

# Ovarian Sertoli-Leydig cell tumor with heterologous elements of gastrointestinal type associated with elevated serum alpha-fetoprotein level: an unusual case and literature review

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## ABSTRACT

Here we describe the case of a 19-year-old woman with a poorly differentiated ovarian Sertoli-Leydig cell tumor and an elevated serum alpha-fetoprotein level. The patient presented with diffuse abdominal pain and bloating. Physical examination, ultrasound, and magnetic resonance imaging revealed a right ovarian tumor that was histopathologically diagnosed as a poorly differentiated Sertoli-Leydig cell tumor with heterologous elements. Her alpha-fetoprotein serum level was undetectable after tumor resection. Sertoli-Leydig cell tumors are rare sex cord-stromal tumors that account for 0.5% of all ovarian neoplasms. Sertoli-Leydig cell tumors tend to be unilateral and occur in women under 30 years of age. Although they are the most common virilizing tumor of the ovary, about 60% are endocrine-inactive tumors. Elevated serum levels of alpha-fetoprotein are rarely associated with Sertoli-Leydig cell tumors, with only approximately 30 such cases previously reported in the literature. The differential diagnosis should include common alpha-fetoprotein-producing ovarian entities such as germ cell tumors, as well as other non-germ cell tumors that have been rarely reported to produce this tumor marker.

## CASE REPORT

### CASE REPORT

A 19-year-old woman complaining of diffuse abdominal pain and sudden abdominal bloating was referred to our institute because of her family history. Her elder sister died of ovarian cancer at the age of 11. Her past personal medical history was unremarkable, except for an etonogestrel implant, inserted 7 months before for the prevention of pregnancy. She reported irregularities in her menstrual cycle since then.

On physical examination, a 10-cm-sized, mobile, ovoid-shaped, and painless pelvic mass was detected. Other physical assessments were negative. Routine laboratory data, cancer antigen 125 (CA-125), cancer antigen 19.9, carcinoembryonic antigen, and human chorionic gonadotropin ( $\beta$ -hCG) were within their respective normal ranges. The only altered laboratory finding was an increased level of alpha-fetoprotein (AFP) of 46.3 ng/ml (normal range < 7 ng/ml).

### Imaging findings

Pelvic transabdominal and transvaginal ultrasounds were performed, and we found a complex-appearing tumor of the right ovary, predominantly solid with focal cystic components and measuring 82 × 75 × 65 mm (Fig. 1). Color Doppler revealed flow in the solid areas (Fig. 1C); a small amount of pelvic ascites was also present (Figs. 1A-C; white asterisks). The uterus and the left ovary were unremarkable.

Since the patient was young and the ultrasound examination revealed a suspicious ovarian mass, magnetic resonance imaging was performed, and a tumor confined to the right ovary was seen (Figs. 1 and 2). The tumor was well defined and encapsulated, with the exception of a protruding eccentric anterior segment (Fig. 2B; white arrow).

The solid component demonstrated intermediate T2 signal, T1 signal intensity similar to muscle, and avid contrast uptake on fat-suppressed gadolinium-enhanced T1-weighted images (Figs. 2, 3A, and 3B; white-bordered arrows). The cystic areas had low signal on T1-weighted images, high T2 signal, and did not enhance (Figs. 2, 3A, and 3B; white thin arrows). Diffusion-weighted images (b1000 s/mm<sup>2</sup>) showed high-signal intensity in the solid components of the tumor, and apparent diffusion coefficient images showed low-signal intensity of these components; therefore, restricted diffusion was present (Figs. 3C-D; white-bordered arrows). A small amount of ascites was detected only in the pelvis (Figs. 2A-C; black asterisks). No inguinal, pelvic, and para-aortic lymphadenopathies, nor peritoneal implants or hepatic metastases, were detected.

### Management and follow-up

Because the tumor had magnetic resonance imaging features suggestive of malignancy, and seemed resectable, fertility-sparing salpingo-oophorectomy, partial omentectomy, and peritoneal biopsies were performed. The pelvic and abdominal cavities were explored during surgery, and neither peritoneal implants nor lymphadenopathies were found.

The right ovary, weighing 140 g and with the longest axis measuring 8 cm, exhibited an area of rupture on the surface associated with tumor extravasation (Fig. 4A), as described on gross examination of the pathologic specimen. The ovarian mass was a tan-grayish variegated tumor, predominantly solid with small cystic areas (Fig. 4B). The histopathologic examination revealed a tumor composed of 2 cell populations: Sertoli cells and Leydig cells. Leydig cells were seen isolated, or in small aggregates, admixed in between Sertoli cell aggregates. Sertoli cells were arranged in various patterns, forming hollow tubules and cords in an edematous stroma (Fig. 5A). There were areas of marked edema producing a microcystic pattern (Fig. 5B), and in very few areas, the Sertoli cells were arranged to form slit-like spaces giving a retiform appearance (Fig. 5C). Poorly differentiated areas of immature Sertoli cells growing in a predominantly diffuse pattern with small tubular differentiation and a high mitotic index (with an average of 26 mitoses in 10 high power fields) were identified (Fig. 5D). A small focus of heterologous elements was found, with mature intestinal differentiation (Fig. 5E). Due to the reduced dimension of this focus, with the longest axis

measuring 0.9 mm, the immunohistochemical study with AFP was inconclusive regarding this small focus of heterologous components, and no expression was found in the Sertoli cells or in the Leydig cell components. There were foci of necrosis. No lymphovascular invasion was documented. The multiple peritoneal tissue biopsies received were negative for tumor cells.

The diagnosis of poorly differentiated Sertoli-Leydig cell tumor (SLCT) with heterologous gastrointestinal epithelium was made.

