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Surgical compliance with guidelines for paratesticular rhabdomyosarcoma (RMS). Data from the European Study on non-metastatic RMS

To the Editor,

The superficial site of paratesticular rhabdomyosarcoma (RMS) usually makes early diagnosis and complete primary excision of the tumor feasible. Surgery of paratesticular RMS represents the cornerstone of treatment, and all studies recommend radical orchiectomy with high ligation of the spermatic cord via an inguinal incision [1,2]. Patients with suspected or definite residual tumor after initial surgery require primary re-excision (PRE) in order to achieve a microscopic tumor-free margin (complete resection). PRE should be performed through an inguinal approach when an incomplete removal of the mass or cord has left residual tumor. This may require hemiscrotectomy if the initial approach was undertaken through a scrotal approach [3]. A primary combined inguinal and scrotal approach is sometimes indicated if the tumor is too large to deliver into the inguinal incision without risk of rupture.

The current European rhabdomyosarcoma trial (EpSSG RMS 2005) stipulates these guidelines. In this study, patients with an initial scrotal approach who do not undergo hemiscrotectomy with a PRE must be upstaged and receive more aggressive chemotherapy.

On behalf of the EpSSG RMS 2005 Trial Management Committee, the surgical panel has recently evaluated the compliance of the initial surgical approach to protocol guidelines. Patients with non-metastatic paratesticular RMS enrolled between October 2005 and December 2011were analyzed. The aim was to assess the data quality and possible discrepancies concerning surgery in this site, taking into consideration the high number of centers as well as pediatric surgeons and urologists involved (148 institutions from 15 countries). Since the study is still open, the analysis does not consider the effects on the outcome of the patients.

Data were available in 112 of 124 enrolled patients. The evaluation of compliance of the initial surgical approach to protocol guidelines was carried out in 108 cases, since 4 patients were excluded due to errors in tumor stage or site.

An inguinal approach was initially done in 81 (75%) of 108 patients. However, an orchiectomy with high ligation of the cord was performed in 62 (57%) of 108, and only 55 of 62 achieved an initial complete excision. Residual tumor was left in 7 patients mainly due to large tumor burden (>5 cm). Nineteen of 81 patients underwent incomplete surgery through the inguinal approach. Four were biopsied, 7 had a tumorectomy without orchiectomy, and 7 had an orchiectomy. All subsequently underwent PRE and achieved complete resection.

An initial trans-scrotal approach was done in 27 (25%) of 108 cases: 5 had a biopsy of the tumor, 9 a tumorectomy without orchiectomy, and 13 an orchiectomy. Thereafter, 19 of 27 patients underwent PRE with hemiscrotectomy (15) or without hemiscrotectomy (4). The other 8 did not undergo any further operation and were treated with more aggressive chemotherapy.

Review of these 108 cases has prompted us to share some observations:

1) Paratesticular RMS represents a favorable subgroup of RMS. The superficial location of the tumor usually allows for a complete resection at diagnosis and is generally considered an easy site to access for the surgeon. In this analysis, however, we have observed a variety of initial surgical approaches and operations. Probably these depend on the initial symptoms and clinical characteristics of the patient. Lack of suspicion for the possible existence of a tumor may influence the initial approach.
2) The guidelines for the surgical approach to paratesticular RMS are very similar to North American and European protocols and many papers describe the benefits of a correct inguinal approach. In rare cases when tumors are huge and delivery through the groin is difficult or risks tumor rupture, an inguino-scrotal incision is considered acceptable (keeping the tunica vaginalis intact). Alarmingly only 57% of patients underwent initial radical tumorectomy and orchiectomy with high ligation of the spermatic cord via an inguinal incision. Not all of these achieved complete removal of the tumor. Although the role of hemiscrotectomy has been debated [4], and data on the

outcome of the patients of this ongoing study are not available, previous reports support our guidelines. Clinical studies emphasize the value of PRE in cases with doubtful residual tumor and /or after an inappropriate surgical approach. Patients who obtain complete local control are treated with less intensive chemotherapy.

3) This analysis shows that surgical approaches and procedures were unexpectedly varied at this site. Although many cases with inappropriate primary surgery were salvaged with PRE, this is not considered satisfactory surgical management. Reasons for these inappropriate approaches are complex. Benign scrotal pathology is far more common than malignant disease, and therefore it is not always easy to differentiate the rare tumor from the far more common benign pathology. The time pressure brought on by the potential differential diagnosis of testicular torsion can lead to inadequate preoperative assessment. Patients with scrotal masses are managed in many different hospitals and initially operated on in these centers, unlike patients with RMS located in other anatomical sites who are mainly managed in oncologic institutions.
4) How can compliance with treatment protocols be improved? Dissemination of information to all surgeons who operate on these patients is pivotal. The spread of this information in each country could improve the knowledge of the disease in this particular site and increase awareness that the correct surgical approach represents the cornerstone of successful treatment.

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Letter to the Editor

To the Editor:

We wish to express a cautionary note regarding the recent study by Hassan and colleagues, "Needle core vs open biopsy for diagnosis of intermediate- and high-risk neuroblastoma in children" [1]. While the diagnostic yields of needle core and open biopsies may be similar in the hands of experienced surgeons, radiologists, and pathologists, the choice of procedure has much wider ramifications beyond tumor-naming.

First, although needle core biopsies are usually adequate to identify a tumor as neuroblastic in origin, they do not provide sufficient information about higher-order architecture and regional histologic variation that could affect International Neuroblastoma Pathology Classification and, thus, the assignment of risk grouping and treatment protocol [2]. These neoplasms are highly heterogeneous, and core biopsies can easily miss, for example, a nodule of poorly-differentiated neuroblastoma within an otherwise stroma-rich nodular ganglioneuroblastoma, thus incorrectly ascribing favorable histology to an unfavorable histology tumor and placing the patient on a suboptimal therapeutic regimen. Also, the biopsy tissue is used for ancillary studies equally important for clinical management, including analysis of karyotype and DNA ploidy, *MYCN* amplification, and other molecular studies. It is unclear from the manuscript what criteria were used to define "adequacy," and whether pathologists were involved in assessing this for the study cohort. Nonetheless, despite the inclusion of double the number of open biopsy cases, every single one had enough material for "complete characterization," whereas almost a third of the needle core biopsies were lacking.