PEDIATRIC SURGERY

International

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Springer

Pediatr Surg Int (1993) 8: 244–247

Pediatric Surgery International

Original article

Testicular tumors in childhood: a national report

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Accepted 19 May 1992

Abstract. The authors report preliminary data of an Italian multicentric, retrospective study of primary testicular tumors (PTT) in childhood involving 20 Italian departments of pediatric surgery. Forty cases of PTT were observed during a 10-year period (1977–1987), with yolk-sac tumors being the most frequent (45%). Most testicular tumors were first perceived as a unilateral testicular mass. Scrotal ultrasonography represents the main diagnostic investigation, together with serum marker levels.

Key words: Neoplasms – Primary testicular tumors – Germ-cell tumors – Gonadal stromal tumors – Lymphangiomas

Introduction

Primary testicular tumors (PTT) are very uncommon in childhood, accounting for 0.5%–4% of all solid neoplasms in boys under 15 years of age [2, 3, 25]. Because of the rarity of these neoplasms, there still is considerable confusion as to the appropriate surgical treatment of malignant PTT. Although the initial form of therapy is radical inguinal orchiectomy in all patients, the use of retroperitoneal lymph-node dissection remains controversial. Some authors believe that if serum α-fetoprotein (αFP) and β-human chorionic gonadotropin (βHCC) levels regress to normal values postoperatively, retroperitoneal lymph-node dissection can be avoided without compromising long-term survival [4, 6–8, 10–12, 15]. In contrast, other authors advocate retroperitoneal node dissection as a necessary additional treatment in all cases of malignant PTT [12, 18].

The controversy about diagnostic and therapeutic parameters induced us to carry out a multicentric, retrospective study of these neoplasms. This paper reviews all the patients with PTT observed during a 10-year period (1977–1987) in 20 Italian departments of pediatric surgery, with special reference to the surgical treatment of malignant tumors.

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Materials and methods

Forty cases of PTT were observed in 20 Italian departments of pediatric surgery during a 10-year period (1977/1987). The following parameters were taken into consideration:
(1) the histopathologic findings, following the classification of Hays [9]; (2) the interval between the onset of symptoms and diagnosis; (3) significant associated diseases and/or the presence of neoplasms in family members; (4) clinical features; (5) diagnostic procedures (laboratory, imaging); (6) surgical treatment; (7) follow-up; (8) monitoring of serum marker levels; and (9) postoperative imaging studies (ultrasonography and/or CT if necessary).

Results

Histology

Figure 1 lists PTT according to histologic types and shows that in our series there was a high rate of yolk-sac tumors (45%), followed by mature teratomas (25%), teratocarcinomas (7.5%), Leydig-cell tumors (7.5%), Sertoli-cell tumors (7.5%), embryonal carcinomas (2.5%), lymphangiomas (2.5%), and epidermoid cysts (2.5%).

Clinical features

The interval between the onset of symptoms and diagnosis varied from 2 days to 52 weeks. Figure 2 outlines the initial symptoms and signs: in our series, most testicular tumors were first perceived as a unilateral testicular mass (95%), sometimes associated with scrotal pain (15%), hydrocele (7.5%), or acute scrotum (2.5%).

An association was also found between PTT and precocious puberty (5%), undescended testes (7.5%), and the presence of neoplasms in other family members (10%).

