

## Hodgkin's Lymphoma presenting as a recurrence of primary disease at osseous site: Review of Literature

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### Abstract

Hodgkin's lymphoma at an osseous site is not a common presentation. Here we are reporting a case of recurrence of Hodgkin's lymphoma at osseous site with review of literature.

**Keywords:** Hodgkin's lymphoma, osseous.

### 1. Introduction

Hodgkin's Lymphoma (HL) is a systemic disease with most common presentation in the lymph node [12,3]. Ten percent of Hodgkin's Lymphoma originates from extra nodal regions, but primary extra nodal presentation of HL occurs in less than 0.25% of patients [4]. Bone involvement occurs in 10-20% cases [5]. There are very few case reports of primary solitary osseous HL [5-7].

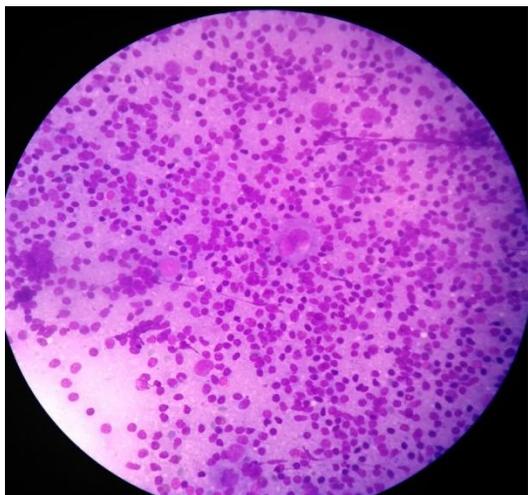
The clinical, radiological and histological features of osseous Hodgkin's Lymphoma can mimic other medical conditions, thereby making the diagnosis difficult, often leading to delay in treatment [5]. We present a case of bone involvement in HL presenting as recurrence of disease at osseous site.

### 2. Case summary

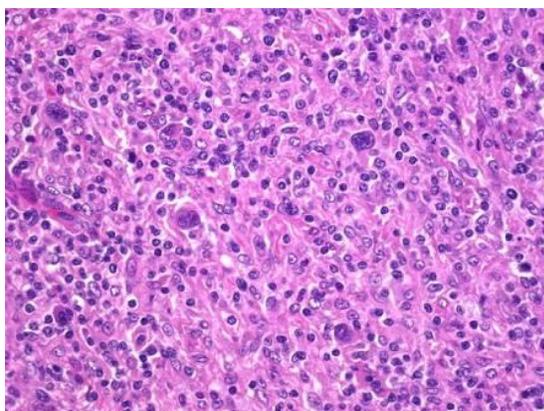
A 35 years old male, known case of HL who underwent complete chemotherapy for bilateral cervical lymphadenopathy; now presenting in Orthopedics OPD of SVBP Hospital, LLRM Medical College, Meerut with swelling over right shoulder noticed 2 months back. Right shoulder radiography was performed which showed osteolytic lesion.

Patient was sent to Department of Pathology for FNAC. On examination, the swelling was 3x3cm, diffuse, firm to hard, fixed and tender. At the same time, a right cervical lymph node of size 0.5x0.5cm was also seen that was firm, mobile and non-tender. Fine needle aspiration cytology from both the shoulder swelling and cervical lymph node was performed. Smears from right shoulder swelling showed Reed-Sternberg (RS) cells over a reactive background and a provisional diagnosis of Hodgkin's Lymphoma was made. (Figure 1) Smears from cervical lymph node showed necrosis only.

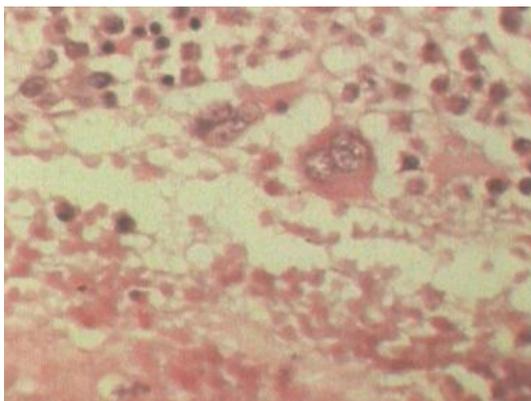
Biopsy of lesion from shoulder showed cells bearing characteristic morphology of RS cells dispersed against a reactive background (Figure 2). Bone marrow aspiration and biopsy was performed from posterior superior iliac spine for staging of the disease. Aspiration was negative for lymphoma cells while biopsy showed presence of RS cells, hence Stage IV disease (Figure 3). Therefore, a diagnosis of HL presenting as a recurrence of primary disease at osseous site was made and treatment chemotherapy was started with ABVD. However, the patient died one month after the treatment.



**Figure 1: FNAC right shoulder**



**Figure 2: biopsy from right shoulder**



**Figure 3: Bone marrow biopsy**

### 3. Discussion

Langley *et al* [5] stated that Hodgkin's Lymphoma frequently presents as a progressive, painless enlargement of the peripheral lymph nodes, particularly around the cervical region.

