Case Report

Steroid Cell Tumor NOS and Xanthogranulomatous Oophoritis: A Rare Association Mimicking Ovarian Malignancy

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Abstract

Steroid cell tumor, NOS is an infrequently reported ovarian tumor, rarely presenting without endocrine symptoms. Xanthogranulomatous inflammation of ovaries is an extremely rare finding which often mimics malignancy on imaging. This case report presents a unique co-incidental finding of these two rare entities as incidental findings.

A 44 year old female with previous history of total abdominal hysterectomy for leiomyoma presented with abdominal discomfort. Clinical examination revealed bilateral adnexal mass. Imaging suggested a possibility of bilateral ovarian mucinous cystadenocarcinoma. CEA, CA19-9 and CA125 were within normal limit. No signs and symptoms of endocrine manifestation were seen. Bilateral salpingo-oophorectomy specimen was sent for histopathological examination. Grossly, Left ovary showed a single yellow nodule measuring 1cm in diameter. Right ovary showed a gray tan cut surface.

Microscopic examination revealed a left ovarian nodule composed of diffuse sheet of large tumor cell with abundant eosinophilic cytoplasm, round nucleus with a prominent nucleolus in a background of extensive vascular congestion. No reinke crystals, mitosis, necrosis, hemorrhage or nuclear atypia was seen. The tumor cells were immunopositive for Vimentin, Pan CK and Inhibin. Adjacent left ovarian stroma and right ovary showed features suggestive of XI. A diagnosis of Steroid cell tumor, NOS with xanthogranulomatous oophoritis was offered.

Steroid cell tumor and xanthogranulomatous oophoritis, both are rare entities which may mimic malignancy. Awareness of these entities alone or in combination allows a clinician to avoid radical surgeries in such cases.

Keywords:

Ovary, tumor, inflammation, immunohistochemistry, Oophoritis

Introduction

Steroid cell tumor is a rare ovarian tumor, characterized by endocrine manifestation. On rare occasions, they may present without endocrine manifestations. [1] Diagnosis of steroid cell tumor after previous hysterectomy in the absence of endocrine manifestation is seldom reported. [2]

Xanthogranulomatous inflammation is a rare form of chronic inflammation, commonly reported in kidneys and gallbladder. Ovarian involvement is extremely rare. [3] It often masquerades as malignancy on clinical examination and imaging. [4]

Coexistence of both these rare entities is exceedingly rare and has not been reported till date. Our case reports a unique combination of two very rare entities coexisting together as incidental findings in a patient with a history of hysterectomy.

Case Report

A 44 year old female presented to gynecology department with abdominal discomfort. Patient had previously undergone total abdominal hysterectomy for leiomyoma. Bilateral adnexal mass was found on clinical examination. No signs of virilization or hirsutism were observed. Ultrasonography reported a bilateral adnexal cystic lesion (ORADS IV) with intermediate risk of malignancy. Contrast enhanced MRI suggested a possibility of bilateral ovarian mucinous cystadenocarcinoma. Tumor marker studies showed a normal CarcinoEmbryonic Antigen of 1.38 ng/ml, normal CA-19-9 of 7.6 U/mL and normal CA-125 of 5.79 U/mL. The thyroid function test revealed a normal TSH of 2.72 uIU/ml, a normal T3 of 1.39 ng/ml and slightly raised T4 of 10.82 ug/dl (normal range, 4.60-10.50 ug/dl). Patient showed no signs and symptoms of endocrine manifestation.

Bilateral salpingo-oophorectomy specimen was sent for histopathological examination. Grossly right and left ovary measured 4x3.7x1 cm and 3.5x4x1.1 cm, respectively. The left ovary showed a single yellow nodule measuring 1cm in diameter. The right ovary showed a gray tan cut surface.