The serum AFP level was undetectable after tumor resection.

Adjuvant chemotherapy treatment options were discussed in the multi-disciplinary gynecology oncology group, and the patient was considered for a bleomycin, etoposide, and cisplatin (BEP) regimen. Thus far, the patient has undergone 4 months of therapy without major complications.

## DISCUSSION

We report a rare case of a poorly differentiated ovarian SLCT with foci of heterologous gastrointestinal epithelium that presented with a slight elevation of serum AFP levels. Since the early 1980s, only approximately 30 cases of SLCTs associated with elevated levels of AFP have been reported in the literature [1-24].

### *AFP in ovarian tumors - differential diagnosis*

AFP is a major plasma glycoprotein produced by the liver, yolk sac cells, and fetal gastrointestinal tract [1,4,5]. It can also be produced by Sertoli cells, Leydig cells, and heterologous components, such as hepatocytes and/or gastrointestinal epithelium [1,4].

Commonly, elevated serum levels of AFP have been detected in hepatocellular carcinoma and ovarian germ cell tumors, such as yolk sac tumor (endodermal sinus tumor) and embryonal carcinoma, among others [25]. Therefore, the presence of a probably malignant unilateral ovarian mass in a young woman, in whom the only abnormal laboratory finding was the presence of elevated serum levels of AFP, led to the suspicion of a yolk sac tumor. The presence of a yolk sac tumor was the first diagnosis considered, since it is strongly associated with the production of AFP and is the second most frequent type of malignant germ cell tumor (after dysgerminoma), often occurring mixed with other entities [26]. Moreover, as in the case we present, it occurs mainly in the second and third decades of life as a unilateral, mainly solid tumor frequently associated with cystic areas. However, this type of germ cell tumor frequently exhibits intra-abdominal spread, invasion of the surrounding structures, and lymphatic metastases, none of which were present [26]. Although AFP serum levels are frequently elevated in embryonal carcinoma without admixed yolk sac component, this rare type of tumor generally presents as an aggressive large solid mass frequently associated with elevated β-hCG serum levels [27].

Another germ cell tumor that should be included in the differential diagnosis is immature teratomas [28]. The finding of an elevated level of serum AFP has been reported in 33-65% of patients with immature teratomas [29,30]. Although the lack of demonstrable fat and calcifications with ultrasound and magnetic resonance imaging makes this diagnosis improbable, it should not be ruled out in a young woman with a unilateral heterogeneous mass, since scanty fatty tissue may not be identified on imaging studies.

On rare occasions, non-germ cell tumors of the ovary have been described to produce AFP [25]. These include histological types of epithelial tumor, sex cord stromal tumors, and metastatic tumors to the ovary [25].

The diagnoses of an ovarian epithelial tumor and ovarian metastasis were very unlikely in this clinical setting, since they occur mainly in older age groups. There have been sparse cases reported of AFP-producing epithelial tumors. These include hepatoid carcinoma, ovarian serous carcinoma, ovarian clear cell carcinoma, ovarian endometrioid carcinoma, ovarian undifferentiated carcinoma, ovarian mucinous carcinoma, and ovarian malignant mixed Müllerian tumor [25, 31-42].

Based on the literature reviewed, to our knowledge, the only case of an AFP-producing epithelial tumor occurring in the first 2 decades of life was a clear cell carcinoma that simulated a yolk sac tumor in a 17-year-old patient, reported by Bahri et al. [36]. Ovarian metastases producing AFP have been primarily described in older women with gastric or hepatocellular carcinoma [43,44]. The histological types of sex cord stromal tumors that have been reported to be associated with raised serum AFP are granulosa cell tumors and SLCTs. Although only sporadic cases of juvenile granulosa cell tumors associated with high serum AFP have been described in the literature, this was a diagnosis that could not be ruled out clinically [25,45]. Similar to this case, juvenile granulosa cell tumors generally occur in women aged less than 30 years of age and almost always present as a unilateral, solid ovarian mass with variable cystic/hemorrhagic components.

Despite the few reports of ovarian SLCTs associated with raised levels of AFP, they are the most common AFP-producing non-germ cell tumor of the ovary [25].

#### Etiology and demographics

SLCTs are more likely to occur in young women, as in our case, with approximately 75% of cases occurring in women 30 years of age or younger [46,47]. However, a few cases have been reported in postmenopausal women [48,49]. SLCTs are unilateral in more than 98% of cases, and 80% are confined to the ovary at the time of diagnosis [27,46]. They contain variable portions of Sertoli cells and Leydig cells, and according to the degree of differentiation of these cells, SLCTs are further divided into 4 histologic subtypes: well differentiated, intermediately differentiated, poorly differentiated, and retiform variant [46]. These last 3 categories may have variants that contain heterologous elements [46]. Heterologous elements are reported to be present in 20% of SLCTs and are represented by endodermal and mesenchymal elements [50]. Heterologous elements of the

endodermal type may be hepatocyte cells or gastrointestinal mucin-secreting elements, whilst heterologous mesenchymal elements may be in the form of cartilage and skeletal muscle. Most previous reports have demonstrated AFP expression by immunohistochemistry in Sertoli cells only, in Leydig cells only, in both Sertoli and Leydig cells, in hepatoid cells, and in heterologous gastrointestinal epithelium [1-25].

In our case, the specific type of AFP-producing cell could not be found.

#### Clinical and imaging findings

About 30-50% of SLCTs produce androgens (testosterone and a variety of androgenic precursors), which are responsible for androgen excess signs, such as oligomenorrhea, amenorrhea, and virilizing symptoms [27,47]. Many are non-functioning, and a small subset of tumors are estrogenic [27,47,51].

