Case Report

Paratesticular Pleomorphic Rhabdomyosarcoma: A Report of Two Cases

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Pleomorphic rhabdomyosarcoma (RMS) is a rare tumor with an aggressive behavior, described mainly in adulthood. Herein we present two cases of paratesticular pleomorphic RMS in 71- and 16-year-old patients with metastases at initial diagnosis. Histological, immunohistochemical, and ultrastructural findings were essential to confirm diagnosis. Few months after radical orchiectomy, both patients died before or just after starting adjuvant chemotherapy.

1. Introduction

Rhabdomyosarcoma (RMS) is a highly malignant mesenchymal tumor thought to originate from immature striated muscle. It is characterized by the presence of cells having an identifiable striated muscular differentiation with rhabdomyoblasts cells [1]. Twenty percent of all cases of RMS arise from the genitourinary system [2]. The pleomorphic form is typically more frequent in adults and has a poor prognosis [1].

Herein, we report two cases of pleomorphic RMS diagnosed in an adolescent and adult patient with multiple metastases at initial diagnosis.

2. Case N 1

A 71-year-old man presented to our urology department for a 2-year history of testicular swelling and a painless mass in the left scrotum. The patient felt uncomfortable with that mass with a feeling of heaviness. His past medical history was unremarkable. The physical examination showed a 20 cm solid mass occupying the left scrotal region with a buried penis (Figure 1).

The mass seemed to be independent from the testis and ultrasonography confirmed that it was an extratesticular solid mass. A contrast enhanced computed tomography (CT) revealed multiple pulmonary lesions and retroperitoneal lymph nodes. The diagnosis of paratesticular tumour with multiple metastases was made and surgical excision with adjuvant chemotherapy was decided. The patient underwent left high inguinal orchiectomy. The testis was macroscopically normal and two tumours of 15 and 4 cm were localized in paratesticular region (Figure 2). The total weight of specimen was 1.2 Kg. On macroscopic examination, a white, encapsulated partially calcified tumour was observed. Microscopic histopathologic examination revealed a malignant mesenchymatous proliferation made of pleomorphic cells, arranged in sheets and lobules with cross striations. Mitoses were very numerous. Elsewhere, architecture was made of round, polygonal cells, and large multinucleate cells with strongly eosinophilic and vacuolar cytoplasm and with vesicular chromatin. Large areas of necroses were present. The sarcomatoid proliferation invaded muscular and surrounding fat. The testicular parenchyma was normal. Immunohistochemical study showed that tumour cells were positive for desmin and negative for alpha-smooth-muscle actin. The PS100 showed a positivity of the tumoral cells for desmin, the actin smooth muscle, and myogenin attesting the striated muscular nature with this proliferation. Final pathological diagnosis was paratesticular pleomorphic RMS.

Two months after surgery the patient died from cachexia before starting any adjuvant therapy.
3. Case N 2

A 16-year-old boy presented to our department for scrotal swelling evolving for 6 months. He complained of scrotal enlargement and of uncomfortable heavy sensation. He denied any scrotal trauma and his history was unremarkable. Physical examination revealed an enlarged, tender, right scrotum measuring 18 cm with unrecognizable testis. Ultrasonography revealed that the mass originates from the paratesticular region. CT scan showed a solid process in the right scrotum compressing the two testicles with large retroperitoneal lymph nodes. The diagnosis of paratesticular tumour was considered and a right radical inguinal orchietomy was performed.

Pathologic microscopic examination revealed a malignant mesenchymal proliferation made out of fusocellular pleomorphic architecture arranged with spindle-shaped cells. Elsewhere, architecture was diffuse; made of polygonal, racket-shaped cells, and large multinucleate cells with strong hematoxylinophilic cosin staining, vacuolar cytoplasm, and with ovoid circular core provided with vesicular chromatin. There were also many zones of necroses. Mitoses were very numerous. This sarcomatoid proliferation invaded muscular and fat fabric of vicinity. The testicular parenchyma was normal. The immunohistochemical study using the muscular markers and the PS100 showed a positivity of the tumoral cells for desmin, actin smooth muscle, and myogenin attesting the striated muscular nature with this proliferation. The final pathologic diagnosis was pleomorphic RMS involving paratesticular soft tissues.

An adjuvant chemotherapy protocol using vincristine, actinomycin and ifosfamide was started one month postoperatively and radiotherapy to retroperitoneal lymph nodes was planned thereafter. Unfortunately, the patient died 4 months after surgery.

