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Prikaz slučaja – Case Report
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PARATESTICULAR RHABDOMYOSARCOMA IN THE PEDIATRIC POPULATION – CASE REPORT

PARATESTIKULARNI RABDOMIOSARKOM U PEDIJATRIJSKOM UZRASTU – PRIKAZ SLUČAJA

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Summary **Introduction**: Paratesticular rhabdomyosarcoma is a rare and aggressive intrascrotal tumor. Its superficial localization allows for early detection, an adequate diagnostic approach, and timely therapy, all of which are associated with a favorable outcome.
Case outline: We present the case of a 13-year-and-2-month-old boy with a painless tumor mass in the left hemiscrotum, present for approximately two months prior to admission. He had no symptoms until 2–3 days before admission, when pain in the left hemiscrotum occurred during physical activity. No other complaints were reported. Initial evaluation included a scrotal ultrasound, which showed heteroechoic paratesticular tumor change of the left testis. MRI of the scrotum and pelvis, performed both natively and post-contrast, revealed a spherical, expansive extratesticular lesion located between the testis, which it displaced cranially, and part of the epididymal tail, which it displaced caudally. The lesion measured 35 mm × 36 mm × 37 mm. Laboratory findings, including tumor markers (beta-hCG, alpha-fetoprotein and LDH), were within physiological limits. Additionally, abdominal ultrasound, chest X-ray and the chest CT showed no pathological changes. Following the decision of the oncology board, a radical orchiectomy with complete excision of the paratesticular tumor mass was performed. Histopathology with immunohistochemistry confirmed a high-grade primary malignant mesenchymal tumor with rhabdomyoblastic differentiation, consistent with rhabdomyosarcoma. Accordingly, chemotherapy was initiated following the standard IVA regimen used in Europe (Ifosfamide, Vincristine, Actinomycin). During the 2-year follow-up period from the end of treatment, the patient showed no signs of recurrence of the underlying disease.
Conclusion: Paratesticular rhabdomyosarcoma most commonly presents similarly to other intrascrotal tumors. Due to its aggressiveness and tendency for rapid spread, early diagnosis and treatment are essential. Multimodal therapy—including surgery, chemotherapy, and occasionally radiotherapy—improves survival prospects and contributes to a more favorable outcome.

Keywords: paratesticular rhabdomyosarcoma, surgery, chemotherapy, pediatrics

Sažetak **Uvod**: Paratestikularni rabdomiosarkom je redak i agresivan intraskrotalni tumor. Njegova površinska lokalizacija omogućava rano prepoznavanje, adekvatan dijagnostički pristup i blagovremenu terapiju koja je povezana sa povoljnim ishodom.
Prikaz slučaja: Prikazan je dečak starosti 13 godina i 2 meseca sa bezbolnom tumorskom masom u levom hemiskrotumu, prisutnom unazad oko 2 meseca od prijema. Bez tegoba do 2-3 dana pre prijema, kada se javlja bol u levom hemiskrotumu prilikom aktivnosti. Bez ostalih tegoba. Inicijalno se pristupilo ultrazvučnom pregledu skrotuma gde je uočena heteroehogena paratestikularna tumorska promena u levom hemiskrotumu. Na pregledu na magnetnoj rezonanci skrotuma i male karlice, nativno i postkontrasto, uočena sferična, ekspanzivna lezija smeštena ekstratestikularno položena između testisa koga kranijalno potiskuje i dela repa epidimidimisa koga potiskuje kaudalno, ukupnih dimenzija 35 mm x 36 mm x 37 mm. Laboratorijski nalazi, uključujući i tumor markere (beta hCG, alfa fetoprotein i LDH) su bili u fiziološkim granicama. Takođe, ultrazvuk abdomena, rengenski snimak grudnog koša i CT grudnog koša su bili bez patoloških promena, to jest nije utvrđena zahvaćenost regionalnih limfnih žlezda, niti postojanje udaljenih metastaza. Nakon odluke onkološkog konzilijuma, učinjena je radikalna orhidektomija sa ekstirpacijom kompletne paratestikularne tumorske mase. Patohistološki uz potvrdu imunohistohemije utvrđeno je da promena odgovara *high grade* primarnom malignom mezenhimalnom tumoru sa rabdomioblastičnom diferencijacijom, odnosno rabdomiosarkomu. Shodno tome, primenjena je hemoterapija po standardnom režimu koji se koristi u Evropi, *IVA* (*Ifosfamide*, *Vincristine*, *Actinomycin*). Tokom perioda praćenja od 2 godine od završetka lečenja, pacijent nije pokazao znake recidiva osnovne bolesti.
Zaključak: Paratestikularni rabdomiosarkom se najčešće prezentuje kao i ostali intraskrotalni tumori. Agresivnost tumora i sklonost ka rapidnom širenju zahtevaju ranu dijagnostiku i lečenje. Multimodalna terapija, koja uključuje hirurgiju, hemoterapiju, nekada i radioterapiju, donosi bolje izglede za preživljavanje i povoljniji ishod.

