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ORIGINAL ARTICLE

D. Harms · L.R. Kock

Testicular juvenile granulosa cell and Sertoli cell tumours: a clinicopathological study of 29 cases from the Kiel Paediatric Tumour Registry

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Abstract Testicular Sertoli cell tumours (SCT) and juvenile granulosa cell tumours (JGCT) are rare in childhood. This study was designed to investigate the clinical picture, morphology and disease course in a comparatively large series of cases (total number = 29). Of 198 cases of childhood testicular tumour documented in the Kiel Paediatric Tumour Registry 18 were cases of infantile SCT (9.1%) and 11 of JGCT (5.6%). The average age at the time of diagnosis was 4.2 months for infantile SCT and 0.4 months for JGCT. SCT and JGCT often showed infiltrative growth into adjacent testicular tissue, dense cellularity and considerable proliferation activity. Immunohistochemically all cases expressed vimentin intermediate filaments in both tumour types. Next in frequency of expression were cytokeratins (SCT: 7/16; JGCT: 7/10) and smooth-muscle actin (SCT: 9/15; JGCT: 4/10). Follow-up studies (24/29) showed that in cases of tumour manifestation in infancy and after complete tumour removal (usually orchiectomy) no local recurrences and no metastases occurred. The most important conclusion for diagnosis and therapy is that despite infiltrative growth, incomplete differentiation, dense cellularity and considerable proliferation activity, after surgical excision infantile SCT and JGCT have a good prognosis. Adjuvant chemotherapy or more extensive operations with lymphadenectomy are thus not indicated.

Key words Testicular tumours · Juvenile Sertoli cell tumour · Juvenile granulosa cell tumour · Tumours of infancy

Introduction

Sex cord-stromal tumours of the testis form a subgroup among the nongerminative testicular tumours which includes juvenile granulosa cell tumours (JGCT), Sertoli cell tumours (SCT) and Leydig cell tumours [26]. Testicular sex cord-stromal tumours are rare. They account for approximately 4% of testicular tumours in adults [45], but in childhood the proportion of sex cord-stromal tumours in testicular neoplasms is higher, amounting to approximately 19% [25]. Due to the rarity of testicular tumours in this age group, sex cord-stromal tumours in childhood are something of a curiosity. Consequently the number of personal observations in the published case series is small. The largest compilation of cases from one institution is that of Lawrence et al. [21] and comprises 14 JGCT taken from the consultation files of R.E. Scully.

In the Kiel Paediatric Tumour Registry 29 cases of "infantile" testicular JGCT/SCT were collected over a period of 14 years and 2 months. Tumours with a predominant granulosa cell tumour pattern (11 cases) were classified as JGCT, while stromal-rich lesions presenting with a cord-like or tubular pattern (18 cases) were categorized as (juvenile) SCT. This comparatively large series is the basis of this study, which focussed on the relative frequency of these rare tumours, clinical data including follow-up, light microscopy and immunohistochemistry.

Materials and methods

The files of the Kiel Paediatric Tumour Registry were searched for all testicular neoplasms in the period from 1 January 1980 through 28 February 1994. Only patients who were younger than 16 years were included. One hundred and ninety-eight tumours were classified according to the WHO classification [26]. They are listed in Table 1. Among them were 35 sex cord-stromal tumours, which were divided into 18 cases of SCT, 11 JGCT, 3 large cell calcifying Sertoli cell tumours (LCCSCT) and 3 Leydig cell tumours. In cases in which a diagnosis of SCT or JGCT had been made, questionnaires were sent to the submitting hospitals, asking for clinical data. Data we were interested in included the age of patient at diagnosis, localization, size and macroscopic appearance of the tumour, type of treatment and outcome. Follow-up data, including information on the clinical outcome, were available for 24 of 29 patients (82.8%).

Paraffin sections and paraffin blocks were available in most cases. The slides were stained with haematoxylin and eosin,

D. Harms (✉) · L.R. Kock
Department of Paediatric Pathology, Michaelisstrasse 11, D-24105 Kiel, Germany
Tel.: (49) 431-597-3451, Fax: (49) 431-597-3486

Table 1 Childhood testicular tumours diagnosed between 1 January 1980 and 28 February 1994, age limits 0–180 months (*LCCSCT* large cell calcifying Sertoli cell tumour)

Tumour groups and types	Number of cases	Percentage of cases	Age (months)		
			Average	Range	Median
Germ cell tumours					
Seminoma	4	2.0	71.8	13–171	57.4
Yolk sac tumour	62	31.3	19.7	6–104	16
Mature teratoma	23	11.6	37.1	5–155	24
Immature teratoma	23	11.6	8.1	2–14	7
Tumours of more than one histological type	18	9.1	102.1	0–180	161.5
Total	130	65.7			
Sex cord-stromal tumours					
Granulosa cell tumour	11	5.6	0.4	0–3	0
Sertoli cell tumour	18	9.1	4.2	0–14	4
LCCSCT	3	1.5	101.3	65–133	106
Leydig cell tumour	3	1.5	52	3–119	34
Total	35	17.7			
Other tumours					
Haemangioendothelioma, benign	1	0.5	19		
Leukaemic infiltration	22	11.1	89.7	14–166	78.5
Metastasis of neuroblastoma	1	0.5	10		
Paratesticular rhabdomyosarcoma with testis infiltration	8	4.0	74.1	4–154	52.2
Epidermoid cyst	1	0.5	155		
Total	33	16.6			
Testis tumours, total	198				