Here, we present an unusual case in which the only altered laboratory finding was a slightly elevated serum level of AFP (46.3 ng/ml; normal range < 7 ng/ml). Both androgen hormones and tumor markers (CA-125; carcinoembryonic antigen;  $\beta$ -hCG) were in the normal range. The patient presented with sudden abdominal pain and swelling, which are unspecific signs but very common complaints of hormonally inactive tumors.

There are no SLCT-specific imaging features.

Ultrasound remains the primary imaging modality for the assessment of adnexal masses [52]. Since transabdominal ultrasound is useful in the identification and characterization of large masses and those positioned superiorly and laterally in the pelvis, and transvaginal ultrasound provides excellent visualization of adnexal masses, both approaches were performed [52]. Ultrasound reflects the gross pathologic characteristics of the tumor and usually reveals a well-defined hypoechoic mass or a predominantly solid heterogeneous mass with multiple cystic areas, as in the case presented here. Color Doppler imaging may help detect virilizing SLCTs that may be small and difficult to visualize using transvaginal ultrasound [47,53,54].

Computed tomography imaging usually reveals a soft tissue density adnexal mass with heterogeneous or homogenous avid contrast enhancement by the solid components of the tumor [29]. Calcification is uncommon [29,47].

With magnetic resonance imaging, the solid components of the tumor display different signal intensity on T2-weighted imaging, depending on the amount of fibrous stroma. However, a strong low-signal intensity on T2-weighted images is not commonly encountered [46,47,55,56]. Cystic portions usually display with high signal on T2-weighted images and with low signal on T1-weighted images, but sometimes hardly noticeable high-signal intensity on T1-weighted images can be seen [55]. Solid components tend to show avid homogenous or heterogeneous contrast uptake on gadolinium-enhanced images, whereas cystic areas are non-enhancing [29].

### Treatment and prognosis

Due to the lack of standardized protocol guidelines, there is no uniform therapeutic approach to these tumors [57]. The type of surgical approach depends on patient age and preference, tumor stage, and tumor differentiation. Most of these tumors are unilateral and confined to the ovary at the time of clinical diagnosis. Hence, in patients at reproductive age, with stage Ia/Ib disease, who prefer fertility preservation, unilateral salpingo-oophorectomy with or without the exploration of the contralateral ovary is appropriate [58].

In women with intermediate or poorly differentiated stage Ic disease, unilateral salpingo-oophorectomy with formal staging surgery is a pertinent treatment [58]. The patient in this report was involved in the decision-making process and expressed her wish to preserve fertility. She had Ic stage disease (rupture of the capsule), so unilateral salpingo-oophorectomy plus standard staging surgery (partial omentectomy and several peritoneal biopsies) were performed.

According to Brow et al. and Thrall et al., lymphadenectomy may be omitted from the surgery staging procedure of sex cord-stromal tumors due to the rarity of lymph node metastases in these types of tumors [59,60].

Women who do not wish to preserve their fertility or with advanced stage disease (stage II or higher) should be considered for radical surgery with total abdominal hysterectomy and bilateral salpingo-oophorectomy or cytoreductive surgery [58].

Management of SLCTs after surgery is not clearly defined and is based on limited studies [57,58].

Adjuvant chemotherapy is recommended for patients with advanced disease, intermediate and poor tumor differentiation, retiform pattern, and presence of heterologous elements [58]. A retrospective study of 21 patients treated in 11 centers conducted by Sigismondi et al. concluded that the prognosis of patients with well differentiated SLCT is excellent without adjuvant chemotherapy. In contrast, patients with advanced stage or with intermediate or poorly differentiated tumors appear to benefit from adjuvant chemotherapy [51]. In this case, the patient underwent adjuvant chemotherapy with the BEP regimen (bleomycin, etoposide, and cisplatin) because of poor tumor differentiation and the presence of heterologous elements. BEP is an effective first-line chemotherapeutic regimen [57,58]. However, toxicities and lack of durable activity associated with BEP may limit its use [58,61].

A retrospective review of 44 patients with sex cord-stromal tumors concluded that taxanes may be an effective treatment, and combination therapy with taxanes and platinum warranted further investigation [61]. Currently, a randomized phase II trial conducted by the Gynecologic Oncology Group of the United Cancer Institute is comparing the effectiveness of paclitaxel and carboplatin versus BEP in the treatment of advanced and recurrent sex cord-stromal tumors.

SLCTs are typically associated with a good prognosis.

Disease behavior correlates with the stage and with the degree of differentiation at the time of diagnosis [47,62].

Young and Scully reviewed 207 cases of SLCTs, and clinical malignancy was identified in 18%. All well-differentiated tumors were benign; however, 11% of the intermediately differentiated tumors, 59% of poorly differentiated tumors, and 19% of tumors with heterologous elements were malignant [62]. Sigismondi et al. reported 5-year survival rates of 100% for patients with well-differentiated tumors and 77.8% for those with intermediate and poorly differentiated tumors. Patients with stage I SLCTs had a 5-year overall survival rate of 92.3%, whereas that for patients with stage > I disease was 33.3% [51].

In contrast to other sex cord-stromal tumors, such as granulosa cell tumors, which may have late recurrence of the disease, malignant SLCTs tend to recur 2 to 3 years after the initial diagnosis [57, 63]. Recurrence is usually confined to the pelvis and abdomen, and typically presents without distant metastases. Without exception, when reviewing the cases reported in the literature of AFP-producing SLCTs, including the case we present here, serum AFP fell to undetectable levels postoperatively. There are few reports of AFP-producing SLCT recurrences, and to our knowledge, all cases had raised AFP levels at the onset of the ovarian mass [3,22]. Nonetheless, there is limited information on surveillance strategies and the prognosis of patients with AFP-producing SLCTs.

