Case Report

A Rare Case of Humerus Synovial Sarcoma Originated from Bone Tissue: Case Report

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Abstract

Synovial sarcoma (SS) is a malignant mesenchymal neoplasm that is relatively common in the distal extremities. Primary SS of bone is an extremely rare finding. Here in this report, we present a 44-year-old male patient referred with bone and later bone fracture that was finally diagnosed with primary SS of thumerus. So far, 13 documented cases of primary SS of the bone have been reported. The current case is the second known case of primary SS of humerus. Our case was treated with both neoadjuvant and adjuvant chemotherapies associated with surgical tumor removal and prosthesis implantation. Follow-up of the case demonstrated significant remission but with late metastasis and subsequent advanced chemotherapy regimens.

Keywords: Humerus, intraosseous, synovial sarcoma

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Introduction

Synovial sarcoma (SS) is a malignant mesenchymal neoplasm with variable epithelial differentiation. Epidemiologic studies declared that SS is more common in young adults and can arise at almost any site. SS account for almost 5%–10% of all soft tissue sarcomas.[1] Differential diagnostic considerations for a musculoskeletal tumor largely depends on patient's age and site of incidence. Based on previous evidence, the sarcomas that typically arise within soft parts actually occur primarily within bone.[2]

SS is relatively common in the distal extremities, including hand, and is the most common soft-tissue sarcoma of the foot. Primary SS of bone is an extremely rare finding, and very few cases have been reported so far. As a result, regarding the rare prevalence of this tumor, biopsy and cytology studies must be performed in each case for a definite diagnosis.[3] SS is a misnomer as the source of this tumor is not from synovial tissue but its histologic manifestation is similar to synovial tissue.

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Here in this report, we presented a 44-year-old male patient referred with bone mass and later atypical humeral fracture that was finally diagnosed with primary SS of humerus.

Case Report

The presented patient was a 44-year-old male, married and had two children. He was a manual worker and had a low socio-economic level. He was referred to our medical center due to severe radicular pain in his right arm and also a palpable mass in the medial side of the arm. The patient declared to have mild pain in the neck and right shoulder from two years before his admission.His pain had proper response to common analgesics without any additional associated symptoms, so he did not call a physician. Within the past one year, he noticed gradually progressive swelling in the proximal of the humerus mostly in the medial and axillary side. Swelling was located between medial arm and the trunk.He had no severe pain and reported mild changes in the dimensions of the swelling

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site after physical exercises. No other symptoms including restricted range of motion in the arm or shoulder were reported. No positive points were detected in his past medical history, drug history, or familial history. Approval from the ethics committee was obtained in 10 November of 2021.

He attended a physician before admission in our hospital and a humerus mass was detected in radiography [Figure 1], ultrasonography (to evaluate the mass about soft tissue invasion and its echogenicity) and magnetic resonance imaging (MRI) studies, respectively. Reports of ultrasonography showed hetero-echo mass with visible margins in the anterior muscular side of the humerus with the size of 46 × 40 mm and 12.25 mm deep from the skin. The mass had inner vascularity with coarse calcification areas. No pathologies were observed in the brachial artery and basilic or cephalic veins. These findings initially were suggestive of chronic hematoma with calcifications but further imaging studies using MRI without contrast were performed that demonstrated organized hematoma or hemorrhagic sarcoma such as malignant fibrous histiocytoma (MFH). Tissue sampling was recommended by the radiologists.

The patient underwent tissue sampling using incisional biopsy and indicated chronic inflammation and fibrosis with dystrophic calcification. During our complementary work ups, our patient reported sudden reduction in the affected limb's forces and grip power who subsequently was admitted to our medical center. At the time of admission, he had normal vital signs. But, in right arm inspection and palpation, a palpable nonmobile mass measured 8cm in 12cm was recorded.

Single-cortex fracture of the humerus was observed using X-ray imaging. The results of MDCT-scan (multidetector computed tomography) with contrast showed right humeral malignant tumor infiltration associated with pleural base nodules suggestive of metastasis. Immediately, tumor incisional biopsy was done (both biopsies were taken by single surgeon). The single-cortex fracture of the humerus primarily was treated by conservative methods and using u-slab splint. Figure 2 shows post-operative X-ray of the patient (both specimens were reviewed by pathologists who were experts in bone malignancy).

