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Osteosarcoma Variant Resembling Renal Cell or Adrenocortical Carcinoma

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Introduction

The evaluation of bone tumors often requires quite one imaging modality, including radiography, CT, MRI and PET. Conventional radiography is usually the first step in the diagnostic process, representing an essential tool in the evaluation of the aggressiveness of the lesion and the response of the host bone. It provides important information regarding lesion site and edges, bone matrix mineralization, cortical involvement and periosteal reaction; thus, it remains the cornerstone for differential diagnosis of skeletal tumors and tumor-like lesions

Primary bone cancers may affect the bone cells. These cancers are all termed “sarcoma”. A sarcoma may be a cancer that originates from cells that construct the connective tissues (supporting tissues) of the body. This includes cells of the bone, muscle, cartilage, ligaments, etc.

Types of bone cancers include osteosarcoma, Ewing's sarcoma, and spindle cell sarcoma. Osteosarcoma of the foot is usually not amenable to limb-sparing surgery because of the poor compartmentalization of the tumor in the foot and the subsequent need to amputate to achieve sound oncology margins. In all of our cases, below-knee amputation was seen to be a satisfactory form of treatment with no evidence of local recurrence in those who had survived for more than 2 years. A high death rate from this condition reflects the high grade nature of these tumors. We cannot discuss the effect of chemotherapy on these patients due to the long period during which patients were treated also because the differences in type and dose of chemotherapy employed. However, with modern chemotherapy regimens, it may be possible to improve the survival of patients having this tumor.

Pleomorphic lip sarcoma is the third, least common and least understood subtype of lip sarcoma. A single characteristic genetic disease has not yet been identified; instead, complex changes are seen with chromosomal duplications, gains, losses, and rearrangements. By histology, pleomorphic lip sarcoma resembles a non-adipocytic soft tissue sarcoma called Malignant Fibrous Histocytoma (MFH), also referred to as Undifferentiated Pleomorphic Sarcoma (UPS), with high cellularity and additionally, presence of pleomorphic lip blasts and occasional multinucleated giant cells. An epithelioid histologic variant resembling renal cell or adrenocortical carcinoma has also been described. Patients with pleomorphic lip sarcoma most ordinarily present with disease within the lower extremity and infrequently at other sites, including the retro peritoneum and mediastinum. Disease progression is much more aggressive compared to the other lip sarcoma subtypes, with a higher (30%–50%) frequency of distant metastasis to visceral organ sites, including lung, bone and liver. Tumors are highly immune to all current treatment modalities.

Thermal ablation techniques also are increasingly getting used within the palliative treatment of painful metastatic bone disease. Currently, external beam radiotherapy is that the standard of look after patients with localized bone pain thanks to metastatic disease. Although the bulk of patients experience complete or partial relief of pain following radio therapy, the effect isn't immediate and has been shown in some studies to be transient in more than half of patients. For patients who aren't eligible or don't answer traditional therapies (i.e. radiation therapy, chemotherapy, palliative surgery, bisphosphonates or analgesic medications), thermal ablation techniques are explored as alternatives for pain reduction.

Several multi-center clinical trials studying the efficacy of RFA within the treatment of moderate to severe pain in patients with metastatic bone disease have shown significant decreases in patient reported pain after treatment. These studies are limited however to patients with one or two metastatic sites; pain from multiple tumors is often difficult to localize for directed therapy. More recently, Sarcoma has also been explored as a potentially effective alternative because the area of destruction created by this system are often monitored more effectively by CT than RFA, a possible advantage when treating tumors adjacent to critical structures. This orderly spectrum is a smaller amount likely to use when comparing tumors of differing histologic groups. FDG avidity of several benign tumors are often as high as or above a number of the malignancies. For example, giant cell tumors of bone are repeatedly reported to be

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more avid than grouped chondro sarcomas. No significant difference between the FDG uptake of high-grade malignancies and benign aggressive lesions, and a different study found a trend toward higher uptake in the benign aggressive lesions than the malignancies. Benign aggressive lesions like giant cell tumors of bone and giant cell reparative granuloma, are often locally destructive but don't typically metastasize with the resultant death of the patient. Additional samples of tumors which will exhibit deceptively high FDG avidity include chondro blastomas, osteo blastomas, osteoid osteomas, Langerhans cell histio cytosis, chondro myxoid fibromas, brown tumors, fibrous dysplasias, fibroxanthoma, desmo plastic fibromas, and small bone cysts. Nevertheless, individual tumor types can have widely varying FDG uptake.

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