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Initial surgery for localized rhabdomyosarcoma: a report from the Italian Soft Tissue Sarcoma Cooperative Group Studies

La chirurgia iniziale del rabdomiosarcoma localizzato: l'esperienza del Gruppo Cooperativo Italiano Sarcomi delle Parti Molli

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Summary

Objective. Analysis of the initial surgical approach in patients with localized rhabdomyosarcoma, enrolled in Italian Studies (1979-2003).
Methods. Surgical records of 587 patients were evaluated. As primary surgery, the excision was recommended if microscopically complete, otherwise biopsy was preferable.
Results. The initial biopsy, performed in almost 50% of patients in RMS-79 protocol, has been adopted more often in the protocol RMS-88 and 96. However, the "complete resection" rate has remained quite stable through the years, as well as the percentage of microscopic residue. The macroscopic residue disease has been more important in RMS-88 study than in RMS-96. Most GUnBP tumors and about 10% of HNnPM and other site were successfully excised. A complete resection was rarely attempted in orbit, HNPM, and GUBP sites; regarding the extremities tumors, 62.5% of patients in RMS-79 study underwent a complete conservative excision, while most patients in RMS-88 and 96 underwent biopsy only.
Conclusion. Initial surgical approach has become more conservative, avoiding mutilations, but the incomplete resection rate is still high. A satisfactory initial excision was obtained in GUnBP RMS (mainly paratesticular). In other localizations the feasibility of a complete resection was slightly better for T1a RMS; a careful preoperative evaluation is always necessary.

Key words

Soft Tissue Sarcoma • Rhabdomyosarcoma • Surgery

Parole chiave

Sarcoma delle parti molli • Rabdomiosarcoma • Chirurgia

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Riassunto

Scopo. *Analisi dell'approccio chirurgico iniziale in pazienti con rabdomiosarcoma localizzato (Studi Italiani 1979-2003).*
Materiali e Metodi. *Valutazione dei dati chirurgici di 587 pazienti. L'exeresi iniziale era raccomandata se microscopicamente completa, altrimenti la biopsia era preferibile.*
Risultati. *La biopsia iniziale, eseguita nel 50% dei pazienti del protocollo RMS-79, è stata adottata in un numero maggiore di pazienti nei protocolli successivi. Tuttavia, il tasso di resezione incompleta si è mantenuto alto, come pure quello dei residui microscopici. La percentuale di residui macroscopici è stata maggiore nel protocollo RMS-88 rispetto a quella del 96. La maggior parte dei tumori GUnVP e circa il 10% di quelli TCnPM e altre sedi è stata asportata completamente. Per quanto riguarda i tumori dell'orbita, TCPM e GUVP, l'exeresi radicale è stata tentata raramente. Nel protocollo RMS-79, il 62,5% dei pazienti con tumore agli arti ha ottenuto una resezione radicale; nei successivi, la maggior parte dei pazienti è stata sottoposta solo a biopsia iniziale.*
Conclusioni. *La chirurgia iniziale è diventata più conservativa, evitando le mutilazioni, ma le exeresi incomplete sono ancora numerose. Un'asportazione iniziale soddisfacente è stata ottenuta nei RMS GUnVP (prevalentemente paratesticolari). In altre localizzazioni le exeresi radicali sono state più frequenti nei RMS T1a; un'attenta valutazione pre-operatoria è sempre necessaria.*

Introduction

RMS account for approximately half of all sarcomas in childhood and represent about 6% of all pediatric malignancies [1].