National Comprehensive Cancer Network surveillance guidelines for sex cord-stromal tumors recommend physical examination, review of symptoms, and measurement of tumor markers if they were initially elevated every 2-4 months during the first 2 years and every 6 months thereafter [63,64]. There are insufficient data to support the periodic use of radiographic imaging. Computed tomography imaging and measurement of tumor markers are recommended when there is suspicion of recurrence [63,64].

### TEACHING POINT

Although rare, AFP-producing non-germ cell tumors must be considered in the presence of an ovarian mass associated with an elevated serum AFP level. In a young woman, the 2 major diagnoses that need to be taken into account are SLCTs and juvenile granulosa tumors, which must be distinguished clinically, radiologically, and pathologically from AFP-producing germ cell tumors, such as yolk sac tumors, immature teratomas, and embryonic carcinomas, as the diagnosis affects the treatment plan for the patient.

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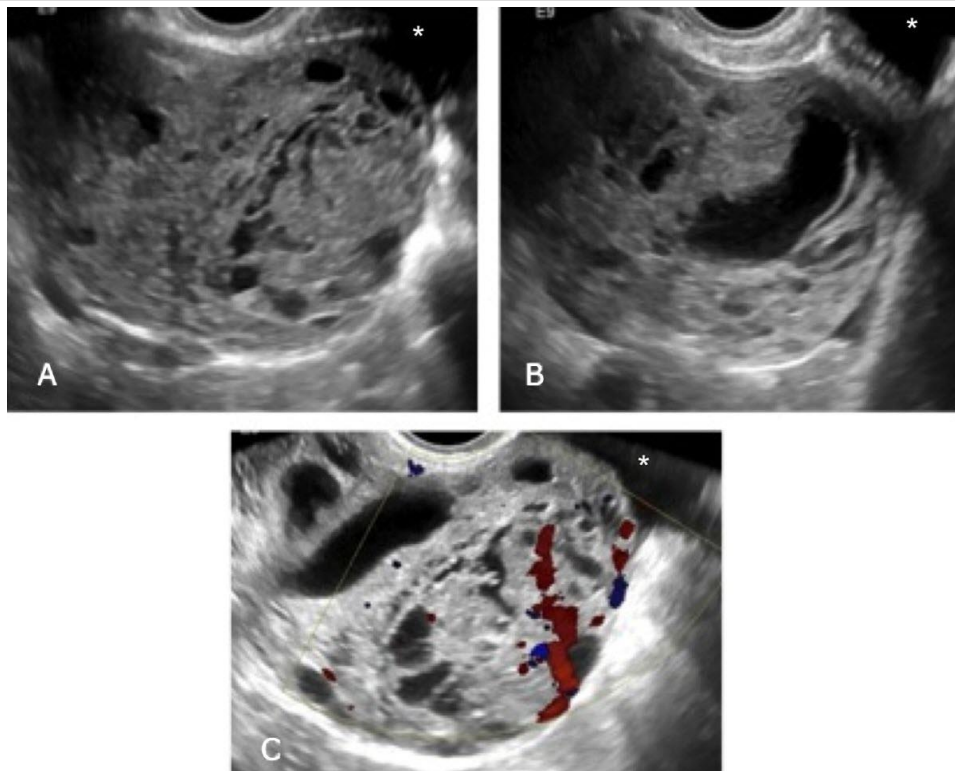
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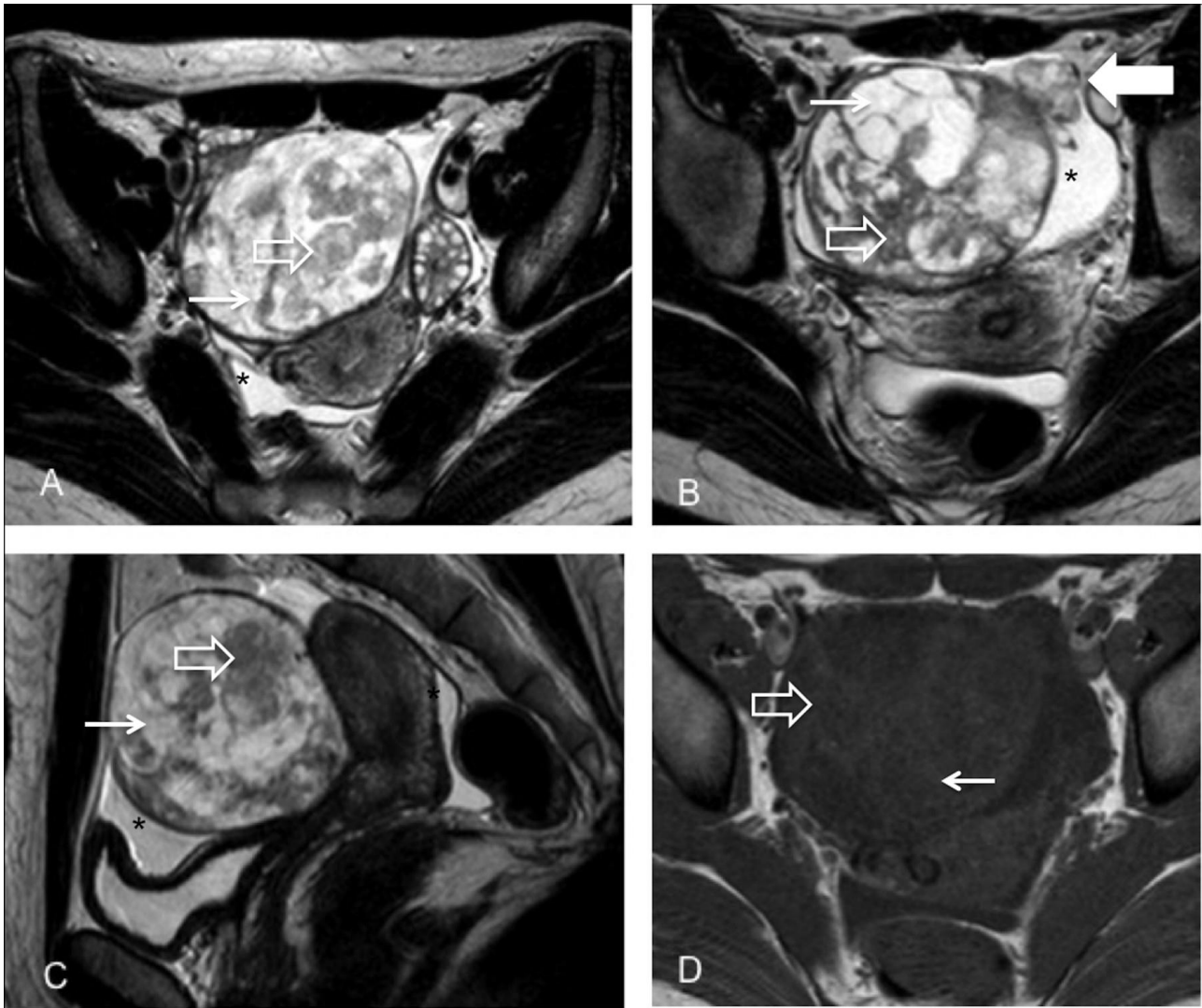
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FIGURES



