

CASE # 3

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History: 65 year old woman presented with abdominal fullness, pressure and early satiety. An ultrasound and abdominal CT revealed a right ovarian mass and at resection it measured 20 x 18 x 12 cm. An H&E slide is from the ovarian tumor. (Case contributed by Dr. Yibin Zhang, Citrus Valley Medical Center, Queen of the Valley Campus. West Covina).

Gross and Microscopic: The right ovary measured 20 x 18 x 12 cm. Left tube and ovary were within normal limits. The uterus had an endometrial polyp, adenomyosis and leiomyoma but no tumor seen. No tumor in 31 lymph nodes.

The tumor is composed predominately of sheets of primitive round to oval cells with fine granular chromatin, small nucleoli, and brisk mitotic activity. Focally the tumor shows spindled anastomosing columns (figure 1). The tumor has areas of well-differentiated mucinous glands with gastrointestinal type epithelium (figure 2), cartilage and fetal type skeletal muscle (figure 3). In rare areas there was an irregular network of elongated, slit like tubules (figure 4).

Figure 1:

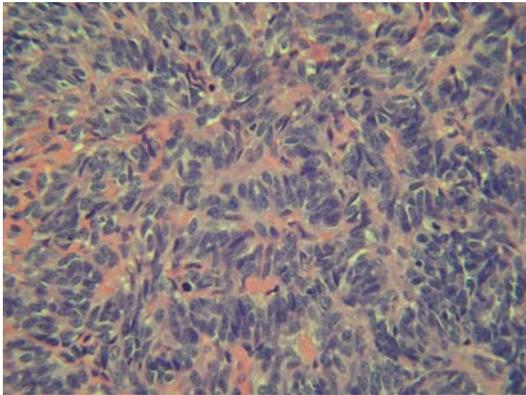


Figure 3:

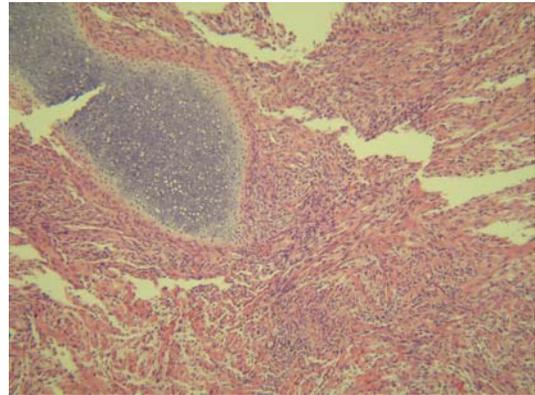


Figure 2:

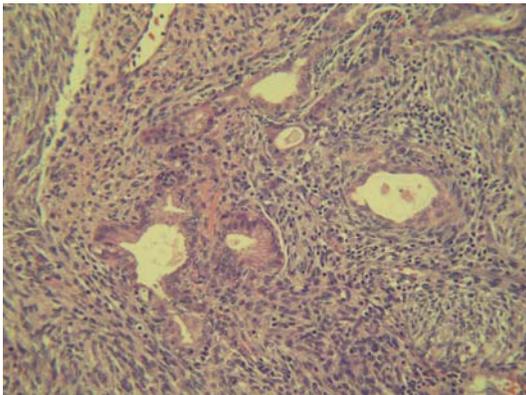
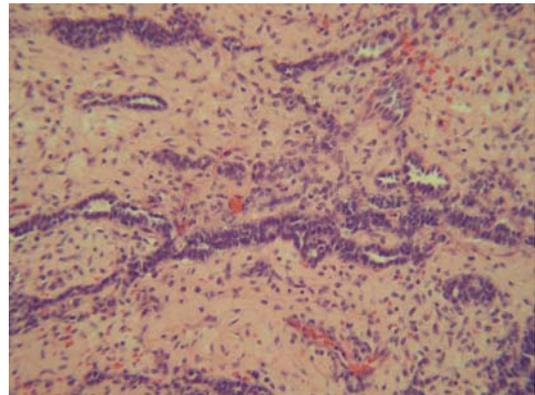


Figure 4:



Immunohistochemistry: Stromal component positive for keratin, inhibin (focally), calretinin, melan A and rare scattered clusters positive for CD10 and CD99 also showed focal positivity. The tumor is negative for EMA, chromograin, and snaptophysin.

Diagnosis: Poorly differentiated Sertoli-Leydig cell tumor with heterologous elements.

Discussion: The majority of this tumor was poorly differentiated with both areas of spindle cells resembling stromal elements and other areas that resembled poorly differentiated carcinoma. However, well-differentiated heterologous elements (mucinous glands, skeletal muscle, smooth muscle, and few islands of cartilage) seemed to be an integral component. The histologic differential of this tumor with a biphasic pattern includes carcinosarcoma (MMT), a teratoma with dedifferentiation, and Sertoli-Leydig cell tumor with heterologous elements. In this patient's age group (65 yrs) a carcinosarcoma would be much more likely, but usually in MMT the epithelial and mesenchymal components are high grade. In this tumor the glandular elements are all well-differentiated and the mesenchymal elements although immature, are cytologically bland. The immunohistochemistry was helpful as MMTs are usually positive for EMA and keratin and negative for the stromal marker inhibin (Costa, 1992). In teratomas, there is usually an ectodermal component and they have not been reported to have the gonadal stromal elements seen in this tumor. The tumor was categorized as a Sertoli-Leydig cell tumor with heterologous elements. The positive Calretinin and Melan A are also supportive (Steward, 1999). See table below for IHC role in differential diagnosis.

Sertoli-Leydig cell tumors (SLCT) are rare and represent only about 1% of all ovarian tumors. They are almost always unilateral and present in women of reproductive age (ave age ~25 yrs) but have been seen in children and rarely in post-menopausal women (Oliva, 2005). Hormonal symptoms, usually androgenic, are seen in about 50% of the patients. The histologic features of the tumor are quite variable and they enter into the differential diagnosis of many more common ovarian tumors. WHO classifies SLCT as well-differentiated, intermediate-differentiated, poorly differentiated and retiform pattern. Heterologous elements can be seen in either the intermediate or poorly differentiated tumors, but is not a feature of the well-differentiated cases. The prognosis is generally good but there is data to suggest tumors with a prominent retiform component have a somewhat poorer prognosis than similar tumors without this pattern. More aggressive behavior is also associated with the poorly differentiated tumors, those with heterologous mesenchymal elements and rupture.

TABLE 2. Sex cord-stromal tumors of the ovary and their mimics: Differential immunohistochemical profiles

	Keratin	EMA	Inhibin	Calretinin	Melan-A	CD99	CD10	Chromogranin
Sex cord-stromal tumor	+	-	+	+	+	+/-	+/-	-
Endometrioid carcinoma	+	+	-	-/+	-	-/+	-/+	-
FATPWO	+	-/+	-/+	+	-	-	+	-
Carcinoid tumor	+	-	-	-	-	-	-	+
Endometrioid stromal tumor	-/+	-	- (*)	- (*)	-	- (*)	+	-
Steroid cell tumor	+/-	-	+	+	+	-	+/-	-

(*) Except in sex cord areas.

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