RMS arise from immature mesenchymal cells, and can be found anywhere in the body, including those sites where normally there are no striated muscles [2]. The histological classification divides them into three main groups: those with favorable prognosis (botryoid and spindle cells RMS), those with intermediate prognosis (embryonal RMS), and those with unfavorable prognosis (alveolar RMS). Following an international definition, primary sites are divided into: head-neck parameningeal (HNPM), head-neck non parameningeal (HNnPM), orbit, genito-urinary bladder prostate (GUBP) and genito-urinary non bladder prostate (GUnBP) (paratesticular site, vagina, uterus), extremities (limbs with shoulder girdle and thigh), other sites (trunk, abdominal wall, intrathoracic and intra-abdominal sites, pelvis, perineal and paravertebral regions). The clinical features depend mainly on the site of the tumor origin, its extension to contiguous organs or tissues and the presence of metastases.

In non-metastatic RMS, all the cooperative studies have demonstrated the crucial role of disease extent at diagnosis in predicting survival, as defined by the clinical TNM and surgical Intergroup Rhabdomyosarcoma Study (IRS) system. Other important prognostic factors are primary site, histology, and local control obtained during treatment.

These tumors are the object of many multi-institutional clinical trials in Europe and the USA, and their treatment is multimodal, including surgery (S), radiotherapy (RT) and chemotherapy (CT) in order to achieve the local control and to eradicate both micro- and macroscopic metastases [2]. The clinical pattern of RMS and the good response to CT explain the less aggressive surgical management adopted in children, comparing it with that in adults. Surgery represents an important step in the therapeutic plan of RMS, and a correct initial approach increases the possibility of a complete remission.

The purpose of this study was the analysis of the initial surgical approach in patients with non-metastatic tumors, enrolled in the National Cooperative Studies of the Italian Association of Pediatric Hematology and Oncology (AIEOP RMS-79, 88 e 96). The compliance to the protocol guidelines was evaluated, and the results obtained are discussed taking into account the main clinical factors.

Materials and methods

From October 1979 to December 2003, 587 valuable patients were enrolled from 35 Italian Pediatric Oncology and Pediatric Surgery Centers: 132 in RMS-79 study, 215 in RMS-88 and 240 in the ongoing RMS-96. Patients' ages ranged from 0 to 18 years (median 61.8 months), but until 1996 it varied between 0 and 16 years. The sites were the following: orbit 74, HNnPM 64, HNPM 114, GUBP 61, GUnBP 98, extremities 53, other sites 123.

Patients were staged according to the TNM clinical system: T1 indicated a neoplasm confined to the anatomic site of origin, T2 a tumor involving contiguous organs or structures, or with neoplastic effusion to adjacent serous cavity (pleural or peritoneal). T1 and T2 tumors were also divided, on the basis of size, into substages "a" and "b", indicating tumors ≤ 5 cm and tumors > 5 cm, respectively. Tx was used when the minimal required characteristics to define the primary tumor were unknown. As to lymph node involvement, N0 indicated no clinical involvement, N1 extension to regional nodes and Nx was used when the clinical involvement was unknown. Similarly, M0 indicated absence of distant metastases, M1 disseminated disease, and Mx absence of data.

The post-surgical staging system, used in all the studies and proposed by the IRS, was based on the results of the initial surgical approach: Gr.I - localized tumor, microscopically resected and no lymph node involvement; Gr.II - tumor grossly resected, but with microscopic residue +/- regional lymph node involvement; Gr.III - biopsy only or excision with macroscopic-residue disease; Gr.IV - distant metastases at diagnosis

The multimodal treatment, recommended in the 3 protocols, has not substantially changed over the years. An initial excision was indicated only when it could be microscopically complete and without major anatomic, functional mutilations, such as orbital exenteration, head-neck dissection, permanent colostomy, ileostomy or urinary diversions, cystectomy, prostatectomy, hysterectomy, limb amputation, resection of muscle groups or neurovascular structures with functional impairment. The tumors had to be excised surrounded by a layer of normal tissue and without rupture. Excisions with microscopic or macroscopic residue, requiring high dose CT and RT, had to be avoided.

If a complete excision was not feasible, the suggested option was a surgical biopsy in order to obtain a diagnosis (tru-cut biopsy was accepted only for selected cases, and the fine-needle aspiration biopsy was discouraged). The delayed surgery had to be planned after the neoadjuvant CT.

