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MONODERMAL TERATOMAS OF THE OVARY: A CASE SERIES EXPLORING THE RARE STRUMA OVARII AND OVARIAN CARCINOID TUMORS

KEY WORDS: monodermal teratoma, struma ovarii, ovarian carcinoid tumor

Sonia Mary Thomas*

Assistant Professor Department Of Pathology Pushpagiri Institute Of Medical Sciences And Research Centre, Thiruvalla, Kerala, India, Pin 689101 *Corresponding Author

Lillykutty Pothen

Professor Department Of Pathology Pushpagiri Institute Of Medical Sciences And Research Centre,Thiruvalla,Kerala,India,Pin 689101

ABSTRACT

Background: Rare monodermal ovarian teratomas, including struma ovarii and ovarian carcinoid tumor, present diagnostic and management challenges due to diverse clinical features and malignant potential. **Case Series:** We present three cases (two struma ovarii, one ovarian carcinoid) encountered unusually within a two-year period with no other cases observed before or after. **Discussion:** Accurate diagnosis of monodermal teratomas, often benign but with malignant potential or hormone-related symptoms, relies on histopathology and molecular diagnostics. Advances in imaging, NGS, and minimally invasive surgery aid diagnosis and better outcomes. Management ranges from surgery to RAI and somatostatin analogs in select cases. **Conclusion:** This case series illustrates the varied presentation, pathology, diagnosis, and treatment of monodermal teratomas. Further research into molecular markers and personalized therapies is crucial for improving outcomes in malignant or recurrent cases.

INTRODUCTION

Ovarian germ cell tumors include the more common mature teratomas and rarely monodermal teratomas like struma ovarii and ovarian carcinoid. Struma ovarii, mainly composed of thyroid tissue and comprising 2-3% of ovarian teratomas, primarily affects middle-aged women. While mostly benign, 5-10% can turn malignant into thyroid carcinomas. Clinical presentation varies complicating diagnosis and requiring imaging/biochemical tests.[1, 2] Ovarian carcinoid tumors, are extremely rare comprising 0.1% of ovarian tumors. Though often benign, they can metastasize and rarely cause carcinoid syndrome (flushing, diarrhea, bronchospasm).

Their rarity makes diagnosis challenging, often incidental on imaging or surgery for other gynecological issues. Preoperative identification relies on ultrasonography and MRI, with histopathology confirming the diagnosis.[3, 4, 5]. Treatment typically involves surgical resection, with additional therapies needed for malignant cases. Regular follow-up is essential due to recurrence risk [6].

This case series details three rare monodermal ovarian teratomas (one struma ovarii, two carcinoids) clustered within two years which clustered over a two year period with no instances prior or after.

This report aims to provide insights into their clinical presentation, pathology, and management, contextualized within current literature. Additionally, it highlights recent molecular diagnostic advances with potential to improve the management of these unique ovarian tumors.

Case Presentation

Case 1 : A 34-year-old nulliparous woman with thyrotoxic symptoms (hair loss, anxiety, unexplained weight loss) was referred to OBG. Examination was unremarkable, but ultrasound showed a bulky right ovary with polycystic features. TSH was <0.005, and T4 was 9.19. Tc99m scintigraphy showed increased right pelvic activity. Right salpingo-ovariotomy and infracolic omentectomy were performed. Gross examination revealed a 7x5x3 cm multiloculated ovarian cyst with yellow fluid and three tan solid omental nodules (largest 4x4 cm) (Fig. 1).

Histopathology confirmed colloid-filled thyroid follicles in both ovary and omentum, diagnosing struma ovarii with peritoneal strumosis (Fig. 2).

