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Case report

A rare sweat gland tumor in an ovarian teratoma: Spiradenocylindroma case report

Mariah Vesely [a], Atousa Ordobazari [b], Jane Messina [c], Mahsa Chitsaz [b], Vernon K. Sondak [d], Monica Avila [e,*]

[a] Lake Erie College of Osteopathic Medicine, Bradenton, FL, USA
[b] Department of Pathology, H Lee Moffitt Cancer Center, Tampa, FL, USA
[c] Department of Pathology and Cutaneous Oncology, H Lee Moffitt Cancer Center, Tampa, FL, USA
[d] Department of Cutaneous Oncology, H Lee Moffitt Cancer Center, Tampa, FL, USA
[e] Department of Gynecologic Oncology, H Lee Moffitt Cancer Center, Tampa, FL, USA

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ABSTRACT

Background: Spiradenocylindroma is a benign tumor of skin adnexal origin with overlapping features of two distinct neoplasms: spiradenoma and cylindroma. This cutaneous tumor typically presents on the head and neck and extracutaneous presentations are uncommon. The presentation described below involves a spiradenocylindroma within a mature ovarian teratoma is very rare.
Aim: The aim of this article is to portray the diagnostic process of this unusual spiradenocylindroma presentation.
Case presentation: A 65 year-old female with a left adnexal mass underwent ultrasonography and magnetic resonance imaging (MRI) which showed a left ovarian multiseptated lesion, with mural calcifications and projections into the mass. Excisional surgery was performed and histopathological examination revealed a spiradenocylindroma.
Conclusion: Spiradenocylindroma is rare, hard to identify, and often misdiagnosed. Our study described the process of diagnosis and depicts the rare presentation of this lesion arising within a mature teratoma.

1. Introduction

Spiradenocylindromas are considered cutaneous adnexal neoplasms in which tumors adnex normal structures on the skin including sweat glands, sebaceous glands and/or hair follicles (Plotzke et al., 2022 Jan). They are typically considered a benign lesion containing distinct characteristics of both a spiradenoma and cylindroma (Facchini et al., 2022; Bostan et al., 2023). The origins of these types of lesions are disputed initially considered to arise from both eccrine and apocrine sweat glands. More recently it has been suggested they likely arise from an epithelial stem-cell origin. These lesions are hard to identify and often misdiagnosed (Facchini et al., 2022; Miceli and Spiradenoma, 2022; Daneshbod et al., 2022 Apr; Yazganoglu et al., 2011 Sep; Liu et al., 2023 Aug).

Histologically, spiradenomas are characterized by large, sharply demarcated lobules containing two distinct types of cells. Towards the perimeter of the lesion, the cells are smaller, darker, basaloid cells with small, hyperchromatic nuclei, and towards the center of the lesion the cells appear larger, more pale, and with larger, more pale, ovoid nuclei (Miceli and Spiradenoma, 2022; Tran et al., 2020 Jun; Jones et al., 2016 Jul; Li et al., 2021). In the tumor, the larger, more central cells arrange around a lumen forming a duct-like structure and large cystic spaces, often containing finely granular eosinophilic material. Lymphocytic infiltration of these spaces differentiates spiradenomas from cylindromas (Miceli and Spiradenoma, 2022; Jones et al., 2016 Jul). Further histopathological defining characteristics include the presence of elongated tubules, wide vessels containing thrombi, squamous differentiation, and perivascular fibrinoid material (Jones et al., 2016 Jul; Li et al., 2021). If a lesion shows overlapping features of both cylindroma and spiradenoma comprising 10 % of tumor volume, it is termed spiradenocylindroma.

Histologically, cylindromas are composed of numerous smaller, irregularly-shaped, tightly packed adjacent cellular aggregations arranged in a jigsaw-like pattern, surrounded by a prominent PAS-positive

* Corresponding author at: 12902 Magnolia Dr, CSB-1 GYN PROG, Tampa, FL 33612, USA.
E-mail addresses: MVesely50116@med.lecom.edu (M. Vesely), Atousa.Ordobazari@moffitt.org (A. Ordobazari), Jane.Messina@moffitt.org (J. Messina), Mahsa.Chitsaz@moffitt.org (M. Chitsaz), Vernon.Sondak@moffitt.org (V.K. Sondak), Monica.Avila@moffitt.org (M. Avila).