Since 1990, the primary re-excision (PrE) has been utilized for those cases in which residual disease was suspected after an initial excision; it consisted in the removal of the area of the previous operation before other treatments, with the aim to obtain a radical excision without mutilations. This procedure was recommended particularly for trunk and extremities and in children below age 3 years , in whom local treatment cannot be generally completed with RT. Particular surgical recommendations were addressed to the paratesticular RMS. These had to be removed by a radical inguinal orchiectomy, with resection of the entire spermatic cord: if the scrotum appeared to be involved, a hemiscrotectomy and the exploration of the inguinal lymph

nodes had to be associated. The regional lymph node biopsy was recommended in all sites only if the primary tumor had been removed. Moreover for extremity RMS, the biopsy was always recommended, because of the high rate of lymph node involvement. In paratesticular RMS, the retroperitoneal lymph node exploration was suggested only in case of a suspected swelling at the imaging evaluation.

Results

An initial excision was attempted in 260/587 patients: 67/132 (50.8%) in the RMS-79 Study, 90/215 (41.9%) in the RMS-88, 103/240 (42.9%) in the RMS-96. In the first protocol, about 50% of patients underwent excision and about 50% just a biopsy; in the following protocol, the rate of initial biopsy raised. The excisions were largely conservative: only in the RMS-79 did two patients undergo a mutilating excision, which was radical in one case and left microscopic residue in the other. During the last two studies no mutilating initial excisions were performed.

In RMS-79, apart from the 2 patients who underwent an initial mutilating excision, 65 had an initial surgical approach to get a complete resection, but only in 31/65 (47.7%) this was successful; in 52.3% of patients, residues were left (microscopic in 16/65 and macroscopic in 18/65).

In RMS-88 an initial excision was attempted in 90/215 patients (41.9%), with completeness achieved in 29 (32.2%), 7 of whom with a PrE; in 25/90 patients microscopic residues were left and in 36/90 they were macroscopic.

In RMS-96, the initial excision was carried on in 103/240 patients (42.9%). It was radical in 48/103 cases (46.6%), through a PrE in 23, resulting in microscopic residue in 37 and with macroscopic residue in 18.

Overall, during the 3 Studies considered, less than 50% of the patients who underwent an initial excision effectively obtained complete resection of the neoplasm. Considering the whole population, only 108/587 cases (18%) obtained the local control through complete initial surgery (Tab. I).

Role of tumor site

Considering the tumor site, the complete excisions in the RMS-79 study was 31/132 (24%): 1/10 (10%) HNnPM, 2/11 (18.2%) GU BP, 14/19 (73.7%) GUnBP, 10/16 (62.5%) extremities, 4/30 (13.3%) other sites.

In RMS-88, the microscopically-complete resections were 29/215 (14%); 22/29 GUnBP tumors (75.9%) obtained a complete excision, while the other sites had lower percentage of radical excision: orbit 2/22 (9.1%), HNnPM 2/28 (7.1%), extremities 1/14 (7.1%), other sites 2/53 (37.7%). None of the HNPM or BP RMS had a complete excision.

In RMS-96, an initial complete excision was obtained in 48/240 cases (20%): 2/26 HNnPM (7.7%), 40/50 GUnBP (80.0%), 2/23 extremities (8.7%), 4/40 other sites (10.0%) (Tab. II).

Role of tumor size and invasiveness

The results of the initial surgery on the basis of tumor dimensions and local invasiveness (T in the TNM system) were evaluated only in the RMS-96 study.

Of the 240 patients enrolled, 121 had tumors ≤ 5 cm and 119 tumors > 5 cm. Seventy patients (58.3%) with ≤ 5 cm tumors underwent an initial excision, and 34 (48.6%) obtained a complete resection. The group included mostly paratesticular tumors and two cases with tumors of extremities.

