SERTOLI CELL ADENOMA IN MORRIS SYNDROME

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SUMMARY

An unusually large Sertoli cell adenoma was detected in a patient with Morris syndrome and there is a description of the uncommon hormonal data found in this particular case which are not typical for this kind of neoplasia.

The patient is alive and well at the present time which is now three years after surgical intervention.

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Sertoli cell adenomas are distinctive tumors that are the most frequent type of neoplasia found in the immature testis of patients with testicular feminization syndrome (T.F.S.) although it is difficult to assess their true incidence ([1, 2, 9, 10]). The unusual case discussed herein is one of a tumor of exceptionally large proportions ([3]) which we present together with the correlated hormonal data.

CASE REPORT

A 60-year-old female patient, 169 cm tall and weighing 74 kg, was admitted to our hospital for hepatopathy and a pelvic mass was noted upon physical examination. She was happily married and had had an apparently normal sexual life, although she had never menstruated. The scalp hair was distributed normally while pubic hair was very scanty and axillary hair was totally absent. Breast and nipple development appeared to be normal and external genitalia were normal as well. The vagina was 7 cm in length and ended in blind pouch without a cervix. The patient related that her aunt had never menstruated either, and we also found a sister and a niece affected by the same syndrome (Fig. 1).

Karyotypic analysis of a peripheral blood sample revealed a 46 XY pattern. Hormonal studies (Table I) showed high levels of testosterone, estradiol, 17 ketosteroids and luteinizing hormone as compared to those of normal males while the urinary estrogens and the 17 hydroxy corticosteroids were in the normal range. During surgery a large tumor mass, measuring $27 \times 21 \times 14$ cm (Fig. 2) whose weight was not determined, was found in place of the left gonad and subsequently removed. Simultaneously the right gonad, which was the size of a normal testis, was also removed. The tumor mass was solid, grey-tan in color and had a smooth external surface. Uterus and fallopian tubes were absent and there was no evidence of metastatic disease.

Histological examination was performed on various peripheral and central samples of the tumor. The peripheral samples were surrounded by a thin layer of connective tissue which was externally coated with another layer of mesothelial cells. The tumoral tissue was identical in both the central and peripheral areas of the mass: it consisted of seminiferous tubules, characteristic of those found in the immature testis, which were surrounded by Sertolian elements having a relatively large and clear cytoplasm (Fig. 3). The tubular basal membrane seemed to be of normal thickness (Fig. 4). Occasional groups of Leydig cells, in which Reinke crystals were

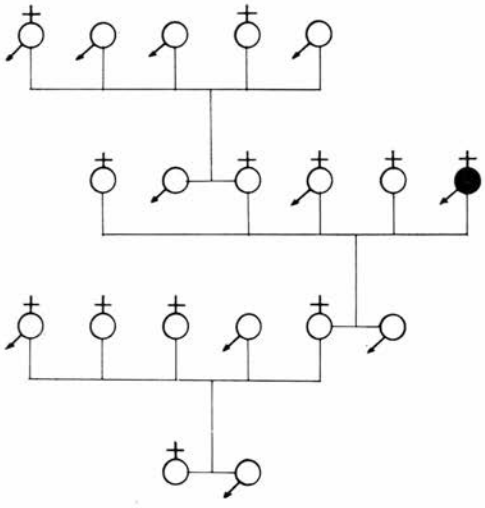

Fig. 1. — Patient's family history.

Fig. 2. — Sertoli cell adenoma. Gross appearance of the tumor encompassing the left gonad, and the right gonad.

not present (Fig. 5), were found in the interstitial space. Areas of recent necrobiosis were observed which might have been related to a thrombosis. There was no evidence of either mytosis or atypic nucleus. Since the presence of small groups of Leydig elements is frequently found in Sertoli cell adenomas, the histological diagnosis was therefore one of a "well-differentiated Sertoli cell adenoma" [4]. The right testis was also characterized by immature Sertoli tubules which showed evidence of interstitial fibrosis.

