

Sex Cord Tumor with Annular Tubules

輪狀細管形成을 隨伴한 卵巢의 未分類 性索間質細胞種(Sex-cord tumor with annular tubules)에 關한 比較形態學的 研究

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INTRODUCTION

In 1970, Scully (1970) reported 13 cases of a distinctive neoplasm that was characterized by the formation of simple and complex annular tubules and designated as a "sex cord tumor with annular tubules(SCTAT)". Six of 13 cases were associated with Peutz-Jeghers syndrome(PJS).

Scully maintained that the sex cord tumor with annular tubules arose from granulosa cells but grew in a pattern more characteristic of Sertoli cells. Hart et al. (1980) and Crissman and Hart(1981) considered SCTATs unassociated with the PJS to be granulosa cell tumors.

Tavassoli and Norris(1980) suggested a Sertoli cell origin of the SCTAT unassociated with the PJS. Recently Young et al. (1982) maintained that the SCTAT is a distinctive ovarian neop-

lasm having morphologic features intermediate between those of the granulosa cell tumor and those of the Sertoli cell tumor with a capability of differentiating in both directions.

The purpose of the present study is to define the nature of the SCTAT on the basis of light and electron microscopic study of 3 cases of the SCTAT. As a preliminary procedure to understand the processes involved in the differentiation of the gonads, a review of indifferent gonads and testes and ovaries at various stages of development was done because knowledge of their development is essential to define the nature and histogenesis of the SCTAT. A review of cryptorchid testes was also done.

MATERIALS AND METHODS

Three cases of sex cord tumor with annular tubules were obtained from three institutions. Case 1 was sent from Choong-Ang Hospital for consultative opinion. Case 2 was on file at the Department of Pathology, Korea University Medical School. Case 3 was on file both at Sacred Heart Hospital, Choong-Ang University and Incheon Gil Hospital.

Two cases were reported previously (Kim et al., 1978 and Park, 1982).

The original gross specimens(Case 1 & 2) and metastatic tumor (case 3) were available for review. The original microscopic slides from

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Clinicopathologic Features and Follow-up Data for Three Patients with SCTAT

Patient	Age (years)	Symptoms and signs	Site and size of tumor	Multi-focal	Calci-fied	Endo-metrium	Follow-up	
							Metastasis (time after initial operation)	Last known status (time after initial operation)
1	17	1. Low abd. distension and palpable mass and menorrhagia for 8 months 2. Colicky abd. pain for 2 hours	Left ovary 16×13× 10cm	—	—	NA	None	Alive, NED (3 years)
2	11	1. Abd. distention, mass and pain for 5 months 2. Bilateral enlargement of breasts and axillary and pubic hairs	Left ovary 25×15× 15cm	—	+	NA	Lost to follow-up	
3	24	1. Abd. mass for 3 months 2. Amenorrhea for 7 months	Left ovary 22×14× 12cm	—	—	NA	1. Mesenteric metastasis (12×10×10cm, 5 years and 9 months) 2. Left retroperitoneal metastasis (9.5×8×5cm, 6 years and 10 months)	Retroperitoneal metastasis (7 years and 10 months)

* NA : Not available NED : No evidence of disease

both primary and metastatic tumors were available for review. Multiple new sections were made from paraffin-block-embedded specimens in cases 1 & 2 as well as fixed, wet specimens in all cases. Sections of paraffin-embedded tissue were stained with hematoxylin-eosin (H & E) and periodic acid-Schiff (PAS).

For embryological consideration the materials consisted of 91 cases of testis and 83 cases of ovary at various stages of development and 4 cases of indifferent gonad, which were collected from registry of congenital malformation in department of pathology, college of medicine, Seoul National University, and personal collections of senior author (Ahn, G.H.).

An electron microscopic study was performed

on all the three neoplasms; two from primary tumors and one from metastatic tumor. Samples of tissue were removed from formalin, fixed in glutaraldehyde, postfixed in 2% buffered osmium tetroxide, and embedded in epon after dehydration in graded alcohols and propylene oxide. Thin sections were cut on a LKB ultramicrotome, and stained with lead citrate, and examined in a Hitachi 500 electron microscope.