Ključne reči: paratestikularni rabdomiosarkom, hirurgija, hemoterapija, pedijatrija

INTRODUCTION

Paratesticular rhabdomyosarcoma accounts for 7–10% of all genitourinary tract rhabdomyosarcomas in the pediatric population, with a peak incidence between 1 and 5 years of age, as well as a second peak around 16 years of age. It is the third most common localization, following rhabdomyosarcomas of the prostate and urinary bladder (1,3).

The superficial location of this tumor enables early detection, and the majority of patients present with localized disease at the time of diagnosis (60–80%). It most commonly presents as a painless scrotal mass (2).

Scrotal ultrasonography is the initial diagnostic modality for paratesticular rhabdomyosarcoma. Contemporary treatment approaches include surgery, chemotherapy, and radiotherapy (3).

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CASE PRESENTATION

A 13-year-and-2-month-old boy was admitted to the Department of Urology of the University Children's Hospital in Belgrade due to a tumor-altered left hemiscrotum. According to the patient, enlargement of the left hemiscrotum had been present for approximately two months prior to admission. He denied trauma and reported no symptoms until 2–3 days before admission, when intermittent pain in the left hemiscrotum occurred during physical activity. No other systemic complaints were reported.

On admission, the patient was in good general condition, eupneic at rest, afebrile, acyanotic, anicteric, and hemodynamically stable, without peripheral lymphadenopathy or signs of hemorrhagic syndrome.

The abdomen was soft, non-tender, without guarding, peritoneal irritation, or organomegaly. Hernial orifices were free, and the lumbar regions were non-tender to percussion.

Local examination revealed enlargement of the left hemiscrotum. Clinically, a firm, painless paratesticular mass measuring approximately 4–5 cm was palpable in the left hemiscrotum, with a cranially oriented structure morphologically corresponding to the left testis, which was also painless.

The right testis was in a normal position, with preserved morphology and no tenderness. The skin of both hemiscrotums was normally colored.

Laboratory findings were within normal limits. Tumor markers (β-hCG, alpha-fetoprotein, and LDH) were not elevated.

IMAGING DIAGNOSTICS

Initial scrotal ultrasound showed the right testis in normal position, measuring 35 × 20 mm, with homogeneous echotexture and normal vascularization. In the left hemiscrotum, an extratesticular ovoid lesion measuring 33 × 37 × 44 mm was observed. The lesion was well circumscribed, clearly demarcated from the left testis, heteroechoic, with small areas of punctate calcifications and marginal cystic changes, and demonstrated sparse but visible vascularization. The mass was adjacent to the epididymis, from which it could not be clearly separated. The epididymis itself showed normal echogenicity and preserved vascularization.

The left testis measured 37 × 20 mm, was displaced cranially, positioned transversely, and showed normal echogenicity and vascularization.

