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Case Report

Extraskeletal Osteosarcoma Recognized following Acute Traumatic Injury

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Keywords
Extraskeletal osteosarcoma · Trauma · Radiation

Abstract
Extraskeletal Osteosarcoma (ESOS), a rare entity accounting for less than 2% of all soft tissue sarcomas. Known risk factors for development include: middle aged and elderly patients, a history of radiation, and a controversial link to trauma. The typical presenting symptoms, if any, are tenderness and swelling. In trauma patients, these symptoms often mask the ESOS diagnosis and are assumed to be hematoma or other traumatic diagnosis. Easy misinterpretation of what appears to be obvious traumatic injury, can lead to delays in accurate diagnosis and appropriate treatments.

A condensed poster presentation of this case was reported on February 12, 2018, at Southeastern Surgical Congress Annual Meeting: Kiosk 2 – Cancer, Tampa, FL, USA.

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Introduction

Extraskeletal Osteosarcoma (ESOS) is a seldom seen occurrence, historically defined by Allan and Soule in 1971 with three characteristics: first, the presence of a uniform morphological pattern of sarcomatous tissue, excluding possibility of mixed mesenchymal tumor, next the production by sarcomatous tissue of malignant osteoid, bone, or both, and finally the exclusion of osseous origin [1]. The study further went to suggest the prevalence of ESOS as 1.2% in all soft tissue sarcomas, a figure which has been heavily referenced in the literature [1, 4, 6, 7]. To provide more evidence to the rarity of this pathology, Wurlitzer reported that ESOS comprised only 3.6% of all total osteosarcomas [2]. Though, a large recent study produced by Thampi et al. reviewed information from the SEER database ranging from 1973–2009 and evaluated 4,173 cases of high grade osteosarcoma with an incidence in this review of ESOS at 6.1% [8]. The overall entity over ESOS is rare on its own, however this case necessitates presentation due to the large size and the exceedingly rapid growth of the tumor, in a patient who could be considered at risk for developing ESOS. Through literature review it appears that this case is not only unique due to the extremely size of 26 cm, recognized following an acute traumatic event, but also due to the rapidity of its growth to reach this size over five months. Whereas both trauma and radiation therapy have previously been described in literature with a possible link to developing ESOS, this patient's history and presentation placed her in now what can be assumed to be a heightened risk for development of ESOS [1, 3–5, 7].

Here, we present one case of a 70-year-old female who presented to the Surgical Clinic within less than a month of minor trauma to her thigh. Her evaluation was initially suspicious for hematoma and was asked to return for follow up in 6 months if symptoms were not improved. However, she returned just three months later with a painful, rapidly enlarging, mass at the site that obviously necessitated further investigation. After further imaging, biopsy, and removal, pathology was significant for a 26 cm Extraskeletal Osteogenic Sarcoma.

Case Report

Patient is a 70-year-old female with past medical history significant for prior breast malignancy status post breast conserving surgery, adjuvant chemotherapy and radiation, and five years of anastrozole treatment, who presented to the Surgical Clinic for evaluation of assumed tender hematoma on the medial aspect of her left thigh. Patient reports that painful swelling appeared on medial portion of proximal left thigh after low grade trauma one month prior when the site in question collided with exercise equipment. She stated that she was experiencing mild discomfort, especially with ambulation, and that the tender area was enlarging. She was planned for follow up in six months for her presumed traumatic hematoma. However, patient returned to clinic three months later as pain was persistent, the "hematoma" continued to enlarge, and was beginning to affect her mobility. At that time, patient underwent ultrasound (US) and computed tomography (CT) to rule out any underlying pathology and evaluate the extent of the hematoma in order to proceed with incision and drainage. The US was pertinent for report of "massive hematoma on medial left thigh, 17.8 cm × 12.4 cm × 12.1 cm, comprised of multiple septations." Her extremity (CT) scan, revealed a significant mixed solid and cystic mass within the Sartorius muscle of the medial left thigh with multiple nodules and septations (Fig. 1). The mass measured approximately 12.2 cm × 12.5 cm × 17.5 cm, and was noted to abut the Superficial Femoral Artery and be in contact with the Saphenous vein. No skeletal involvement was visualized. Given these findings, the diagnosis of hematoma was

discarded in favor of neoplasm. She underwent Magnetic Resonance Imaging (MRI), where reports measured a well-defined, T2 intense mass, 11 cm × 13 cm × 19 cm, in the left Sartorius muscle with mixed components of nodules and multiple septa, as well as subacute and chronic hemorrhages, and a diffuse, thick capsule (Fig. 2). The mass compressed the Vastus Medialis muscle and displaced the adductor compartment, as well as abutted the Femoral Artery and Femoral Vein, and displaced and compressed the Greater Saphenous Vein, with no documented bone involvement. Due to these findings, the patient underwent biopsy of the mass which revealed a myxoid sarcoma. The decision was made with the patient to proceed with surgical excision of the mass, and pathology results of the excision were significant for a 26 cm Extraskeletal Osteogenic Sarcoma. She underwent Positron Emission Tomography (PET), which was negative. Her post-operative course was complicated by a 10 cm seroma formation at the surgical site. A Jackson-Pratt drain was placed, which had continued daily outputs of greater than 600 mL, so she went to the operating room for evacuation and closure. She developed a site infection and seroma recurrence, then returned to the operating room for debridement and full thickness skin graft with negative pressure wound vac therapy. Prior to beginning chemotherapy, patient had worsening pain and respiratory complaints, thus she elected to pursue palliative therapy and shortly thereafter expired.