Table 2 Types of antibodies used (*M* monoclonal, *P* polyclonal)

Antibodies against	Source	Dilution	Reference
Vimentin (M)	Dakopatts	1:20	Miettinen et al. [23]
Cytokeratin Kl-1 (M)	Immunotech	1:200	Moll et al. [24]
Desmin (M)	Dakopatts	1:30	Miettinen et al. [23]
Actin (M)	Enzo Diagnostics	1:2	Tsukada et al. [42]
α-Smooth muscle actin (M)	Dakopatts	1:20	Skalli et al. [36]
Epithelial membrane antigen (M)	Dakopatts	1:320	Pinkus and Kurtin [30]
Proliferating cell nuclear antigen (M)	Dakopatts	1:40	Hall et al. [15]
Ki-S5 (M)	Department of Pathology, Kiel*	1:1	Kreipe et al. [20]
Ki-S1 (M)	Department of Pathology, Kiel*	1:40	Kreipe et al. [19]
Neuron specific enolase (P)	Dakopatts	1:200	True [41]
S-100 protein (P)	Dakopatts	1:1000	Barwick [2]
α-Fetoprotein (P)	Dakopatts	1:800	Jacobsen et al. [16]

* Supplied by Professor H. Kreipe

Giemsa, Goldner, Bielschowsky (reticulin) and periodic acid-Schiff (PAS). Each slide was examined several times and the various tumour components were noted. If paraffin blocks or paraffin sections were available, additional immunohistochemical analyses were carried out, using the alkaline phosphatase-anti-alkaliline phosphatase method according to Cordell et al. [7]. The following immunohistochemical stains were used: vimentin, cytokeratin (Kl-1), desmin, epithelial membrane antigen, muscle specific actin, α-smooth muscle actin (SMA), neuron specific enolase (NSE), S-100 protein (S-100) and α-fetoprotein. Furthermore, we used the proliferation markers Ki-S1, Ki-S5 and anti-proliferating cell nuclear antigen (anti-PCNA) to detect proliferating cells. Table 2 summarizes the types of antibodies used, their sources, dilutions and references.

Results

Between 1 January 1980 and 28 February 1994 tumour specimens (mostly from orchiectomies) from 198 boys below 16 years of age were examined. Germ cell tumours formed by far the largest group in this age group

Table 3 Important clinical and morphological features of 18 boys with Sertoli cell tumours and 11 boys with juvenile granulosa cell tumours (L=retroperitoneal lymphadenectomy, *Ch* chemotherapy, *NED* no evidence of disease)

		Sertoli cell tumour	Juvenile granulosa cell tumour
Number of cases		18	11
Age at diagnosis/surgery (months)	average	4.2	0.4
	range	0–14	0–3
	median	4	0
Side involved	right	7	2
	left	10	8
	unknown	1	1
Size of tumour (cm)	average	1.9	2.0
	range	0.7–4.0	1.0–2.2
	median	1.7	2.0
Treatment	orchiectomy	16	7
	orchiectomy+L	0	1
	orchiectomy+L+Ch	1	0
	enucleation	1	1
	unknown	0	2
Follow-up	number of cases with NED	16	8
Months after first diagnosis	average	39.3	46.1
	range	9–96	7–117
	median	33.5	41
Number of cases lost to follow-up		2	3

(130 cases, 65.7%). Yolk sac tumours (YST) were most frequent (62 cases, 31.3% of all tumours and 47.7% of the germ cell tumours). The median age at diagnosis was 16 months for patients with YST.

The group of sex cord-stromal tumours included 35 cases, corresponding to 17.7% of all testicular neoplasms. SCT was most frequent with 18 cases (51.4% of sex cord-stromal tumours), followed by JGCT with 11 cases (31.4%). The median age at diagnosis of patients with SCT was 4 months (range: 0–14 months), while patients with JGCT had already been treated within the first few days of life in most cases (range: 0–3 months). The extremely rare LCCSCT and Leydig cell tumours (three cases each) were diagnosed at a median age of 106 and 34 months, respectively. Whereas two of three Leydig cell tumours and one of three LCCSCT were bilateral, the much more frequent SCT and JGCT were always unilateral.

The third group of testicular tumours (33 cases) comprises histogenetically unrelated tumours and includes leukaemic infiltrations (22 cases) and secondary testicular involvement by paratesticular rhabdomyosarcoma (8 cases).

