Case Report

Sertoli-Leydig Cell Tumor of Ovary: A Rare Case Report with Heterologous Elements and Focal Marked Anaplasia

Abstract

Sertoli-Leydig cell tumor (SLCT) of the ovary is an extremely uncommon neoplasm accounting for <0.5% of all primary ovarian neoplasms. These tumors belong to the category of sex cord-stromal tumors. The tumor has variable clinical and histopathological presentations complicating the diagnosis and therefore the treatment. The presence of heterologous elements is seen in one-fifth of these already rare neoplasms. Herein, we report a case of a 28-year-old female presenting with irregular menses, features of virilization, and abdominal pain. Histopathological examination revealed marked focal anaplasia in this tumor of, otherwise, intermediate differentiation along with the presence of heterologous elements. Reporting of such elements is imperative for adequate treatment and deciding follow-up.

Keywords: *Anaplasia, heterologous elements, Sertoli-Leydig cell tumor*

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Introduction

Sertoli-Leydig cell tumor (SLCT) of the ovary (also known as androblastoma/arrhenoblastoma) is a rare sex cord-stromal neoplasm, which comprises <0.5% of all primary ovarian neoplasms.[1] Most of the cases present in the second and third decades of life. Around half of the cases present with androgenic activity; however, they are less common in cases with heterologous elements.[2] The tumor is generally unilateral and around 80% of the cases are diagnosed in stage Ia.[2] Prognosis of SLCT is overall favorable but is related to the stage and grade of tumor. Intermediate-grade tumors are clinically malignant in about 10% of cases. The presence of heterologous elements also portends poor prognosis. Recurrence is usually within 2 years[3,4] and occurs in the peritoneal cavity. On histopathology, marked anaplasia is described in cases with intermediate differentiation[5] and no adverse prognostic importance has been ascribed to these cases; however, a very few cases have been reported and larger studies are still lacking.

Case Report

A 28-year-old female presented in the gynecology department in AIIMS, Jodhpur, with complaints of oligomenorrhea, hypomenorrhea, hirsutism, acne, and pain in abdomen. The patient is a gutka chewer for 6–7 years. No comorbidities or family history of malignancy were present. Medical and surgical history was unremarkable. Preoperative serum testosterone level of the patient was 520.14 ng/dl (normal range 15–70 ng/dl). Ultrasonography revealed an echogenic mass measuring 7.2 cm × 6 cm in the right ovary. Left ovary and uterus were unremarkable. Ascitic fluid of the patient was examined and it was negative for malignant cells.

Right salpingo-oophorectomy was done, in which the right ovary was seen to be replaced by a solid cystic tumor with intact capsule [Figure 1]. Grossly, the right ovary was enlarged measuring 9 cm × 6 cm × 5 cm. The cut-section showed a solid cystic tumor replacing the entire ovary. The solid areas are gray-yellow, showing multiple foci of hemorrhage, and the cysts were filled with thin clear yellow fluid [Figure 2]. Microscopically, the tumor was composed of open and closed tubules, alveolar pattern, along with cellular lobules and cords composed of darkly staining cells. The cells had scant-to-moderate amount of cytoplasm, round-to-oval nucleus, and 0–1 nucleoli. Admixed clusters and nests of Leydig cells were noted within

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the tumor. Delicate fibrous stroma was seen at places in which Leydig cells were found in small clusters, cords, and lying singly [Figure 3]. Many mitotic figures were identified. Heterologous elements in the form of glands lined by mucinous epithelium were seen [Figure 4]. Focal area showed marked anaplasia in the form of many large, bizarre, and multinucleated cells [Figure 5]. Cystic spaces were also noted. No capsular breach was identified.

On immunohistochemistry, the tumor cells were immunoreactive for calretinin. The cells were nonreactive for epithelial membrane antigen. MIB1 labeling index was approximately 30%. However, areas with anaplasia show a MIB1 labeling index of 80%–90% [Figure 6].

Based on the above findings, a final diagnosis of SLCT with intermediate differentiation along with focal anaplasia and heterologous elements of gastrointestinal type was made.