RESULTS

Clinical Findings

The major clinicopathologic features and follow-up data are presented in table 1. All of the

patients were women whose age at the time of diagnosis of their ovarian tumors were 17, 11 and 24 years. All the patients presented with an abdominal mass and/or distention for 3~8 months' duration. One patient was admitted to the hospital because of colicky abdominal pain due to torsion of the tumor. All the patients presented with symptoms and signs suggesting endocrinologic manifestations including menorrhagia(case 1), bilateral enlargement of breasts and axillary and public hairs (case 2) and amenorrhea (case 3). The endometrium was not available for review in any of the cases. Review of medical records (case 1, 2 and 3) and upper G-I and barium enema examination (case 1) failed to demonstrate any evidence of intestinal polyposis or other evidences of the Peutz-Jeghers syndrome.

Operative Findings and Initial Treatment

At laparotomy, large unilateral left-sided ovarian tumors were identified in all the cases. Ascites was absent, and there was no evidence of metastasis.

Initial treatment consisted of unilateral salpingo-oophorectomy in all 3 cases. None of the patients received adjunctive radiation therapy or chemotherapy.

Pelvic mass was palpated in all the patients. Follow-up ranging from 3 years to 7 years and 10 months was available in two cases. One patient showed no evidence of tumor for three years and the other had one metastatic tumor removed and was alive with metastatic SCTAT 7 years and 10 months postoperatively.

Pathologic Findings

Gross Pathologic Findings

The tumors ranged from 16cm to 25cm in greatest dimension. The outer surfaces were smooth and glistening without evidence of rupture. The cut surfaces were yellowish or light yellow predominantly solid and nodular in two cases. The cut surface of case 3 was

described as gray white. Case 1 showed cleft-like spaces measuring up to 7cm in diameter. Case 2 and 3 contained multiple small cystic areas. The cysts contained light yellow serous fluid. Hemorrhage, necrosis and hyaline degeneration were noted in case 2 and fibrous trabeculations were identified in case 1.

Microscopic Findings

The tumors consisted of round circumscribed nests of tumor cells with central hyaline acidophilic bodies and thick prominent basement membranes. Two patterns of arrangement were noted; simple annular tubules were a minor component and consisted of paired rows of columnar tumor cells around central single hyaline acidophilic bodies. The nuclei were arranged in antipodal fashion around the acidophilic body and at the periphery of the nest. The paired rows of tumor cells formed blind tubules without any true lumen.

The predominant pattern was complex annular tubules, which were characterized by communicating blind tubules encircling multiple hyaline acidophilic bodies. Many epithelial nests were surrounded by thick basement membranes. In areas the thick basement membranes were continuous with the hyaline bodies. The hyaline bodies coalesced to obscure the cellular elements in areas. Some of the large cell nests formed round or elongated cysts containing pale acidophilic fluid, simulating macrofollicles as seen in the granulosa cell tumor, in all the cases, especially in case 2. Solid cellular nests without hyaline bodies and thick basement membranes in case 2 showed palisading of nuclei at the periphery, simulating a granulosa cell tumor. The nuclei of the tumor cells were round, ovoid or irregular with rare central grooves. Nucleoli were small and indistinct. The cytoplasm was abundant and pale and often contained lipid vacuoles. Focal necrosis and dystrophic calcification were noted in case 2.

Between the tumor cell nests ovarian stromal tissue or hyalinized fibrous tissue was noted. In case 2 focal collection of lutein cells was noted.

Mitotic figures were rare, ranging from 0~1 per 10 high power fields. The SCTAT with metastasis was similar to those without metastasis. Angioinvasion or stromal invasion was not seen in any of the cases.

Embryological Considerations

Indifferent Gonad:

Sections of 2 mm long embryo (Streeter age group X) showed large round cells with nucleoli along the dorsal mesentery; interpreted as primary germ cells. Serial sections of a human embryo of Streeter age group XVI showed small nodular elevations in medioventral region of mesonephros consisting of proliferating coelomic epithelium and large cells with distinct nucleoli. The coelomic epithelium proliferated and infiltrated into the underlying mesenchyme. The epithelial cells were arranged in a cord-like pattern, within which primary germ cells were noted. In the embryo of the Streeter age group XVII the genital ridge enlarged more than that of the Streeter age group XVI and presented a distinct elevated mass ventromedial to mesonephros.

Serial sections of two embryos of Streeter age group XVIII were available for review: one case had been sectioned coronally and the other case transversely. The genital primordium formed a predunculated mass into the coelomic cavity ventromedial and slightly inferior to the mesonephros from which it was well demarcated.

