

# OVARIAN SERTOLI-LEYDIG CELL TUMOR WITH A PREDOMINANT RETIFORM DIFFERENTIATION

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**SUMMARY:** In the present study, light microscopic features of a tumor localized in the right ovary of a 17 years old-girl is presented. The patient had symptoms of virilization such as clitoral hypertrophy and defeminization such as amenorrhea. The dimension of the tumor was 14 cm, and microscopically it was a moderately differentiated sertoli Leydig cell (SLC) tumor made up largely of retiform areas.

## INTRODUCTION

Sertoli-Leydig cell (SLC) tumors comprise less than 0,5 % of all ovarian tumors. They're most frequently seen in young patients and between ages 20-30, but can be seen in all age groups. These tumors frequently have a virilizing character, but some do not have demonstrable endocrine effect of this type and some others have estrogenic effects (6,8).

SLC tumors don't have specific characters. Bilateral involvement is seen in 3 % (6,8).

Retiform differentiation is encountered in 10 % of SLC tumors. Areas showing retiform differentiation are composed of blunt papillae with hyalinized or edematous cores and irregular cleft-like spaces. These formations are lined by cuboidal cells. This appearance resembles the retia of mature gonads (7).

Serov and scully (10) classified the SLC tumors as well, moderately and poorly differentiated and according to the presence of heterologous elements, a fourth group is also included. In this classification, retiform differentiation is not mentioned. But some authors categorize them as a different group (5).

## MATERIAL AND METHOD

The first case diagnosed as "retiform type SLC tumor" in the Department of Pathology, İstanbul Medical Faculty is examined histopathologically. 19 sections for histologic examination from different parts of the tumor are taken.

Sections from paraffin blocks are stained with hematoxylin-Eosin (HE) stain.

## CASE (Biopsy prot. 13279/90)

17 years-old girl having a months-period of amenorrhea, complained of an abdominal mass and sense of suprapubic discomfort since the last two months. The clinical diagnosis was "a right ovarian tumor" and a right ooforectomy was performed.

In gross examination, the tumor was 14 x 11 x 11 cm. in this longest dimensions and had a well-vascularized capsule. The external surface of the tumor was grayish-pink in color. The mass had a soft consistency. In its cut sections grayish solid areas and between them, various cystic spaces of variable size containing a yellowish serous fluid were seen.

Microscopically, in some areas of the tumor small tubules with lumina or cord-like structures of Sertoli cells were seen. Sertoli cells of this type were surrounded by fusiform cells (Fig 1). These mesenchymal areas were moderately cellular. In a few sections, Leydig cells with vesicular nuclei and acidophilic cytoplasm, and few in number, interspersed singly or in small groups were seen (Fig 2). Areas with these features constituted a very small portion of the tumor. In all other parts of the tumor, irregular tubules of various sizes and cystic spaces lined by cells having vesicular nuclei and ill-defined cytoplasmic contours were seen in a loose, myxoid stroma (Fig 3). These cells, elsewhere, had the tendency of lining a surface and papillae with a loose edematous stroma. Cells lining all these different formations were cuboidal or flattened. They were usually monolayered, but in some areas, were pseudostratified (Fig 4).

With these microscopic findings, our diagnosis was "moderately differentiated SLC tumor with a predominant re-

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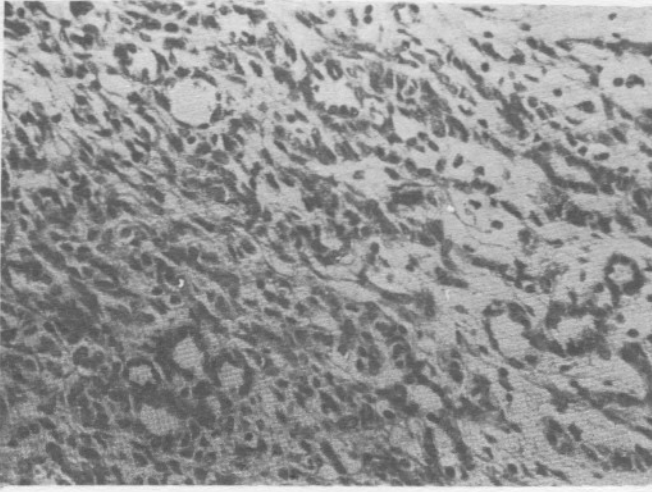


Figure 1 - Sertoli cells forming tubules and cord-like structures and in between them a stroma formed of fusiform cells (Biopsy No: 13279/90, H.E.x310)

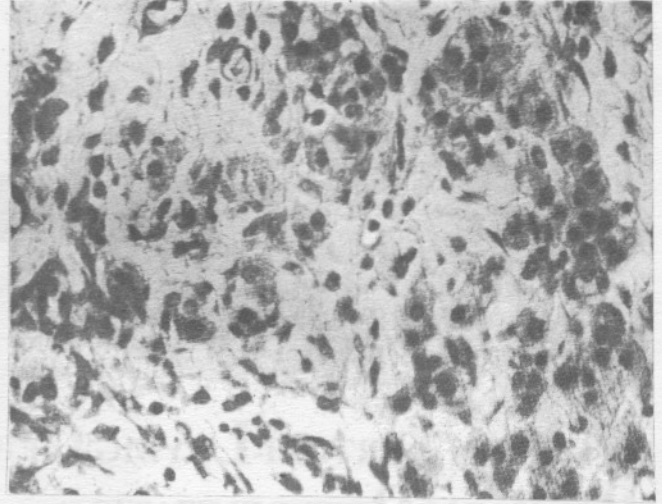


Figure 2 - Scattered single or small groups of Leydig cells (Biopsy No: 13279/90, H.E x 500)

tiform differentiation".

