Paratesticular embryonal rhabdomyosarcoma
- Case report

G. Gluck¹², B. Spiridonescu², Monica Hortopan³, Manuela Chiriță³, I. Sinescu¹²

¹ „Carol Davila” Medicine University, Bucharest
² Center for Urology and Renal Transplantation, Fundeni Clinical Institute, Bucharest
³ Pathology Department, Fundeni Clinical Institute, Bucharest

Abstract

Introduction and Objectives. Paratesticular rhabdomyosarcoma represents 17% of all malignant intrascrotal tumors in children under 15 years old.

Materials and Methods. A 15 years old male patient presented in our department with a painless right testicular swelling that progressively increased in size over the last two months. Physical examination detected a firm, painless right scrotal tumor measuring about 7/5 cm, no-adherent to the scrotal wall. Tumor markers (beta-human chorionic gonadotropin, alpha-fetoprotein, lactate dehydrogenase) were negative. The ultrasound revealed a tumor mass in the right testis, additional high performance imaging investigation being required. The MRI described a large tumor in the right testis occupying almost the entire testicle, measuring 63/58/45 mm, with heterogeneous periphery contrast flow, small central areas of necrosis and bilateral inguinal lymph nodes up to 16 mm. Chest CT scan reported no pathological lesions.

Results. Surgery was indicated - right inguinal orchiectomy being performed with placement of silicone testicular prosthesis. Final histopathological exam described an adenomatoid tumor (mesothelial benign tumor) measuring 8/6 cm without invasion of the testicular parenchyma, immunohistochemistry being required. The IHC investigation put the final diagnosis of embryonal rhabdomyosarcoma of the epididymis (Ki67 positive for 25%). The tumor staging was pT1BcN1M0 and the patient included in I IRS (Intergroup Rhabdomyosarcoma Study). He was referred to Pediatric Oncology and Radiotherapy Department where he is now under chemotherapy with IFVA – Ifosfamide, Vincristine and Cosmegen (dactinomycin) and Filgrastim well tolerated by the patient. There are 3 types of testicular rhabdomyosarcomas described in the literature: embrionar (most commonly), alveolar and anaplastic. Prognostic factors involved in patient’s survival are related to: tumor size, staging, surgical resectability, patient age, presence of the retroperitoneal lymph nodes and metastasis.

Conclusions. At the moment there is no standardized treatment for this pathology, although studies underlined the importance of chemotherapy and radiotherapy as complementary treatments to surgery. Consequently, a significant growth in survival over 20 years from 25 % to 75% was stated when combined treatment was performed.

Keywords: rhabdomyosarcoma, testicular, orchiectomy

Correspondence to: Dr. Manuela Chiriță M.D.
Fundeni Clinical Institute, Urology and Renal Transplantation Center
258 Fundeni Str., sector 2, code 022328, Bucharest, Romania
Tel/Fax: 021 300 75 70
E-mail: manuchirita@yahoo.com
Clinical cases

Introduction
Testicular rhabdomyosarcoma counts for 17% of all rhabdomyosarcomas and is the most frequent type of sarcoma found in children with an annual incidence of 4-7 cases in a million\(^1\). In most of the cases it develops from the mesenchymal elements of the spermatid cord, epididymis and testicular tunics, forming a painless scrotal mass found in children and young adults.

Materials and Methods
A 15 years old patient, with no medical history, presents in our department with a painless right testicular mass which appeared during the last 2 months. The clinical examination revealed a 7/5 cm firm, mobile, painless right testicular mass. Tumor markers (β-HCG, AFP, LDH) showed no elevation. The scrotal ultrasound raised the suspicion of testicular tumor requiring further investigations.

The abdominopelvic MRI describes a tumor mass involving the whole right testicle of 63/58/45 mm, with irregular contrast enhancement, small central areas of necrosis and bilateral superficial inguinal adenopathies up to 16 mm. Thoracic scan was normal.

Results
The patient underwent right radical inguinal orchiectomy with scrotal prosthesis.