Clinical features varied in the two main groups of sex cord-stromal tumours (Table 3). In Sertoli cell tumours the tumour was diagnosed in descended testes in all 18 cases. In most enlargement of a scrotal testis was the only clinical symptom, in some cases accompanied by an inguinal hernia or a hydrocoele. Seventeen of these patients were treated by unilateral radical orchiectomy; in 1 patient the tumour was enucleated. In addition to the orchiectomy, in 1984 one patient was treated by retroperitoneal lymphadenectomy (without metastases) and polychemotherapy. No postoperative treatment was given in any of the other cases. Nevertheless all 16 patients for whom follow-up data are available were alive and well up to 8 years after surgery.

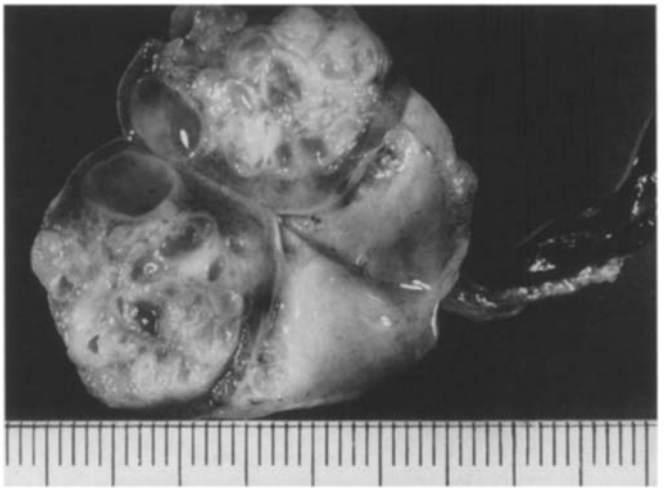
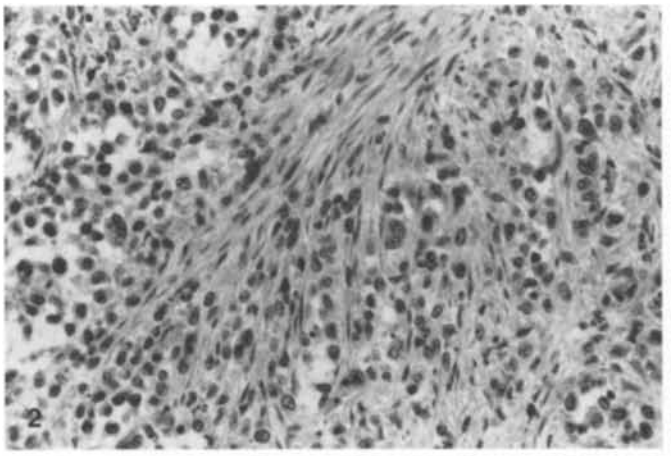


Fig. 1 Juvenile testicular granulosa cell tumour. Multicystic lesion with a greyish-white cut surface. Case number 25, 1-day-old newborn (photograph: Professor N. Böhm, Freiburg)

