

## CASE REPORT

# JUVENILE EWING SARCOMA PRESENTING AS A PELVIC MASS

Tahira Naru, Fauzia Haq Nawaz and Javed Rizvi

### ABSTRACT

A teen aged girl with rapidly developing pelvic mass, associated with weight loss and left leg weakness, was evaluated and found to have Ewing's sarcoma of non-osseous origin from pelvis, which is a rare situation. She was treated by a multidisciplinary approach by surgery, chemotherapy and rehabilitation by physiotherapy to effective response.

**KEY WORDS:** Pelvis mass. Juvenile. Ewing sarcoma. Non-osseus.

## INTRODUCTION

Ewing's sarcoma is a highly malignant tumor of osseous or non-osseous origin. It is the second most common bone tumor of childhood and adolescence. More than half of all patients present in the second decade of life and is uncommon in adults older than 30 years. There is a male preponderance of approximately 1.5:1. Ewing's sarcoma rarely occurs in black and Chinese children.<sup>1</sup>

The Ewing's sarcoma usually arises in the metaphysis or diaphysis of the long bones of extremities. Pelvic area, ribs and scapulae are less frequent sites although any bone can be involved. The lungs, bones and bone marrow are the most common sites of metastasis.<sup>1, 2</sup> This report describes a case of extra osseous Ewing sarcoma of the pelvic in teen-age female.

## CASE REPORT

A 15-year-old Pakistani girl presented in the clinic in August 2004 with lower abdominal pain, palpable abdominal mass and left leg weakness. Since 2 months, the abdomen had increased in size with lower abdominal discomfort and significant loss of appetite and weight. It was followed, a few weeks later, by sudden onset of left lower limb weakness and muscular atrophy with difficulty in walking and severe pain starting from back of gluteal region to mid calf.

She was dehydrated and emaciated. Her vital signs and general examination were normal. On abdominal examination, a mass was palpated from left lateral pelvic wall, reaching to just below the epigastrium, which was firm, well circumscribed and non-tender. Liver and spleen were not palpable and shifting dullness was absent. There was no lymphadenopathy. Specific neurological examination of the lower limbs revealed left leg muscular atrophy with less sensory motor power and foot drop.

Complete blood counts, coagulation profile, renal and liver functions were normal. Laboratory investigation revealed hemoglobin of 10.9 gm/dl, hematocrit of 40.1%, total leukocyte

count of 10,000/mm<sup>3</sup>, platelet count of 275,000 /mm<sup>3</sup> and ESR of 39 mm/hr. Tumor markers CA-125 (73.63 iu/ml) and LDH (1100 iu/L) were raised. Serum human choriongonadotrophin and alpha feto-protein were in normal range (<2.010 and 1.76 respectively). Chest X-ray showed no evidence of pleural effusion or any other active lesion.

Ultrasonography of liver, spleen, pancreas and gall-bladder were normal. No para-aortic lymph nodes were seen. Due to pressure effect of the mass, there was bilateral pelviccalyceal dilatation by 1 cm. A solid mass of 17.9 x 9.9 cm was seen in lower abdomen with possibility of uterine sarcoma. Both ovaries were normal.

Doppler ultrasound showed high vascular tumor with possibility of sarcoma. MRI of pelvis revealed a solid mass of 18.0 x 12.0 cm, arising from pelvis and left adnexa, involving the upper part of vagina, obturator internus and gluteus minimus muscles (Figure 1a and 1b).

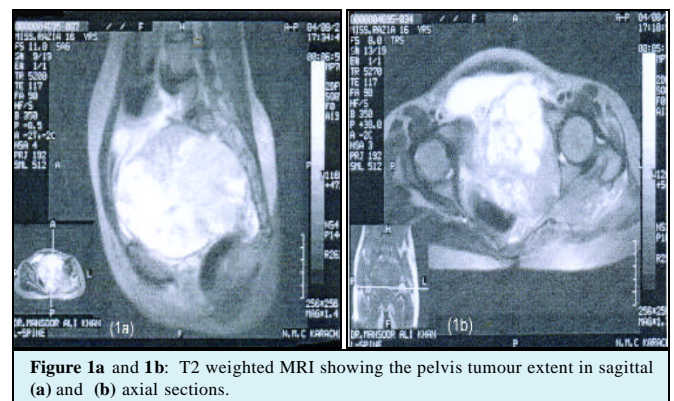
Because of retroperitoneal nature of tumor and suspicion of uterine sarcoma laparotomy was performed. Pelvic peritoneal washing were obtained but there was no free fluid in peritoneal cavity. A large retroperitoneal mass of 18 x12 cm was arising from left lateral pelvic wall to hollow of sacrum, and densely adherent to the posterior wall of uterus. Both ovaries were normal.