The genital primordium consisted of round or ovoid cells arranged in a cord-like pattern and scattered primary germ cells. The latter were slightly larger than the epithelial cells, showing prominent nucleoli and mitotic figures. The epithelial cells had moderate amounts of faintly acidophilic or clear cytoplasm, an indistinct

cytoplasmic margin and round or ovoid nuclei with finely dispersed chromatin and indistinct nucleoli. These crescentic strips of gonadal tissue were in juxtaposition to the mesonephros but quite distinct from it. The covering coelomic epithelium proliferated with multistratification and cord-like downward growth continuous with gonadal tissue. In the deeper portion (away from coelomic epithelium) the cordlike arrangement was more apparent in one case, suggesting "rete blastema".

Development of Testis: Ninety-one cases of testis were available for review; they ranged from 8th to 40th week of development. Serial sections were available in 6 cases.

Serial sections of embryo of an Streeter age group XXIII showed prominent straight testicular tubules separated by mesenchymal tissue. In the subcoelomic cellular layer germ cells and sex cords disappeared and were replaced by loose textured spindle cells. The straight tubules were continuous with rete cords.

Two types of cells were noted in the testicular tubules: elongated cells with ovoid nuclei and spheroidal cells with clear cytoplasm. The latter cells were not seen in the rete cords. In 16th week of development the testis had seminiferous tubules similar to those of a neonate. In general Leydig cells showed changes that were more prominent than those observed in seminiferous tubules. They were most prominent during the 16th to 18th week of development, especially during the 18th week. Subsequently they gradually underwent degeneration or atrophy, having clear or vacuolated or scanty amounts of heavily staining cytoplasm and pyknotic nuclei. During the 36th to 40th week of development few islands of interstitial cells were noted. As the degeneration and atrophy of interstitial cells progressed the seminiferous tubules enlarged and elongated and tubular lumens began to appear.

Early in development very small numbers of primary germ cells were noted but later many primary germ cells were noted in the seminiferous tubules.

Development of Ovary: Eighty-three ovaries were available for review; they ranged from the 8th to the 42nd week of development. Serial sections of embryo of Streeter age group XXIII showed elongated structure attached to the ventromedial side of the mesonephros consisting of epithelial cells and primary germ cells. These cellular layer merged imperceptibly with coelomic epithelium. Scattered connective tissue septa were noted. In 12th week of development the ovarian tissue was undifferentiated and consisted of massive sex cords and sparsely disposed mesenchymal cells. In 17th week of development the cellular zone with primary germ cells persisted: the cellular aggregates consisted of diffuse or cordlike arrangement of epithelial and primary germ cells. In the deep portion of the gonad the germ cells showed degenerative changes. At this stage fetal stroma invaded the medulla. In 18th week of development primary follicles appeared for the first time in the medulla in 6 out of 9 ovaries. As the fetal development progressed the stroma invaded into the ovary. The oogonia were enlarged with vacuolation and eventually degenerated. In 25th week of development primary follicles appeared throughout the entire thickness of the ovary except for the outermost layer which contained them in the 30th week of development. In the 27th week of development some of the primary follicles showed multiple layering. Three ovaries of 37th, 38th and 40th week of development contained maturing follicles with antrum and one ovary of 37th week of development contained an atretic follicle.

Pathologic Finding of Cryptorchid Testis: Six cases of cryptorchid testis were examined: the alterations varied widely from small atrophic

testes with small seminiferous tubules to relatively well preserved seminiferous tubules. In the extremely atrophic testis the tubular basement membrane was markedly thickened and tubules contained only Sertoli cells. Interstitial cells were relatively prominent. But blind or annular tubules were not encountered.

Ultrastructural Findings

Electron microscopic study of three cases of the SCTAT unassociated with the PJS showed similar findings. Columnar epithelial-type tumor cell nests were surrounded by multiple parallel layers of basement membrane-like material. The single or multiple hyaline eosinophilic bodies as seen on light microscopic examination consisted of basement membrane-like material. The small round aggregation of basement membrane-like material was connected or surrounded by rough endoplasmic reticulum in case 3. The tumor cells consisted of two types of cells: light cells with relatively fewer cytoplasmic organelles and dark cells with compact cytoplasmic organelles. Basically the two types of tumor cell were similar. The nuclei were ovoid or elongated with deep indentations, which were more prominent in dark cells. The light cells were the predominant cells and dark cells were located usually at the periphery of the cell nests. Case 1 consisted nearly exclusively of light cells. The epithelial-type tumor cells had straight cell borders with occasional interdigitations and were connected by distinct desmosomes in case 1 and 3 and by less numerous desmosomes in case 2. True lumen and microvilli were observed in case 1. Microfilaments were found in all the cases and Charcot-Bottcher crystals were readily identified in all the blocks in case 2 and 3 but in case 1 a few Charcot-Bottcher crystals were noted in only one of 7 blocks submitted. The Charcot Bottcher crystal consisted of closely packed electron-dense longitudinal fibrils, which were usually located close