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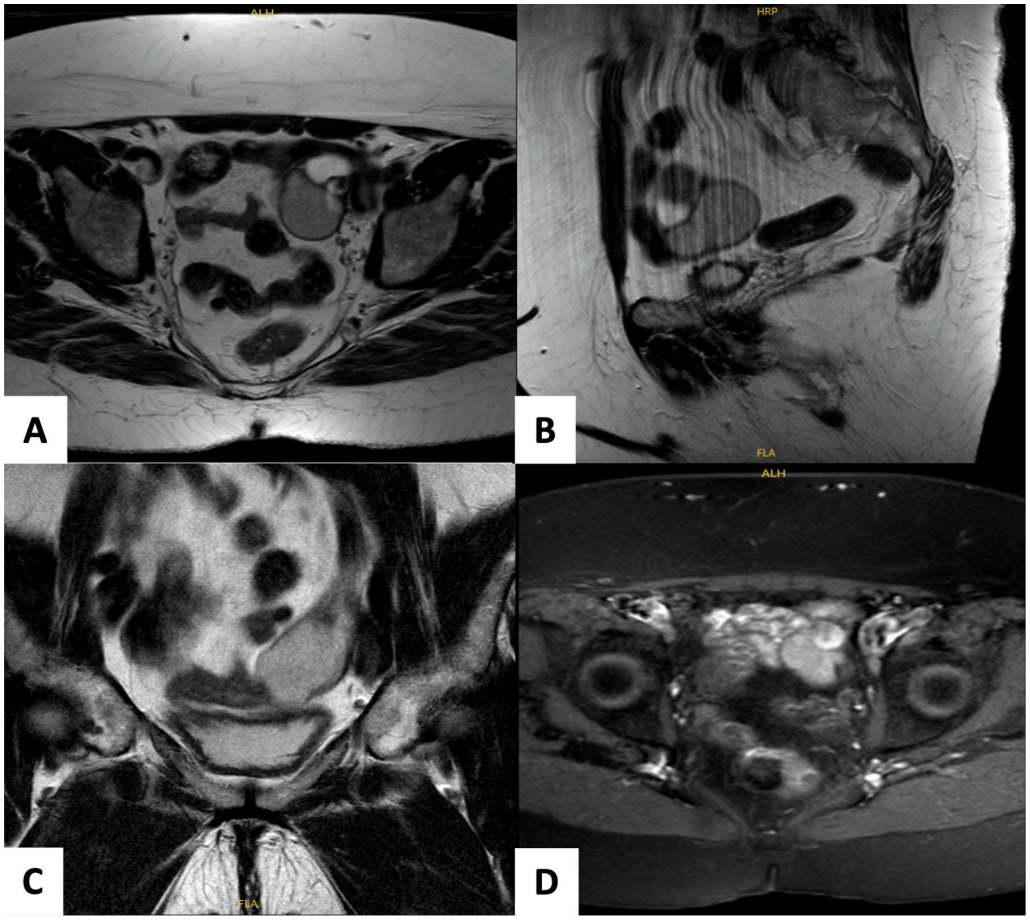

Fig. 1. A) MRI Pelvis Axial T2 TSE image of mass; B) MRI Pelvis Sagitial T2 TSE image of mass; c) MRI Pelvis Coronal T2 TSE Sense image of mass; D) MRI Pelvis Axail FS T1 image of mass.

basement membrane. This basement membrane matrix material occurs within the tumor as hyaline droplets (Liu et al., 2023 Aug; Jones et al., 2016 Jul; Michal et al., 1999 May; Silvestri et al., 2020). Like spiradenomas; cylindromas are composed of 2 distinct cell types. The more peripheral, undifferentiated cells are characterized with a smaller, darker nucleus, while the more central, differentiated cells contain a larger, more pale nucleus. Unlike spiradenomas, cylindromas contain very few, if any, lymphocytes (Jordão et al., 2015 Mar).

