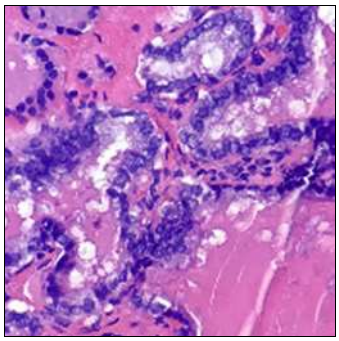

Fig 4: PTC in struma stained by hematoxylin and eosin, 400x

Discussions

Cystic teratomas are the most common neoplasm of the ovary and the most common type of germ cell tumor [2, 5]. Annual incidence rate is 1.2 to 14.2 cases per 100,000, accounting for 15-20% of ovarian tumors.[1,5] Histologically, they are divided into monodermal, immature, and mature teratomas ("dermoid cysts") [5]. Of the subtypes, the mature cystic teratoma is the most common, occurring in 20% of teratoma cases on adults and 50% of teratoma cases in children [5]. In adults, they are most common in the second to third decades of life [5]. They are typically benign and are the most common benign tumor observed in women under 45 years of age [2, 5]. Cystic teratomas are usually asymptomatic, though can have minimal symptoms or can induce ovarian torsion or compressive symptoms depending on size and location [5]. Benign cystic teratomas have an excellent prognosis after surgery, with some recurrence risk within 2-10 years after surgical intervention [5].

Struma ovarii is a rare type of teratoma which is composed of at least 50% of thyroid tissue [1-4]. While 20% of teratomas contain thyroid tissue, less than 5% qualify as struma ovarii [1-3]. A struma ovarii can fall into either the monodermal teratoma or the mature cystic teratoma subtype, depending on the percentage of thyroid tissue. Struma ovarii accounts for 2.7% of all ovarian teratomas, and most commonly occurs in pre-menopausal women between the ages of 30 and 50 [4]. Typically presenting as a unilateral adnexal mass, they are asymptomatic, with 41.2% of them being discovered incidentally during routine imaging [2, 4]. 5-8% of occurrences are associated with clinical hyperthyroidism [1, 4]. On ultrasound, the most specific characteristic is the struma pearl [2]. Among struma ovarii, the majority are benign, with less than 5% undergoing malignant transformation which most commonly arises within the fifth decade of life [1, 2, 4].

Rarely, mature cystic teratomas can undergo malignant transformation [3, 5]. Malignant transformation has been

estimated to have occurred in 0.17-2% of cases, with patients between 40 and 60 years old having a higher incidence [3, 5]. A teratoma size of greater than 10 cm has been suggested to confer an increased risk of malignant transformation [5]. When transformation occurs, transformation to squamous cell carcinoma is the most common, occurring in 50% of cases [3, 5]. Among struma ovarii, papillary thyroid carcinoma or follicular carcinoma may arise [1, 2, 4]. Metastasis are uncommon, with metastatic spread to the peritoneum, mesentery, and omentum being more typical [1, 2].

Cystic teratomas that have undergone malignant transformation having varying prognosis dependent on factors such as staging and vascular invasion [5]. 5-year survival ratings for stages I-IV have been estimated at 50%, 25%, 12%, and 0% respectively [5]. Prognosis of malignant struma ovarii is typically good, with a 25-year survival of 84%, though the exact percentage is contentious [1, 3]. Some studies contain reports of 44% of patients living with no recurrence, 22% living with persistent disease, and 15% of patients dying from metastatic disease [2].