Discussion

Extraskeletal Osteosarcoma (ESOS) is a unique diagnosis in the medicine field. Since it was originally described by Wilson in 194 and further defined by Allan in 1971, a limited number of cases have been reported in literature [1, 9]. With ESOS accounting for only approximately 1.2% of all soft tissue sarcomas, and only 3.6% of all osteosarcomas, the occurrence is proven to be low [1, 2]. The reported occurrences of ESOS overall have shown similar predisposing factors, all of which occurred in our patient: middle aged or elderly [1–4, 7, 8, 10], involvement of lower extremities [2, 4, 7, 8, 11, 12], history of radiation therapy [1–5, 7, 10], and a possible link to traumatic events [1, 3–7, 11–13]. Tumor size has also been reported as an important prognostic factor. Unfortunately our patient had a significantly large tumor, measuring 26 cm at time of surgical excision. In the report published by Bane et al., comparing patients with a tumor size of <5 cm to size >5 cm, tumor size was an important predictive value of patient survival ($p < 0.001$). Only one tumor related death occurred in the <5 cm group out of 7 patients, whereas in comparison to 14 deaths in a >5 cm population of 16, at time of study production [4]. To add further evidence to the significance of tumor size in ESOS survival, Berner et al. also compared tumor size, with their variable of study being 10 cm. According to their published data, a one year survival rate of 91% was appreciated in patients with a tumor size <10 cm, in comparison to 54% one year survival rate in those patients with tumor size of >10 cm [10].

Treatment of ESOS is also rather controversial, with traditional treatment restricted to local surgery alone, or with the possibility of chemotherapy and radiation [10]. A previous study reported their confirmation of no appreciable difference observed in survival rates between local resection and amputation for treatment, as well as that adequate surgery was a significant prognostic factor for survival with no reported influences on survival of patients who underwent radiation therapy [10]. Treatment of ESOS with chemotherapy historically has been unable to confirm a consistent positive response, unlike the success of chemotherapy for intraskeletal osteosarcoma [4, 10, 12]. Though some recent studies produced evaluating

multi-agent chemotherapy may provide beneficial results of therapy, these studies require further investigation [7, 10].

Aside from the overall uniqueness of this entity's general occurrence, the size at resection was uniquely large, and the time frame after the initial traumatic event to grow to this large size at resection, was exceedingly quick and rather unprecedented. Through literature review of multiple other published studies on ESOS, few reported cases of larger size were discovered [1, 2, 4–6, 10–15]. The literature search performed for the production of this study, produced one extremely large ESOS reported at 50 cm by Chung and Enzinger, though median size reported for the overall study was only 7 cm. As well as two studies identified 30 cm ESOS. One report was per Bane (median 10 cm), though specific patient details regarding history or duration of symptoms were not reported by Bane (or Chung). The other report of a 30 cm ESOS by Fine and Stout was present for 9 months, but patient history and median size of the study were not provided [4, 11, 14].

Multiple cases of ESOS occurrences have been recognized after traumatic events [1, 4–7, 11–13]. For which, the recognition, questionably the development, has been primarily seen years later, which is in stark contrast to our patient's rapid development in only five months. The report by Sood et al details the development of a 17 cm mass, eighteen months post traumatic event; Bane details three trauma related ESOS diagnoses, one following repeated chronic trauma, the others diagnosed 25 and 36 years after the inciting traumatic event [4, 13]. The landmark study produced by Allan et al., details a traumatic relationship in six of their twenty – six patients in question, the time frame from trauma to diagnosis ranged from 10 months to 12 years, with one patient sustaining chronic trauma to the site [1]. Sordillo also reports 6 out of 48 patients with ESOS and an identified history of trauma, with a period ranging from 2 years to 15 years from event to diagnosis, two of which sustained chronic trauma [5]. A history of traumatic events prior to diagnosis was reported by Chung et al though no details regarding size at intervention for this subset of patients was reported [11].

Conclusion

Through extensive literature review, there is no question that Extraskeletal Osteosarcoma is a rare medical occurrence. With this notion, it has been proven by this case in particular due to the many risk factors that in the acute setting following a traumatic event to the lower extremity, in an elderly patient with a history of radiation, with an unexplained mass, concern should be elicited for the diagnosis of ESOS. The case presented here displays the unusually acute and rapid growth of a small, mass recognized after trauma into a 26 cm pathologically diagnosed ESOS over five month course. After literature review, such rapid expansion of ESOS after initial presentation, could not be found. Thus, it is with this occurrence that we implore physicians to practice strict regular follow up visits after traumatic events to the lower extremities, especially in middle aged or elderly patients with past medical histories significant for radiation or questionable presentation, and to have a low threshold for additional interventions if suspicious.

