EWING’S SARCOMA IN A 52 YEAR-OLD WOMAN WITH LEG PAIN

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INTRODUCTION

Ewing Sarcoma (ES), often referred to as Ewing’s sarcoma family tumors (ESFTs), is the second most common primary sarcomatous tumor. ES are aggressive tumors with a tendency toward recurrence and pronounced propensity toward early hematogenous metastasis to lungs and bone. In 90% of cases, ESFTs cells harbor the translocation t(11;22)(q24;q12), and in the remaining 10% of cases the variant translocation t(17;22)(q22;q12). Although peak incidences are between the ages of 10 and 20 years, patients at younger or older age account for almost 30% of the cases.

IMAGING STUDIES AND IMMUNOHISTOCHEMICAL SLIDES:

CASE PRESENTATION

CHIEF COMPLAIN:
Right thigh pain

HISTORY OF PRESENTING ILLNESS:
A 52-year-old woman presented with a two week history of pain in her right posterior thigh radiating to the knee. No constitutional symptom was present.

PHYSICAL EXAMINATION:
The strength, sensory function, range of motion and reflexes were normal.

INITIAL DIAGNOSTIC IMPRESSION:
The initial diagnostic impression was hamstring tendinopathy. Despite anti-inflammatory medication and physical therapy the pain persisted. On subsequent reassessment, a radicular component was present.

INVESTIGATIONS AND FINAL DIAGNOSIS:
- Lumbar spine MRI and CT scan demonstrated an irregular shaped complex right presacral mass with thick enhancing wall and central necrosis extending into right S2 neural foramen associated with lysis bone destruction.
- Pathologic evaluation revealed a small, round blue cell neoplasm suggestive of a primitive neuroectodermal tumor positive only for vimentin and CD-99. Fluorescence in situ hybridization analysis was positive for the Ewing Sarcoma gene translocation (t(11;22))
- Staging workup with a PET/CT did not demonstrate abnormal area of uptake and the tumor staged as III T2b, N0, M0.

TREATMENT:
There was significant response after four cycles of therapy with Vincristine, Adriamycin, Cytoxan with, Mesna rescue alternating with Ifosfamide and Etoposide (IE). The involvement of S1 and S2 nerve roots represented a significant surgical challenge. As a result, the patient received radiation followed by additional cycles of chemotherapy. Post treatment CT demonstrated a near complete resolution with minimal residual infiltrates. PET imaging demonstrated normalization of FDG uptake within the pelvic mass.

POST-TREATMENT COURSE:
6 weeks post adjuvant chemotherapy the patient presented with acute shortness of breath. CT scan of the chest showed a new mass within right lower lobe measuring 5 cm, representing internal development of metastatic disease. PET-CT demonstrated areas of increased FDG uptake within both pulmonary hila.

REFERENCES


DISCUSSION

- This case emphasizes the importance of timely establishing a correct diagnosis in patients whose symptoms fail to respond to conservative therapy so that targeted therapy can be started in order to optimize the potential benefit from treatment. Because cancer is rarely in the differential diagnosis for a healthy patient with leg pain, the average time from initial symptoms to actual diagnosis is about 8 months.
- This case also illustrates complex issues with localized pelvic ES in an older patient who initially responded well to combined chemotherapy and radiation therapy with complete resolution of the tumor. Despite adequate control of the local disease, multimodal therapy did not appear to effect metastasis.
- Although sarcomas are notorious for metastasis into lungs and bone, the utility of CT and MRI for detecting subclinical recurrence or metastases has not been established during the treatment or the follow up. Only the primary site is evaluated approximately every 10-12 weeks during therapy.

CONCLUSIONS

- The impact of chemotherapy on metastasis of ES patients older than age 30 is unclear. At this time, patients should be offered participation in a clinical trial when available.