**Figure 1:** A 19-year-old female patient with a poorly differentiated Sertoli-Leydig cell tumor of the right ovary with heterologous gastrointestinal epithelium. Pelvic transvaginal ultrasound with a 10-MHz endocavitary probe. A heterogeneous complex-appearing mass, predominantly solid with focal cystic areas was detected in the right ovary (1A and 1B). The tumor exhibited abundant and low resistance flow within the solid areas on color Doppler imaging (1C). These sonographic characteristics, combined with a small amount of pelvic ascites (white asterisks; 1A, 1B, and 1C), suggested a malignant nature.



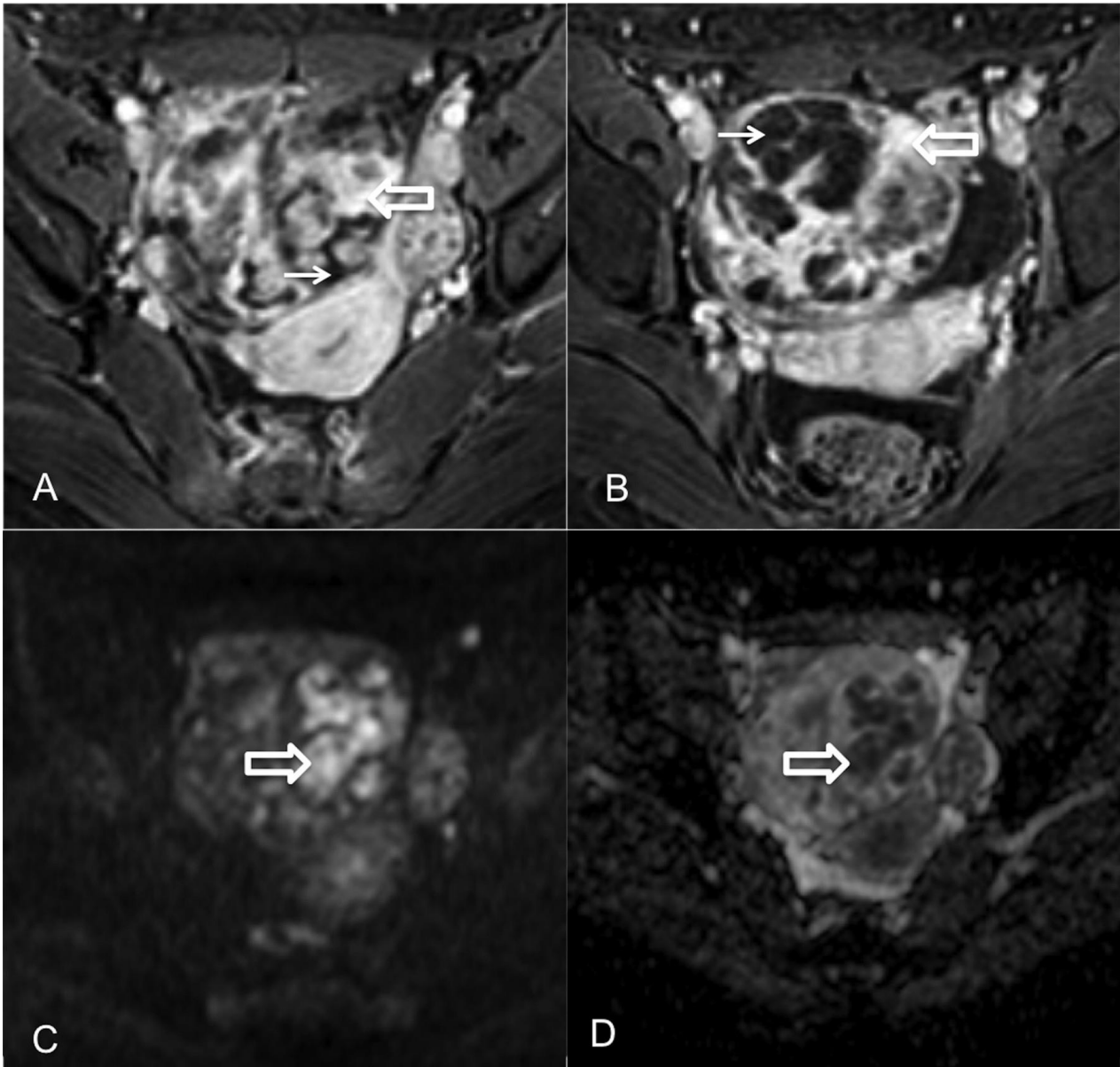
**Figure 2:** A 19-year-old female patient with a poorly differentiated Sertoli-Leydig cell tumor of the right ovary with heterologous gastrointestinal epithelium.