Results of pathological studies demonstrated high-grade spindle cell sarcoma (mostly SS) with no lymphovascular invasion. Immunohistochemistry (IHC) staining was also recommended. The results of IHC showed positive results for EMA, BCL2, S-100, CD99, and also Ki-67 markers that were suggestive for high-grade SS.Laboratory data indicated anemia (hemoglobin (Hb) = 10.9 g/dl) with decreased MCV (mean corpuscular volume) and MCH (mean corpuscular hemoglobin) (77.8 FL and 27.3 pg, respectively). Based on consultations with oncologists, radiotherapy and chemotherapy were initiated for the patient due to suspicious involvements of axillary lymph nodes. During chemotherapy, he experienced a sudden severe pain in the right limb and complete humeral fracture was detected in the radiography; subsequently, proximal part of the humerus due to suspicion of bone involvements was removed. After humerus bone resection, the samples were analyzed by pathologists. The results of pathological examinations indicated SS (monophasic) score 3 with free margins and also no lymphovascular invasion. Pathologic stage classifications also showed T1NxMx. The patient underwent the third surgical procedure for complete bone removal and prosthesis implantation associated with bone allograft for proximal of humerus [Figures 3 and 4].

In the follow-up, the patient's condition and satisfaction improved. Physical examinations of the upper limb showed limited range of motion in abduction (40°–50°) and forward flexion but no other restrictions were observed. He is still under treatments with chemotherapy. Unfortunately after 17 months follow-up and under judicious chemotherapy regimens, pulmonary metastasis was detected and chemotherapy regimens were justified consequently.

Discussion

In the current report, a 44-year-old patient was presented with primary SS of humerus. The initial signs and symptoms were

