Prikaz slučaja – Case Report
https://doi.org/10.46793/PP260102022M

EXTRAOSSEOUS EWING'S SARCOMA OF THE PELVIC REGION IN THE PEDIATRIC POPULATION – A CASE REPORT

EKSTRAOSEALNI JUINGOV SARKOM KARLICE U PEDIJATRIJSKOM UZRASTU – PRIKAZ SLUČAJA

Saša Milivojević[1,2], Đorđe Topalović[3]
[1]Univerzitetska dečja klinika, Beograd, Srbija
[2]Medicinski fakultet, Univerzitet u Beogradu, Beograd, Srbija
[3]KBC dr Dragiša Mišović, Beograd, Srbija

ORCID iD: Saša Milivojević https://orcid.org/0000-0002-4152-8165
Đorđe Topalović https://orcid.org/0009-0001-3982-4694

Summary

Introduction: Primary extraosseous Ewing sarcoma is a rare malignant soft tissue tumor, accounting for a small proportion of the Ewing sarcoma family. Pelvic localization in the pediatric population is exceptionally uncommon. Treatment requires a multimodal approach including chemotherapy, surgery, and radiotherapy when indicated, while prognosis depends on patient age, tumor size, and the possibility of complete resection.

Case outline: We present the case of a 13-year-old boy admitted with acute urinary retention, accompanied by impaired urination, constipation, and lumbar pain lasting one month. Ultrasound and MRI revealed a large solid–cystic pelvic mass associated with bilateral grade II hydronephrosis. Tumor biopsy followed by histopathological, immunohistochemical, and FISH analyses confirmed the diagnosis of extraosseous Ewing sarcoma, with diffuse FLI-1 and NKX2.2 positivity.

Treatment was initiated according to the Euro-Ewing 2012 protocol. After six VIDE chemotherapy cycles, a significant reduction in tumor size was achieved. Therapy was continued with the VAI protocol with the aim of further tumor reduction and potential surgical resection. However, despite the initial favorable response, rapid disease progression occurred, leading to abandonment of surgical treatment and initiation of radiotherapy. Due to aggressive local tumor growth, infiltration of surrounding structures, and distant metastatic dissemination, a fatal outcome ensued.

Conclusion: Extraosseous Ewing sarcoma of the pelvic region is an extremely rare and aggressive malignancy in children. Definitive diagnosis relies on histopathological and immunohistochemical evaluation. Although multimodal treatment is essential, pelvic localization is associated with limited therapeutic options and an unfavorable prognosis.

Keywords: extraosseous Ewing's sarcoma, pediatrics, surgery, chemotherapy, radiotherapy

Sažetak

Uvod: Primarni ekstraosealni Juingov sarkom predstavlja redak maligni tumor mekih tkiva i čini manji deo ukupne familije Juingovih sarkoma. Karlična lokalizacija ovog tumora u pedijatrijskom uzrastu izuzetno je retka. Lečenje podrazumeva multimodalni pristup koji uključuje hemoterapiju, hirurgiju i po potrebi radioterapiju, dok prognoza zavisi od uzrasta, veličine tumora i mogućnosti kompletne resekcije.

Prikaz slučaja: Prikazan je slučaj trinaestogodišnjeg dečaka primljenog zbog retencije urina, uz prisutne tegobe otežanog mokrenja, defekacije i bola u lumbalnoj regiji u trajanju od mesec dana. Ultrazvučni i MRI pregled pokazali su veliku solidno-cističnu tumorsku masu male karlice sa bilateralnom hidronefrozom II stepena. Nakon biopsije, histopatološka, imunohistohemijska i FISH analiza potvrdile su dijagnozu ekstraosealnog Juingovog sarkoma, uz difuznu FLI-1 i NKX2.2 pozitivnost.

Započeto je lečenje prema protokolu Euro-Ewing 2012, sa inicijalnim dobrim terapijskim odgovorom i značajnom redukcijom tumorske mase nakon šest VIDE ciklusa. Uprkos tome, došlo je do brze progresije bolesti, te je odustalo od hirurškog lečenja i uvedena je radioterapija. Zbog agresivnog lokalnog rasta tumora, infiltracije okolnih struktura i diseminacije bolesti, nastupio je letalni ishod.