Cylindromas, spiradenomas, and spiradenocylindromas are typically found in the head and neck region, especially the scalp. (Bostan et al., 2023; Daneshbod et al., 2022 Apr; Yazganoglu et al., 2011 Sep; Rajan and Ashworth, 2015 Sep; Rajan et al., 2016). Solitary spiradenomas are often described as painful lesions (Bhat et al., 2019). Single lesions are associated with *MYB* gene mutations (Merlo et al., 2022 Jan 26). Multiple lesions are seen in Brooke-Spiegler syndrome, an autosomal dominant hereditary disorder with mutation(s) in the *CYLD* gene at chromosome 16q12-13 (Rajan and Ashworth, 2015 Sep; Kazakov et al., 2009 May). These patients may also demonstrate membranous type basal cell adenomas of the salivary gland, which are histologically identical to cylindromas (Antonescu and Terzakis, 1997 Aug). Multiple lesions on the scalp have been coined“turban tumors” (Jordão et al., 2015 Mar). Mucous membrane presentations involving the anus have been described and extracutaneous presentations include case reports in the kidney(Facchini et al., 2022). All of these lesions are benign, but lesions of longstanding may rarely undergo malignant transformation. Malignant tumors arising from spiradenomas, cylindroma, or spiradenocylindroma are termed spiradenocarcinoma, cylindrocarcinoma, or spiradenocylindrocarcinoma respectively.

The presentation of either of these lesions found within a mature ovarian teratoma is exceedingly rare and to our knowledge has not previously been described. This case report describes the process of identifying this lesion and arriving at such an uncommon diagnosis.

2. Ethics

This study is conducted in accordance with the Declaration of Helsinki and “good clinical practice” guidelines. Written informed consent was obtained from the patient prior to the study.

3. Case and results

We present the case of a 65 year-old female with a history of recurrent squamous cell carcinoma of the neck and scalp status post multiple resections and radiation therapy who came to our attention in 2023 with a left adnexal mass. This patient did not have any of the known associated benign lesions or syndromes described above.

She reported a one year history of intermittent pelvic pain. In January 2023, the patient reported worsening soreness, numbness and bloating in her right lower quadrant accompanied by diarrhea. She also noted early satiety with pelvic pain radiating to her groin.

Pelvic endovaginal ultrasound noted a complex left adnexal cyst with mural calcifications and internal septations measuring 5.7x4.3 cm.

A follow-up pelvic MRI was ordered showing a multiseptated, pelvic mass with mural calcifications and projections into the mass likely arising from the left ovary. It was noted to have solid enhancing components within the superior aspect of the mass measuring 1.9 cm x 2.6 cm. The mass measured 5.5 cm x 4.2 cm x 5.1 cm. [Fig. 1]. The patient had normal pre-operative tumor markers including CA125, CEA and

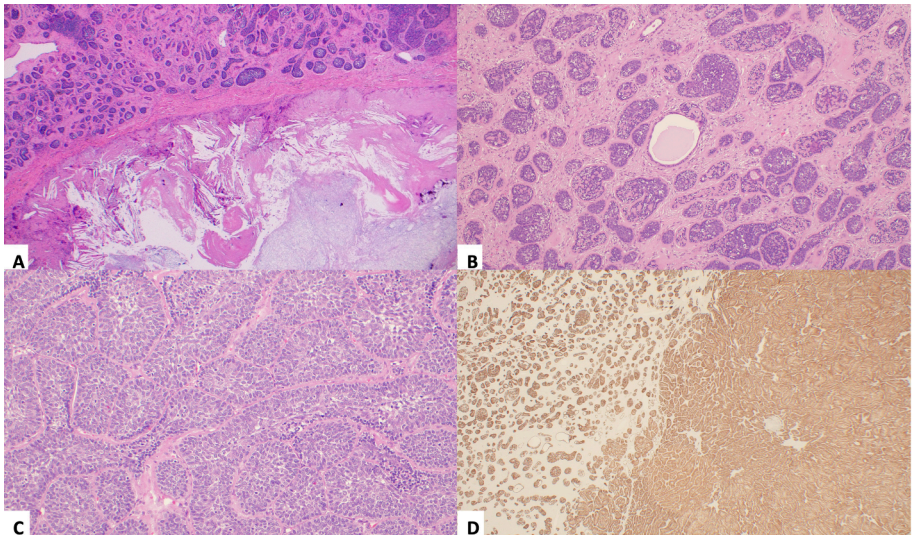

Fig. 2. A) 2x. H&E. Ovary with mature teratoma, demonstrating mature squamous epithelium and keratinization (black arrow) with basaloid epithelial proliferation of adnexal histology; B) 4x. H&E. The basaloid proliferation demonstrates variably-sized nests with focal jigsaw configuration, containing a mixture of small hyperchromatic cells and larger cells, with surrounding hyalinized stroma. Some nests contain basement membrane-like material; C) 40x. H&E. Mixture of small and large cells within jigsaw nests, indicating overlapping features of spiradenoma and cylindroma; D) 2x. P63. highlights the adnexal neoplasm, indicating cutaneous/ectodermal derivation.