Study done by Gandhi *et al* [6] and Kammerer *et al* [7] defined Classic HL as a well-established, proliferative neoplasm of the lymph node that is composed of mononuclear Hodgkin cells and multinucleated RS cells in variable proportions, along with neutrophils, eosinophils,

histiocytes, fibroblasts, collagen fibers, non-neoplastic lymphocytes and plasma cells.

Zucca [8] and Guermazi *et al* [9] concluded that extra-nodal forms of HL are rare, accounting for less than 1% of all HL cases. Bhagwati *et al* [3] and Newcomer *et al* [10] stated that HL is a systemic disease, with 10-20% patients demonstrating bone involvement throughout the disease progression. However, patients presenting with primary HL of bone are unusual as seen in studies done by Langley *et al* [5] and Oshikawa *et al* [11]. In contrast, study done by Felt *et al* [15] showed that bone involvement is a relatively common finding in HL and is not an independent adverse prognostic factor.

Hodgkin's disease may involve any organ, but the most common extra nodal locations of HL are digestive tract, lungs, central nervous system and bone tissue as seen in study done by Guermazi *et al* [9]. Chan *et al* [12] found that the incidence of skeletal HL varies from 9-14% during the course of the disease with up to 30-50% at post-mortem. Singh *et al* [16] reported osseous lesions at the time of relapse in pediatric HL with vertebra and pelvis as most frequently involved sites, commonly with osteolytic picture.

Chan *et al* [12] in their study found that at the time of diagnosis, osseous involvement is uncommon and even in late stages, only 9-35% of cases have any bony involvement. It is therefore, extremely rare for patients to present with primary HL of bone. If there is no associated extra-osseous involvement, the condition is referred to as primary osseous HL (POHL). It is termed primary multifocal osseous HL; if more than one osseous site is involved. Sato *et al* [13] reported a case of primary multifocal osseous lymphoma in a six year old girl presenting with multifocal osteolytic lesions without systemic symptoms or identifiable non-osseous primary tumor.

Langley *et al* [5] presented skeletal involvement in three different ways: 1. Primary osseous HL (either solitary or multifocal), 2. Simultaneously in osseous and non-osseous sites, or 3. Recurrence of disease at osseous sites. Our case belongs to third category i.e., recurrence of disease at osseous site. There are very few case reports of primary solitary osseous HL by Langley *et al* [5], Gandhi *et al* [6] and Kammerer *et al* [7]. In a study done by Langley *et al* [5], mean age of the reported patients with solitary osseous HL has been about 35 years and most commonly involved bones were the humerus, vertebrae, iliac crest and femur. In our case also, patient was about 35 years and right humerus was involved.

Clinically, the primary form of extra-nodal HL presents as intermittent pain that lasts for months. Local swelling and palpable mass occur. Common symptoms of lymphoma such as weight loss and fever may be helpful for

diagnosis. Ostrowski *et al* [14] concluded from their study that unlike skeletal expansion of Hodgkin's disease (which is painless), primary bone Hodgkin's disease is painful. Griguolo *et al* [15] also reported a case of 30 year old man with bone pain and multifocal HL bone involvement initially misdiagnosed as chronic recurrent osteomyelitis.

Location of solitary primary HL in bone is non-specific. Ostrowski *et al* [14] found primary bone HL lesions in spine, pelvis, ribs, mandible, femur, tibia and scapula. Primary HL lesions in the bone cannot be correlated with specific parts of bone, bone types, age or gender. This makes diagnosis difficult. Therefore, repeated investigations and clinical suspicion of unusual presentations may be helpful in order to establish a correct diagnosis.

According to Koseoglu *et al* [16], bone involvement in the later stages of Hodgkin's disease is an expected phenomenon but it is very rare in early stages of disease. Radiologically, bony lesions of Hodgkin's disease may be lytic, sclerotic or mixed. One study conducted by Ostrowski *et al* [14] showed that 75% were lytic, 13.6% sclerotic and 11.4% mixed. In our study, bone lesion was lytic.

Biswas *et al* [17] reported a case of osseous HL which highlights the importance of clinical suspicion of unusual presentation of lymphohematopoietic tumors of the bone especially in developing countries, where chronic granulomatous disease is preponderant.

In all reported cases, the correct diagnosis was only reached after extensive and repeated investigations and review of the histology. Our case also highlights the difficulty in diagnosing this rare form of Hodgkin's disease.

#### 4. Conclusion

This case indicates that osseous presentation of Hodgkin's disease presents difficulty in reaching a definitive diagnosis, especially when patient fails to respond to initial therapy. Imaging may not be sufficient to enable a clear diagnosis; in order to improve the success rate of diagnosis, extensive pathological examination is necessary.

#### Conflict of interest

The authors have no conflict of interest.

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