Zaključak: Ekstraosealni Juingov sarkom karlične regije je izuzetno redak entitet u pedijatrijskom uzrastu. Definitivna dijagnoza se postavlja histopatolški i imunohistohemijski. Multimodalni pristup u terapiji uključuje hirurgiju, hemoterapiju i radioterapiju u zavisnosti od resektibilnosti tumora. Karlična lokalizacija tumora nosi lošiju prognozu.

Ključne reči: ekstraosealni Juingov sarkom, pedijatrija, hirurgija, hemoterapija, radioterapija

INTRODUCTION

Ewing sarcoma is a malignant tumor that most commonly arises from bone tissue, whereas primary extraosseous Ewing sarcoma represents a rare form originating from soft tissues and accounts for a smaller proportion of the Ewing sarcoma family of tumors (up to 20%). Localization of extraosseous Ewing sarcoma in the pelvis is extremely rare (1).

Most cases of extraosseous Ewing sarcoma are reported in patients between 10 and 30 years of age (2).

Treatment modalities for this tumor include surgery and chemotherapy for resectable tumors, while radiotherapy is used in tumors that are not amenable to surgical treatment (3,4).

The prognosis of the disease depends on multiple factors; younger age and complete tumor resection are considered the main predictive factors for a better outcome (5). Poorer outcomes are observed in patients older than 14 years, those with tumor volumes greater than 200 cm³, and patients with lung and bone marrow metastases (6).

CASE OUTLINE

A thirteen-year-old boy from a socially disadvantaged family was referred to the University Children's Hospital in Belgrade from a regional health center due to urinary retention. The patient

Korespondencija / Correspondence: Đorđe Topalović, KBC dr Dragiša Mišović, Heroja Milana Tepića 1, 11040 Beograd
E-mail: djotopalovic@gmail.com | Primljen/Received: 2.1.2026 | Revidiran/Revised: 22.3.2026 | Prihvaćen/Accepted: 3.4.2026

reported symptoms lasting for one month, including lower back pain, difficulty voiding, and absence of bowel movements for several days. He denied other systemic symptoms.

Bladder catheterization yielded 2250 mL of urine, after which the patient experienced relief. Following bladder emptying, the abdomen remained mildly distended above the level of the thorax, soft, and non-tender on palpation. A digital rectal examination revealed a tumor mass in the small pelvis.

Subsequent abdominal and pelvic ultrasound demonstrated a pelvic mass, prompting hospital admission. Due to the inability to void spontaneously, a permanent urinary catheter was placed.

Laboratory analyses revealed thrombocytopenia, elevated urea levels, and increased creatine kinase values, while other parameters were within reference ranges. Tumor markers (alpha-fetoprotein, beta-hCG, neuron-specific enolase) were within normal limits. Plasma fibrinogen was mildly elevated.

IMAGING DIAGNOSTICS

Initial abdominal and pelvic ultrasound revealed a mass located posterior to the urinary bladder. The lesion was described as encapsulated, heteroechoic, solid-cystic, with multiple internal septations. Tumor dimensions were 115 × 110 × 145 mm (AP × LL × CC). The largest cystic area, centrally located, measured 100 × 50 mm. The cystic content was hypoechoic (turbid), while the solid components showed minimal vascularization.

The urinary bladder had smooth walls with a minimal amount of appropriately echogenic content.

Pelvic magnetic resonance imaging demonstrated an extensive tumor mass located presacrally, precoccygeally, and anterior to the L5 vertebral body, posterior to the urinary bladder and prostate, medial to the iliopsoas muscles and iliac vessels, and posteromedial to the seminal vesicles (Figure 1). The anterolateral left wall of the mass appeared adherent to the right posterolateral wall of the rectum and distal sigmoid colon, from which it could not be clearly separated. The rectum was deformed and significantly compressed, displaced anterolaterally to the left, while the proximal sigmoid colon remained free.