Microscopic examination revealed the left ovarian nodule composed of diffuse sheet of large tumor cell with abundant eosinophilic cytoplasm, round nucleus with a prominent nucleolus in a background of extensive vascular congestion (Fig 1). No reinke crystals, mitosis, necrosis, hemorrhage or nuclear atypia was seen. The tumor cells were diffusely immunopositive for Vimentin, Inhibin and Pan CK (Fig 2). Adjacent left ovarian stroma and right ovary showed extensive chronic inflammation admixed with sheets of foamy macrophages and fibrosis, suggestive of xanthogranulomatous inflammation (Fig 3). Michaelis-Gutmann bodies or epithelioid cell granulomas were absent. Ziehl Neelsen stain for acid fast bacilli was negative. Areas of stromal hyperthecosis (Fig 4) were also seen.

A diagnosis of Steroid cell tumor, NOS with xanthogranulomatous oophoritis was offered.

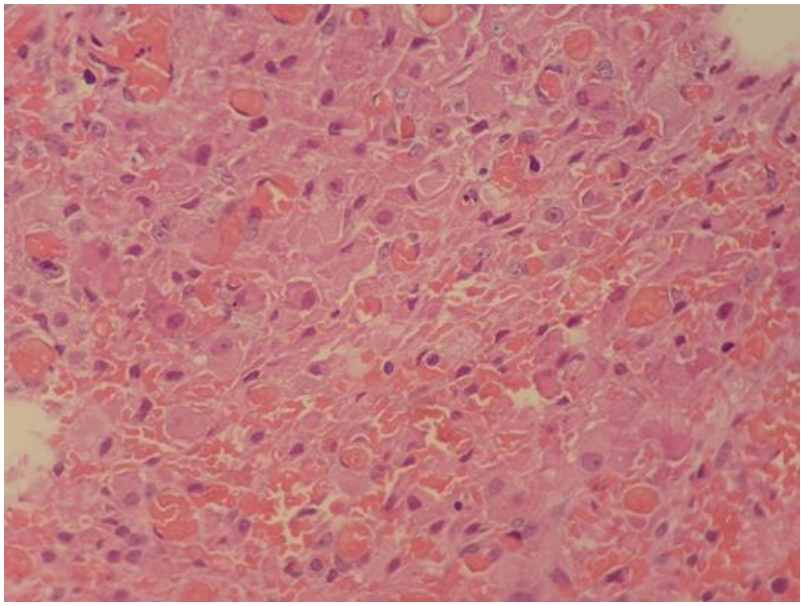

Figure 1: Steroid Cell Tumor, NOS- Tumor is composed of Diffuse sheet of large tumor cell with abundant eosinophilic cytoplasm, round nucleus with a prominent nucleolus in a background of extensive vascular congestion. (HE 400x)

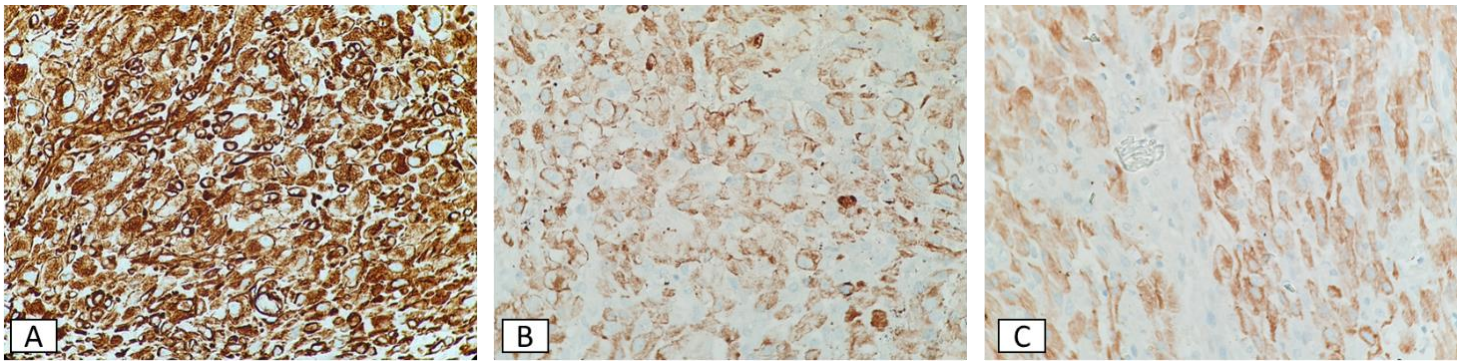

Figure 2: These tumor cells were diffusely immunopositive for Vimentin (A), Inhibin (B) and Pan CK (C).