4. Discussion

Malignant tumours of the paratesticular region are uncommon and are mainly sarcomas. The most common histological subtypes of sarcomas are leiomyosarcoma (32%), rhabdomyosarcoma (RMS, 24%), liposarcoma (20%), and malignant fibrous histiocytoma (MFH, 13%) [3].

RMS is a highly malignant tumour arising from striated muscle cells and is associated with early and wide spread metastasis [1, 3]. Common metastatic sites include lungs, lymph nodes, and bone marrow [4]. The usual sites of origin are the head and neck, extremities, and soft tissues; only 15–20% of the cases arise from the genitourinary tract [1].

RMS is more frequent in males than in females with a ratio of 3:1 [5] and occurs mainly in young patients with a mean age of 6 years. There are many subtypes of RMS and all of them may occur in the paratesticular region; however, the commonest subtypes seen are embryonal, alveolar, pleomorphic, and mixed tumours. Pleomorphic RMS is the rarest type and is described mainly in adults and has a poor prognosis, whereas the embryonal type is known to have excellent prognosis [3]. Only spread cases for pleomorphic paratesticular RMS have been reported in adult patients [6–9].

Clinically paratesticular tumour presents as an intrascrotal mass, usually large, sometimes reaching up to the external inguinal ring and compressing the testis and epididymis. In large tumours it may be clinically indistinguishable from testicular tumours. The usual clinical presentation is a painless testicular enlargement. Pain has been reported only in 7% of cases [3], sensation of swelling is more frequent, and a hydrocele can be occasionally present. This could explain why diagnosis can be made late in adults and the tumour could be misdiagnosed as hydrocele; however, ultrasonography can easily demonstrate its solid component and its origin.

Extension of paratesticular tumours to retroperitoneal lymph nodes is common and occurs in up to 30% of patients and lung metastases are present in diagnosis in 14% of cases [10]. Usually a CT is realized to evaluate lymph nodes and distant metastases.

Diagnosis of pleomorphic RMS is histopathological. Diagnosis of this tumour is based on morphologic, immunohistochemical, and ultrastructural findings that identify
Table 1: IRSG Presurgical Staging Classification

<table>
<thead>
<tr>
<th>Stage</th>
<th>Sites</th>
<th>Tumor (T)</th>
<th>Size</th>
<th>Node (N)</th>
<th>Metastases (M)</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Orbit, head and neck (excluding parameningeal) GU: nonbladder/nonprostate</td>
<td>T₁ or T₂</td>
<td>a or b</td>
<td>N₀, N₁, or Nₓ</td>
<td>M₀</td>
</tr>
<tr>
<td>II</td>
<td>Bladder/prostate, extremity, cranial, parameningeal, other (includes trunk, retroperitoneum, and so on)</td>
<td>T₁ or T₂</td>
<td>a</td>
<td>N₀ or Nₓ</td>
<td>M₀</td>
</tr>
<tr>
<td>III</td>
<td>Bladder/prostate, extremity, cranial parameningeal, other (includes trunk, retroperitoneum, and so on)</td>
<td>T₁ or T₂</td>
<td>a</td>
<td>N₁ or Nₓ</td>
<td>M₀</td>
</tr>
<tr>
<td>IV</td>
<td>All</td>
<td>T₁ or T₂</td>
<td>a or b</td>
<td>N₀ or N₁</td>
<td>Mₓ</td>
</tr>
</tbody>
</table>

Note. Tumor: (a) ≤ 5 cm in diameter in size; (b) > 5 cm in diameter in size; Node: N₀, regional nodes not clinically involved; N₁, regional nodes clinically involved by neoplasm; M₀, no distant metastasis; M₁, metastasis present.

Abbreviation: GU: genitourinary.

5. Conclusion

Paratesticular pleomorphic RMS is a rare and aggressive tumour. The diagnosis is made after histological examination. Immunohistochemical and ultrastructural findings could be necessary in some cases. Retroperitoneal lymph node dissection causes a big morbidity and is not recommended. Computed tomography is effective in staging the disease.

The first step of treatment of paratesticular RMS consists of a high inguinal orchidectomy. Adjuvant chemotherapy should be added, with or without radiotherapy, for distant metastases diseases.

Metastatic paratesticular pleomorphic RMS has a poor prognosis.
References