In the group of > 5 cm tumors, 33 patients (27.7%) underwent an initial excision, which was eventually complete in 14 (42.4%). The tumors were mostly GUnBP, and in three patients they were intra-thoracic, intra-abdominal and involving the neck (Tab. III).

In the group of 48 patients who underwent a complete excision, a PrE was performed in 24: 19/24 had ≤ 5 cm tumors, and 18/24 cases were paratesticular (75%); 9/18 underwent a hemiscrotectomy because a transscrotal surgical approach had been conducted as a first operation: the histology did not demonstrate any evidence of residue or scrotal infiltration.

Analyzing the tumor invasiveness, 113 patients had T1 and 127 T2 tumors. Sixty-seven patients (59.3%) with T1 tumors underwent an initial excision that was complete in 35 (52.2%). Thirty-six patients (28.3%) with T2 tumors had an excision, which was radical in 13

Tab. I. Initial surgical approach (October 1979-December 2003).

	RMS-79 132	RMS-88 215	RMS-96 240	Total 587
Biopsy	65	125	137	327
Conservative excision	65	90	103	258
Complete (after PrE)	31	29 (7)	48 (23)	108 (30)
Microscopic residue	16	25	37	78
Macroscopic residue	18	36	18	72
Mutilating excision	2	—	—	2
Complete	1	—	—	1
Microscopic residue	1	—	—	1

Tab. II. Complete excisions by tumor site.

	RMS-79	RMS-88	RMS-96	Total
Orbit	0/23	2/22	0/29	2/74
HNnPM	1/10	2/28	2/26	5/64
HN PM	0/23	0/43	0/48	0/114
GU BP	2/11	0/26	0/24	2/61
GUnBP	14/19	22/29	40/50	76/98
Extremities	10/16	1/14	2/23	13/53
Other Sites	4/30	2/53	4/40	10/133
Total	31/132	29/215	48/240	108/587

Tab. III. Complete excisions by tumor size (Protocol RMS-96 up to December 2003).

	≤ 5 cm	≥ 5 cm	Total
Orbit	0/13	0/0	0/13
HNnPM	1/12	1/2	2/14
HN PM	0/1	0/4	0/5
GU BP	0/2	0/3	0/5
GUnBP ♂	28/28	10/10	38/38
GUnBP ♀	1/5	1/3	2/8
Extremities	2/6	0/2	2/8
Other Sites	2/3	2/9	4/12
Total	34/70	14/33	48/103

Tab. IV. Complete excisions by tumor invasiveness (Protocol RMS-96 up to December 2003).

	T1	T2 or Tx	Total
Orbit	0/11	0/2	0/13
HNnPM	2/11	0/3	2/14
HN PM	0/0	0/5	0/5
GU BP	0/2	0/3	0/5
GUnBP ♂	27/27	11/11	38/38
GUnBP ♀	2/7	0/1	2/8
Extremities	2/6	0/2	2/8
Other Sites	2/3	2/9	4/12
Total	35/67	13/36	48/103

(36.1%). It is important to note that all the neoplasms excised in HNnPM, extremities and other sites were "not invasive" (T1), while among the 13 completely excised "invasive" tumors, 11 were paratesticular (Tab. IV).

Initial Biopsy

Regarding the 137 patients in the RMS-96 study who underwent an initial biopsy (Tab. V), 117 required surgery; in 18 the biopsy was carried on through a "trucut" and in 2 through a fine needle aspiration biopsy due to an emergency situation.

Discussion

Since the introduction of multimodal therapy for the treatment of RMS, the role of surgery has been deeply modified. The surgeon does not have to make any effort to remove the tumor initially, but has a role in a complex treatment program, alongside the pediatric oncologist and the radiotherapist.

The initial surgery should be planned only if microscopic residues are not left in the bed of the resection, and the procedure is not mutilating. It is very well known that non radical excisions do not have true advantages in the outcome of the disease and those with microscopic residues make treatment more difficult due to the absence of a measurable target.

