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LETTER TO THE EDITOR

Male Health

A giant adult paratesticular rhabdomyosarcomar

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Dear Editor,

Rhabdomyosarcoma (RMS) is a rare malignant neoplasm that can involve the bladder, prostate, vagina, or paratesticular area. The peak incidence of paratesticular RMS occurs at 2–5 years of age. Herein, we report a case of adult paratesticular RMS that is relatively uncommon.

A 20-year-old man presented with a mass in the left scrotum that had been progressively growing over a period of 1 month. The patient had found a pea-size neoplasm in the scrotum during bathing 1 month previously. Subsequently, it grew quickly in the scrotum without causing pain and fever. The physical examination resulted in the detection of a soft mass with mild tenderness in the left scrotum. The ipsilateral spermatic cord was obviously thickened when the testis was not palpable. The transillumination test was negative. Serum β-human chorionic gonadotropin and α-fetoprotein levels were normal. A scrotum magnetic resonance imaging (MRI) scan revealed a huge inhomogeneous mass with soft tissue and watery signals surrounding the left testicle (**Figure 1a**). A radical inguinal orchiectomy was performed to obtain a comma like mass (**Figure 1b**). It was soft, flexible, and smooth, measuring 13 cm × 9 cm × 8 cm. The sectioned specimen included a clear boundary of testicle measuring 4 cm × 2.5 cm × 2.5 cm; it surrounded neoplastic tissue with a fish-flesh appearance and hemorrhage and necrosis on the peripheral edge (**Figure 1c**). Histologically the mass consisted of diffuse small round and short spindle cells and deeply stained nuclei; the mitotic count was high (**Figure 1d**). Immunochemical staining indicated Vimentin (+), Myogenin (+), MyoD1 (+), S-100 (−), CK (−) and Desmin showing focal myotube-like differentiation. Taking these findings together, the mass was diagnosed as paratesticular embryonal rhabdomyosarcoma. To clearly evaluate tumor stage we performed computed tomography (CT), which implied the presence of multiple retroperitoneal and cervical lymph nodes and lung metastasis. A bone scan did not indicate any abnormalities. There were no obvious symptoms of metastatic disease. The patient accepted VAC (vincristine, dactinomycin, and cyclophosphamide) and IE (ifosfamide and etoposide) chemotherapy (CCT) on an alternating basis for four cycles. Serious liver damage occurred at the beginning of therapy but was cured after treatment. When the patient finished the first two rounds of chemotherapy a CT scan revealed obviously shrinking of the lymph and lung metastases, which indicated partial remission.

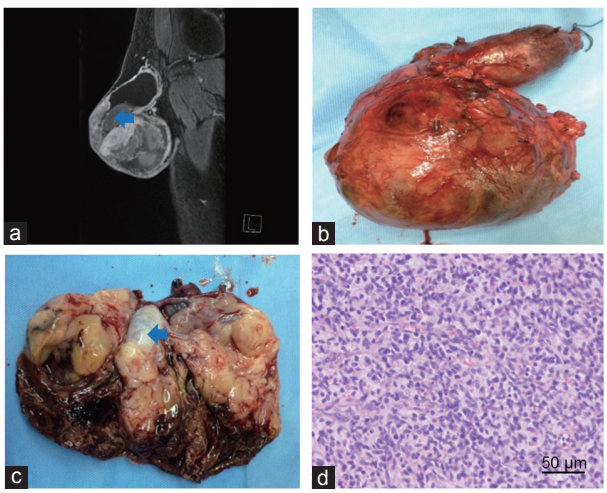

Figure 1: (**a**) Magnetic resonance sagittal image (T1-WI). The blue arrow indicates the testis surrounded by hypointensity above and heterogeneous intensity with nonhomogenous enhancement below. (**b**) Specimen obtained from inguinal radical orchiectomy. (**c**) Dissected specimen. The blue arrow indicates the testis. (**d**) Photomicrograph of a histological slide showing primitive small round blue cells.

Although most patients with paratesticular RMS are children, another age peak occurs in adolescence. Notably, the survival rate is even worse in adults with similar tumors.[1] Patients typically present with a unilateral and painless small mass in the inguinal canal or scrotum, which can easily initially be misdiagnosed as a benign tumor. Indeed, our patient initially ignored the pea-size mass because it was painless. In addition to patient history and physical examination, MRI is usually chosen to investigate the primary mass because ultrasound may not be able to deliver clear images of deeply located tumors.[2] Sagittal MR images revealed that the tumor surrounded the testis, which was consistent with paratesticular RMS arising from mesenchymal elements of the spermatic cord, epididymis, and tunics.[3] However, it was not possible to make a precise diagnosis using radiographic images. In contrast to other sites of RMS, biopsy is not recommended for paratesticular sites because of the high risk of tumor spread.

Radical inguinal orchiectomy is the primary method used for tumor dissection and obtaining sufficient tissue for pathological detection. However, the optimal management of paratesticular RMS still remains unclear because of the rarity of this disease in adults. In addition to complete resection of the primary tumor, CCT, radiotherapy and retroperitoneal lymph node dissection (RPLND) have also been

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reviewed in the literature.[2,4] A remarkable improvement in tumor progression-free and overall survival using CCT has been reported in children, usually involving various combinations of vincristine, dactinomycin, and cyclophosphamide.[5] However, the role of CCT in adults remains controversial. Hawkins *et al*.[6] concluded that CCT was of no benefit in the treatment of patients with RSM aged >21 years. Ferrari *et al*.[7] retrospectively reviewed 171 adults with RMS and found that the overall rate of response to CCT was 85%, which was similar to that reported in children. This finding was consistent with our case. Concerning the intergroup Rhabdomyosarcoma Study (IRS) IV stage of our case, we did not perform RPLND. Opinions regarding the use of RPLND remain inconsistent.[8,9] Most of the time, RPLND is used as part of the staging process, because a CT scan may not be sensitive enough and lead to decrease in identifying retroperitoneal lymph node involvement. Based on the results obtained in the IRS-III trial, patients experienced poor outcomes if they were treated with RPLND followed by CCT.[10]

In conclusion, because the outcomes of adult RMS patients are even worse than those of children, the diagnosis of a scrotal mass should be carried out even more carefully in adults to exclude this malignant disease. When RMS is suspected, regular ultrasound may not be sufficient, and MRI should be used. In addition to surgical treatment, chemotherapy involving the VAC and IE regimens also plays an important role in improving the outcomes of late stage patients. Chest and abdominal CT is usually used for the assessment of partial remission during chemotherapy. The role of RPLND in the treatment of adult RMS still requires additional clinical evidence to confirm its efficacy.

COMPETING INTERESTS

The authors declare no competing interests.

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