The JGCT were discovered in scrotal testes as a unilateral testicular mass. Their localization was striking, in that 8 of 11 tumours were located in the left testis and only two in the right (in one case the localization was unknown). Eight patients were treated by unilateral radical orchiectomy; in one of these patients additional ex-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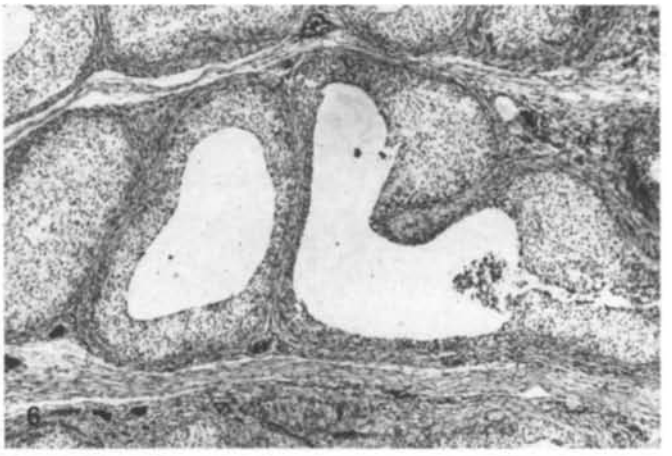
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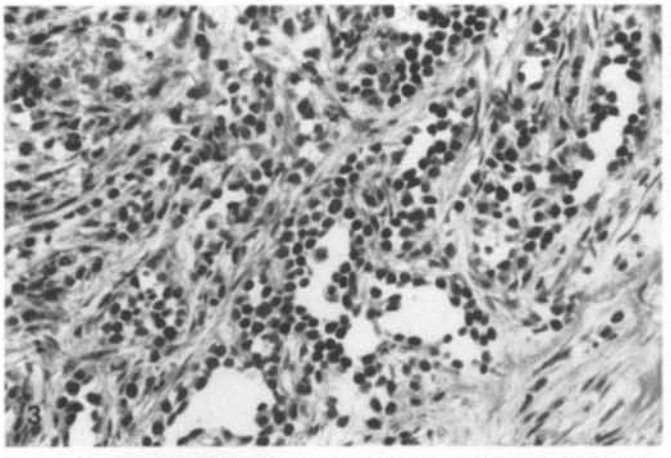
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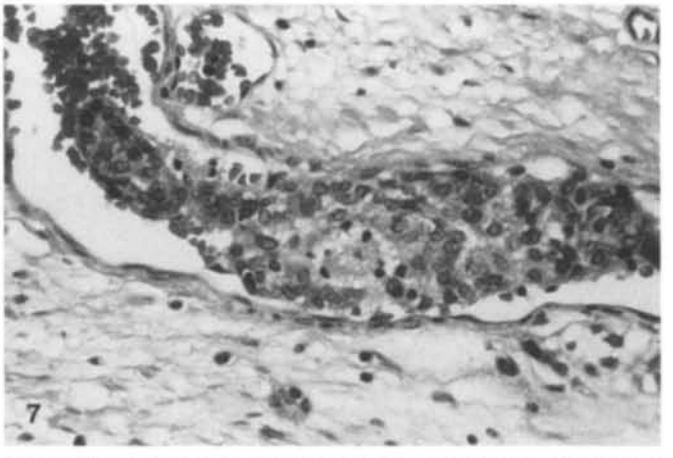
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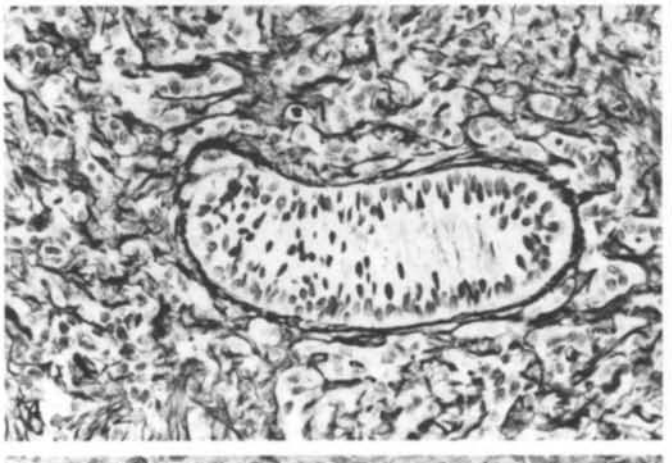
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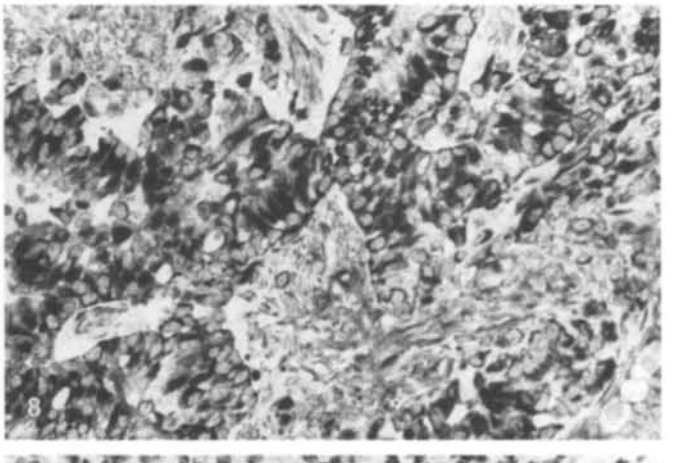
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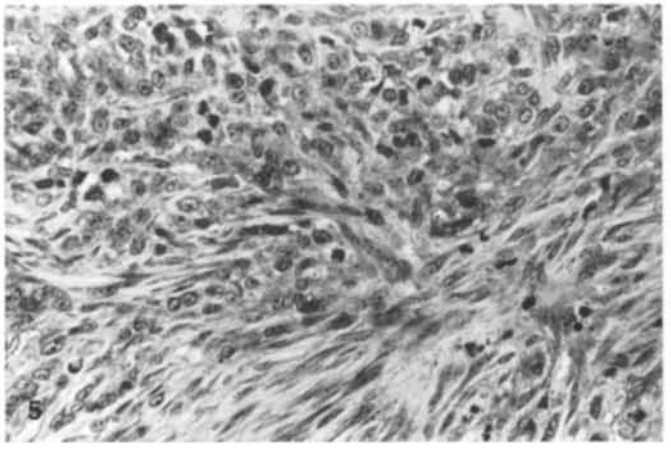
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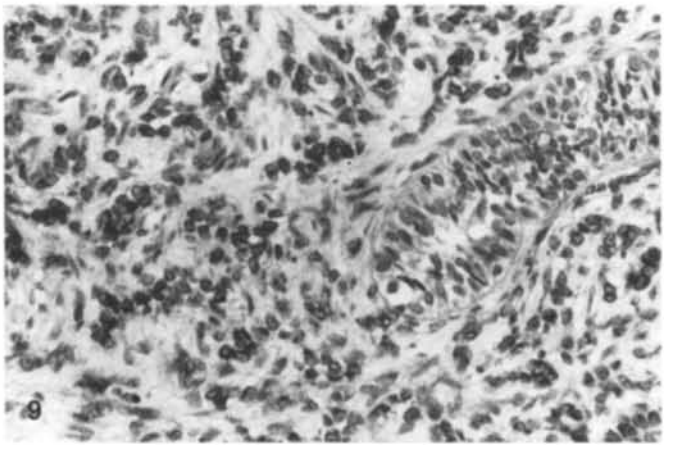
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Table 4 Immunohistochemical findings (0 negative, 1+ 0–10% of tumour cells stained, 2+ 10–50% of tumour cells stained, 3+ >50% of tumour cells stained, *EMA* epithelial membrane antigen, *NSE* neuron specific enolase, *PCNA* proliferating cell nuclear antigen)