The caudal portion of the mass was located between the obturator internus muscles (which appeared intact), above the level of the pubococcygeal muscle, which showed pathological signal intensity on the right. The lesion was oval-shaped, measuring up to 145 × 100 × 150 mm (AP × LL × CC), with mixed solid-cystic architecture. Solid components were predominantly peripheral, hypointense on T1W/T2W sequences, with small linear and punctate T1W hyperintense areas suggestive of hemorrhage, showing diffusion restriction. The cystic-necrotic components were multiloculated with numerous thin irregular septa, containing dense fluid of proteinaceous content.

Post-contrast imaging showed enhancement of the capsule, fibrovascular septa, and heterogeneous enhancement of the solid tumor components.

The urinary bladder was empty, containing a urinary catheter, displaced anteriorly and elongated, with its dome located just below the umbilicus.

The prostate was of homogeneous structure, compressed between the pubic symphysis anteriorly and the tumor mass and rectum posteriorly.

The seminal vesicles were displaced anterolaterally, more pronounced on the right, without signal abnormalities.

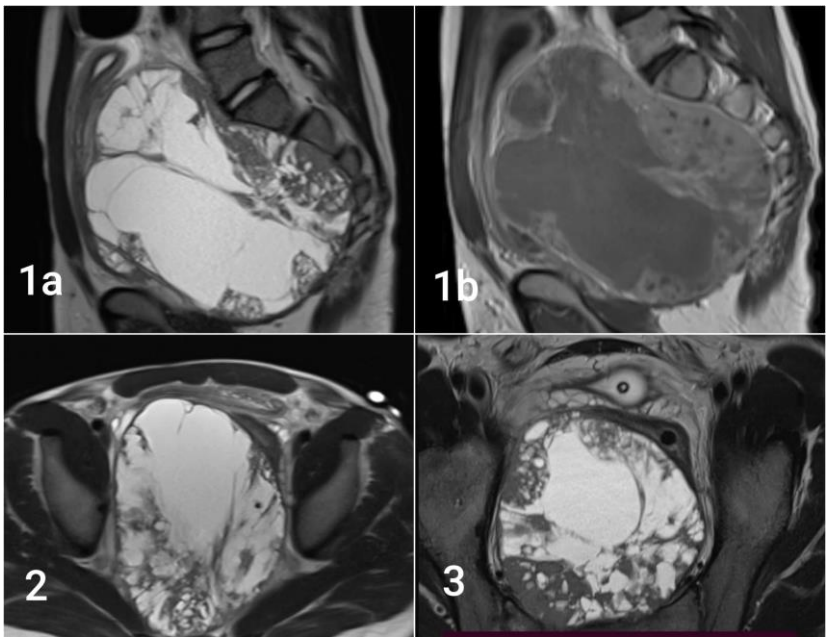
Both kidneys were normally positioned, with craniocaudal diameters of up to 11 cm on the right and 12 cm on the left, preserved parenchymal thickness, and maintained corticomedullary differentiation. Dilatation of the collecting systems and ureters resulted in bilateral grade II hydronephrosis.

Bone and vascular structures were preserved.

Chest CT showed no evidence of pulmonary or mediastinal infiltration. Bilateral reactive axillary lymph nodes were observed, the largest on the right measuring 23 × 12 mm.

Skeletal scintigraphy demonstrated relatively uniform distribution of the osteotropic radiopharmaceutical, with no focal areas of increased osteoblastic activity suggestive of metastatic deposits.

Following complete diagnostic evaluation, the patient was referred to a multidisciplinary board, where a decision was made to perform a biopsy of the pelvic tumor mass.



Slika 1. Snimak abdomena i male karlice magnetnom rezonancom, sagitalni (1a, 1b), transverzalni (2) i koronalni (3) presek.
Figure 1. Abdominal and pelvic MRI scan in sagittal (1a, 1b), transversal (2) and coronal (3) plane.