The progress of the surgical approach, as demonstrated in literature, is also evident in the surgical data of the Italian Studies, especially those of RMS-96.

The initial surgical approach has become more conservative, with the progressive reduction in the number of excisions with macroscopic residual diseases in the three Italian Studies. Debulking procedures are discouraged for their high intra-operative risk, and because they do not offer advantages in term of results compared to a simple biopsy.

The rate of excision with microscopic residual disease is instead still relevant, even in the last study. While in particular sites, such as HNPM or GUBP, the biopsy is inevitable, for some other sites, such as limbs, HNnPM and trunk, the feasibility of a complete resection has been over-estimated: the choice between excision or biopsy is based on clinical-radiological findings, but might also be influenced by the surgeon's skills and the desire to remove immediately the tumor. In these sites the excision at diagnosis may develop into microscopic residual disease and the patients are necessarily given RT and CT. In this situation the possibility to perform a PrE has to be taken into account to obtain or confirm the radicality of the operation, without mutilations. This procedure, introduced by Hays at the end of 80s [3]

Tab. V. Biopsies on the basis of site and technique employed (Protocol RMS-96 up to December 2003).

	FNAB*	Tru-cut	Incisional biopsy	Total
Orbit		1	15	16/29
HNnPM		1	11	12/26
HN PM		7	36	43/48
GU BP		2	17	19/24
GUnBP			4	4/50
Extremities	1	2	12	15/23
Other Sites	1	5	22	28/40
Total	2	18	117	137/240

* FNAB: fine-needle aspiration biopsy

and progressively developed in the last 15 years, has improved the results of primary surgery in many cases, both in the RMS-88 and RMS-96. It has allowed radicality in those cases in which the mass had been removed without following the classic oncologic criteria, either because the lesion had been considered benign (very usual in adolescents and young adults) and also because the patient had to be treated for an emergency situation, such as torsion of the testis, through a transcrotal approach. The analysis emerging from our experience and the data of the literature widely confirm the usefulness of PrE [4].

When the primary surgical operation leaves microscopic residues and a PrE is not feasible, CT and RT must be given at the usual doses.

Patients in Gr.I, in whom the tumor has been removed without microscopic residue, at diagnosis or within 6 weeks with a PrE, are at lower risk of developing relapse and have a better prognosis, in terms of overall survival and event free survival.

The tumor site has been the most important factor to predict a possible complete resection. On the basis of our experience, only for paratesticular tumors, is a primary complete resection safely feasible. In the RMS-96 Study, only 48 out of 103 initial resections planned (46.6%) had been radical, and in 38/48 cases (79.2%) the tumor was paratesticular. This site is considered the most favorable for a radical excision and also for a PrE, in case of possible residue on the tumor bed. The local relapse observed in one patient who had a first operation through a scrotal incision and a PrE with resection of the vaginal layer, without performing the hemiscrotectomy, provides evidence fact that a PrE with resection of all the soft tissues belonging to the testis is mandatory [5]. Hemiscrotectomy is always recommended; however, recent reports question its usefulness in case of a radical transcrotal incision [6 7]. Larger series are needed to confirm these data.

The rate of complete resections has been moderately better in T1a tumors, comparing them with those with T2b tumors, especially for the lesions occurring in limbs and in other sites. In our series, a resection was carried on in 58.3% of patients with ≤ 5 cm tumors, and it resulted radical in half of these patients; in larger tumors, the resection, attempted in 27.7% of cases only, resulted complete in a very low percentage. Similar results have been obtained, if we consider the T-status: 51.3% of patients with T1 tumors underwent a resection, which resulted complete in 50% of them; the resection, performed in 28.3% of patients with invasive tumors, was radical in 1/3 of the cases.

In cases in which a complete excision is not feasible, the biopsy should be the best choice; a delayed surgery is preferable, after the shrinkage of the tumor obtained with CT. At that time a more aggressive surgical approach is encouraged, if necessary, to achieve local control.

