Post functional reconstruction of modified pollicization technique and posterior interosseous artery flap in primary bone NHL of the hand: A rare case report

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ABSTRACT

Introduction: Primary Non Hodgkin Lymphoma (NHL) of the bone is rare malignancy accounting less than 1–2% of adult NHL and less than 7–10% of primary bone tumors. Most primary bone lymphomas are NHL with diffuse large B-cell subtype. It is frequently misdiagnosed due to its rarity, uncommon presentation and indistinctive radiographic features. Case Report: A 40-year-old man presented with the left 2nd metacarpal tumor complained of pain, swelling with difficulty in both active and passive movements. The diagnosis of bone NHL established from clinical finding, radiographic examination and biopsy. Immunohistochemical result showed positivity of LCA and CD20, with high proliferation index of Ki-67, suggesting B-cell lineage of lymphoma. Functional hand reconstruction with modified pollicization technique and posterior interosseous artery flap were performed. The follow-up for the possibility of a local tumour recurrence and metastasis was evaluated after 8 courses of RCHOP regimen chemotherapy. Conclusion: Primary NHL of the bone especially in hand is very rare and challenging condition for orthopaedic surgeons. The triple diagnosis of clinical, radiographic and biopsy examinations were very useful to establish solid diagnosis of bone NHL. Therefore, prognosis, nonetheless, is much better than that of other malignant bone tumours. Awareness and knowledge in decision making of hand reconstruction and post-operative chemotherapy planning are needed to achieve satisfactory outcome.

Keywords: Hand reconstruction, NHL of the bone, Pollicization technique, Primary bone lymphoma

INSTRUCTION

Lymphomas commonly occur in the lymph nodes, and extranodal sites are a less frequent origin of lymphomas. Primary bone lymphomas (PBLs) are rare, even though secondary involvement of the bone marrow is a common event in systemic lymphomas. Primary bone lymphoma was first described by Oberling in 1928 [1]. Parker and...
Jackson reported a PBL series in 1939 [2] under the designation reticulum cell sarcoma of bone, establishing PBL as a distinct entity. The lymphoid and B-cell origin was subsequently proved by immunohistochemistry [3, 4]. Primary bone lymphoma is defined as a lymphoma that is confined to the bone or bone marrow without evidence of concurrent systemic involvement. According to the 2002 World Health Organization classification of tumors of soft tissue and bone, the criteria for a diagnosis of PBL are (1) a single skeletal tumor without regional lymph node involvement, and (2) multiple bone lesions without visceral or lymph node involvement [5].

Beal et al [6] reported a series of primary bone diffuse large B-cell lymphomas that included 82 patients. In that series, the frequency of different bone involvement was femur (27%), pelvis (15%), tibia/fibula (13%), polyostotic (13%), humerus (12%), spine (9%), other (5%), mandible (2%), radius/ulna (1%), scapula (1%), and skull (1%). Rarely, small bones of the hands and feet are involved in primary bone diffuse large B-cell lymphomas.

The clinical presentation depends upon the rate of the tumour cell proliferation and on the initial localization. The patients generally present with localized bone pain and, less frequently, with a soft-tissue swelling or a palpable mass. On conventional radiology, PLB has a widely variable imaging manifestation which consists of either a ‘lytic destructive pattern’ or a ‘blastic sclerotic pattern’ [7].

CASE REPORT

A 40-year-old active engineer Indonesian man complained of pain and swelling around his left index finger with difficulty in both, active and passive movements since five months before admitted to the hospital. The pain worsened over the past 1 month with VAS (Visual Analogue Scale) within 6–7 without any history of trauma or history of fever, sweats, weight loss or fatigue. On examination of his left hand we found a diffuse swelling and palpable mass in the left 2nd metacarpal region (Figure 1). From general physical examination, clinically the patient was well, no neck or axillary lymph nodes were palpable. There was no palpable hepatosplenomegaly or lymphadenopathy. The chest radiograph was normal. Both the WBC and the CRP levels were within the normal range. The only medications which were taken at the time of the presentation were non-steroid anti-inflammatory drugs (NSAIDS). The X-ray result showed an ill-defined osteolytic lesion, with cortical breakage and soft tissue involvement in the left 2nd metacarpal (Figure 2). Screening with skeletal survey, chest X-ray, CT scan of the whole abdomen and the chest were performed and showed no lymph node involvement. Magnetic resonance imaging (MRI) of the left hand showed that the lesion was hypointense on T1-weighted images and hyperintense on T2-weighted images (Figure 3).

A malignant lesion was suspected and an open bone biopsy was performed, large cell type of Non Hodgkin Lymphoma was revealed and immunohistochemical result showed positivity of LCA and CD20, with high proliferation index of Ki-67, suggesting B-cell lineage of lymphoma (Figure 4).