Due to high vascularity of tumor, partial debulking was performed. Hemostasis was secured. Frozen section of specimen revealed malignant undifferentiated anaplastic sarcoma with differential diagnosis of Ewing's sarcoma or lymphoma. Postoperatively, she was kept in Special Care Unit (SCU) and received packed cell transfusion and further hospital course was unremarkable.

Department of Obstetrics and Gynecology, The Aga Khan University Hospital, Karachi-74800.

**Correspondence:** Dr. Fauzia Haq Nawaz, Department of Obstetrics and Gynecology, The Aga Khan University Hospital, P.O. Box 3500, Karachi-74800. E-mail: fuzia.nawaz@aku.edu.

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**Figure 1a and 1b:** T2 weighted MRI showing the pelvic tumour extent in sagittal (a) and (b) axial sections.

The diagnosis of Ewing's sarcoma was supported by the identification of glycogen within the tumor cells and by the demonstration of the MIC2 gene product on the tumor cell membrane. The immunohistochemical marker (vimentin and S-100) were positive. Bone scan and bone marrow biopsy were performed, which were negative for metastasis.

Emphasis was given on limb physiotherapy with various exercises. On 6<sup>th</sup> postoperative day, she received first cycle of multi-agent chemotherapy of IVAC (Ifosfamide, vincristine, adriamycin, cyclophosphamide). She was discharged on 10<sup>th</sup> postoperative day. On follow-up visit, the pelvic mass was regressing, so patient and her parents were counseled about compliance for further chemotherapy cycles. After 9 cycles of IVAC, tumor was not palpable abdominally. A repeat ultrasound revealed no pelvic mass. On follow up visits, she was doing well.

## DISCUSSION

Ewing's sarcoma is the second most common malignant bone tumor in young age group, and is commonly diagnosed in the second decade with male preponderance. The annual incidence rate averages less than 2 cases per million children.<sup>1,3</sup> In addition to primitive neuroectodermal tumor of the uterus and mesenchymal lesion of the uterine cervix, there are reports of pelvic mass of osseous and non-osseous origin in either gender.<sup>4-6</sup>

A delay in early symptoms and diagnosis is quite common, particularly of pelvic tumors in which this mass is not palpable until it is quite large.<sup>1,7</sup> The most important and earliest symptom is pain, which may radiate to the limbs, especially with tumor in the vertebral or pelvic region. Neurological signs such as nerve root sign or cord compression are present in one-half of patients with the involvement of axial skeleton. Patient with metastatic Ewing's sarcoma may also present with systemic signs and symptoms such as weight loss and fever.<sup>1</sup>

This patient presented with lower abdominal pain, large palpable pelvic mass, left leg weakness, weight loss, and fever. Laboratory studies in this case showed an increased erythrocyte sedimentation rate, elevated lactate dehydrogenate (LDH), and absence of metastasis. The major prognostic factors in Ewing's sarcoma are: age, site (pelvis, sacrum), volume of tumor (more than 10 cm), and presence of metastasis at the time of diagnosis.<sup>7</sup>

MRI imaging is preferred method in assessing the involvement of soft tissue and bone marrow. Molecular and cytogenetic analysis should be considered as the standard practice in the diagnostic evaluation of Ewing's sarcoma.<sup>1</sup>

With the advent of modern chemotherapy, control of local disease has become more important with increased long-term survival rates. A recent study of Ewing's sarcoma of the pelvis also found an increased 5 years survival rate for patients who had resection of their primary tumor. The patients with non-

metastatic Ewing's sarcoma, treated with chemotherapy and operative resection had a 75% overall 5-year survival rate, compared with 25% overall survival rate where surgery was not performed. Surgery eliminates the residual tumor that are left behind after chemo- or radiation therapy.<sup>4,7</sup>

The development of multi-agent chemotherapy has increased the survival rate of patients with Ewing's sarcoma from 10 % to 70%. More recently, Ifosfamide has emerged as a very effective chemotherapeutic agent, especially in patients resistant to other drugs. With multidisciplinary treatment, long-term survival can be achieved in 70 to 80% of patients presenting with non metastatic disease.<sup>7-9</sup> In this case, laparotomy was planned first due to the suspicion of uterine sarcoma, and the mass being retroperitoneal. With this approach, not only histological confirmation of Ewing's sarcoma was possible, but also more than half of the tumor volume was reduced and further tumor reduction was done by means of multiagent chemotherapy.

Surgical resection, multiagent chemotherapy and radio-therapy are the mainstay of treatment. The treatment plan should be individualized for each patient, which should be based on age, location, stage, size of the tumor and response to therapy.

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