Antibody	Sertoli cell tumour					Juvenile granulosa cell tumour				
	Number of cases	0	1+	2+	3+	Number of cases	0	1+	2+	3+
Vimentin	16	0	1	10	5	11	0	0	8	3
Cytokeratins (Kl-1)	16	9	4	3	0	10	3	7	0	0
Desmin	16	15	1	0	0	10	10	0	0	0
EMA	15	15	0	0	0	9	9	0	0	0
Actin	16	6	9	1	0	10	8	2	0	0
Smooth muscle actin	15	6	7	2	0	10	6	1	3	0
NSE	16	15	1	0	0	9	9	0	0	0
S-100	15	13	1	1	0	9	7	1	1	0
α-Fetoprotein	15	15	0	0	0	11	11	0	0	0
Ki-S5	12	0	2	10	0	10	0	0	10	0
Ki-S1	13	0	1	12	0	8	0	0	8	0
PCNA	12	0	3	6	3	7	0	0	7	0

ploratory excisions of retroperitoneal lymph nodes were carried out. One patient was treated only by tumour enucleation, and in two cases the exact type of treatment was unknown. As far as we know no postoperative treatment was given in any of the cases of JGCT. All eight patients with follow-up information were alive and well between 7 months and nearly 10 years after surgery (average: 46.1 months).

The gross pathology of the lesions also differed. The SCTs ranged from 0.7 cm to 4.0 cm, with an average tumour size of 1.9 cm (Table 3). The cut surface of the tumour was usually solid and appeared greyish-white to yellowish in colour. The smallest JGCT was 1.0 cm, the largest 2.2 cm with an average greatest diameter of 2.0 cm (Table 3). However, the most common appearance was that of a multicystic tumour with a greyish-white to yellowish cut surface (Fig. 1).

Microscopically, the SCT were solid with tumour cells in cords (Fig. 2) or in tubule-like structures (Fig. 3). The cords and tubules did not contain germ cells, especially spermatogonia, which were regularly observed in normal seminiferous tubules adjacent to the tumour tissue. Sometimes primitive Sertoli cells and fibroblast-like cells were arranged irregularly with no recognizable pattern. In these areas abundant reticulin fibres were observed (Fig. 4). The Sertoli cells were round to slightly elongated with hyperchromatic nuclei and sparse, slightly eosinophilic cytoplasm, which was usually PAS negative. The nucleoli were small, but detectable at higher magnification. Most tumours showed strong mitotic activity with up to 38 mitotic figures per 10 high-power fields (HPF; Fig. 5). Usually the mitotic figures were regular, but some atypical ones were also observed. The stroma was composed of spindle cells with moderate cellularity and some areas of hyalinization. The tumours were only incompletely surrounded by a pseudocapsule and showed infiltrative growth, but no destructive growth pattern, with preexisting seminiferous tubules entrapped in the peripheral parts of the tumour (Fig. 4).

The JGCT were partially cystic and partially solid (Fig. 6). The solid components were composed of groups or cords of tumour cells, which contained few reticulin fibres. The follicles surrounded by granulosa cells varied in size and shape and appeared to be empty, but sometimes contained a slightly eosinophilic watery or viscid fluid. The tumour cells were round or, especially at the periphery, slightly elongated with sparse cytoplasm, which was partly vacuolized and focally PAS positive. Occasionally some tumours contained extracytoplasmic eosinophilic globular bodies with intensive PAS positivity, analogous to the small Call-Exner bodies of ovarian granulosa cell tumours. The nuclei were round to ovoid in shape and small to medium in size, while the nucleoli were very small and inconspicuous. Nuclear grooves did not occur. Mitotic counts ranged from 3 per 10 HPF to