to the nucleus and measured from 0.5 to 3 μ in length. Lipid droplets were rarely found in case 2 and 3 but were frequently found in case 1. Lysosomes were small and rare in case 2 and 3 but many large lysosome were noted in some of the tumor cells in case 1. Golgi apparatus was indistinct. Rough endoplasmic reticulum was moderate in amount in the light cell but abundant in the dark cells. Some of tumor cells showed concentric whorls of rough endoplasmic reticulum. Moderate numbers of mitochondria were noted, which contained transverse laminar or tubular cristae in all 3 cases and showed varying degrees of swelling. Smooth endoplasmic reticulum was less abundant than rough endoplasmic reticulum and showed dilatation in some cells.

DISCUSSION

The annular tubules, simple and complex, found in the tumor are similar to atrophic tubules lined only by Sertoli cells in cryptorchid testes, testicular tubular adenomas (Halley, 1963) or ring-like tubules (Halley, 1963). In atrophic tubules definite lumen is usually identified in contrast to solid nature of the tubules of the SCTAT, in which the neoplastic nuclei lie at the periphery and pale cytoplasm obliterates the lumen. Similar solid tubules may be found in Sertoli-Leydig cell tumors, granulosa cell tumors and gynandroblastomas. But an annular pattern with thick basement membrane is unusual in those tumors mentioned above.

The solid tubules of the SCTAT are similar to the testicular tubules in the embryo of Streeter age group XXIII, suggesting male rather than female sexual differentiation. In the ovary of Streeter age group XXIII prominent sex cords were not identified; absence of such a male differentiation was the criteria for identification of the gonad as an ovary. In this context of

sexual differentiation in the embryo the SCTAT resembles testicular differentiation. In other words if we assume that sex cord stromal tumors recapitulate differentiation of sex cord and/or gonadal mesenchyme as suggested by Teilum (1958), the SCTAT seems to recapitulate the differentiation of testis in embryonic period. Tubular structures with fat droplets in the cytoplasm are similar to those seen in canine Sertoli-cell tumor (Scully and Coffin, 1952).

The ultrastructural findings of three cases of SCTAT are similar to those observed by Tavassoli and Norris (1980). The Charcott-Bottcher crystals were identified in all the three cases unequivocally identifying the Sertoli differentiation of the SCTAT. The ultrastructural findings of normal Sertoli cells and granulosa cells are similar but the most characteristic feature of normal adult human testicular Sertoli cells is Charcott-Bottcher crystals (Bawa, 1963; Bloom and Fawcett, 1968; Weiss and Greep, 1977). Charcot-Bottcher crystals are found in these cells near the nucleus (Bloom and Fawcett, 1968), but was not identified in some of the reported cases of Sertoli cell tumors (Jenson and Fehner, 1969; Roth, et al, 1974; Goellner and Myers, 1975; Ramzy and Bos, 1976). But other ultrastructural findings of the latter are similar to those in present series of cases. Charcot-Bottcher crystals were also identified in a Gonadoblastoma (Ishida et al., 1976). Crissman and Hart (1981) reported granulosa cell nature of the SCTAT on the basis of an electron microscopic study of three cases, in which they could not find any Charcot-Bottcher crystals. Young et al (1982) also could not find any microfilaments or Charcot-Bottcher crystals in one case of SCTAT. The absence of the crystals and microfilaments in the cases of Young et al., (1982) and Crissman and Hart (1981) may have been due to limited sampling for electron microscopic study. A few

