



### An Unusual Cause of Urinary Incontinence: Ewing's Sarcoma

#### Üriner İnkontinansın Alışılmadık Bir Nedeni: Ewing Sarkomu

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#### ABSTRACT

Urinary incontinence in children can be originated mostly from urinary tract infections, but constipation, neurologic disorders, obstruction and tumors can also be considered among other causes. Pelvic tumors may present with back pain, bladder or bowel dysfunction. Ewing's sarcoma is among the small round-cell tumors of the childhood and potentially can arise from any part of the body. Here, we report an 11-year-old male presented with urinary incontinence and diagnosed as Ewing's sarcoma after 6 weeks' delay. Clinicians should suspect from pelvic tumors in the presence of urinary incontinence especially associated with low back pain.

**Key words:** Ewing's sarcoma, urinary incontinence, childhood

#### ÖZET

Çocukluk çağında üriner inkontinans en sık idrar yolu enfeksiyonu nedeniyle görülmekle birlikte diğer nedenler arasında konstipasyon, nörolojik hastalıklar, obstrüksiyon ve tümörler de bulunmaktadır. Pelvik tümörlü hastalar bel ağrısı, mesane ve barsak fonksiyonlarında bozukluk yakınmalarıyla başvurabilir. Ewing sarkomu insanda tüm vücut bölgelerinden kaynaklanabilen bir tümördür. Bu yazıda, üriner inkontinans yakınmasıyla başvuran ve 6 haftalık bir gecikme sonrasında Ewing sarkomu tanısını alan 11 yaşında bir erkek hasta sunulmaktadır. Klinisyenlerin, üriner inkontinansla başvuran hastalarda özellikle bel ağrısı da eşlik ediyorsa pelvik tümörleri de ayırıcı tanıda düşünmeleri gerekmektedir.

**Anahtar kelimeler:** Ewing sarkomu, üriner inkontinans, çocukluk çağı

#### INTRODUCTION

Ewing's sarcoma (ES) is among the small round-cell tumors of the childhood and potentially can arise from any part of the body. Pain and swelling at the site of tumor are the cardinal symptoms at presentation of the patients with ES<sup>1</sup>. Among the most frequent symptoms of pelvic tumors low back pain, bladder or bowel dysfunction can be seen at the time of diagnosis<sup>2,3</sup>. Here, we report an unusual case of ES localized to

the pelvic region and invading the spinal canal, presenting with urinary incontinence. We did not encounter any case of ES presenting with urinary incontinence neither in English nor in Turkish literature. Pediatricians and pediatric surgeons should pay special attention to patients presenting with urinary incontinence especially with accompanying low back pain for it can arise from a pelvic tumor.

## CASE

An 11-year-old male presented with urinary incontinence that have been present for 6 weeks. In patient history it was learnt that, the patient had experienced incontinence both in day and night and clean intermittent urinary catheterization had been recommended in another center. With continuing complaints and additional back pain the patient admitted to our hospital after 2 weeks. Family history was unremarkable. Physical examination was normal except for pain in suprapubic palpation. Laboratory findings revealed hemoglobin 12.8 g/dL; hematocrit 37.9%; white blood cell count  $7.6 \times 10^6 / \mu\text{L}$ ; platelets  $350 \times 10^6 / \mu\text{L}$  and in serum biochemistry there was no abnormality. Urine analysis showed pyuria with nitrite and leukocyte esterase positivity. *Proteus mirabilis* of 100000 CFU/ml was cultured in urine. Magnetic resonance imaging (MRI) showed a 7.5 cm solid mass lesion in anterior of the sacrum and inferior to the bladder with invasion to the spinal canal. Tru-cut biopsy from the mass revealed small round cell tumor with CD99 positivity consistent with Ewing's sarcoma. Thoracic computerized tomography (CT) showed bilateral metastatic pulmonary nodules. PET-CT revealed an increased FDG accumulation in sacrum. Bone marrow biopsy showed no infiltration of the disease. The patient treated according to the Euro-Ewing 99 protocol<sup>4</sup>. After 6 courses of VIDE (vincristin, ifosfamide, doxorubicin, etoposid) chemotherapy there was no residual tumor in primary and there were millimetrical nodules in both lungs. External beam radiotherapy of 5040 cGy was delivered to primary tumor region. The patient was also treated with 1200 cGy whole-lung irradiation. After 7 courses of maintenance therapy consisting of vincristin, actinomycin-D and cyclophosphamide the treatment was stopped in complete remission. The patient is well without tumor recurrence for 14 months and still needs intermittent urinary catheterization and takes cefixime for prophylaxis of urinary tract infections.

## DISCUSSION

Ewing's sarcoma accounts for 1% of all childhood malignancies, and is the second most common malignant bone tumour in pediatric age group<sup>3</sup>. Ewing's sarcoma is diagnosed frequently in the second decade of life like in our patient. The most frequent localizations affected by the disease are pelvis, diaphysis of the femur, tibia and ribs<sup>1</sup>. Pain and swelling are the cardinal symptoms at presentation. Back pain, bowel or bladder incontinence are not surprising in pelvic masses as in the case of our patient. Urinary incontinence in childhood can arise from many other reasons including urinary tract infection, constipation, neurologic disorders, obstruction and tumors. A tumor anywhere along in urinary tract can block the normal urine flow, leading to overflow incontinence or spinal injury caused by tumor can interfere with nerve signals involved in bladder control, causing urinary incontinence<sup>5</sup>. In the present case, mass lesion in anterior of the sacrum showed an invasion to the spinal canal and unfortunately caused persistent urinary incontinence. The disorder continued even after complete remission for the ES probably because of damage to the sacral nerves.

Pelvic tumors can sometimes present and/or be diagnosed with delay to the some extent. In our case, the mass in the pelvis was missed in another center because the chief complaint at the time of presentation was urinary incontinence. With additional complaint of low back pain, an MRI of the pelvis depicted the tumor. The masses in pelvic region are best imaged with MRI, which ensures the extent of the soft-tissue mass and relation with spinal canal<sup>6</sup>. Histopathological diagnosis of ES is based on the presence of small round tumour cells and immunohistochemical studies including CD99. A reciprocal translocation between chromosomes 11 and 22, the (11;22) (q24;q12) is present in approximately 85% of the patients<sup>7</sup>. Although we did not make cytogenetic study, morphological appearance and CD99 positivity were sufficient for the diagnosis of ES.

Survival rates in ES have steadily improved in recent years, and the increase can be attributed to advances in treatment strategies including surgical techniques, chemotherapy regimens and irradiation schemes. Recent clinical studies which combined these modalities have shown survival rates of near 80%<sup>8,9</sup> although results from our country were slightly inferior<sup>10</sup>. Local control of the tumor in patient with ES is tried to be ensure with surgery and/or radiotherapy. In pelvic tumours, resection is an option in case of no neurovascular or pelvic organ involvement exists. If surgery is not feasible, radiotherapy alone can be used for local treatment. Poor outcome in pelvic tumours can be associated with the delay in diagnosis resulting in large tumour size and increased presence of metastatic disease at presentation. Fortunately, in case of our patient the diagnostic delay was not more than 6 weeks and the tumor response to multimodal treatment was complete.

Clinicians must be alert about urinary incontinence especially with accompanying low back pain in pediatric age group and make further investigation. The possibility of pelvic tumor in such instances should be keep in mind.

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