The pathology specimen showed adenomatoid tumor (benign mesothelial tumor) measuring 8/6cm, restricted to the testicular parenchyma, but the immunohistochemistry exam showed 25% positive Ki67 embryonal rhabdomyosarcoma.

Final tumor staging was pT1bN1M0, the patient being enrolled in the IRS I group. Adjuvant chemotherapy was initiated with Ifosfamide, Vincristine, D-Actinomycin (FVA) and profilactic Filgrastine with good tolerance.

Discussions
Rhabdomyosarcoma represents 17% of all scrotal malignant tumors in under 15 years old children. Three types of testicular rhabdomyosarcomas are described in literature: embryonal, alveolar and anaplastic. Prognostic factors that influence survival are related to size, invasion, resectability, age and presence of retroperitoneal adenopathies and distant metastasis. Multimodal treatment is needed and includes surgery (radical inguinal orchiectomy), chemotherapy and radiotherapy. Testicular biopsy for diagnostic purposes or scrotal orchiectomy is not recommended due to the high risk of dissemination.

Chemotherapy as first line is indicated in patients with unresectable tumors for downstaging.

The IRS proposed a new staging for patients with rhabdomyosarcoma taking into consideration the tumor site, resectability and positive lymphnodes as shown in the table below.

Radiotherapy is indicated in patients with distant metastasis. It has not been proven any advantage of radiotherapy in IRS group I patients[2]. Despite that, radiotherapy can be beneficial after surgery in case of residual tumor (R1/R2). Hermans et al. underline the fact that IRS group II patients undergoing retroperitoneal lymphnode dissection and adjuvant chemotherapy have better long time results, offering also an accurate staging[3].
Table 1 – IRS staging

<table>
<thead>
<tr>
<th>Group</th>
<th>Extent of Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group I</td>
<td>Localized disease, excised</td>
</tr>
<tr>
<td>Group Ia</td>
<td>Confined to site of origin</td>
</tr>
<tr>
<td>Group Ib</td>
<td>Infiltrative, beyond site of origin; negative lymph nodes</td>
</tr>
<tr>
<td>Group II</td>
<td>Total gross resection with regional disease spread</td>
</tr>
<tr>
<td>Group IIa</td>
<td>Localized tumor with microscopic residual disease</td>
</tr>
<tr>
<td>Group IIb</td>
<td>Regional disease with positive lymph nodes, excised No microscopic residual disease</td>
</tr>
<tr>
<td>Group IIc</td>
<td>Regional disease with positive lymph nodes Grossly resected with microscopic residual disease</td>
</tr>
<tr>
<td>Group III</td>
<td>Gross residual disease</td>
</tr>
<tr>
<td>Group IIIa</td>
<td>Localized or regional disease, Biopsy</td>
</tr>
<tr>
<td>Group IIIb</td>
<td>Localized or regional disease, Resection (debulking of more than 50% of tumor)</td>
</tr>
<tr>
<td>Group IV</td>
<td>Distant metastasis</td>
</tr>
</tbody>
</table>

Lymphnode metastasis are reported in 1/3 of patients with a 20% rate of pulmonary, liver and bone metastasis at diagnosis. Tumor spread is mostly lymphatic following the iliac and paraaortic route, but also hematogenous to the liver and lungs. Until now there is no standardized treatment for such disease, but studies undergone on large groups of patients showed the importance of multimodal therapy – surgery followed by chemotherapy and radiotherapy. As a consequence in the last two decades there has been a significant improvement in survival, from 25% to 75%.

Conclusions

Rhabdomyosarcomas are aggressive tumors, requiring precise diagnosis and treatment. Physical examination and testicular ultrasound should be routinely performed in all cases of scrotal tumor masses.

Testicular rhabdomyosarcoma treatment involves a multimodal approach, combining surgery (radical orchiectomy) with chemotherapy and radiotherapy according to the IRS group, histopathological subtype, risk and prognostic factors.

References