Statement of Ethics

Informed consent was obtained over the telephone in the presence of a witness from the patient's brother, given her death during production, for publication of this case report, "Extraskeletal Osteosarcoma Recognized following Acute Traumatic Injury," and any accompanying images. A copy of the consent obtained is available for review by the Editor of this journal.

Ethical approval to report this case was obtained from Duke LifePoint Conemaugh Memorial Medical Center Institutional Review Board, ID 17-29.

Disclosure Statement

The authors of this article declare that they have no conflicting interests regarding the publication of this paper.

Author Contributions

Contributors to conception and design, acquisition of data, and interpretation of data: KRC.
Manuscript writing and drafting: KRC, SLM.
Revising it critically for important intellectual content: KRC, SLM.
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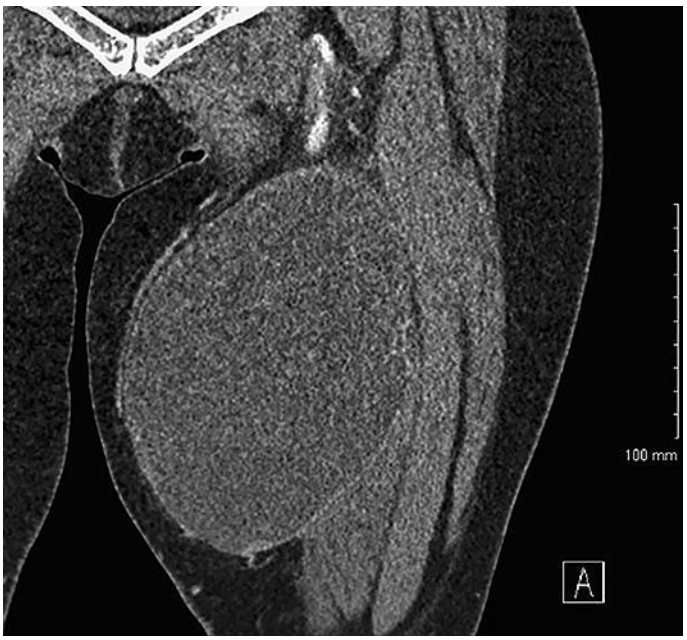

Fig. 1. Computed Tomography coronal view of mass on patient's left medial thigh. Given patient worsening symptoms, she underwent CT imaging which revealed a significant mixed solid and cystic mass within the Sartorius muscle of the medial left thigh with multiple nodules and septations, measuring approximately 12.2 cm × 12.5 cm × 17.5 cm, and abutting the Superficial Femoral Artery and in contact with the Saphenous vein. No skeletal involvement was visualized.

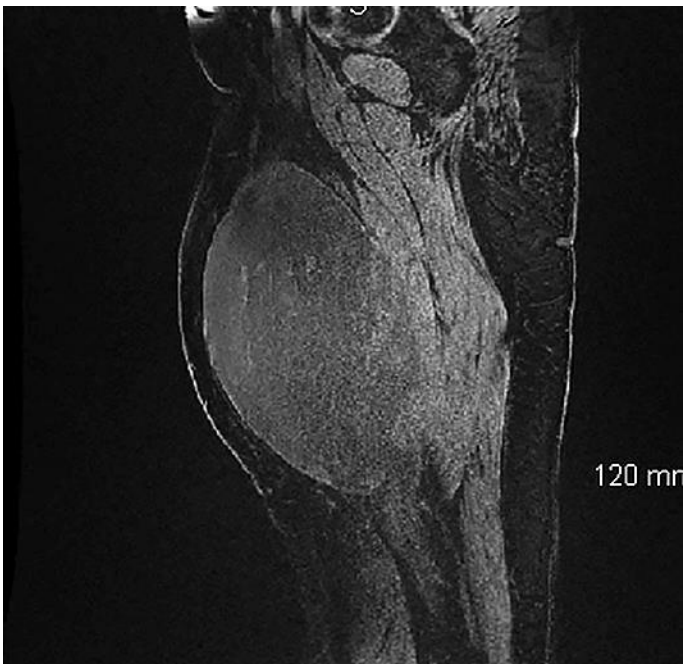

Fig. 2. Magnetic Resonance Imaging sagittal view of mass on patient's left medial thigh. Due to patient findings on CT scan, she proceeded with MRI which demonstrated a well-defined, T2 intense mass, 11 cm × 13 cm × 19 cm, in the left Sartorius muscle with mixed components of nodules and multiple septa, as well as subacute and chronic hemorrhages, and a diffuse, thick capsule. The mass compressed the Vastus Medialis muscle and displaced the adductor compartment, as well as abutted the Femoral Artery and Femoral Vein, and displaced and compressed the Greater Saphenous Vein, with no documented bone involvement.