CA19-9. CT chest was negative for any evidence of metastatic disease.

The patient was consented and underwent robotically-assisted bilateral salpingo-oophorectomy with potential for surgical staging pending frozen section diagnosis. Excision of the left tube and ovary revealed a 6 cm in diameter mass with solid and cystic areas. Frozen section from a solid area was diagnosed as "epithelial neoplasm with basaloid features."

Final histopathological examination revealed evidence a mature teratoma, with epithelia from both ectodermal and endodermal origin (mature skin, thyroid gland, respiratory epithelium) and mesenchymal elements (mature fat, cartilage). Approximately 60 % of the tumor volume was composed of a basaloid epithelial neoplasm with jigsaw-like confirmation and two cell types [Fig. 2]. Immunohistochemical stains of this neoplasm demonstrates a profile with positive AE1/AE3, CAM5.2, p63, CD117, and ER (10 %, moderate). The Ki-67 proliferative index was focally increased up to 30 %, but P53 appeared wild type, and all other hereditary tests were negative. Thus, a diagnosis of spiradenocylindroma arising in mature teratoma was rendered. Given the patient's age and lack of history of other skin tumors in this lineage, she does not fulfill the criteria for Brooke-Spiegler syndrome. She was dispositioned to continue annual benign gynecology visitations.

4. Conclusions

Spiradenocylindroma is a benign cutaneous adnexal neoplasm that has not been previously described within gynecologic adnexal neoplasms.

It is characterized by the coexistence of a spiradenoma and a cylindroma in the same lesion (Bostan et al., 2023; Miceli and Spiradenoma, 2022). Historically, spiradenomas and cylindromas were thought to originate from eccrine glands with newer data suggesting an apocrine or pluripotent stem cell origin (Bostan et al., 2023; Daneshbod et al., 2022 Apr; Yazganoglu et al., 2011 Sep; Liu et al., 2023 Aug).

Upon histological examination of this lesion, the lesion was categorized as spiradenocylindroma due to the pathognomonic tubule morphological appearance typical of both a spiradenoma and cylindroma. The spiradenoma portion included characteristic lymphocytic infiltration whereas the cylindroma portion did not. An adenoid cystic carcinoma diagnosis was considered, but ruled out due to the lack of true "biphasic" myoepithelial and ductal differentiation. Immunohistochemical stains of this neoplasm demonstrated a focally increased Ki-67 proliferative index to 30 % and found to be P53 wild type. MYB protein gene staining typical of isolated lesions was negative. Given the absent clinical history of multiple cutenaous lesions, CYLD gene staining was deferred. Further, there was no evidence of overt features of malignancy, including sheet-like growth, anaplasia, or frank invasion.

Most spiradenocylindromas are cutaneous neoplasms found on the face and scalp (Bostan et al., 2023; Daneshbod et al., 2022 Apr). Less commonly, they are found on other cutaneous or mucosal surfaces such as the mandible, vulva or anus. Prior extracutaneous cases have been reported in the kidney. The specific development of spiradenocylindroma inside a mature teratoma in the ovary is very rare and to our knowledge this is the first time it has been described in the literature.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

CRediT authorship contribution statement

Mariah Vesely: Writing – review & editing, Writing – original draft, Data curation. **Atousa Ordobazari:** Writing – review & editing, Methodology. **Jane Messina:** Writing – review & editing, Writing – original draft, Methodology. **Mahsa Chitsaz:** Writing – review & editing, Resources. **Vernon K. Sondak:** Writing – review & editing. **Monica Avila:** Writing – review & editing, Writing – original draft, Supervision, Project administration, Methodology, Data curation, Conceptualization.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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