BIOPSY

Following infraumbilical laparotomy, exploration confirmed a large, previously described tumor mass with a smooth wall and fluctuation on palpation, predominantly cystic in nature. Needle aspiration evacuated approximately 150 mL of serohemorrhagic fluid. The tumor wall tissue was friable and showed profuse bleeding.

Adequate hemostasis was achieved after biopsy, and tissue samples were sent for histopathological analysis. The postoperative course was uneventful.

HISTOPATHOLOGICAL DIAGNOSIS

Histopathological examination revealed features of a small round cell tumor. Immunohistochemical and FISH analyses were most consistent with a tumor from the Ewing sarcoma family. Alpha-SMA positivity is not characteristic of Ewing sarcoma, and the EWSR1 (22q12) translocation is not specific; however, other small round cell tumors with this translocation exhibit different immunohistochemical profiles. The closest immunohistochemical overlap was with desmoplastic small round cell tumor; however, the absence of desmin positivity and diffuse FLI-1 and NKX2.2 positivity favored the diagnosis of Ewing sarcoma.

After receipt of the histopathological diagnosis, the patient was admitted to the Hematology and Oncology Department for further evaluation and treatment. A multidisciplinary oncologic-surgical board decided to initiate chemotherapy in an attempt to achieve maximal tumor reduction without mutilating surgery.

CHEMOTHERAPY

Treatment was initiated according to the Euro-Ewing 2012 protocol. The patient received six VIDE cycles (vincristine, ifosfamide, doxorubicin, etoposide) without complications or episodes of febrile neutropenia requiring blood product transfusion.

After the first cycle, a positive clinical response was observed, allowing removal of the urinary catheter and restoration of spontaneous voiding.

Following the sixth cycle, protocol-based evaluation demonstrated a reduction of the tumor mass to approximately half of its initial size.

Given the favorable initial response, the possibility of maximal surgical tumor reduction without mutilation was considered after an additional chemotherapy cycle (vincristine, adriamycin, ifosfamide – VAI).

However, despite initial success, rapid disease progression occurred after exhaustion of all available chemotherapy modalities according to the Ewing sarcoma treatment protocol (within 16 months of treatment initiation). As a result, maximal surgical reduction was abandoned, and radiotherapy was introduced as the treatment modality.

RADIOTHERAPY

Following consultation for radiation therapy, the patient was transferred to the radiation oncology department, where radiotherapy of the primary tumor site was administered according to protocol.

FOLLOW-UP

The patient was regularly monitored throughout chemotherapy and radiotherapy.

Despite initial tumor reduction, rapid disease progression occurred within 16 months of therapy initiation.

The primary tumor mass significantly enlarged, infiltrating the sacrum and posterior rectal wall with intraluminal extension, resulting in obstructive symptoms accompanied by vomiting and constipation. Hartmann's colostomy was indicated but declined by the patient and his parents.

Metastatic dissemination to the liver, lungs, and bones was also observed.

After exhaustion of all available therapeutic options, the patient died 22 months after initiation of treatment for extraosseous pelvic Ewing sarcoma.

DISCUSSION

Extraosseous pelvic Ewing sarcoma in the pediatric population is an extremely rare entity, with very few reported cases in the literature.

Clinically, extraosseous Ewing sarcoma is characterized by rapid tumor growth with or without local pain and nonspecific radiological features (1). Our patient presented with lumbar pain, urinary retention, and constipation caused by compressive effects of the tumor mass.

Diagnosis largely relies on histopathological and immunohistochemical analyses. Although FLI-1 positivity may be found in other soft tissue tumors, it remains one of the more specific markers for the Ewing sarcoma family. More than 85% of patients harbor the EWS–FLI-1 translocation. Fluorescence in situ hybridization (FISH) is particularly valuable when histological and immunohistochemical findings are inconclusive (7). In the present case, the final diagnosis of Ewing sarcoma was established based on histopathological findings supported by FISH analysis, demonstrating EWSR1 gene rearrangement. However, analysis for specific fusion transcripts (EWSR1/FLI-1) was not performed.