Charcot-Bottcher crystals were noted in only one out of seven blocks in case 1 of this study, suggesting a paucity of these crystals in some of the cases. The main controversy over the origin of the SCTAT was whether SCTAT is a Sertoli cell tumor (Norris and Tavassoli, 1980) or a variant of granulosa cell tumor (Hart, et al., 1980, Crissman and Hart, 1981). Scully (1981) believes that SCTAT is a distinct tumor which may show differentiation toward both Sertoli cell and granulosa cell tumor even though Charcot-Bottcher crystals were identified in a few of the SCTAT in the series by Tavassoli and Norris (1980). In 1982 Young et al reviewed 74 cases of SCTAT and assumed that SCTAT had morphologic features intermediate between those of granulosa cell tumor and those of the Sertoli cell tumor. Focal solid nests of tumor cells consistent with granulosa cell tumor were found in 1 of 3 cases of present series, 12 out of 47 cases of the SCTAT without the PJS in the series by Young et al. (1982), 4 out of 27 cases by Tavassoli and Norris (1980), and 5 out of 6 cases by Hart, et al. (1980). Histopathologically SCTAT may be defined as a tumor showing predominant Sertoli differentiation with minor component like granulosa cell tumor. One may assume that the SCTAT is a tumor with morphologic features intermediate between those of granulosa cell tumor and those of the Sertoli cell tumor just because of small area of granulosa cell tumor-like component in the background of predominant or nearly exclusive Sertoli differentiation.

But one can also argue that a minor component like granulosa cell tumor is not unusual in the tumors showing predominant Sertoli differentiation and that the SCTAT is a variant of Sertoli cell tumor. With the definite evidence of Sertoli differentiation (Charcot-Bottcher crystal), one can classify SCTAT as an annular and membranous

variant of Sertoli cell tumor or Sertoli cell tumor, annular tubular type. Structures resembling granulosa cell tumor are not unusual in Sertoli-Leydig cell tumor. Morris and Scully (1958) mentioned about component like the granulosa cell tumor in Sertoli-Leydig cell tumor in their book as follow: "In the solid islands, small cystic spaces resembling the Call-Exner bodies of granulosa cell tumors may be present (Fig. 45 of 47)". Later, Scully (1979) documents about the focal component like granulosa cell tumor in Sertoli-Leydig cell tumor as follow: "... on occasion, scattered nests of cells containing Call-Exner bodies suggest focal granulosa cell differentiation".

We propose the classification of the SCTAT as a Sertoli cell tumor, annular tubular type. The term "sex cord tumor with annular tubules" is appropriate for the tumor under discussion: its distinctive morphology of the SCTAT, absence of any Leydig cell element in the present series and previous reports (Scully, 1970; Hart et al., 1980; Tavassoli and Norris, 1980; Crissman and Hart, 1981), frequent association with the PJS, and occasional association with adenoma malignum (Young, et al., 1982) warrant its classification as a distinct entity. The term SCTAT should be retained but used parenthetically like Sertoli cell tumor, annular tubular type (SCTAT).

The present review of embryonic and fetal, indifferent gonads, ovaries, and testes supports concept of Gillman (1948) that coelomic epithelium gives rise to granulosa cells and Sertoli cells. The absence of primary germ cells in the coelomic epithelial covering also denies origin of germ cells from coelomic epithelium. Many pathologists believe that the frequent occurrence of tumors showing combinations of granulosa cells, theca cells, Sertoli cells, and Leydig cells can best be explained on the basis of their origin from the primitive gonadal stroma or

mesenchyme (Busby and Anderson, 1954; Norris and Chorlton, 1974; Novak et al., 1971), even though there is not total agreement among embryologists that Leydig, Sertoli, granulosa, and theca cells have a common origin.

Other pathologists prefer to leave open the possibility that these cell types are ultimately derived from coelomic epithelium and favored the term sex cord mesenchyme tumor (Morris and Scully, 1958) or sex cord stromal tumors (Serov et al., 1973).

The SCTAT may occur in two clinical settings: one associated with the PJS and the other without the PJS. The SCTATs with the PJS were characteristically multifocal, bilateral, very small or even microscopical in size, and calcified. The SCTATs without the PJS were unilateral and usually large (Young et al., 1982). Concerning the nature of the lesion, Scully (1970) believed that the SCTAT with or without associated PJS should be considered a true neoplasm, although he acknowledged that it was difficult to separate hamartomatous lesion from true neoplasm among SCTAT. Hart, et al., (1980) and Tavassoli and Norris (1980) considered SCTAT with the PJS a hamartomatous lesion and SCTAT without PJS a true neoplasm with low grade malignant potential. However, clear separation of the SCTAT on the basis of presence or absence of the associated PJS is too artificial: the histopathologic features of the SCTAT with the PJS and the SCTAT without the PJS are overlapping or are quite similar. Our three cases were not associated with the PJS, presented with unilateral grossly visible mass, and showed calcification in one case. One patient had bilateral enlargement of breasts and axillary and pubic hairs probably related to the SCTAT. The other patients presented with menorrhagia (case 1) and amenorrhea (case 3), which were probably manifestations

of hyperestrinism by ovarian tumors. These symptoms were relieved by excision of the SCTAT (case 1 & 3).