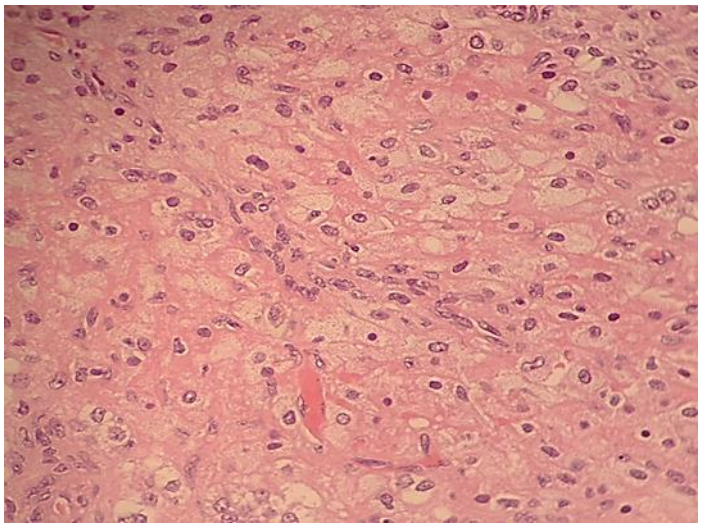

Figure 3: Xanthogranulomatous Oophoritis - Ovarian stroma showed extensive chronic inflammation admixed with sheets of foamy macrophages and fibrosis. (HE 400x)

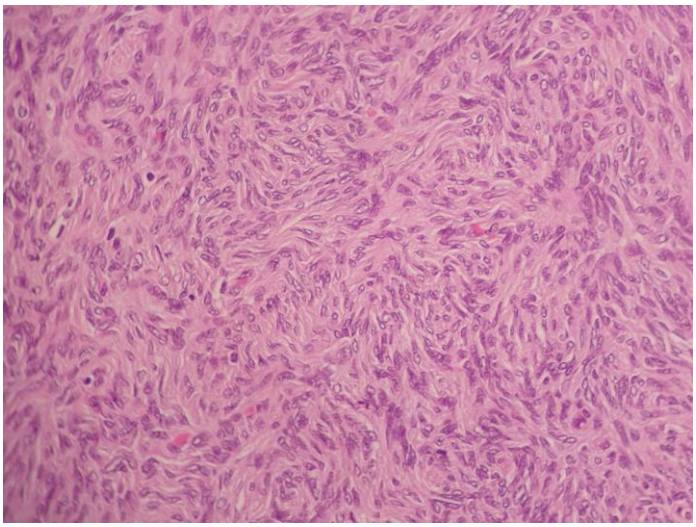

Figure 4: Stromal Hyperthecosis (HE 400x)

Discussion

Steroid cell tumor is a rare tumor accounting for less than 0.1% of all ovarian tumors. (1) According to WHO classification of female genital tract, 5th edition, it is classified under pure stromal tumor and further subtyped into Steroid cell tumor, NOS and Steroid cell tumor, Malignant. [5]

Steroid cell tumor, NOS can present in any age group. However, tumor usually present in middle age with an average age of 43. [1] They may present with symptoms associated with an ovarian mass or abdominal distension and bloating. They frequently present with endocrine manifestation, with androgenic symptoms and estrogenic symptoms in more than 50% and 10% of patients, respectively. Rarely, patients may present without endocrine symptoms. In such cases, it is often an incidental finding. [1-2]

They are most commonly unilateral but rarely may be bilateral. The size of tumor ranges in size from less than 1cm to more than 45cm with mean size of 8.4cm. [1-2] They are well circumscribed with solid yellow cut surface. [1-2,6]