The patient's recovery after the operation was uneventful. Postoperative hormonal patterns showed significant reductions in the levels of

Table 1. — *Peripheral plasma hormonal patterns.*

	Normal values in males	Before surgery	10 days later	2 years later
Testosterone	4-9 ng/ml	38	0.5	0.2
Estradiol	20-50 pg/ml	90	30	20
Urinary estrogens	7-25 μg/24 hr	21	5	4
17 Hydroxy corticosteroids	5-23 mg/24 hr	17.2	9.2	8.5
17 Ketosteroids	8-28 mg/24 hr	47.2	3.8	2.5
LH	5-20 mU.I./ml	37	60	37
FSH	5-20 mU.I./ml	—	80	38

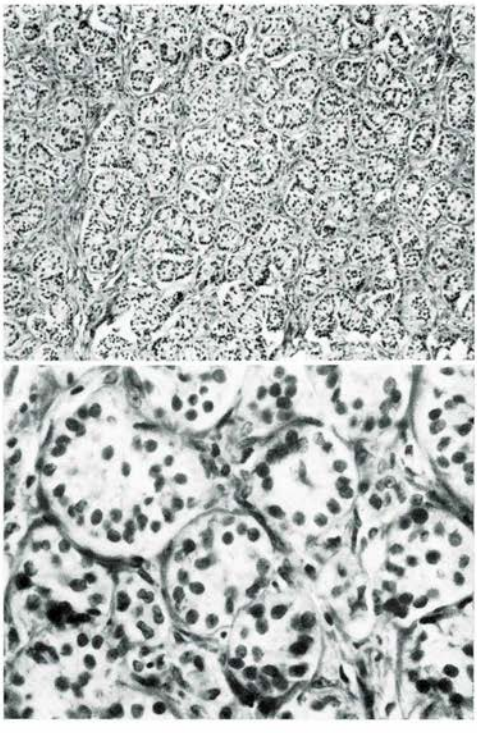

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Fig. 3. — Histological examination of the tumor. Sertoli cells surrounding the seminiferous tubules – note the immaturity. (H.E. ×100).

Fig. 4. — The tubular basal membrane. (H.E. ×400).

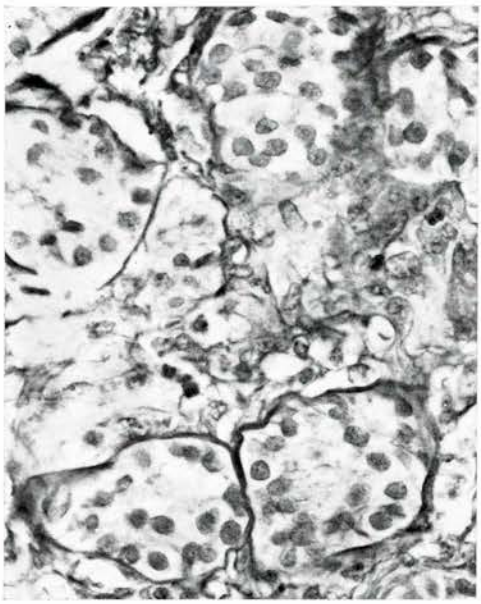

Fig. 5. — Leydig ceil lacking Reinke crystals. (V. Gieson ×400).

testosterone, estradiol, urinary estrogens, 17 hydroxy corticosteroids and 17 ketosteroids and increased levels of LH and FSH (Table 1). (Preoperative FSH level had not been determined).

The patient is well and free of active disease three years following surgery and hormonal patterns are normal (Table 1).

DISCUSSION

The incidence of testicular tumors in patients with T.F.S. appears to be approximately 25 percent ([5]). However, only between zero and 8 percent of these tumors have malignant characteristics ([6, 7, 8]). As far as benign neoplasias are concerned, Sertoli cell adenomas are the most common but they rarely reach the dimensions seen in this case ([3]). In other similar reports the hormonal data were not presented. Whereas in patients with small Sertoli cell adenomas normal levels of testosterone, estradiol, urinary estrogens and 17 ketosteroids have been reported, in this case the high levels of hormones may be related to the presence of Leydig cells in such a large tumor mass. In fact, the hormone levels decreased considerably immediately after gonadectomy. Two years after surgery the even lower hormone levels together with a clinical examination of the patient which included laparoscopical observation confirmed that the neoplasia was histologically benign.

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