Pelvic magnetic resonance imaging.

Axial T2-weighted images (2A and 2B), sagittal T2-weighted image (2C, and axial T1-weighted image (2D) showed a heterogeneous, well defined tumor of the right ovary. The tumor was well encapsulated, with the exception of a protruding eccentric anterior segment (white arrow; 2B). Solid components showed intermediate T2 signal (white-bordered arrows; 2A-C) and T1 signal intensity similar to muscle (white-bordered arrow; 2C), while other components had high T2 signal (thin arrows; 2A-C) and low-signal on T1-weighted images (thin arrow; 2D), suggesting a cystic nature. A small amount of ascites was identified in the pelvic recesses (black asterisks; 2A-C).

(Philips Intera Pulsar 1.5T: T1-weighted images [TR = 604; TE = 10]; T2-weighted images [TR = 3500; TE = 100]).





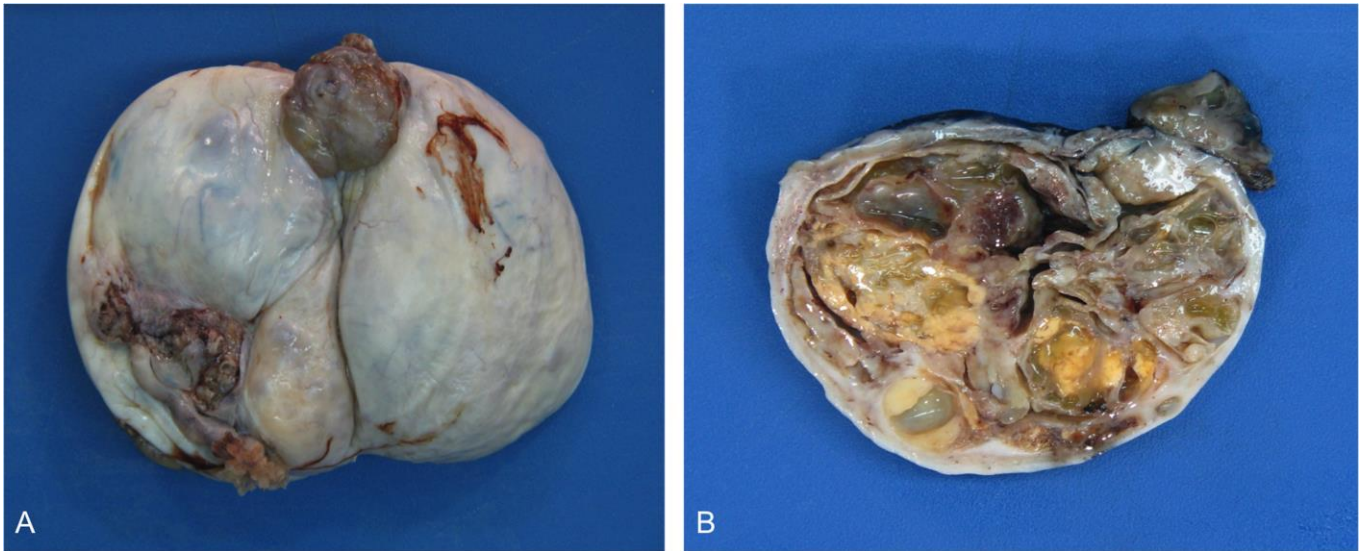
**Figure 3:** A 19-year-old female patient with a poorly differentiated Sertoli-Leydig cell tumor of the right ovary with heterologous gastrointestinal epithelium.