There is an increased incidence of ovarian pathology in patients having a history of hysterectomy for benign conditions reaching upto 3.8%. [7] In literature, there are only four reported cases of ovarian Steroid cell tumor diagnosed after previous hysterectomy. Ours is the fifth case. [2,8-9]

These tumors are characterized by diffuse pattern of large cells with abundant eosinophilic (lipid poor) to pale vacuolated (Lipid rich) cytoplasm, round nucleus and a prominent central nucleus. They may also rarely demonstrate pseudo-glandular, cords, nest or follicular pattern. Stroma varies from scant to cellular with fibrous bands. Adjacent stroma can show features of stromal hyperthecosis like our case. [1]

The majority of tumor have benign behavior with low rates of metastasis and or recurrence. [1,6]

According to Hayes et al [1], following histological features favors malignant behavior: size > 7cm, nuclear atypia of grade 2 or 3, 2 or more mitosis per 10 high power fields, necrosis and hemorrhage. None of these findings were present in our case. Thus, predicting a benign nature of tumor in this patient.

Steroid cell tumor needs to be differentiated from Leydig cell tumor which contains crystal of reinke. Other common differential are ovarian clear cell carcinoma, pregnancy luteoma and metastasis from kidney, breast or gastrointestinal site. [10] Use of Immunohistochemistry in such scenarios aids into confirming the diagnosis. Steroid cell tumor cells are positive for Inhibin, Calretinin, Melan A and Vimentin. [11]

Xanthogranulomatous inflammation is a rare form of chronic inflammation, commonly reported in kidney and gallbladder followed by bones, stomach and testes. Female genital tract is rarely affected. [3-12]

It presents clinically with non-specific symptoms such as weight loss, anorexia, fever, lower abdominal pain, vaginal bleeding or fever. [13] It presents as an adnexal mass when it affects ovary or fallopian tube like our case. Age at the time of diagnosis vary from 23-72 years with rare occurrence in pediatric patients. [3]

Xanthogranulomatous oophoritis often confound clinician as it mimics ovarian malignancy both clinically and on radiology. [4,14-15] Diagnosis is confirmed by histopathological examination. Its common differential is malakoplakia, which is distinguished by the presence of Michaelis-Gutmann bodies. [13] Other differential diagnosis includes infective condition like tuberculosis and fungal infections, which are excluded by special stains and culture.

The exact pathogenesis xanthogranulomatous inflammation is still an enigma. Multiple factors proposed as possible causes are long standing Pelvic inflammatory disease, endometriosis, ineffective antibiotic treatment, intrauterine devices, chemotherapy for breast cancer, prolonged antibiotic use, leiomyoma, abnormal lipid metabolism and ineffective clearance of bacteria by phagocytes. [3-4,13] Punia et al [16] reported it as a late sequel of long standing pelvic inflammatory disease treated inadequately for staphylococcus. On rare occasions, it may coexist with malignancy. Rare association of xanthogranulomatous inflammation with endometrial carcinoma and gallbladder carcinoma has been documented in female genital tract and gallbladder respectively. [17,18] But association of ovarian tumour with xanthogranulomatous inflammation has not been documented till date to the best of our knowledge.

Xanthogranulomatous inflammation may play a role in pathogenesis of malignancy. More studies are required to understand the pathogenesis of xanthogranulomatous inflammation and to find its association with malignancy. Further studies are required to elucidate this notion which currently at best is speculative.

Surgery is offered as primary treatment for both entities. [1-16]

Patient had an uneventful follow up and is presently doing well.

Conclusion

Steroid cell tumor and xanthogranulomatous oophoritis, both are rare entities. Co-existence of such two rare entities together as incidental finding has not been reported to the best of our knowledge till date. This case reports a rarity of two incidental findings masquerading as ovarian malignancy. Awareness of these entities alone or in combination allows a clinician to avoid radical surgeries. It prompts a pathologist to gross such lesions more extensively if xanthogranulomatous inflammation is seen to rule out co-existing malignancy for better management of these patients.

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