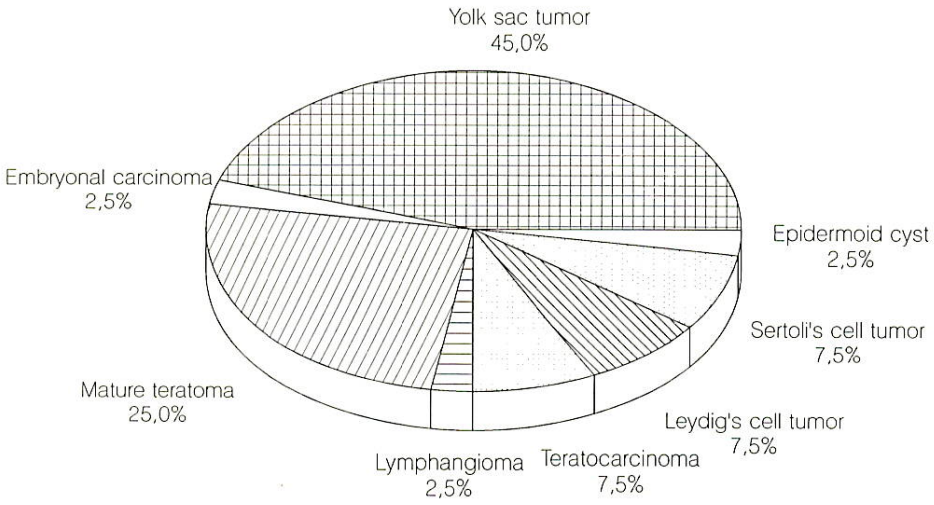

Fig. 1. Distribution of histopathologic types

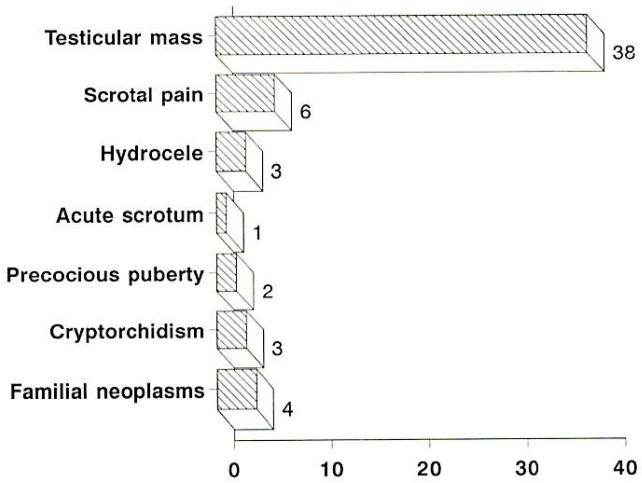

Fig. 2. Clinical features

Specifically, the cases of precocious puberty were found in association with a Leydig-cell tumor and a yolk-sac tumor; all the cases of undescended testes were observed in patients with yolk-sac tumors; and neoplasms were present in family members of 3 patients with yolk-sac tumors and 1 with a Leydig-cell tumor.

Diagnostic procedures

The following laboratory and imaging procedures were performed, depending on clinical and/or logistic circumstances: (1) echography of the testis (40%), associated in 12 patients with echography of the abdomen; (2) CT of the abdomen (15%); (3) needle-biopsy (7.5%); (4) preoperative assessment of serum marker levels: αFP (52.5%), βHCG (25%), and carcinoembryonic antigen (CEA) (10%); and (5) specific hormone assays: follicle-stimulating hormone, luteinizing hormone, and testosterone (7.5%).

Tables 1 and 2 show the laboratory and imaging procedures and relate the results of each investigation to the different histologic tumor types.

Treatment

All patients underwent surgical treatment.

Thirty-five had inguinal orchiectomy immediately followed by retroperitoneal radical lymph-node (RLn) resection in 6 cases; RLn biopsy or "look" was performed in a further 4 patients. Radical resection and biopsy were performed on ipsilateral retroperitoneal lymph nodes and provided histologically negative results in all 10 cases.

In 6 cases, unilateral or bilateral lymph-node dissection was performed after chemotherapy or radiotherapy; 4 of these patients had inconclusive CT scans for the presence of retroperitoneal lymph-node metastases and/or high serum levels of αFP, while histological findings were negative; 2 of these showed clinical evidence of a retroperitoneal mass, as confirmed by positive techniques and histologic specimens.

Two patients underwent excision of the tumor without orchiectomy. Of these 2, 1 had a Leydig-cell tumor and the other had a mature teratoma. Both patients are alive and well at the end of the study period, with postoperative follow-up periods of 36 and 60 weeks, respectively.

Three patients underwent scrotal orchiectomy: of these, 1 had an inguinal recurrence and for this reason underwent retroperitoneal and inguinal lymph-node dissection after chemotherapy. Chemotherapy, which was not examined in this study, was given after surgery to 12 patients with yolk-sac tumors. Six of these underwent retroperitoneal lymph-node dissection after chemotherapy, as previously reported. In most cases various association of bleomycin, vincristine, cis-platinum, actinomycin D, cyclophosphamide, and adriamycin were used. One patient who presented with "giant" retroperitoneal lymph-node metastases [20] underwent radiotherapy.