**Pelvic magnetic resonance imaging.**

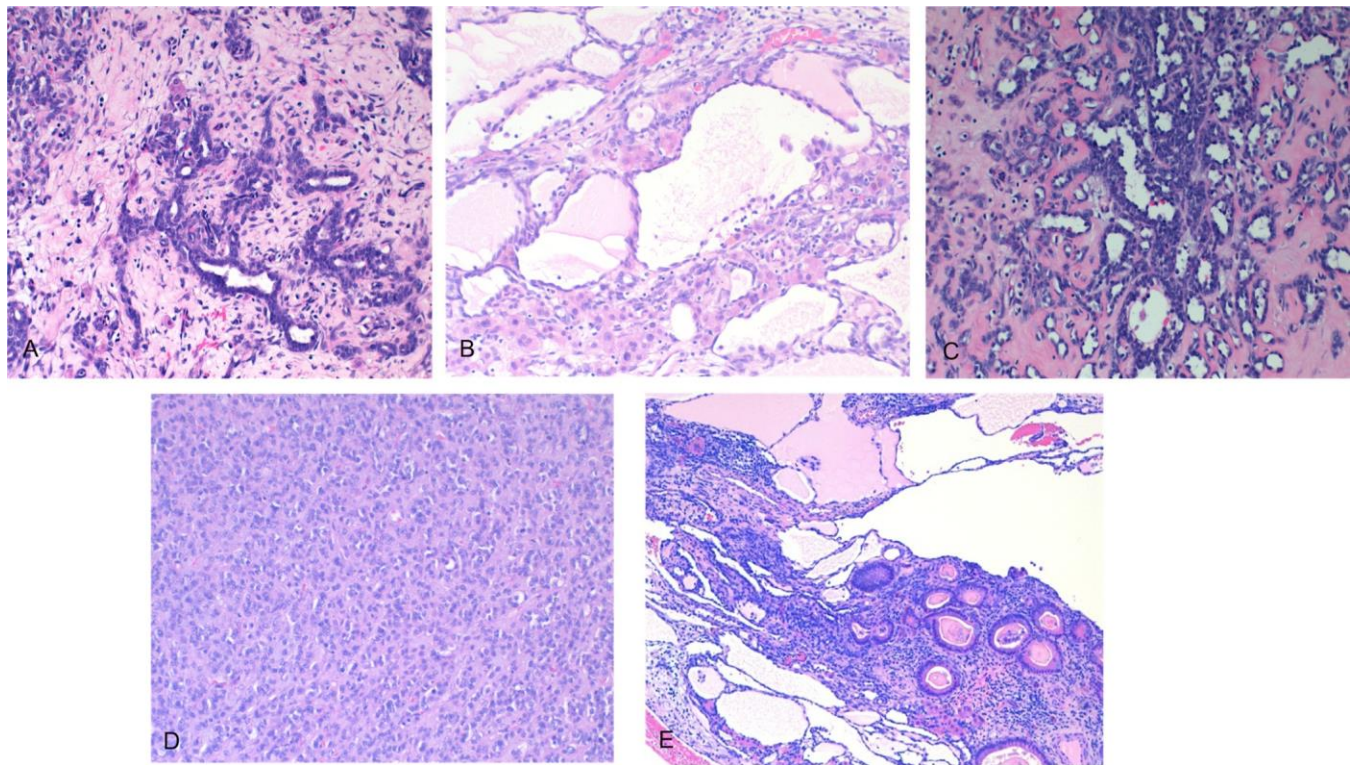
Axial gadolinium-enhanced fat-suppressed T1-weighted images showed avid contrast uptake by the solid components of the tumor (white-bordered arrows; 3A and 3B), without enhancement of the cystic components (thin arrows; 3A and 3B).

Axial diffusion-weighted image (b1000s/mm<sup>2</sup>) showed high-signal intensity of the solid components of the tumor (white-bordered arrow; 3C), while the apparent diffusion coefficient map showed low-signal intensity of the same components (white-bordered arrow; 3D). These findings were consistent with restricted diffusion.

(Philips Intera Pulsar 1.5T: dynamic contrast-enhanced image obtained with a 3-dimensional gradient-recalled echo T1-weighted sequence after the administration of 0.1 mmol/kg of gadopentetate dimeglumine at a rate of 2 mL/sec acquired 2 minutes after injection; axial diffusion-weighted image [TR = 2882; TE = 67; flip angle 90° performed with b values of 1000 s/mm<sup>2</sup>]).



**Figure 4:** A 19-year-old female patient with a poorly differentiated Sertoli-Leydig cell tumor of the right ovary with heterologous gastrointestinal epithelium. Macroscopic examination revealed an enlarged ovary (140 g and 8 cm in length at the longest axis) with a smooth gray-whitish surface with rupture and tumor extravasation (4A). On cut sections, the ovary was replaced by a variegated solid and multicystic tumor (4B).



**Figure 5:** A 19-year-old female patient with a poorly differentiated Sertoli-Leydig cell tumor of the right ovary with heterologous gastrointestinal epithelium. Microscopic examination of hematoxylin- and eosin-stained sections (200 $\times$ ). The tumor was composed of Sertoli cells arranged in hollow tubules and inter-anastomosing cords in an edematous stroma (5A), and in some areas, marked edema produced a microcystic pattern of the Sertoli cell component, intermingled with Leydig cells (5B). In addition, areas of slit-like spaces of the Sertoli cell component were observed in a background of hyalinized stroma (5C). There were also poorly differentiated areas composed of immature Sertoli cells growing in a predominantly diffuse pattern with small tubular differentiation and a high mitotic index (5D). A small focus (0.9 mm) of heterologous elements was found, composed entirely of mature intestinal gland (5E).