Pelvic tumors generally have a poorer prognosis due to difficult local disease control and close proximity to deep pelvic structures (8).

Treatment requires a multimodal approach combining surgery and chemotherapy, with radiotherapy used for unresectable tumors (3,4). Our patient is discovered in end-stage disease, therefore neo-adjuvant treatment was not sufficient to prevent tumor re-growth.

Patients with extraosseous Ewing sarcoma have a higher incidence of metastases compared to those with osseous Ewing sarcoma, with 30–40% presenting with metastases at diagnosis (9).

In our patient, late presentation, large tumor size, aggressive tumor biology, and exhaustion of all treatment modalities resulted in a fatal outcome.

Given the often subtle and nonspecific presentation, awareness is essential. Parents should be advised to seek medical evaluation in the presence of persistent or progressive pelvic or abdominal pain, unexplained constipation or urinary symptoms, palpable abdominal or pelvic mass, general symptoms such as fatigue or unexplained weight loss. Any symptoms persisting beyond a few weeks, especially if worsening, warrant imaging (ultrasound or MRI) and further evaluation. Early detection significantly improves the likelihood of localized disease and favorable outcome.

CONCLUSION

Extraosseous Ewing sarcoma of the pelvic region is an extremely rare entity in the pediatric population. Due to nonspecific clinical presentation and radiological features, definitive diagnosis relies on histopathological and immunohistochemical evaluation. A multimodal therapeutic approach includes surgery, chemotherapy, and radiotherapy, depending on tumor resectability. Pelvic localization is associated with a poorer prognosis. Favorable outcomes are influenced by early diagnosis, smaller tumor size, tumor molecular characteristics, and complete resection of the primary lesion. Continued reporting of such rare cases and incorporation of contemporary evidence are essential to refine diagnostic pathway and optimize outcomes.

References

1. Gurung S, Thapa S, Gautam S. Extraosseous Ewing sarcoma in a pelvic region: A case report. JNMA J Nepal Med Assoc. 2022;60(251):638–640. doi: 10.31729/jnma.7523
2. Iwamoto Y. Diagnosis and treatment of Ewing's sarcoma. Jpn J Clin Oncol. 2007;37(2):79-89. doi: 10.1093/jjco/hyl142
3. Bailey K, Cost C, Davis I, Glade-Bender J, Grohar P, Houghton P et al. Emerging novel agents for patients with advanced Ewing sarcoma: a report from the Children's Oncology Group (COG) New Agents for Ewing Sarcoma Task Force. F1000Res. 2019;8:F1000 Faculty Rev-493. doi: 10.12688/f1000research.18139
4. Dunst J, Schuck A. Role of radiotherapy in Ewing tumors. Pediatr Blood Cancer. 2004;42(5):465-70. doi: 10.1002/pbc.10446
5. Ahmad R, Mayol BR, Davis M, Rougraff BT. Extraskeletal Ewing's sarcoma. Cancer. 1999;85:725–731
6. Ladenstein R, Pötschger U, Le Deley MC, Whelan J, Paulussen M, Oberlin O, et al. Primary disseminated multifocal Ewing sarcoma: results of the Euro-EWING 99 trial. J Clin Oncol. 2010;28(20):3284-91. doi: 10.1200/JCO.2009.22.9864
7. Lee SY, Lim S, Cho DH. Personalized genomic analysis based on circulating tumor cells of extra-skeletal Ewing sarcoma of the uterus: A case report of a 16-year-old Korean female. Exp Ther Med. 2018;16(2):1343–9. doi: 10.3892/etm.2018.6323
8. Mankin HJ, Hornicek FJ, Temple HT, Gebhardt MC. Malignant tumors of the pelvis: an outcome study. Clin Orthop Relat Res. 2004;(425):212–7. doi: 10.1097/00003086-200408000-00030
9. Zheng C, Zhou Y, Luo Y, Zhang H, Tu C, Min L. Case report: primary Ewing sarcoma of the penis with multiple metastases. Front Pediatr. 2021;8:591257. doi: 10.3389/fped.2020.591257