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# CASE STUDY ON EWING SARCOMA

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**Abstract:**
*Ewing's sarcoma is the 2nd most common primary sacral tumor. Ewing's sarcomas are rare, aggressive tumors with a tendency concerning recurrence subsequent resection and early metastasis. Although peak incidents are between the ages of 10 and 20 years, patients of younger or older age account for almost 30% of the cases. We report the case of a 37-year-old healthy male who presented with an 8-month history of pain in his right posterior thigh that was unable to be comforted by non-steroidal anti-inflammatory medicine and physical therapy. Magnetic resonance imaging shown an irregular right presacral mass and core needle biopsy shown a small, round blue cell neoplasm. Staging workup was normal and an open biopsy was positive for the ES translocation. The patient was treated with 5 cycles of vincristine, adriamycin and cytoxan with mesna rescue, alternating with ifosfamide and etoposide. 4 month post-treatment the patient presented with a recurrent tumor. This case emphasizes the importance of timely establishment of initial diagnosis, early metastasis in treatment responsive patients and under-utilization of positron emission tomography-computed tomography (PET-CT) during the treatment to detect sub-clinical metastasis. Doctor changed the chemotherapy medicines to cyclophosphamide and carboplatin after reoccurrence of tumor.*

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## INTRODUCTION:

Ewing's sarcoma (ES) is the 2nd most common bone tumor after osteosarcoma in children and adolescents. ES are aggressive tumors with a tendency towards recurrence following resection and pronounced proclivity toward early hematogenous metastases to lungs and bone. No hereditary or congenital syndromes, environmental or known risk factors have been associated with the occurrence of ES. In 90% of cases, Ewing's sarcoma family tumor (ESFT) cells harbor the translocation t(11;22)(q24;q12), and in the remaining 10% the variant translocation is t(21;12)(22;12). Although peak incidence occurs between the ages of 10 and 20 years, patients of younger or older ages account for almost 30% of the cases. Poor prognostic factors include tumor ≥8 cm, pelvic primary, presence of metastases and age >15 at the time of diagnosis. Older patients with sarcoma also have a higher risk of thromboembolism.

**Case report:**

A 37-year-old healthy male presented with an 8-month history of pain in his right posterior thigh. The pain originated in the patient's right buttock and released to the back of the knee without a radicular component. Motor strength, sensory function and reflexes were normal. Musculoskeletal checkup was within normal limits without tenderness of the hips or back.

Even though the use of non-steroidal anti-inflammatory medications and physical therapy the pain persevered. On consequent reassessment, a radicular component was present. Magnetic resonance imaging (MRI) revealed a large soft tissue intensity mass seen in right hemipelvis extending into gluteal region through greater sciatic notch. The mass measures approx. 19 × 15cm. Diffuse marrow contribution of right hip bone only sparing articular surfaces of sacroiliac joints. Deposits also seen in consensual femoral shafts. Within the pelvis, mass is shifting urinary bladder and recto-sigmoid.

After I/V contrast administration, the shows diffuse enhancement. Mild hip joint effusion and trochanteric bursitis also seen.

Tissue biopsy shown fibrocollagenous and adipose tissue fragments showing foci of round blue cell tumor with hyperchromatic nuclei and slight to clear cytoplasm. The tumor cells are frequently arranged around blood vessels showing pseudorossette pattern. Scattered areas show crushed artifact. These cells show diffuse strong nuclear positivity for FLI-1 and are negative with LCA immunohistochemical stains. Findings are consistent with Ewing's sarcoma.

## DISCUSSION:

The death rate for ES is enormously high when treated with surgery or radiation therapy alone for local control of the disease. Over the past 5 decades, improvements in chemotherapy, surgery and radiation therapy have improved the prognosis of patients with ESFTs. The limited number of patients over the age of 40 and the exclusion of these patients from the majority of trials render these findings tough to generalize.

Tumors of the pelvis have a poorer prognosis when compared with other sites. Whether this is related to the challenge of achieving local control or the proximity to critical deep structures remains to be elucidated. Current treatment recommendations are based on the available literature that is limited by selection bias, small study size, non-standard radiation therapy technique and lack of randomized trials comparing the two management approaches. The role of surgery in treating ES is controversial; however, certain studies advise that resection with chemotherapy and irradiation positively influences patient survival. Radiotherapy is generally applied at doses of 40–45 Gy for microscopic residues and 50–60 Gy for macroscopic disease. Treatment of adult patients follows the same principles. However, tolerability of therapies in adults are taken into account when transferring treatment protocols conceived for patients under the age of 30 years.

Although most cases of ES present as localized disease, overt metastases are accomplished of developing quickly. Microscopic metastatic disease has been postulated to be present at the time of presentation. However, its spread is held in check by as yet anonymous factors secreted by the primary tumor. When the primary tumor is removed or irradiated, the loss of the putative suppressive factors may permit the metastases to grow. The use of chemotherapy in combining with surgery or radiation therapy to treat presumed metastatic disease has significantly improved survival.

Elsewhere specific clinical trials, patients with metastatic disease receive alike therapy to that administered for localized disease, with proper local treatment of metastases, usually radiotherapy. Certain studies have recommended benefit from intensive chemotherapy followed by autologous stem cell rescue, but randomized trials have not yet been performed and the advantage of stem cell transplant remains unproven. Patients with recurrent disease fare poorly, with 5-year survival rates of less than 20% .

Current studies show that, following achieving remission in patients with non-metastatic ES, 30–40% of these patients are probable to develop recurrence of local or metastatic disease . The majority of these studies report a time range of 2–10 years between starting treatment and development of recurrence. Patients relapsing later than 2 years from initial diagnosis have more favorable results.

## CONCLUSION:

This case emphasizes the importance of timely establishment of initial diagnosis, early metastasis in treatment responsive patients. According to current guidelines, initial work-up for staging in a non-metastatic ES is followed by reevaluation of treatment response after 3–6 treatment cycles using focal PET-CT. In treatment responsive ES, local therapy is followed by further chemotherapy. A surveillance follow-up imaging every 2–3 months for the first three years is suggested for localized, non-metastatic ES. The current case has proved complex issues with localized pelvic ES in an older patient who initially responded well to chemo-radiation therapy, with complete resolution of the tumor. Despite adequate control of the local disease, multimodal therapy did not appear to affect metastasis. Although sarcomas are notorious for metastasis into lungs and bone, the utility of PET-CT and MRI for noticing subclinical repetition or metastases has not been established during the treatment period. Only the primary site is evaluated by imaging for treatment response 10–12 weeks in to therapy.

In patients with lung metastases, the resection of residual metastases after chemotherapy, and whole lung irradiation, may grant a survival benefit. Most intensive therapies with additional agents have unsuccessful to markedly increase long-term survival in patients with metastatic disease. The impact of chemotherapy on metastasis of ES patients over the age of 30 remains to be elucidated. A latest intergroup study suggested that the addition of IE to traditional regimens may confer a local control benefit. Similar effects, with the addition of IE on survival advantage, have also been reported by another randomized trial among patients with non-metastatic pelvic ES.

At present, patients should be offered participation in a clinical trial when available. Barring trial participation, multimodality therapy, as described above is recommended.

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