	Ultrasound	Computed Tomography	Magnetic Resonance Imaging
<b>Sertoli-Leydig cell tumor</b>	<ul style="list-style-type: none"> <li>• Unilateral well-defined hypoechoic and heterogeneous mass predominantly solid with multiple cystic areas</li> <li>• Abundant flow within the solid areas on color Doppler imaging</li> <li>• Ascites may be present</li> </ul>	<ul style="list-style-type: none"> <li>• Well defined unilateral soft tissue density mass with avid contrast enhancement by the solid components of the tumor</li> <li>• Ascites may be present</li> <li>• Presence of enlarged lymph nodes - rare</li> </ul>	<ul style="list-style-type: none"> <li>• Solid components -T1 signal intensity similar to muscle; intermediate/low T2 signal depending on the amount of fibrous stroma; avid contrast uptake</li> <li>• Cystic areas - high T2 signal; low-signal on T1-weighted images</li> <li>• Ascites may be present</li> <li>• Presence of enlarged lymph nodes - rare</li> </ul>
<b>Yolk sac tumor</b>	<ul style="list-style-type: none"> <li>• Unilateral mainly solid large mass with cystic areas</li> <li>• Ascites may be seen</li> </ul>	<ul style="list-style-type: none"> <li>• Unilateral large complex mass</li> <li>• Strong enhancement of the solid areas</li> <li>• Sometimes central necrosis and hemorrhage are present</li> <li>• Enlarged lymph nodes, ascites, peritoneal implants, and invasion of the adjacent structures may be present</li> </ul>	<ul style="list-style-type: none"> <li>• Solid component –hypo-intense signal on T1-weighted images; heterogeneous high T2 signal; avid contrast uptake.</li> <li>• Foci of hemorrhage – high T1 signal</li> <li>• Necrosis – high T2 signal</li> <li>• Enlarged lymph nodes, ascites, peritoneal implants, and invasion of the adjacent structures may be present</li> </ul>
<b>Embryonal carcinoma</b>	<ul style="list-style-type: none"> <li>• Unilateral, very large, predominantly solid mass with extensive anechoic areas reflecting necrosis</li> <li>• Ascites may be present</li> </ul>	<ul style="list-style-type: none"> <li>• Unilateral, very large, predominantly solid mass with large low- attenuation areas reflecting necrosis and/or with high attenuation areas reflecting hemorrhage</li> <li>• Avid contrast uptake by the solid portions of the tumor</li> <li>• Ascites, retroperitoneal lymphadenopathies, and peritoneal implants may be seen</li> </ul>	<ul style="list-style-type: none"> <li>• Predominantly low T1 signal mass that may display areas of high-signal intensity due to the presence of hemorrhage</li> <li>• Predominantly high T2 signal intensity mass due to the presence of necrosis</li> <li>• Avid contrast uptake by the solid portions of the tumor</li> <li>• Ascites, retroperitoneal lymphadenopathies, and peritoneal implants may be seen</li> </ul>
<b>Immature teratoma</b>	<ul style="list-style-type: none"> <li>• Heterogeneous predominantly solid adnexal large mass with cystic components</li> <li>• Foci of fat - echogenic foci</li> <li>• Calcifications may be present - highly echogenic foci with posterior acoustic shadowing</li> </ul>	<ul style="list-style-type: none"> <li>• Large heterogeneous mass with enhancing solid areas and cystic components</li> <li>• Presence of fat – foci of low-attenuation</li> <li>• Calcifications- foci of high-attenuation</li> </ul>	<ul style="list-style-type: none"> <li>• Large heterogeneous mass with enhancing solid areas and with cystic components</li> <li>• Presence of fat – high T1 and T2 signal intensity; signal loss on fat sat sequences</li> </ul>
<b>Juvenile granulosa cell tumor</b>	<ul style="list-style-type: none"> <li>• Unilateral, multicystic large mass with solid components</li> <li>• Irregular thin or thick septations</li> <li>• If cystic hemorrhage – heterogeneous echogenicity within the cysts</li> </ul>	<ul style="list-style-type: none"> <li>• Unilateral multicystic large mass with enhancing solid components and septations</li> </ul>	<ul style="list-style-type: none"> <li>• Solid components - intermediate T2 signal; contrast uptake</li> <li>• Thick septations – low signal on T2-weighted images</li> <li>• Cystic areas - high T1 signal due to hemorrhage; fluid-fluid levels</li> </ul>

**Table 1:** Differential diagnosis table of ovarian tumors associated with an elevated serum level of alpha-fetoprotein in a young woman.

<b>Etiology</b>	Type of sex cord-stromal tumor of the ovary Proliferation of variable proportions of Sertoli cells, Leydig cells, and fibroblasts
<b>Incidence</b>	0.5% of all ovarian neoplasms
<b>Gender ratio</b>	Women
<b>Age predilection</b>	75% of cases under 30 years of age
<b>Risk factors</b>	Unknown
<b>Treatment</b>	<ul style="list-style-type: none"> <li>• Unilateral salpingo-oophorectomy (Stage I; women in reproductive age)</li> <li>• Total abdominal hysterectomy and bilateral salpingo-oophorectomy (≥ Stage II)</li> <li>• Adjuvant chemotherapy (≥ Stage II; presence of poorly differentiated and heterologous elements)</li> <li>• Adjuvant chemotherapy + radiotherapy – unknown</li> </ul>
<b>Prognosis</b>	<ul style="list-style-type: none"> <li>• Most of these tumors behave in a benign fashion</li> <li>• Malignant behavior in 10–30% of cases</li> </ul>
<b>Findings on imaging</b>	<ul style="list-style-type: none"> <li>• Ultrasound – Well-defined hypoechoic and heterogeneous mass, predominantly solid with multiple cystic areas and abundant flow within the solid areas on color Doppler imaging</li> <li>• Computed tomography - Soft tissue density mass with avid contrast enhancement</li> <li>• Magnetic resonance imaging - Solid components show intermediate T2 signal and T1 signal intensity similar to muscle; fibrous stroma show low T2 signal; cystic areas have high T2 signal and low signal on T1-weighted images; gadolinium-enhanced images show avid contrast uptake by the solid components of the tumor</li> <li>• Histopathologic findings - Definitive diagnosis</li> </ul>

**Table 2:** Summary table of Sertoli-Leydig cell tumor characteristics

**ABBREVIATIONS**

AFP - Alpha-fetoprotein  
 BEP - Bleomycin, etoposide, and cisplatin regimen  
 β-hCG - Human chorionic gonadotropin  
 CA-125 - Cancer antigen 125  
 SLCT- Sertoli-Leydig cell tumor

**KEYWORDS**

Sertoli-Leydig tumor; Alpha-fetoprotein; Sex cord-stromal tumor; Ovarian tumor; Ultrasound; Magnetic resonance

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