Successful Management of a Rare Case of Humerus Non-Hodgkin's Lymphoma in Rapid Progress

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To the Editor: Primary non-Hodgkin’s lymphoma of bone (PNHLB) accounts for <5% of all primary bone tumors and <1% of all non-Hodgkin’s lymphoma.¹ Only very few cases of PNHLB with a rapid progress have been reported in literature, thus there is still short of imaging proof. Herein, we report a rapid progressing case of PNHLB with the presentation of the clinical, radiological, and histological features of the patient. A 64-year-old woman with severe pain in her right shoulder for 6 months was originally given treatment by another hospital in December 2015. Plain radiograph at her right shoulder disclosed mild osteoarthritis only without any obvious destruction of bone [Figure 1a]. Analgesic therapy was hence proceeded for the pain-relieving purpose, but to little effect. With a rapidly increasing mass reaching the size of 15.0 cm × 15.0 cm after 2 months, accompanied by a worsened pain at her right shoulder, the patient came for consultation in our institution in February 2016. She stated that she did not experience any fever, weight loss, sweating, or other B symptoms, nor was she ever been injured or having underlying diseases. Physical examination revealed a mass sized at 15.0 cm × 15.0 cm in her right shoulder, accompanied by a series of positive signs including limited range of motion, pain in all planes of movement and rising temperature at the local skin. Baseline assessment was performed for further detection including routine laboratory tests (electrolytes, liver and kidney function tests, and complete blood count), tumor markers, immunoglobulin M, immunoglobulin G, serum lactate dehydrogenase, and serum infectious index. Results confirmed that most indicators were within normal range, except that the serum neuron-specific enolase (NSE) was significantly elevated to 36.7 ng/ml (normal: <16 ng/ml). Plain radiographs of the right shoulder demonstrated distantly expansive bone destruction in the upper right humerus [Figure 1b]. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed expansive bone destruction extended within the cortical bone of the upper humerus, accompanied by swelling of the surrounding soft tissue. Bone scintigraphy indicated the skeletal abnormality of the right upper humerus, with high suspicion of malignant tumor [Figure 1c]. Tissue biopsy of the lesion was performed, and the postoperative pathology confirmed a diffuse large B-cell lymphoma [Figure 1d]. Histology revealed a diffuse proliferation of large lymphoid cells with irregular round or pleomorphic nuclei and immunohistochemistry revealed neoplastic cells being B-cell lymphoma 6, CD20, melanoma-associated antigen 1, paired box 5, and NSE positive with 85% Ki-67-positive nuclei. Consequently, a primary bone lymphoma, Ann Arbor stage II, International Prognostic Index score 1 were diagnosed through history taking, laboratory values, pathological studies, and fluoro-deoxyglucose-positron emission tomography/CT (FDG-PET/CT). An effective control with marked regression of the tumor was observed on radiography and FDG-PET/CT after eight cycles of chemotherapy with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP), followed by radiation therapy (50Gy/20f, 2.5Gy/f, 5f/w). FDG-PET/CT demonstrated that the range of locally hypermetabolic lesions of the right humerus and the metabolic activity were both reduced significantly and the enlarged lymph nodes were not seen around the lesion, compared with multiple hypermetabolic lymph nodes revealed by FDG-PET/CT in March 2016 [Figure 1e-1h]. This is an extremely rare presentation of diffuse large B-cell lymphoma. The 2-year follow-up visit found that the patient was doing well, with improved motion, palliative shoulder pain, and no evidence of local progress or distant metastasis. The joint function score assessed on the patient’s shoulder indicated an improvement to 70 points compared with the initial status of 40 points, using Constant-Murley Score system. The physical examination results almost returned to normal.

Patients with PNHLB commonly present with localized bone pain, soft-tissue swelling, or a pathological fracture in a mild progress.²³ This is a rare case of humerus lymphoma occurring with worsened pain and limited range of motion in surprisingly rapid progress. In literature, PNHLB has to be differentiated from the patients with PNHLB who presented with locoregional pain, and the patients who presented with locoregional pain and regional pain. This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms. © 2018 Chinese Medical Journal | Produced by Wolters Kluwer - Medknow

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from neoplasms involving soft tissue, mostly the sarcoma, osteomyelitis, and other malignant tumors.\cite{3,4} In imaging studies, although plain radiographic assessment might reflect the radiologic characteristics of the lesion, CT and MRI are more specific to further assess the extent of the mass within the bone and surrounding soft tissue more accurately.\cite{4‑6} FDG-PET/CT has a complementary role for the diagnosis, especially to stage and estimate the prognosis of PNHLB.\cite{1,4} Pathological results remain to be the “gold standard” to diagnose and definitively confirm the subtype of PNHLB. To date, CHOP-based chemotherapy regimens together with rituximab (R-CHOP) are commonly given to PNHLB patients, but the role of consolidated radiation therapy is not well defined.\cite{2} In conclusion, we present in this report a case with an extremely unusual occurrence of PNHLB of the upper humerus with amazingly rapid progress and that was successfully controlled by chemotherapy and radiotherapy. Such case has not been well defined in literature, and it highlights the significance of early diagnosis and proper treatment for PNHLB.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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