Staging

The following criteria were utilized for staging of our patients: stage I: tumor confined to the testis, completely removed; stage II: retroperitoneal lymph-node metastasis and/or high levels of αFP 2 months after orchiectomy;

Table 1. Preoperative laboratory diagnostic procedures

Laboratory diagnostic procedure	No. of cases	Results		Histologic tumor type	
αFP	21 (52.5%)	not reported	6	teratocarcinoma	1
				mature teratoma	2
				Sertoli-cell tumor	1
				yolk-sac tumor	1
				lymphangioma	1
		positive	7	yolk-sac tumor	
		negative	8	yolk-sac tumor	4
				mature teratoma	2
				Sertoli-cell tumor	2
bHCG	10 (25%)	not reported	2	Sertoli-cell tumor	1
				mature teratoma	1
		positive	2	yolk-sac tumor	2
		negative	6	yolk-sac tumor	4
				mature teratoma	2
CEA	4 (10%)	negative	4	yolk-sac tumor	3
				mature teratoma	1
follicle-stimulating hormone, luteinizing hormone, testosterone	2 (5%)	negative	2	Leydig-cell tumor	1
				yolk-sac tumor	1

Table 2. Preoperative diagnostic imaging procedures

Imaging diagnostic procedure	No. of cases	Results		Histologic tumor type	
Echo testis	16 (40%)	positive	16	yolk-sac tumor	7
				mature teratoma	5
				Leydig-cell tumor	1
				Sertoli-cell tumor	1
				teratocarcinoma	1
				lymphangioma	1
Echo abdomen	12 (30%)	negative	6	yolk-sac tumor	3
				mature teratoma	3
		positive	2	yolk-sac tumor	2
		inconclusive	4	yolk-sac tumor	4
Ct scan abdomen	6 (15%)	positive	2	yolk-sac tumor	2
		inconclusive	4	yolk-sac tumor	4

stage III: diffuse metastases or metastases above the diaphragm. Following these criteria, only 3 patients with retroperitoneal metastases were included in stage II.

Follow-up

Thirty-five patients were followed for 6-months – 11 years postoperatively by monitoring serum levels of tumor markers (αFP, βHCG), chest radiographs, and echography, and so far no evidence of disease has been found. In 1 case ultrasound examination suggested a retroperitoneal mass and the patient underwent CT scanning that excluded the presence of a retroperitoneal metastasis. The follow-up was not completed for 5 patients.

Discussion

Like many authors [1, 3, 5, 7, 12, 13–16], we found a predominance of malignant lesions, with a high rate of yolk-sac tumors. Most testicular tumors were first perceived as a unilateral, painless testicular mass (97.5%), sometimes associated with a hydrocele (12.5%) or acute scrotum (2.5%), that could result in a primary misdiagnosis.

Early diagnosis is important for the successful management of PTT, as demonstrated by the literature and by our data. Our 2 cases of PTT that were included in stage II because of the presence of retroperitoneal lymph-node metastases showed intervals between the onset of symptoms and diagnosis of 30 and 52 weeks, whereas this interval varied from 2 days to 12 weeks for the patients included in

stage I. Hence, in order to prevent delay in diagnosis and treatment, careful clinical and instrumental examination of testicular masses is essential.

Sonographic examination can be extremely helpful in the preoperative evaluation of scrotal masses in children, in particular to differentiate primary testicular masses from extratesticular ones [22]. It is also useful in evaluating tissue characteristics and, in some cases, the benign or malignant nature of the lesion [13, 21].

Despite the absence of reliable sonographic patterns that allow accurate identification of the lesion, it is possible to delineate some features that can be useful in differentiating benign from malignant lesions. Benign tumors usually have a poorly echogenic structure due to their cystic nature, and well-defined margins; malignant tumors, in contrast, are frequently seen as mostly solid, non-homogeneous masses. Necrotic areas, bleeding within the tumor, and the tendency to invade contiguous structures can be detected by sonography [21].

Preoperative assessment and postoperative monitoring of αFP and βHCG levels is mandatory.

Regarding the therapy of PTT, the results obtained indicate that all testicular tumors should be treated by inguinal orchiectomy. Excision of the tumor without orchiectomy, as recommended by other authors [19] for testicular teratomas in prepubertal patients, must be limited, in our opinion, to epidermoid cysts: multifocal microscopic disease or malignant evolution of minimal residues may occur [17, 23, 24, 26].

Retroperitoneal lymph-node dissection is not a necessary additional treatment for testicular tumors, as demonstrated by the negative histologic findings in our series; lymphadenectomy should be restricted to patients with persistently elevated serum αFP levels after inguinal orchiectomy and chemotherapy. The follow-up of our patients varied from 6 months to 11 years, and so far has shown no evidence of disease.

In conclusion, we believe that multi-institutional studies are the best way to evaluate uncommon tumors. Our findings support the possibility that the high variability of diagnostic and therapeutic parameters may indeed be due to poor co-operation between various centers and experts, i.e., the pediatrician, pediatric surgeon, oncologist, endocrinologist, etc. We would emphasize that a correct therapeutic approach to testicular tumors comes from more careful clinical and instrumental examination together with increased experience obtained from more fruitful co-operation among several centers.

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