Fig. 2 Infantile testicular Sertoli cell tumour. Compact tumour tissue with a predominantly trabecular pattern. Slightly polymorphic and hyperchromatic nuclei. Case number 16, 2-month-old infant. Haematoxylin and eosin (H&E), ×195

Fig. 3 Infantile testicular Sertoli cell tumour. Partly cord-like, partly tubular pattern. Case number 5, 6-month-old infant. Goldner, ×195

Fig. 4 Infantile testicular Sertoli cell tumour with an entrapped pre-existing seminiferous tubule. Dense framework of reticulin fibres surrounding small cords of tumour cells. Case number 14, 4-month-old infant. Bielschowsky, ×195

Fig. 5 Infantile testicular Sertoli cell tumour with numerous mitotic figures. Case number 16, 2-month-old infant. Giemsa, ×245

Fig. 6 Juvenile testicular granulosa cell tumour. Microcysts surrounded by granulosa cells and neighbouring fibrous tissue. Case number 24, 5-day-old newborn. H&E, ×40

Fig. 7 Juvenile testicular granulosa cell tumour. Tumour infiltration of a thin-walled venule. Case number 21, 1-day-old newborn. Event-free survival (follow-up period: 9 years and 9 months). H&E, ×245

Fig. 8 Infantile testicular Sertoli cell tumour with intensive expression of vimentin. Case number 18, 11-month-old infant. Immunohistochemistry [alkaline phosphatase-anti-alkaline phosphatase (APAAP) method], ×245

Fig. 9 Infantile testicular Sertoli cell tumour with cytokeratin expression. Case number 14, 4-month-old infant. Immunohistochemistry (APAAP method), ×195

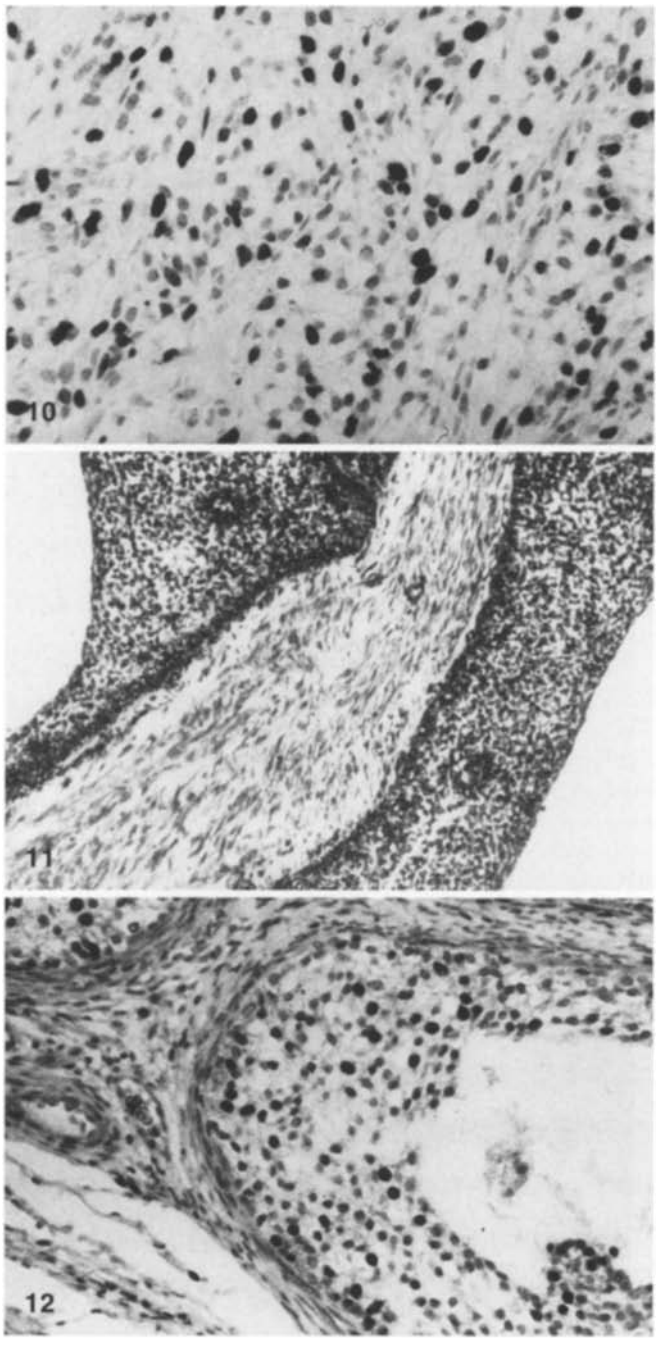

Fig. 10 Infantile testicular Sertoli cell tumour. Demonstration of proliferative activity with the proliferation marker Ki-S5 (*dark nuclei*). Moderate proliferative activity (10–50% positive cells). Case number 5, 6-month-old infant. Immunohistochemistry (APAAP method), ×245