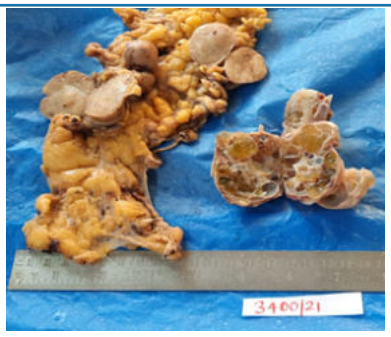

Figure 1: Right ovarian tumor with cystic and solid areas (right) and omentectomy specimen with solid nodules (left)

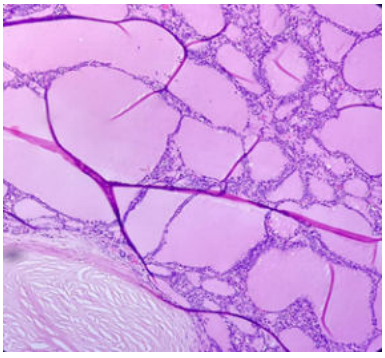

Figure 2: Microscopy of solid-cystic ovarian tumor showing colloid filled follicles lined by benign follicular cells. (H&E, 10x)

Case 2: A 57-year-old woman (P2L2, both LSCS) presented with heavy postmenopausal bleeding. Ultrasound showed a 10.8x5.6 cm lobulated cystic left adnexal lesion with internal echoes, septations, and papillary projections. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed. Grossly, the 9.5x6x5.5 cm left ovarian mass was multiloculated with pultaceous material and a 6.5x4.5x3 cm solid tan-gray area (Fig. 3).

Histopathology revealed a neoplasm with cystic areas lined by stratified squamous epithelium with sebaceous/eccrine glands, and solid areas showing nests, acini, and tubules of cells with salt-and-pepper chromatin and granular eosinophilic cytoplasm, consistent with a monodermal teratoma - Carcinoid tumor(Fig.4).

Figure 3: Hysterectomy specimen with separately sent left adnexal solid-cystic mass.

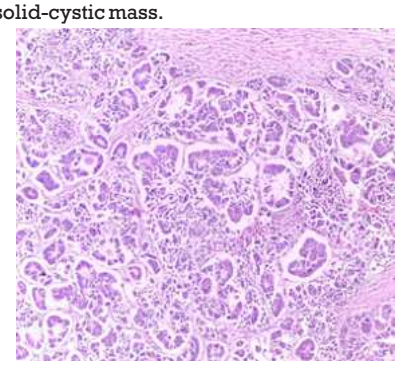

Figure 4: Microscopy showing ovary with carcinoid tumor in characteristic nesting pattern. (H&E, 10x)

Case 3: A 68-year-old female (P3L3, all FTND), with a prior hysterectomy, presented with increased urinary/bowel frequency, abdominal distension, and discomfort. Examination revealed a large (28-32 weeks), mobile cystic mass. Pelvic ultrasound showed a 22x15 cm well-defined cystic lesion with internal echoes and septations, extending to the epigastrium. MRI identified a 22x20 cm cystic left ovarian lesion. Staging laparotomy revealed a 23x21x18 cm multiloculated cystic mass with colloid-filled areas. Microscopy showed cholesterol clefts, foreign body giant cells, thyroid parenchyma with colloid follicles, and degenerative changes (hyalinization, fibrosis, calcification) (Fig. 5).

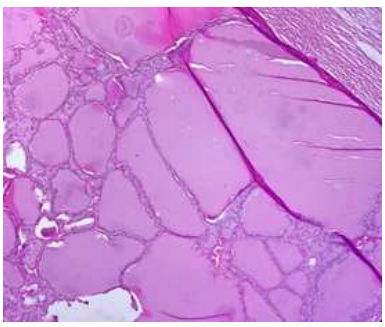

Figure 5: Microscopy of solid-cystic ovarian tumor showing benign colloid filled follicles. (H&E, 10x)

DISCUSSION

Monodermal teratomas of the ovary, including struma ovarii and ovarian carcinoid tumors, are rare germ cell tumors. Understanding their clinical presentation, diagnostic features, genetic basis, and treatment protocols is essential for optimal patient management.

Struma ovarii typically affects women aged 40-60 (mean ~45). Primary ovarian carcinoid has a wider age range (adolescence to elderly, median ~53-55). Our patients of struma ovarii presented at ages 68 and 34 years and carcinoid tumor at 57 years. [7,8]

Struma ovarii often presents with nonspecific abdominal pain, pelvic mass, distension, ascites, or hyperthyroidism; many are asymptomatic. Ovarian carcinoids typically present as unilateral adnexal masses, potentially asymptomatic or with abdominal pain, vaginal bleeding, or carcinoid syndrome. Our struma ovarii cases showed mass effect and thyrotoxicosis, while the carcinoid case had heavy vaginal bleeding.