## DISCUSSION

Rete ovarii is an embryologic remnant which, contrarily to rete testis, does not function. Morphologically, it is composed of irregular anastomosing cleft-like spaces lined by flattened and cuboidal cells (9). Fusiform-shaped cells constitute its stroma which is not in continuity with, but which has a similarity to ovarian stroma. Rete testis, the analogous of this structure in males, tubuli recti and efferent ductules are all lined by an epithelium having the same origin with the Sertoli cells. Epithelium of rete testis differentiates from this epithelium, its cellular appearance varying from cuboidal to flattened (1). SLC tumors, histogene-

sis of which has not yet been clarified, originate from mature or immature cells of testis. And because of the embryologic relationship between the rete epithelium and sertoli cells, retiform differentiation can occur in SLC tumors (7).

SLC tumors occur most frequently in young women (8). Average age in cases with a predominant retiform pattern is reported as 17 (7). Our patient was also 17 years old.

SLC tumors clinically, can be accompanied by some endocrin manifestations. They can cause virilizing and defeminizing symptoms and sometimes, to the contrary, estrogenic manifestations. Some of the SLC tumors do not have any endocrine effect (8). The patient in our case, had clitoral hypertrophy and amenorrhea as symptoms of virilization and defeminization.

SLC tumors are usually unilateral (3,4,11). They have a

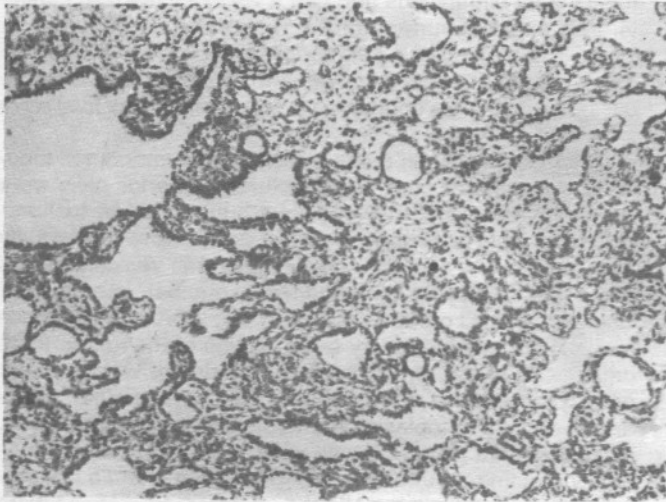


Figure 3 - Clefts and adenoid formations in different sizes in a loose and myxoid stroma (Biopsy No: 13279/90 H.Ex 125)

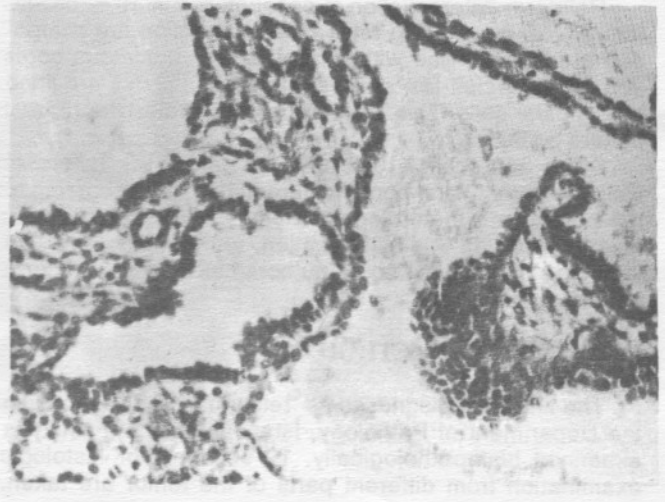


Figure 4 - Cubic and flattened cells paving adenoid formations, cysts and papillary formations in a monolayered form and showing pseudostratification at one side (Biopsy No: 13279/90 H.E x 500).

soft consistency and they contain solid areas yellowish-gray in color, with cystic parts, hemorrhages and necrosis (6). There was a unilateral involvement in our case and the tumor had gross properties similar to those mentioned above.

These tumors are made up of different amounts of Sertoli cells and Leydig cells in different stages of differentiation (6). Retiform differentiation is seen in 10 % of cases (12). These areas may be focal or sometimes dominant (7). Heterologous elements such as glands lined by epithelium of gastrointestinal type, areas of carcinoid tumor, skeletal muscle, cartilage and hepatoid differentiation may accompany these retiform areas (3,10,11,13). Psammoma bodies may also be seen (7). The tumor in our case showed retiform differentiation in large areas and Sertoli cells forming tubules and cords, few Leydig cells and a stroma made up

of fusiform shaped mesenchymal cells. There were neither heterologous elements nor psammoma bodies in our case.

The recognition of retiform type SLC tumors is important for the distinction of this tumor from papillary serous tumors, malignant mixed müllerian tumor with heterologous elements and endodermal sinus tumor. The first two tumors usually occur in an older age group and don't show other features of SLS tumors. Endodermal sinus tumor is a histologically more primitive tumor and stain positively with AFP (7).

The case, as being an example of a SLC tumor with a dominant retiform pattern causes the necessity for us, to search for retiform differentiation and its proportion in all SLC tumor cases in our files and in future, so as to define its influence on the biologic behaviour of these tumors.

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