Young et al (1982) reported that four of the 27 SCTAT with the PJS had "adenoma malignum" of the cervix, which is an important, but rare manifestation of the PJS.

The present cases revealed no evidence of uterine cervical disease clinically; the uterine cervix was not examined pathologically in all 3 cases.

One patient had metastatic tumors and the microscopic appearances of which were similar to that of the original tumor mass, indicating the malignant potential of this tumor, as confirmed by Hart et al. (1980) and Young et al. (1982).

ABSTRACT

A pathologic study was done on 3 cases of sex cord tumor with annular tubules. Ninety-one cases of testis and 83 cases of ovary at various stages of development, 4 cases of indifferent gonad, and 6 cases of cryptorchid testis were analyzed from embryological and comparative morphological view points. All three tumors occurred in young women (11-24 years of age) and were not associated with Peutz-Jeghers syndrome. Three cases presented evidence of hyperestrinism. One case with retroperitoneal metastasis confirmed the low grade malignant potential of this tumor.

Grossly the tumors were solid yellowish unilateral tumors with varying degrees of cystic degeneration. Microscopic examination showed simple or complex annular tubules with prominent basement membrane. Many tumor cells contained lipid in the cytoplasm. In comparison with physiological process of gonadal differentiation, SCTAT showed male sexual differentiation.

Ultrastructural study showed Charcot-Bottcher crystals in 3 cases, indicating Sertoli cell differentiation. True lumen and microvilli were identified in one case.

We propose the classification of SCTAT as a Sertoli cell tumor, annular tubular type(SCTAT) on the basis of ultrastructural study and comparative study.

—국문초록—

輪狀細管形成을 隨伴한 卵巢의 未分類 性索間質細胞腫(Sex-cord tumor with annular tubules)에 關한 比較形態學的 研究

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저자들은 3례의 輪狀細管形成을 隨伴한 卵巢의 未分類 性索間質細胞腫(Sex cord tumor with annular tubes: SCTAT)에 關한 전자현미경적 檢索을 포함한 병리학적 연구결과를 보고하고자 한다. 동시에 胎生學的 및 比較形態學的 測面에서 91례의 嚙丸, 83례의 卵巢, 4례의 無關生殖腺(indifferent gonad) 및 6례의 停留嚙丸을 분석보고한다.

SCTAT 3례 모두 11세에서 24세사이의 젊은 여자에서 발생하였으며 Peutz-Jeghers증후군은 수반되지 않았고 女性호르몬過多症이 있었다. SCTAT 3례중 1례는 첫번 수술후 5년 9개월내지 7년 10개월사이에 後腹膜轉移를 나타내어 SCTAT가 低惡性 潛在能이 있음을 나타내었다.

육안적으로는 黃色 充實性 偏側 卵巢腫瘍으로서 囊性變化가 多少 수반되어 있었다. 현미경적으로 종양세포는 단순한 혹은 복합적인 輪狀細管을 형성하고 현저히 肥厚된 基低膜이 발달되어 있었다. 많은 종양세포 세포질내에는 脂肪이 함유되어 있었다. SCTAT의 형태학적 소견과 生殖腺의 發達過程과 비교하면 SCTAT는 男性分化樣狀을 보였다.

전자현미경적사상 SCTAT 3례 모두에서 Charcot-Bottcher crystal이 관찰되어 SCTAT종양세포가 썬틀리 세포로 분화함을 나타내었다. 內腔 및 微細絨毛는 1례에서 관찰되었다.