The diagnosis of diffuse large B cell lymphoma of the bone was confirmed. The patient underwent functional hand reconstruction with modified pollicization technique and posterior interosseous artery flap to close the wound defect. Adjuvant chemotherapy of eight courses of the RCHOP regiment were completed. The therapeutic effects were assessed both clinically and radiographically after four courses and at the end of the eight courses of chemotherapy. The patient showed a clinically complete remission, and neither relapse nor metastasis has been recognized till his last follow-up at eight months. A continuous follow-up for the possibility of a local tumor recurrence and dissemination of the disease should be
No evidence of distal soft tissue or distal nodal involvement.

In our case, the triple diagnosis of clinical, radiographic and biopsy examinations were very useful to establish solid diagnosis of bone NHL [10]. The other investigations which are done to establish the diagnosis include a skeletal survey, a bone scan and a bone marrow biopsy. CT scan of the whole abdomen and the chest to assess the lymph node involvement and serum LDH estimation are done as a part of the staging procedure [9]. In the younger patients, the differential diagnosis of PLB mainly includes osteosarcoma, Ewing’s sarcoma, and osteomyelitis. In the older patients, bone metastasis of the solid tumours should be considered.

In treating patients with primary bone NHL of the hand, the reconstructive hand surgeon must be creative and must have a wide variety of techniques at his disposal if optimal hand function is to be restored. Although formal pollicization is certainly an option, in our case, the index finger was used in several ways to reconstitute a subtotally resected thumb. Patient factors including age and occupation are important. Co-morbid disease, functional status, patient expectations, and tolerance for donor site morbidity must be considered. The size, location, and involvement of neurovascular structures must be assessed early in the process of establishing the reconstructive plan. Improved preoperative imaging via magnetic resonance imaging has greatly enhanced our ability to make these assessments. Even tough the primary bone NHL described as bone malignancy with the patient’s clinical status of VAS (Visual Analog Scale) within 6-7, we considered the patient factors such as the age, functional status, occupation, and patient expectations to choose the surgery management of the patient with functional reconstruction using modified pollicization techniques beside to choose amputation to the patient’s hand. The procedure was initiated with preservation of native thumb CMC joint and distal half of proximal phalanx. The proximal interphalangeal joint of the middle finger was then transferred in order to reconstruct the metacarpophalangeal joint. The wound defect in dorsal and palmar side was covered with posterior interosseous artery flap (Figure 5).

Integrating the reconstructive plan and chemotherapy into overall treatment strategy is essential to optimize functional outcome. The chemotherapy regimen usually consists of cyclophosphamide (C), doxorubicin (H), vincristine (O), and prednisone (P) with or without rituximab (R) [6]. In a study by Ramadan et al. [6]. Three year progression free survival in patients who received rituximab with cyclophosphamide, doxorubicin, vincristine, and prednisone was 88% compared with 52% for those who received only cyclophosphamide, doxorubicin, vincristine, and prednisone. In our case, the patient received eight courses regiment of adjuvant chemotherapy of RCHOP, 1 cycle of each month.

The functional outcome and radiographic evaluation post-operative chemotherapy showed significant clinical

**DISCUSSION**

Primary bone NHL commonly affects the middle-aged to elderly population, with a median age of 48 years and it shows a male preponderance with a male to female ratio of 3:2. Primary bone NHL of the hand in phalanx and metacarpal area accounts for only 0.2% [8]. The most common presentation is bone pain, and less-frequent presentations include a palpable mass and bone fracture. In order to be diagnosed as PBL the following criteria (known as Coley’s criteria) should be met [9].

- A primary focus in a single bone
- Positive histological diagnosis

![Figure 3: Pre-operative MRI of the left hand showed that the lesion was hypointense on T1-weighted images and hyperintense on T2-weighted images.](image)

![Figure 4: Biopsy result showed positivity of LCA and CD20, with high proliferation index of Ki-67, suggesting B-cell lineage of lymphoma.](image)
remission (Figure 6). We measured the functional outcome using the Disabilities of the Arm, Shoulder, and Hand (DASH) questionnaire and the Enneking score for the hand. The DASH questionnaire is designed to measure total functional disability of the upper limb with a score of 0 representing the least and 100 the most disability, and the Enneking score is a disease-specific but physician based measure. Local recurrence and metastasis were not detected till his end of 8 courses of chemotherapy regiment or followup at 8 months. Both DASH questionnaire measurement and modified Enneking functional scoring system exhibited favorable results, 12.1 for DASH score and 22 for Enneking functional scoring system.

CONCLUSION

Primary NHL of the bone especially in hand is very rare and challenging condition for orthopedic surgeons. The triple diagnosis of clinical, radiographic and biopsy examinations were very useful to establish solid diagnosis of bone NHL. Therefore, prognosis, nonetheless, is much better than that of other malignant bone tumors. Awareness and knowledge in decision making of hand reconstruction and post-operative chemotherapy planning are needed to achieve satisfactory outcome.

REFERENCES


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Author Contributions
Rahadyan Magetsari – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
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EryKus Dwianingsih – Acquisition of data, Analysis and interpretation of data, Revising it critically for important
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Written informed consent was obtained from the patient for publication of this case report.

**Conflict of Interest**
Authors declare no conflict of interest.

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