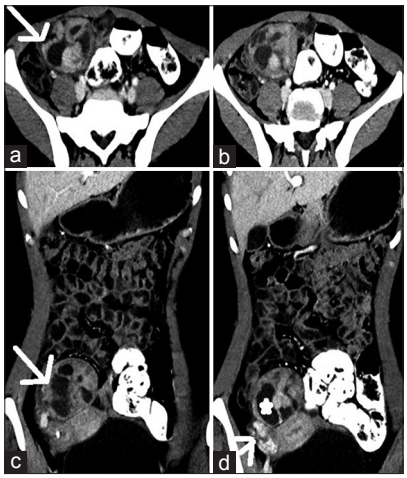

Figure 1: Contrast-enhanced computed tomographic scan of abdomen in axial (a and b) and coronal images (c and d) showing a well-defined complex mass (large arrow) in right adnexa with enhancing solid component and nonenhancing necrotic part (*). Prominent adjacent adnexal vessels (small arrow) with right ovary not separately identified

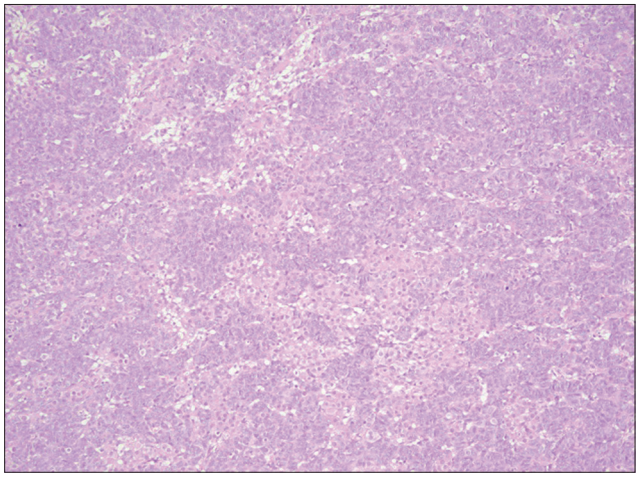

Figure 3: Microscopic examination of tumor showing intermediate differentiation with presence of tubules of immature Sertoli cells and clusters of Leydig cells (H and E, ×10)

Discussion

We, hereby, report a case of SLCT of intermediate differentiation with gastrointestinal-type heterologous elements and marked focal anaplasia.

SLCT is a rare neoplasm and is more frequently seen in young women with a mean age of 25 years.[5] Macroscopically, the size ranges from 2 to 35 cm and may be solid, solid-cystic, or rarely cystic.

Microscopically, well- and moderately differentiated tumors are encountered most frequently.[6] Well-differentiated cases consisted of solid or hollow tubules composed of Sertoli cells which lack significant nuclear atypia or mitotic activity. Leydig cells are seen in delicate fibrous stroma in small clusters, cords, and lying singly. The tumors with intermediate differentiation, as in our case report, comprise cellular lobules of darkly staining Sertoli cells, typically with scant cytoplasm, and admixed in a jumbled fashion with Leydig cells. Nested to alveolar arrangement and solid and hollow tubules lined by Sertoli cells may also be seen. Mitotic figures average 5/10 HPF. Poorly differentiated tumors predominantly have sarcomatoid stroma with minor area of differentiation. Heterologous elements are observed in one-fifth of this already rare neoplasm. They can be

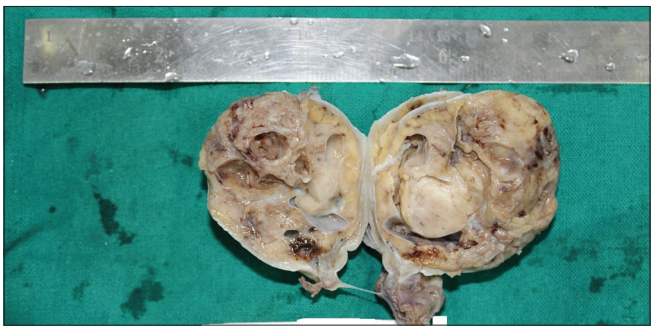

Figure 2: Macroscopic examination reveals an enlarged ovary replaced by a solid cystic tumor. The solid areas appear gray-yellow in color and show multiple foci of hemorrhage. The cystic spaces were filled with thin yellow fluid