이상 3례의 SCTAT에 대한 전자현미경적 檢索을 포

함한 병리학적 檢索과 生殖腺發達過程 및 停留嚙丸과의 比較形態學的 研究結果를 토대로 저자들은 SCTAT를 輪狀細管型 썬틀리세포종[Sertoli cell tumor, annular tubular type(SCTAT)]라고 명명하기를 제의한다. (본연구과정에서 자상한 조언을 주신 하바드의대 Scully 교수, 여러가지로 도와준 서울대학교 의과대학 병리학 교실원여러분, 전자현미경실 김수성기사, 태아의 연속 절편제작으로 수 많은 일요일을 실험실에서 보낸 조한웅기사, 수십번의 타자를 마다하지 않은 정영희양에게 감사한다. SCTAT 2례의 전자현미경적 연구용 종양조직, 임상소견 및 현미경표본을 보내주고 사용하도록 허락해준 고려대학교 의과대학 김인신 조교수, 인천 길병원 박흥래과장, 서울대학교병원 병리과 김철우 전임의(전 한강성심병원 병리과)에게 심심한 사의를 표한다.)

REFERENCES

Bawa, S.R.: *Find Structure of the Sertoli Cell of the Human Testes. J. Ultrastruct. Res.* 9:459-474, 1963.
Bloom, W. and Fawcett, D.W.: *A Textbook of Histology, W.B., Saunders Co., 1968, pp.688-692.*
Busby, T., and Anderson, G.W.: *Feminizing Mesenchymomas of the Ovary. Includes 107 cases of Granulosa, Granulosa-Theca-Cell, and Theca-Cell Tumors. Am. J. Obst. and Gynec.* 68:1391-1420, 1954.
Crissman, J.D. and Hart, W.R.: *Ovarian Sex Cord Tumors with Annular Tubules. An Ultrastructural Study of Three Cases. Am. J. Clin. Pathol.* 75:11-17, 1981.
Gillman, J.: *The Development of the Gonads in Man, with a Consideration of the Role of Fetal Endocrines and the Histogenesis of Ovarian Tumors. Contributions to Embryology, No.210, pp.83-131, 1948.*
Goellner, J.R. and Myers, R.P.: *Sertoli Cell Tumors. Case Report with Ultrastructural Findings. Mayo Clin. Proc.* 50:459-463, 1975.
Halley, J.B.W.: *Germ Cell Loss in Prepubertal Cryptorchid Testes. J. Urol.* 90:211-214, 1963.
Halley, J.B.W.: *The Growth of Sertoli Cell Tumors: A Possible Index of Differential Gonadotrophin Activity in the Male. J. Urol.* 90:220-229, 1963.
Hart, W.R., Kumar, N., and Crissman, J.D.: *Ovarian Neoplasms Resembling Sex Cord Tumors with Annular Tubules. Cancer* 45:2352-2363, 1980.

- Ishida, T., Tagatz, G.E., and Okagaki, T.: *Gnadooblastoma. Ultrastructural Evidence for Testicular Origin. Cancer* 37:1770-1781, 1976.
- Jenson, A.B. and Fechner, R.E.: *Ultrastructure of an Intermediate Sertoli-Leydig Cell Tumors. A Histogenetic Misnomer. Lab. Invest.* 21:527-535, 1969.
- Kim, I.S., Kwak, H.S., Yoon, J.H., and Paik, S.Y.: *Sex Cord Tumor with Annular Tubules-A case report with review of the literature-. Korean J. Path.* 12:239-246, 1978.
- Morris, J. McL. and Scully, R.E.: *Endocrine Pathology of the Ovary. C.V. Mosby Co., St. Louis, 1958, pp. 65, 86-87.*
- Norris, H.J., and Chorlton, I.: *Functioning Tumors of the Ovary. Clin. Obstet. Gynecol.* 17:189-228, 1974.
- Novak, E.R., Kutchmeshgi, J., Mupas, R.S., and Woodruff, J.D.: *Feminizing Gonadal Stromal Tumors. Analysis of the Granulosa-Theca Cell Tumors of the Ovarian Tumor Registry. Obstet. Gynec.* 38:701-713, 1971.
- Park, H.R.: *Sex Cord Tumor with Annular Tubules-A case report with review of the literature-. Incheon Gil Hosp. Med. J.* 2:87-90, 1982.
- Ramzy, I. and Bos. C.: *Sertoli Cell Tumors of Ovary. Light Microscopic and Ultrastructural Study with Histogenetic Considerations. Cancer* 38:2447-2456, 1976.
- Roth, L.M., Cleary, R.E., and Rosenfield, R.L.: *Sertoli-Leydig Cell Tumor of the Ovary, with an Associated Mucinous Cystadenoma. An Ultrastructural and Endocrine Study. Lab. Invest.* 31:648-657, 1974.
- Scully, R.E. and Coffin, D.L.: *Canine Testicular Tumors, with Special Reference to Their Histogenesis, Comparative Morphology, and Endocrinology. Cancer* 5:592-605, 1952.
- Scully, R.E.: *Sex Cord Tumor with Annular Tubules. A Distinctive Ovarian Tumor of the Peutz-Jeghers Syndrome. Cancer* 25:1107-1121, 1970.
- Scully, R.E.: *Tumors of the Ovary and Maldeveloped Gonads. Atals of Tumor Pathology. Second series, Washington, D.C., 1979, Armed Forces Institute of Pathology, p. 191.*
- Scully, R.E.: *Personal communication, Jan. 1981.*
- Serov, S.F., Scully, R.E., and Sobin, L.H.: *International Histological Classification of Tumors, No. 9. Histological Typing of Ovarian Tumors. Geneva, World Health Organization, 1973.*
- Teilum, G.: *Classification of Testicular and Ovarian Androblastoma and Sertoli Cell Tumors. A Survey of Comparative Studies with Consideration of Histogenesis, Endocrinology, and Embryological Theories. Cancer* 11:769-782, 1958.
- Tavassoli, F.A. and Norris, H.J.: *Sertoli Tumors of the Ovary. A Clinicopathologic Study of 28 Cases with Ultrastructural Observations. Cancer* 46:2281-2297, 1980.
- Young, R.H., Welch, W.R., Dickersin, G.R., and Scully, R.E.: *Ovarian Sex Cord Tumor with Annular Tubules. Review of 74 Cases Including 27 with Peutz-Jeghers Syndrome and Four with Adenoma Malignum of the Cervix. Cancer* 50:1384-1402, 1982.
- Weiss, L. and Greep, R.O.: *Histology. 4th ed. McGraw-Hill Book Co., 1977, pp. 1007-1013.*