Fig. 11 Juvenile testicular granulosa cell tumour with strong expression of vimentin. Case number 6, 3-day-old newborn. Immunohistochemistry (APAAP method), ×97

Fig. 12 Juvenile testicular granulosa cell tumour. Demonstration of proliferative activity with the proliferation marker Ki-S5. Moderate proliferative activity (10–50% positive cells). Case number 24, 5-day-old newborn. Immunohistochemistry (APAAP method), ×195

25 per 10 HPF. The tumours were often at least partially surrounded by a pseudocapsule, but in nearly all cases areas without a capsule could be seen. In these areas the tumour tissue infiltrated the adjacent parenchyma. Although destructive growth could not be demonstrated with certainty, one tumour showed two areas with invasion of small thin-walled venules (Fig. 7).

Table 4 shows the immunohistochemical findings in the two groups. All SCT were positive for vimentin (Fig. 8), though the number of positive cells varied. Muscle specific actin was expressed by 10 of 16 tumours and 9 of 15 were positive for SMA. With both antibodies the number of positive cells was usually low. In 7 of 16 tumours a small or moderate number of cells reacted with the cytokeratin antibody Kl-1 (Fig. 9). Staining with the proliferation markers Ki-S1, Ki-S5 and anti-PCNA (Fig. 10) usually revealed between 10% and 50% positive nuclei. The stroma cells frequently displayed coexpression of vimentin, muscle specific actin and SMA.

In all 11 JGCT a moderate (8x) or high (3x) number of tumour cells expressed vimentin (Fig. 11). In 7 of 10 tumours a low number of cells (< 10%) stained positively for cytokeratin as well. Four tumours showed expression of SMA and two additionally expressed muscle specific actin. Staining with the proliferation markers Ki-S1, Ki-S5 (Fig. 12) and anti-PCNA always revealed a moderate number of positive nuclei (10%–50% positive cells). The fibroblast- and theca-like stromal cells surrounding the granulosa cell complexes were usually positive for vimentin and mostly reacted with SMA (9 of 10) and anti-muscle specific actin (7/10) as well.

Discussion

Our study has shown in an unselected series of cases collected in the Kiel Paediatric Tumour registry, that childhood sex cord-stromal tumours accounted for 17.7% of all testicular neoplasms (35/198). SCT and SGCT were by far the most frequent (82.9% of the sex cord-stromal tumours) and appeared at a very early age. From our follow-up studies, it appears that SCT and JGCT can be cured by complete excision of the tumour in all cases where the tumour arises in infancy. Histologically, juvenile SCT and JGCT often show infiltrative growth, "immaturity" and definite proliferative activity, which can simulate malignancy. Immunohistochemically, vimentin intermediate filaments are expressed in all cases; next in frequency, and less intensive, are cytokeratin and SMA.

Sex cord-stromal tumours are neoplasms that arise from the nongerminative specialized gonadal tissue and Teilum [40] has pointed out the homology between sex cord-stromal tumours of the ovary and testis.The most important neoplasms in this group (with the exception of thecomas) are seen in the ovary as well as in the testis. However there are significant differences in their relative frequency, age distribution and biological behaviour. The relative frequency • of 35 sex cord-stromal tumours (17.7%) among 198 unselected testicular tumours in pa-

tients under 16 years of age corresponds with the findings of Mostofi and Price (19%) [25]. There is no information in the literature on the frequency of sex cord-stromal tumours in infancy (including the newborn period). An analysis of our material showed that sex-cord stromal tumours amounted to 39.7% of all testicular tumours in this age group and the percentage of these tumours in children over 1 year of age was low at only 3%. Consequently, despite their general rarity, testicular sex cord-stromal tumours arise remarkably often in infancy. The most frequent tumours in this group are SCT and JGCT. Our data on the relative frequency and age distribution of the various tumour types are representative, since the Kiel Paediatric Tumour Registry is the reference centre for the Cooperative Malignant Testicular Tumour Studies (MAHO) of the German Society of Paediatric Oncology and Haematology (GPOH).

Among the roughly 100 reported cases of SCT in all age groups, there were at least 22 cases showing a malignant course [22, 35]. Testicular GCT analogous to the adult type of ovarian GCT are generally rare and do not appear in childhood. According to Jimenez-Quintero et al. [17], who reported on 19 cases of this adult tumour type, the age at manifestation ranged from 16–76 years (average: 47.7 years). Two of four patients with metastases died of tumour. Testicular JGCT are not known to show malignant behaviour in prepuberty. The youngest patient with a well documented malignant SCT was 8 years old and had metastases in the retroperitoneal lymph nodes [33]. Two years after orchiectomy and removal of the retroperitoneal tumour masses the patient was alive and well. Three additional reports of malignant SCT in the first 2 decades of life have been published [5, 18, 35].