In our experience a biopsy was carried on at diagnosis in 80% of patients: this approach was favored by the impossibility to remove the lesion localized in some specific sites, such as HNPM, GUVP, thorax and abdomen, and by the awareness that an incomplete procedure would not have improved the outcome compared with a simple biopsy.

The biopsy must provide a significant quantity of tissue, to allow the histological diagnosis and the cytogenetic-biomolecular studies, which recently have been gaining more importance for the diagnosis and treatment. Tru-cut biopsies are rarely accepted, since they provide small tissue specimen, inadequate for the various studies requested. They may be done in specific clinical, serious situations. Tru-cut biopsies have obtained good results in the series described by Willman and Hugosson [8 9]. These results are not confirmed in our experience. Fine needle aspiration biopsies are not recommended. This technique may recognize a malignant lesion, but does not identify the histological subgroup [10]; nevertheless some studies are describing successful results [11].

The local treatment of RMS is problematic: on one hand it is necessary to limit permanent damage, particularly important in growing children; on the other, a complete removal of the tumor is required to avoid relapses. The initial surgical approach has become more conservative in the last few years, decreasing the number of mutilating operations and excisions with macro-

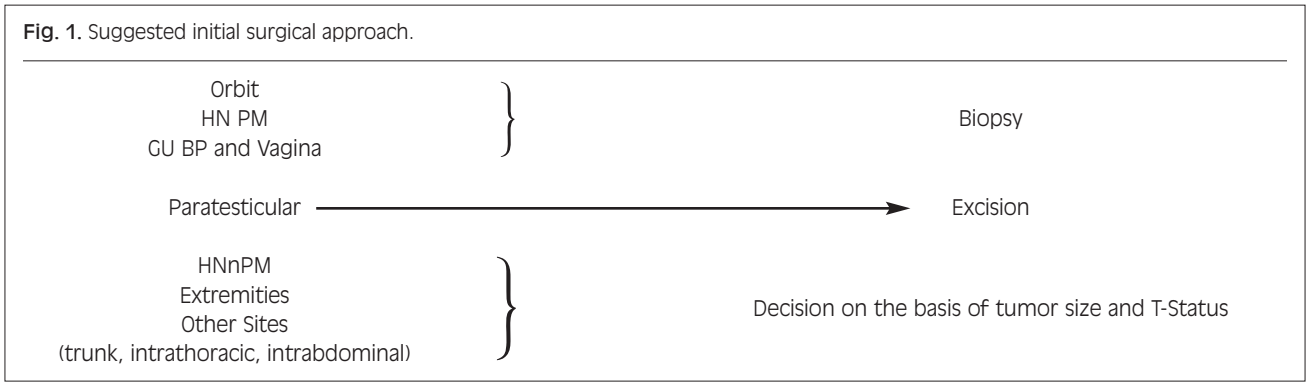

Fig. 1. Suggested initial surgical approach.

scopic residue. However, excisions with microscopic residue are still a problem, probably due to over-estimation of the feasibility of a complete resection. In the past, various scores systems have been proposed to understand whether a radical excision was possible or not, based on tumor site, size and invasivity [12]. If we consider our results, the tumor site is the most important factor to predict the possibility of a complete excision: only the paratesticular site has allowed a higher number of radical excisions. Tumor size and invasiveness must be taken into account in case of lesions of HNnPM, trunk, limbs and other sites (intrathoracic, intrabdominal). Surgical biopsy is the best initial surgical procedure for most tumors, and in particular for orbit, HNPM and GUVP RMS. The possible suggestions on the basis of our experience are summarized in Figure 1. In the future the prognosis will improve if the primary surgical approach is chosen with accuracy, and if there is a deeper collaboration between oncologists, radiotherapists and pediatric surgeons (also those with specific expertise, such as maxillo-facial surgeon, urologist, plastic surgeon, ENT surgeon, orthopedist), whose main target is to make the best decisions in terms of treatment.

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