Struma ovarii imaging shows unilateral complex adnexal masses with mixed cystic/solid components; MRI reveals low T2 and intermediate T1 signal in solid areas due to thyroid tissue/colloid, and CT may show high-density cysts from thyroglobulin. Ovarian carcinoids usually appear as well-defined solid or solid-cystic masses on ultrasound/MRI with solid component enhancement, but imaging is nonspecific. Our patients showed similar radiology findings.

Struma ovarii patients are typically euthyroid or hyperthyroid, with usually normal CA-125. Ovarian carcinoids may secrete vasoactive substances. In our series, one struma ovarii case was hyperthyroid, the other euthyroid, and the carcinoid case had no elevated vasoactive substances. [9-13]

Struma ovarii, mainly thyroid tissue, shares genetic changes with thyroid carcinomas. Malignant cases often have BRAF (35-69%), RAS (10%), RET (5–30%), and TERT promoter mutations, similar to papillary thyroid carcinoma, suggesting shared molecular mechanisms. Somatic uniparental disomy and DNA methylation of tumor suppressor genes (RECQL4, PRDM2) may also contribute. Benign struma ovarii typically lacks these mutations.

Molecular analysis of rare ovarian carcinoids is limited. Studies suggest altered neuroendocrine signaling and epigenetic changes (histone/DNA methylation) promote growth. Rare MEN1 and DAXX mutations may underlie aggressive behavior, potentially leading to recurrence/malignancy biomarkers. [14, 15]

Struma ovarii histologically shows variable colloid-filled thyroid follicles lined by cuboidal to flattened epithelium with round-oval nuclei and clear/oxyphilic cytoplasm, usually scant collagenous stroma (sometimes abundant/edematous with stromal luteinization). Benign forms lack C cells; birefringent calcium oxalate crystals may be present. Patterns include microfollicular, trabecular, solid, and papillary (no papillary thyroid carcinoma nuclei). Rare malignant transformation to papillary/follicular thyroid carcinoma shows primary thyroid cancer features.

Ovarian carcinoid histologically shows uniform polygonal cells in nests/insulae, round-oval monotonous nuclei with "salt and pepper" chromatin and inconspicuous nucleoli. Ample eosinophilic cytoplasm may contain argentaffin granules. Low mitotic activity, variable stroma. IHC positive for chromogranin A, synaptophysin, and CD56. [16, 17] Our cases showed similar findings.

Surgical resection is the main treatment for benign and malignant forms. Malignant struma ovarii often requires thyroidectomy and RAI for metastasis risk. Targeted therapies (BRAF inhibitors for BRAF-mutated, tyrosine kinase inhibitors like sorafenib for RAS-mutated) show promise in malignant struma ovarii. For recurrent ovarian carcinoids, somatostatin analogs (octreotide, lanreotide) are being explored for managing carcinoid syndrome and progression, despite established use in gastrointestinal neuroendocrine tumors. [18, 19].

Prognosis for monodermal teratomas (struma ovarii and ovarian carcinoid) is generally good. Malignant struma ovarii treatment mirrors thyroid carcinoma management, including

total thyroidectomy and RAI to reduce recurrence and manage metastases. [20,21]

Despite advances, gaps persist in understanding and managing monodermal teratomas. Larger multicenter studies are needed to explore the genetic landscape, with molecular profiling of rare subtypes, like strumal carcinoids, potentially uncovering new therapeutic targets.

CONCLUSIONS

In summary, this case series adds to the growing body of literature on monodermal teratomas by highlighting the clinical and pathological features of struma ovarii and ovarian carcinoid tumors. Early detection, accurate diagnosis, and individualized treatment remain key to achieving favorable outcomes. While surgical excision is the cornerstone of treatment, advances in molecular diagnostics and targeted therapies are reshaping the management of these rare tumors. Further research is warranted to explore the potential of these emerging therapies and improve long-term outcomes for patients with malignant or recurrent disease.

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Authors' Contributions

SMT prepared the article abstract, manuscript and gathered the case details along with research on existing information, recent advances and future prospective research directions on this topic.

LP conceptualised the idea of this case series.

All authors read and approved the final manuscript.

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