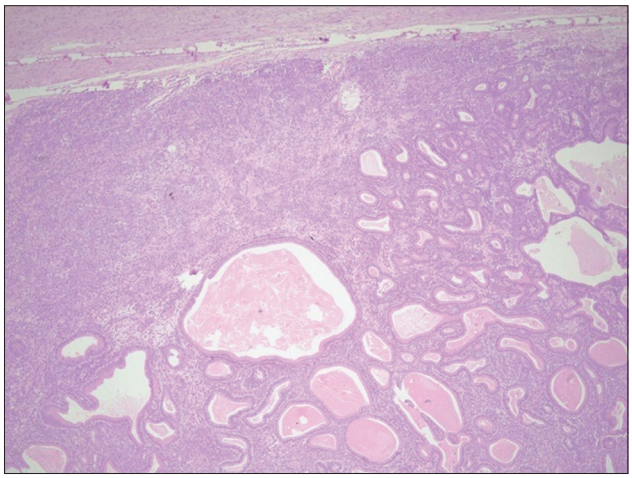

Figure 4: Sertoli-Leydig cell tumor showing heterologous elements lined by mucinous epithelium (H and E, ×10)

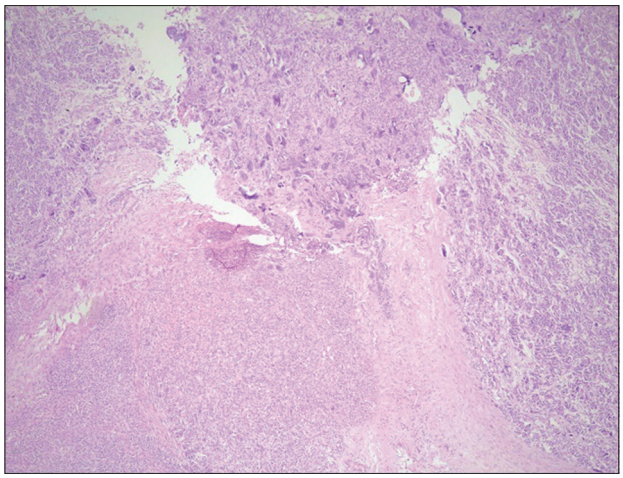

Figure 5: Sertoli-Leydig cell tumor showing marked anaplasia (H and E, ×10)

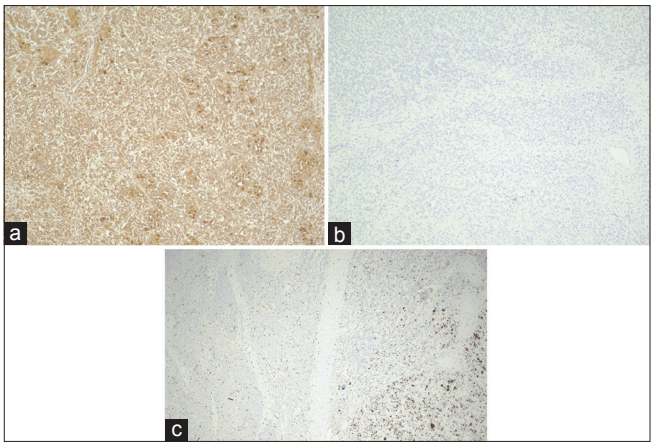

Figure 6: Immunohistochemical examination. (a) The neoplastic cells stained positive for calretinin. (b) The neoplasic cells stained negative for epithelial membrane antigen. (c) MIB1 labeling index is approximately 30%, (Anaplastic focus shows a very high MIB1 labeling index of 80%-90%)

either endodermal elements or mesenchymal elements. The endodermal elements are typically seen in tumors with intermediate differentiation. The mesenchymal elements are seen more in association with poorly differentiated tumors.[7]

The prognosis of SLCT is overall favorable although it is significantly correlated with stage and degree of differentiation.[8,9] Nineteen percent of tumors with heterologous elements were seen to be clinically malignant in a study by Young and Scully.[8]

Adjuvant chemotherapy is recommended for patients with advanced stage, intermediate and poor differentiation, retiform pattern, and presence of heterologous elements.[10]

Although focal anaplasia is described in cases with intermediate differentiation;[5] however, exact course of disease is not known in these cases due to lack of data in this area. Such cases may also behave in a clinically malignant way; therefore, we want to emphasize that more studies are required with proper follow-up of these cases. It might help in deciding the protocols for the use of adjuvant therapy in such cases. SLCT recurs relatively early (within 2–3 years of initial diagnosis;[7,8] therefore, it becomes important to closely follow-up those cases where the tumor has got poor prognostic features. It will help in detecting and treating recurrence early.

Conclusion

SLCT typically has good prognosis. However, adequate sampling and through microscopic examination are required to look for heterologous elements and focal anaplasia as adjuvant therapy might have a role in these patients for a better outcome. Follow-up of these patients is crucial to detect recurrence at an early stage.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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