Management of Bone Sarcoma

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KEYWORDS

• Bone sarcoma • Osteosarcoma • Ewing sarcoma • Chondrosarcoma
• Endoprosthetic reconstruction

KEY POINTS

- Advancements in chemotherapy have been the primary reason for improvements in survival from bone sarcoma in the past 20 years.
- There are currently no chemotherapy agents effective against conventional chondrosarcoma.
- Local recurrence of bone sarcoma is likely related to aggressive tumor biology, but relationship with survival is not fully understood.
- Multiple methods of reconstruction after bone sarcoma resection are available, each with its own benefits and drawbacks.
- Emerging technologies, such as computer-aided surgery, improved imaging, and improved implant design, have potential to improve results of treatment even further in the future.

INTRODUCTION

Incidence and Epidemiology

Bone sarcomas account for approximately 0.2% of new cancer cases in the United States each year. The vast majority of these are either osteosarcoma, Ewing sarcoma, or chondrosarcoma. In 2016, it is estimated that 3300 new cases will be diagnosed; this incidence has been rising on average 0.4% annually over the past decade.[1] More than 27% of new diagnoses are made in patients younger than 20 years; osteosarcoma specifically is reported to be the third most common cancer in adolescence, and eighth

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most common cancer in children overall.[2] Unlike osteosarcoma and Ewing sarcoma, which peak in adolescent age groups, chondrosarcoma incidence increases with age.[3]

It is estimated that 1490 patients will die of bone sarcoma in 2016, representing 0.3% of all cancer deaths.[1] For osteosarcoma, the implementation of multimodal treatment with chemotherapy and surgery has led to a considerable improvement in overall survival, but since that time, survival rates have remained relatively stable. In 2015, cause-specific 10-year survival for patients with localized disease at the time of osteosarcoma diagnosis was 65.8%.[4] Metastatic disease at presentation, which is seen in approximately 24% of patients, lowers this survival rate to 24%.[4,5] Despite improvement in survival of localized disease with modern management, patients with recurrence or metastasis after initial treatment is still associated with a poor prognosis.

Pretreatment Evaluation and Staging

The goal of the preoperative evaluation is to determine the extent of the disease, and allow for optimum treatment planning. Local imaging usually includes orthogonal plain radiographs and MRI of the affected area (**Figs. 1** and **2**). Computed tomography (CT) scan may be helpful in identifying cortical involvement. Imaging of the entire affected bone should be included to identify any skip metastases, the presence of which worsens prognosis.[6]

Evaluation of distant disease is done by using chest CT scan to evaluate for pulmonary metastasis, and Technicium-99 whole-body bone scan and/or PET with fludeoxyglucose F 18 (F^{18}-FDG PET)/CT to evaluate for bony metastases[7,8] (**Fig. 3**). Recent studies have demonstrated that PET/CT is more sensitive than bone scan for detecting metastatic bone lesions, while specificity and diagnostic accuracy were similar. The combination of bone scan and PET/CT provides the highest sensitivity, specificity, and diagnostic accuracy, but this must be balanced with the additional cost. PET/CT scan may have the additional benefit of demonstrating correlation with the aggressiveness of a bone lesion, although is not completely reliable for this purpose.[9,10]

Once biopsy is completed, various staging systems exist. The American Joint Committee on Cancer (AJCC) is most commonly used. For bone sarcoma specifically, an alternative system frequently used is the Musculoskeletal Tumor Society (MSTS) system, described by Enneking in 1980[11] (**Tables 1** and **2**).

Biopsy

When performed appropriately, diagnostic accuracy of surgical incisional biopsy has been shown to be 98%,[12] and as such, surgical biopsy is the preferred method of

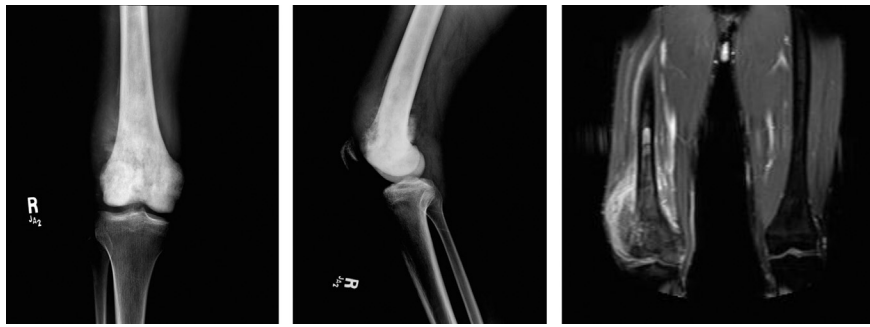

Fig. 1. Orthogonal radiographs and coronal short tau inversion recovery (STIR) MRI scan of conventional osteosarcoma of the right distal femur.