Mullerian abnormalities anomalies have an estimated prevalence of 1% in the general population and 25% among women with infertility [6]. Undescended/maldescended ovaries are a rare congenital condition associated with urogenital malformations including unicornate uterus and renal agenesis [7, 8]. The estimated prevalence of maldescended ovaries are between 0.3 and 2% [7]. The frequency of unicornate uterus is reported to be between 3 and 13%, and are also associated with renal and urinary tract anomalies [6, 8-10]. Up to 42% of patients with unicorunate uterus have concomitant maldescent of the ovary and fallopian tube, whereas the incidence drops to 20% in those with normal uteri [6-8]. Mullerian anomalies such as these are associated with an increase in obstetrical complications such as miscarriages, intrauterine growth retardation, and ectopic pregnancy [8]. Both undescended/maldescended ovaries and fallopian tubes and Mullerian abnormalities are most typically discovered during infertility work-up [6, 11].

Our case was especially unusual given the constellation of rarities than arose. While there are several cases of some of her pathologies reported separately, there has not been a similar case to our patient. Undescended ovary and fallopian tube, unicornate uterus, stuma ovarii, and malignant transformation of a struma ovarii and mature teratoma are all extremely rare conditions that have not been reported to co-occur within a single patient in the current literature. Despite all these conditions, the patient's fertility was not impacted, which deviates from the usual presentation. She was also relatively asymptomatic despite these several concomitant presentations, with incidental discovery of the conditions through urinary tract infection work-up. While she is of the age range for teratoma occurrence, she falls outside the typical age range for malignant transformation.

Conflict of Interest
Not available

Financial Support
Not available

References

1. Salman WD, Singh M, Twaij Z. A case of papillary thyroid carcinoma in struma ovarii and review of the literature. Patholog Res Int. Aug 2 2010;2010:352476. doi:10.4061/2010/352476
2. Wolff EF, Hughes M, Merino MJ, *et al*. Expression of benign and malignant thyroid tissue in ovarian teratomas and the importance of multimodal management as illustrated by a BRAF-positive follicular variant of papillary thyroid cancer. Thyroid. Sep 2010;20(9):981-7. doi:10.1089/thy.2009.0458
3. Pineyro MM, Pereda J, Schou P, *et al*. Papillary Thyroid Microcarcinoma Arising Within a Mature Ovarian Teratoma: Case Report and Review of the Literature. Clin Med Insights Endocrinol Diabetes. 2017;10:1179551417712521. doi:10.1177/1179551417712521
4. Rockson O, Kora C, Ramdani A, *et al*. Struma ovarii: two case reports of a rare teratoma of the ovary. J Surg Case Rep. Dec 2020;2020(12):rjaa493. doi:10.1093/jscr/rjaa493
5. Ahmed A, Lotfollahzadeh S. Cystic Teratoma. Stat Pearls; c2022.
6. Allen JW, Cardall S, Kittijarukhajorn M, Siegel CL. Incidence of ovarian maldescent in women with mullerian duct anomalies: evaluation by MRI. AJR Am J Roentgenol. Apr 2012;198(4):W381-5. doi:10.2214/AJR.11.6595
7. Sinonquel P, Bontinck J, Stevens M. Undescended ovary and fallopian tube presenting as appendiceal mucocele. Facts Views Vis Obgyn. Mar 2018;10(1):47-50.
8. Ombelet W, Grieten M, DeNeubourg P, *et al*. Undescended ovary and unicornuate uterus: Simplified diagnosis by the use of clomiphene citrate ovarian stimulation and magnetic resonance imaging (MRI). Hum Reprod. Apr 2003;18(4):858-62. doi:10.1093/humrep/deg191
9. Grimbizis GF, Camus M, Tarlatzis BC, Bontis JN, Devroey P. Clinical implications of uterine malformations and hysteroscopic treatment results. Hum Reprod Update. Mar-Apr 2001;7(2):161-74. doi:10.1093/humupd/7.2.161
10. Oppelt P, von Have M, Paulsen M, *et al*. Female genital malformations and their associated abnormalities. Fertil Steril. Feb 2007;87(2):335-42. doi:10.1016/j.fertnstert.2006.07.1501
11. Gorgen H, Api M, Delikara N. Undescended fallopian tubes and ovaries: A rare incidental finding during an infertility investigation work up. Acta Obstet Gynecol Scand. Apr 2002;81(4):371-4.

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