LEGENDS FOR FIGURES

- Fig. 1.** (Case 1) Ovarian tumor from 17-year-old girl with menorrhagia without stigmata of PJS. Note predominantly solid appearance with cleft-like cysts.
- Fig. 2.** (Case 1) Complex annular tubules in which communicating tubules encircle hyaline masses. H&E×100.
- Fig. 3.** (Case 1) Simple and complex annular tubules. H&E×100.
- Fig. 4.** (Case 1) Complex annular tubules. Note interconnection between basement membrane and hyaline masses. H&E×100.
- Fig. 5.** (Case 1) Complex annular and solid tubules with thick basement membrane. Solid tubules simulate seminiferous tubules in cryptorchid testis. H&E×200.
- Fig. 6.** (Case 1) Complex annular tubules consisting of sex cord cells with lipid-laden, vacuolated cytoplasm. H&E×200.
- Fig. 7.** (Case 1) Cyst formation in SCTAT. H&E×200.
- Fig. 8.** (Case 2) Solid area without hyaline masses in left-hand side, simulating granulosa cell tumor. H&E×200.
- Fig. 9.** Dorsal mesentery of Streeter age group X, which shows large round primary germ cells with nucleoli. H&E×200.
- Fig. 10.** Indifferent gonad of Streeter age group XVI, showing thickening of the coelomic epithelium and the condensation of the underlying mesenchyme. H&E×100.
- Fig. 11.** Indifferent gonad of Streeter age group XVII, showing prominent genital ridge. H&E×200.
- Fig. 12.** Transverse section through the indifferent gonad of Streeter age group XVIII. There are large primary germ cells between the sex cords. H&E×100.
- Fig. 13.** Gonad of Streeter age group XXIII. Absence of testicular tubular differentiation suggests ovary. H&E×200.
- Fig. 14.** Testis of Streeter age group XXIII. There is enormous amount of interstitial tissue between the arcades formed by the testicular tubules. H&E×100.
- Fig. 15.** Electron micrograph of SCTAT, showing true lumen and microvilli. Note interdigitation of cell border. Uranium acetate and lead citrate×16,800.
- Fig. 16.** Electron micrograph of neoplastic cells and thick basement membrane. Note parallel array of basement membrane encircling nest of tumor cells. Uranium acetate and lead citrate×16,800.
- Fig. 17.** Electron micrograph of Charcot-Bottcher bodies from case 1(c), case 2(a), and case 3(b). Each body consists of closely packed electron-dense longitudinal fibrils. Uranium acetate and lead citrate a, ×42,000; b, ×42,000; c, ×96,000.