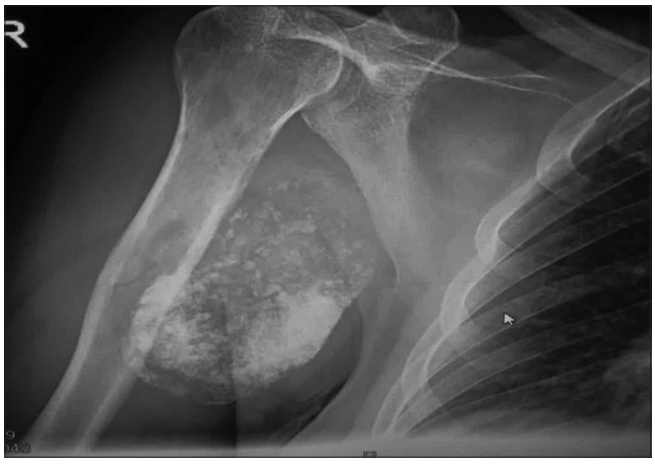

Figure 1: Initial X-ray of the arm by the time of admission

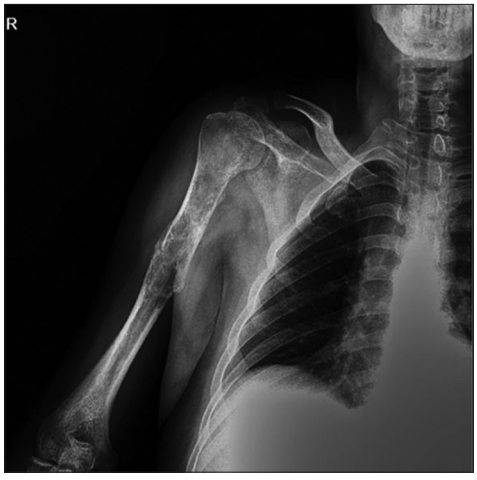

Figure 2: Post-operative X-ray of the right arm

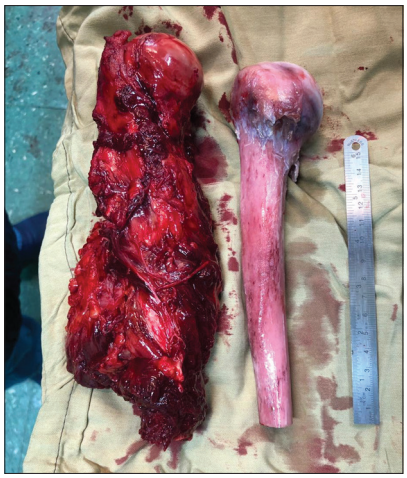

Figure 3: Removed humerus bone and prepared bone allograft

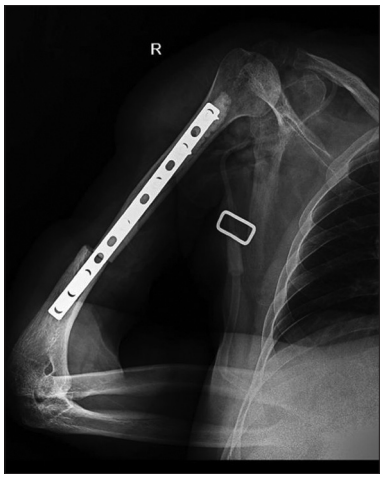

Figure 4: X-ray of the right arm after complete bone removal and insertion of bone allograft and implant

mostly related to shoulder and neck and within one year, a large mass was developed in the extremity that was the result of chronic inflammation and fibrosis with dystrophic calcification. Within months, the diagnosis of SS was made for the patient leading to complete bone resection and prosthesis implantation.

Previously reported cases of SS have been documented in areas such as lung and pleura, heart and pericardium, kidney, prostate, and gastrointestinal tract. This tumor mainly refers to soft-tissue sarcomas, and main pathological characteristics of it include spindle cell sarcoma and the absence of true synovial differentiation. Based on evidence, true primary intraosseous SS that originate within the bone and that is associated with significant intramedullary and cortical bone destruction remains a very rare entity. To date, a total of 13 cases have been reported in the English literature. Most of these cases have been treated with amputations and radical bone resections and some cases received adjuvant chemotherapy.

A recent study by McHugh and colleagues in 2020, reported two cases of primary SS of the bone in a 45-year-old female and a 36-year-old male patients. They explained that both cases had one to two year history of intermittent and gradually worsening pain in the involved limb with no history of trauma or associated diseases. The presented case of 45-year-old female had primary SS of humerus and underwent surgical tumor resection and no local recurrence or distant metastasis was reported.[4] Fujibuchi and colleagues also reported a case of intraosseous SS of the distal ulna that was confirmed by histopathological examination and molecular analysis. They reported that their case had a one-month history of right wrist pain but no other signs or symptoms were noticeable before. This case underwent wide tumor resection and had complete remission within two years of follow-up.[5] Our case also had long-term history of intermittent neck and shoulder pain that was later diagnosed with primary SS.

Horvai and colleagues also reported twocases of SS in Tibia diaphysis in 2019. As they explained, both patients underwent curettage and en bloc resection resulting in full remission.[6]

An important point of the current case was the rare location of primary SS. Within the previously reported cases, most of the tumors were diagnosed in the upper forearm[7,8] or lower limbs.[9,10] Only a recent case of humerus SS was presented by McHugh and others in 2020.[4] Our case is the second documented SS of proximal humerus that was treated with both neoadjuvant and adjuvant chemotherapy after radical tumor resection and prosthesis implantation. Another important aspect of the current case is the clinical manifestations. The patient had radicular neck and shoulder pain for almost two years that progressed and culminated in a medial arm palpable mass. We believe that imaging studies in patients with chronic pain and related symptoms could have beneficial effects. Lack of diagnosis of this tumor is also another issue. As reported, the primary pathological report of our case demonstrated benign tumor but within weeks, following the full humerus fracture, the malignant tumor was identified. Treatment was planned based on pathologic findings, which was successful from surgical standpoint and chemotherapy regimens as well.

Conclusion

Primary intraosseous SS is an extraordinarily rare entity, most commonly arising in the appendicular skeleton, especially the long bones of the lower extremities and tibia. But here we reported a rare case of SS in humerus of a 44-year-old male. Our case had long-term radicular pain in shoulder and neck and was diagnosed with high-grade spindle cell sarcoma and primary SS. Both neoadjuvant and adjuvant chemotherapies were performed for the case associated with surgical tumor removal and prosthesis implantation. We believe that imaging studies in patients with chronic pain and related symptoms could have beneficial effects.

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Conflicts of interest

There are no conflicts of interest.

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