Given the unclear etiology of the extratesticular mass, pelvic and scrotal magnetic resonance imaging was performed (Figure 1). MRI demonstrated a spherical extratesticular expansive lesion in the left hemiscrotum, located between the testis, which it displaced cranially, and the epididymal tail, which it displaced caudally. The lesion measured 35 × 36 × 37 mm, and infiltration of the left testis and epididymis could not be reliably excluded.

On T2-weighted images, the lesion showed heterogeneous signal intensity with a clearly defined hypointense smooth wall measuring 2–3 mm in thickness. On T1-weighted images, it appeared iso- to hypointense, with a hyperintense smooth wall. Post-contrast imaging demonstrated heterogeneous, sparse peripheral enhancement.

The lesion abutted the scrotal coverings without signs of infiltration. The adjacent epididymal segment showed central areas of increased signal intensity, most likely representing edema. The testis was positioned high, transversely oriented, with normal signal characteristics and dimensions of 35 × 25 × 28 mm. The spermatic cord had normal morphology with increased post-contrast vascularization, without signs of infiltration.

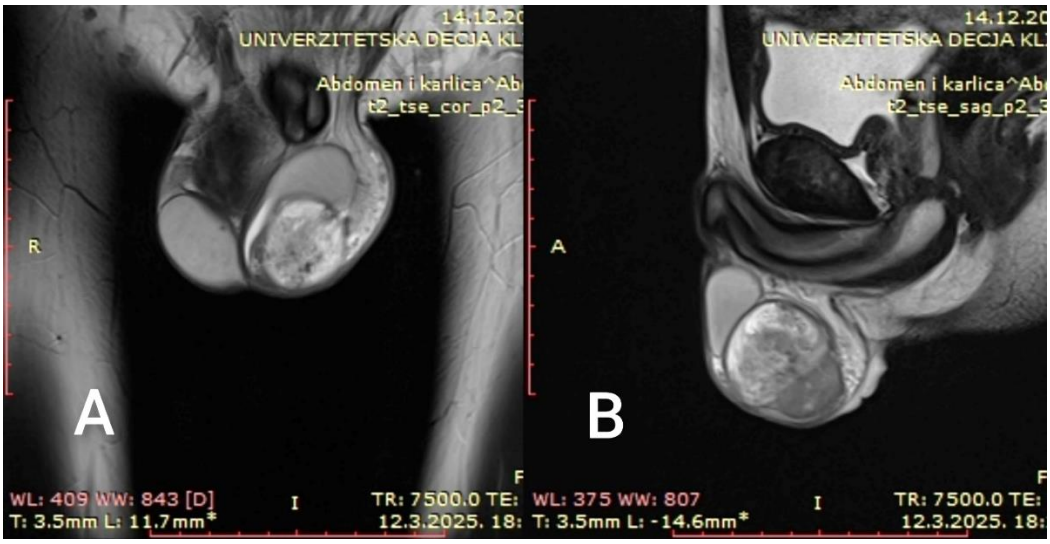
No enlarged inguinal or pelvic lymph nodes were observed.

Abdominal ultrasound, chest radiography, and chest CT were within normal limits, without regional lymphadenopathy or distant metastases.

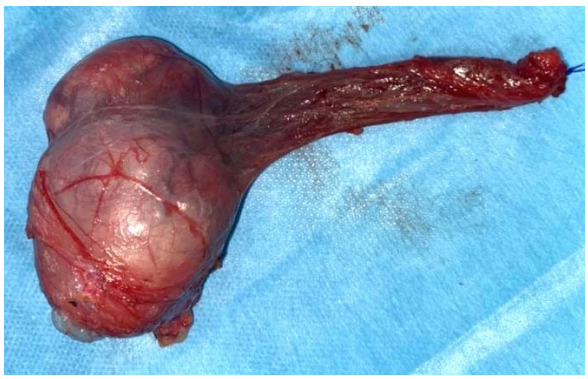
The patient was presented to a multidisciplinary oncologic board, which decided to proceed with tumor extirpation with left-sided orchiectomy.

