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Secondary Angiosarcoma of the Femur after Radiation Therapy for a Primary B-Cell Lymphoma: A Case Report

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Abstract

We present a case of a 68-year-old woman with a complicated history of multiple cancers including diffuse large B-cell lymphoma in the left femur who presented with a pathologic fracture through a radiation induced angiosarcoma of her prior treated bone. She had multiple surgeries on her left femur resulting in a girdlestone resection arthroplasty with a chronic draining sinus communicating with her hip. We present this as a technically difficult surgical case requiring complex decision making and planning. This case highlights the importance of advanced pre-surgical imaging and the benefits of a multidisciplinary approach to cancer patients.

Introduction

Angiosarcoma is a rare, malignant soft tissue sarcoma, accounting for less than 1% to 2% of all soft tissue sarcomas. Interestingly, 40% of angiosarcomas are related to prior radiation history and occur at a rate of approximately 0.9 per 1,000 cases [1-4]. Huang and Mackillop [5] performed a retrospective cohort study of 135 women that suggested those who underwent radiotherapy for breast cancer had a 16-times greater risk of developing angiosarcoma compared to those that did not undergo radiotherapy. A more recent cohort study by Bazire et al. [6] corroborates the increased incidence of sarcomas in patients treated with radiotherapy for breast cancer.

Predisposing factors for the development of angiosarcomas include both radiation and lymphedema. The development of angiosarcoma in patients with chronic lymphedema is best characterized in breast cancer patients following radical mastectomy, and is described as Stewart-Treves syndrome [7]. Given the rarity of angiosarcomas involving bone, there is a paucity of literature pertaining to this topic.

While angiosarcomas can develop in any part of the body, a disproportionate percent involve the skin. In fact, about 50% of angiosarcomas are actually cutaneous angiosarcomas. Furthermore, these are often localized to the trunk, head, and neck. The next most common variants are breast and soft tissue sarcomas at 14.4% and 11.2% respectively. Angiosarcoma involving bone is exceedingly rare. In a review of angiosarcoma by Lahat et al. [8] in 2010, of 222 patients with angiosarcoma only 9 (4.1%) had lesions involving bone [9].

Furthermore, most articles addressing angiosarcoma refer to soft tissue lesions appearing after radiation therapy in management of breast cancer. Our patient, however, developed angiosarcoma after two separate rounds of radiation therapy directed at a primary B-cell lymphoma of her left femur followed by ductal carcinoma of the breast. We realize the limited literature surrounding secondary angiosarcomas of the bone after radiation therapy, and provide our management of a complicated case as follows.

Case Presentation

Our patient had a very complicated history of 4 separate cancers. She initially was diagnosed with lymphoma of her ovary found in 1978 that was treated with resection. In 1998, she suffered a left femoral shaft pathologic fracture that was diagnosed as diffuse large B-cell lymphoma on biopsy. This went on to treatment with femoral IM nailing, CHOP therapy, and 3,600 centigray of radiation to the left femur. Her fracture healed but she continued having hip pain, for which she underwent removal of her nail and conversion to a total hip arthroplasty. Unfortunately, this became infected with MRSA requiring explantation and placement of an antibiotic cement spacer. She underwent several rounds of washouts and eventually went on to a girdlestone due to persistent infection. Antibiotic management during this period was unclear from her records, but she did note having a persistent draining sinus in the proximal portion of her Kocher-style incision. In 2008, she was

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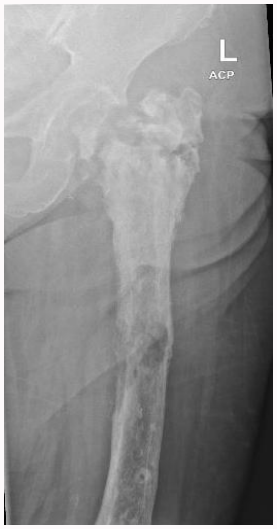

Figure 1: Radiograph of left proximal femur showing evidence of prior girdlestone proximal femoral resection for infection with new aggressive moth-eaten lesion in proximal diaphysis and associated pathologic fracture consistent with her radiation induced angiosarcoma.

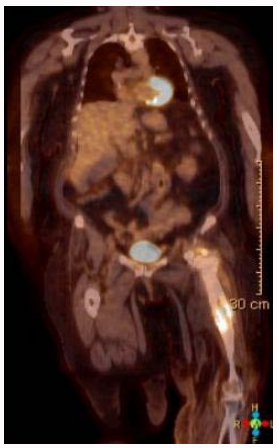

Figure 2: PET/CT for staging showing increased uptake in left femur diaphysis consistent with her radiation induced angiosarcoma of bone without focal uptake in her proximal femur suggesting the cancer was contained to her femur and not affecting her hip joint allowing for a hip disarticulation to achieve negative margins as opposed to hemipelvectomy. Chest imaging showed several indeterminate sub-cm lung nodules, otherwise no evidence of metastatic disease.

diagnosed with ductal carcinoma of her breast which was managed with lumpectomy and 6,000 centigray radiation. Per radiation oncology report the radiation was applied to the entire breast.