The 11 JGCT in our series were all discovered within the first 3 months of life, and 9 of these 11 patients were already operated on within the first few days of life. This age distribution corresponds to previously reported findings [14, 29]. It is not clear why JGCT occur considerably more frequently in the left testis than in the right one. In the ten cases in our series in which the localization of the tumour was known, eight were in the left testis and only two in the right; the series of Perez-Atayde et al. [29] also showed more JGCT in the left testis (5) than in the right (2).

The 18 SCT in our series also appeared in very young children. The median age was 4 months, with a range from 0 to 14 months. These findings are confirmed by Weitzner and Gropp [43], who observed frequent manifestation of this tumour type in the first year of life in 16 of 23 reviewed SCT of the testis in childhood. Consequently it would be better in the future to classify SCT and GCT of early childhood as infantile testicular neoplasms. To prevent misunderstanding, the common terminology (SCT/JGCT in childhood) is retained in this paper.

Clinically, SCT and JGCT presented most frequently with painless enlargement of a testis, which, in individual cases, was accompanied by an inguinal hernia or a hydrocoele. Clinical symptoms of endocrine hyperactivity, including gynaecomastia and sexual precocity, are rare [12, 43] and were not observed in this series. The same is true of the cases of testicular JGCT published by Young et al. [47] and Groisman et al. [14], in contrast to ovarian JGCT, which were associated in approximately 80% of prepubertal patients with sexual (pseudo)precocity [46]. It is noteworthy that 6 of 26 reported testicular JGCT occurred in undescended testes of children with abnormal chromosomes and ambiguous genitalia [6, 11, 31, 39, 47]. In four other cases testicular JGCT were discovered in undescended testes of patients without genital tract abnormalities [1, 9, 21]. The karyotype of these patients was not analysed. All SCT and JGCT in our series were observed in descended testes; all of our patients had normally developed external genitalia and clinically there was no evidence of chromosomal abnormalities.

On conventional light microscopy the diagnostic hallmarks of juvenile testicular SCT are variably primitive tubules and cord-like structures without germ cell precursors [45]. Most tumours show conspicuous cellularity with considerable mitotic activity [28] and proliferative activity, which has been confirmed by analyses with three proliferation markers. Vimentin is the basic intermediate filament of Sertoli cells [3, 8, 10, 14, 32, 34, 37], but in fetal testes coexpression of vimentin and cytokeratins occurs [8, 14, 32, 37], and during early testicular development additional desmin expression can be seen [32]. Thus expression of vimentin and cytokeratins by SCT [27, 34] and, less frequently, of muscle specific actin, SMA and occasionally of desmin, S-100 protein and NSE reflects the immunohistochemical immaturity of SCT, corresponding to the immaturity and incomplete differentiation seen by conventional light microscopy in most cases.

JGCT, in contrast to SCT, which are principally solid tumours, usually present as partially cystic-follicular, partly solid lesions. Histologically, testicular JGCT are very similar to ovarian GCT (see Young and Scully [44]). Like SCT, JGCT display significant mitotic rates, ranging from 1 to 24 per 10 HPF [21] and 3–25 per 10 HPF (this series). In addition, immunoreactions with proliferation markers (Ki-S1, Ki-S5, PCNA) revealed considerable proliferative activity, with between 10% and 50% positively stained nuclei in all cases. Basically, the immunohistochemical profile of JGCT is very similar to that of SCT. The desmin reaction was usually negative in our series, in contrast to recently published data on desmin expression by granulosa cells in JGCT by Perez-Atayde et al. [29]. Desmin expression in granulosa cells may, however, be related to the individual degree of maturation of granulosa cells. The same is true of cytokeratins, since Czernobilsky et al. [10] and Benjamin et al. [3] have demonstrated a gradual reduction of cytokeratins in human ovarian granulosa cells when maturation from primordial to mature follicles takes place.

SCT and JGCT can be considered to be histogenetically related tumours, since SCT may contain foci with a JGCT pattern and vice versa. The growth pattern, the im-

munohistochemical profiles and the prognoses of the two lesions are very similar.

Despite a locally aggressive growth pattern (even with the vascular invasion seen in one case), dense cellularity, considerable mitotic activity and proliferation rates, criteria usually indicating malignancy [38], SCT and JGCT show benign behaviour after complete excision, when tumour manifestation and treatment occur in infancy, since all patients with follow-up data (altogether 24/29 cases) were alive and well.

These data are in accordance with previously reported studies revealing an excellent prognosis for these sex cord-stromal tumours especially in young patients [14, 21, 34, 45]. Complete removal of the tumour, usually by unilateral orchiectomy, is the treatment of choice. Chemotherapy and retroperitoneal lymphadenectomy are not indicated. With regard to the side effects of chemotherapy, Gonzalez-Crussi [13] has warned against additional therapy in this tumour group in childhood. SCT and JGCT of early childhood demonstrate the relativity of criteria of malignancy and are good examples of the "oncogenic period of grace" [4] in this age period.

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