SURGICAL TREATMENT

After adequate preoperative preparation, radical orchiectomy with complete excision of the paratesticular tumor mass was performed under general anesthesia (Figure 2).



Slika 1. Snimak magnetne rezonance skrotuma, koronarni presek (A), sagitalni presek (B).
Figure 1 Magnetic resonance imaging of scrotum, coronal plane (A), sagittal plane (B).



Slika 2. Ekstirpirana paratestikularna tumorska masa i prateći testis.
Figure 2 Excised paratesticular tumor mass and accompanying testis.

An incision was made along the left inguinal crease, and the anterior wall of the inguinal canal was opened. A thickened spermatic cord was identified, carefully dissected from surrounding structures, and ligated and transected at a high inguinal level.

Due to the size of the tumor and partial adhesion to the scrotal coverings, complete extirpation through the inguinal approach was not feasible. Therefore, an additional incision was made in the left hemiscrotum, allowing careful radical dissection and en bloc removal of the paratesticular tumor mass, the left testis, and the funicular pedicle.

The specimen was sent for histopathological examination.

POSTOPERATIVE COURSE

The postoperative course in the urology department was uneventful, without complications.

HISTOPATHOLOGICAL FINDINGS

Microscopic examination revealed left testicular tissue with a well-demarcated paratesticular tumor nodule, partially encapsulated, composed of spindle-shaped cells with storiform and fascicular growth patterns and partially myxoid stroma. Tumor necrosis and foci of degeneration were present. The testicular tissue and spermatic cord were not infiltrated, and resection margins were tumor-free.

Immunohistochemical staining was positive for vimentin, desmin, myogenin, MyoD1, S100, and MDM2.

The findings were consistent with a high-grade primary malignant mesenchymal tumor with rhabdomyoblastic differentiation, i.e., rhabdomyosarcoma.

CHEMOTHERAPY

Following histopathological confirmation of paratesticular rhabdomyosarcoma, chemotherapy was initiated according to the CWS protocol (standard risk group, subgroup B).

FOLLOW-UP

During a two-year follow-up period after completion of treatment, the patient showed no signs of disease recurrence.

DISCUSSION

Paratesticular rhabdomyosarcoma is a rare and aggressive intrascrotal tumor, most commonly involving the spermatic cord, epididymis, and tunica vaginalis. Early diagnosis is crucial for achieving favorable treatment outcomes (4).

Clinically, it presents similarly to other intrascrotal tumors, most often as a firm, painless mass with rapid progression. As tumor markers are usually not elevated, histopathological and immunohistochemical analyses play a key role in definitive diagnosis, with desmin and myogenin positivity supporting the diagnosis (5).

Scrotal ultrasound is the first-line diagnostic modality, while MRI is used for further evaluation of tumor extent and relationships with surrounding structures (5).

CT of the chest, abdomen, and pelvis is essential for staging and detection of lymph node involvement and distant metastases, which most commonly affect retroperitoneal lymph nodes, lungs, and liver (6).

Treatment is multimodal, combining surgery, chemotherapy, and, in selected cases, radiotherapy, leading to improved outcomes (5).

Surgical management is based on radical high inguinal orchiectomy with ligation of the spermatic cord. Postoperative treatment depends on disease stage. In our patient, chemotherapy was administered according to the standard European regimen (IVA followed by VA) for subgroup B (7).

CONCLUSION

Paratesticular rhabdomyosarcoma most commonly presents as other intrascrotal tumors; therefore, early diagnosis and timely treatment are essential due to its aggressive nature and tendency for rapid spread. A multimodal therapeutic approach including surgery, chemotherapy, and occasionally radiotherapy provides better survival prospects and more favorable outcomes.

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