She was ambulatory and living at home when she began having weight bearing pain in her left femur in November 2016. Radiographs at her surgeon's office showed a poorly marginated intramedullary lytic lesion in the left femoral shaft, and biopsy performed on November 25, 2016 revealed an angiosarcoma. On December 05, 2016, she bent over and felt a pop in her left thigh, had an increase in pain, and was no longer able to ambulate (Figure 1). At this time, she was transferred to our institution for management. She was worked up with Positron Emission Tomography/Computed Tomography scanning of her chest, abdomen, and pelvis (Figure 2) as well as MR of her left femur with and without contrast (Figure 3). She was found to not have metastatic disease and her sartorius

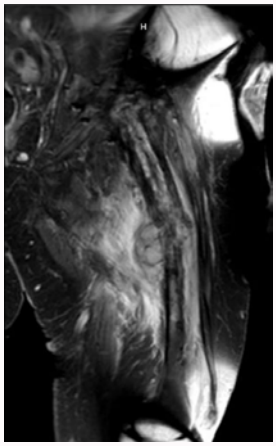

Figure 3: Sagittal view MRI Left Femur with contrast shows aggressive femoral diaphyseal lesion with pathologic fracture and breakthrough of enhancing nodular soft tissue mass with surrounding reactive edema.

and gracilis muscle bellies were disease-free and available to use as a flap. Her case was discussed at our institution's multidisciplinary sarcoma tumor board, and surgical excision by hip disarticulation was recommended. She underwent this procedure on December 15, 2016 and negative margins were achieved. Unfortunately, she went on to have breakdown of the distal portion of her incision and underwent debridement and wound vac placement on January 03, 2017, followed by repeat debridement and wound vac placement on January 05, 2017. The wound was granulating in well and went on to heal secondarily. Cultures taken from her debridement yielded multiple organisms including 3 separate anaerobes. Our infectious disease team was consulted, and she was managed with 6 weeks of zosyn postoperatively. She went on to an extended course of rehab and was able to ambulate with a walker after her treatment. She was offered but refused genetic testing as she did not have any offspring of her own.

Operative Technique

Our preoperative plan was to perform a hip disarticulation. As a quick overview, a hip disarticulation consists of dissecting the proximal femur around the femoral head and neck. After dissection of the femoral head, an amputation is made within the level of the hip capsule. Subsequently, the entire femur and lower extremity are removed as one entity. The patient is subsequently left with an empty acetabulum with no associated lower extremity. For our case, the hip disarticulation started with identifying that the sartorius and gracilis muscle bellies were uninvolved by tumor and appropriates for construct our flaps. Our incision started over the anterior aspect of the iliac crest and extended down the anterior thigh down to just above the adductor hiatus and then went around the back of the thigh to the buttock avoiding the draining sinus, and met up again with itself at the ASIS to make the incision fully circumferential. The sinus tract and involved gluteus maximus was excised. The femoral artery and vein were identified and traced down to the level of the flap. These were ligated just distal to their branch to the Sartorius and gracilis and transected along with the distal muscle bellies. The gluteus maximus musculature was then debrided back to the ilium to excise the entirety of her draining sinus. The hip capsule was resected. The sciatic nerve was identified at its exit point from the sciatic notch and ligated and transected. The hamstrings were then taken off of their origin at the ischium. The short external rotators were freed

from their origins and the remainder of the soft tissues attachments were freed from the femur. The flap was sutured to the outer table of the ilium and acetabulum. Drains were placed in the wound and the incision was closed primarily with addition of an incisional wound vac. After perioperative management the patient went to inpatient rehab, where she started ambulation with a hip disarticulation prosthesis and 2-wheel walker. She was fitted with a prosthetic that allowed her to bear some weight on her disarticulated hip but still required maximum effort with the walker for ambulation.

Discussion

Angiosarcomas while a rare and malignant soft tissue sarcoma have been related to prior radiation and chronic lymphedema. Most current literature addresses the occurrence of secondary cutaneous angiosarcomas in patients with primary breast cancer. During the care of our patient, it became clear that there is a paucity of literature relating to secondary angiosarcoma of the bone. While angiosarcoma of the bone only accounts for about 4.1% of angiosarcoma cases, they are associated with substantial morbidity [8]. Our case highlights the complex diagnostic and management considerations when dealing with a bony angiosarcoma. Further research is needed to better understand the natural history of these lesions and to establish the best strategies for their management.

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