# A diagnostic surprise of Hodgkin's Lymphoma in a 29-year-old long distance runner

**Kaleem Murtaza Buttar** (1)
**Numan Ahmad** (2)

(1) Consultant Family Physician, Healthworks Healthcare Complex, Victoria, Australia.
(2) Fellow Royal Australian College of general practitioners, Melbourne, Australia

**Corresponding author**
Dr. Kaleem Murtaza Buttar, MBBS FRACGP
Consultant Family Physician
Healthworks Healthcare Complex
Kangaroo Flat, Victoria,
Australia
Mobile: +61 413351546
**Email:** drkaleembuttar@gmail.com

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## Abstract

Painless lymphadenopathy enlarging over months is the most frequent presentation in Hodgkin's lymphoma. Bony pain and lesions at presentation is usually not seen in Hodgkin lymphoma. However, in this case study, an uncommon presentation of classical Hodgkin's Lymphoma, involving bone, in a 29-year-old long distance female runner was reported. Patient initially presented with bony pain without classic type B symptoms and lymphadenopathy. A final diagnosis of classical Hodgkin's lymphoma stage IV was confirmed and PET scan evaluation was also done. Patient started on Escalated BEACOPP regimen and four cycles of chemotherapy were completed in 12 weeks and showed good response post treatment. Albeit, its rare, primary osseous Hodgkin's Lymphoma should be considered in the diagnosis of bony pain. The absence of classic type B symptoms and any local lymphadenopathy in this case did make the diagnosis challenging.

Keywords: Hodgkin's lymphoma, bony pain, escalated BEACOPP

## Introduction

Multinucleated Reed-Sternberg cells with inflammatory potency are the characteristic features of Hodgkin lymphoma. Classic Hodgkin lymphoma (CHL) is one of the major types of HL. Hodgkin's Lymphoma is commonly diagnosed in the 20-34 years age group, which accounts for 31% of new diagnoses but it occurs through a wide spectrum of ages ranging from adolescents to the elderly(1). The common mode of presentation of Hodgkin's lymphoma is painless lymphadenopathy enlarging over months. Fevers, night sweats, chill, or sudden weight loss of around 10% is commonly seen in the advanced stage of the disease and that determines the prognosis of the disease. Erythrocyte sedimentation rate (ESR) is usually elevated at the time of diagnosis and acts as a reliable marker to assess the disease severity. Similarly, leukocytosis/neutrophilia and anaemia are observed during advanced stages of Hodgkin lymphoma and have been associated with poor prognosis. Gastro-intestinal tract and pulmonary system are the common extra-nodal sites of involvement in Hodgkin lymphoma. Bony lesions at presentation are usually not seen in Hodgkin lymphoma, although skeletal involvement is seen in the advanced stage of the disease. Primary bone Hodgkin lymphoma is usually rare (2). Hodgkin lymphoma is highly curable and the 5-year relative survival rate is 96.4% in patients between the age group 0-19 years and 89.8% for patients 20-64 years (3). The present case report was an uncommon presentation of classical Hodgkin's Lymphoma in a 29-year-old diabetic female.

## Case Report

In May 2021, a 29-year-old woman developed left sided musculo-skeletal pain the chest region. Initial X-ray investigation was normal. She has type-2 diabetes which is quite well controlled. Subsequently, she developed right sided hip pain which was attributed to her extensive training for her half-marathon. Patient underwent physiotherapy for the symptoms, but the pain increased in intensity, with back pain radiating to the right leg. Initial investigations including X-ray and MRI didn't show any concerning features and were essentially normal. Her left sided rib pain worsened and a repeat X-ray showed lytic lesions in the left lower ribs. She was referred to oncology. A couple of weeks later, in October she presented to emergency with worsening pain and cauda equina symptoms. Repeat MRI this time showed a large expansile mass arising from the sacrum and invading the right posterior ilium, adjacent muscles and Rt S1, S2 and S3 nerve roots of the sacral plexus. Biopsy of the lesion, histological examination and immunohistochemical analysis made the diagnosis of classical Hodgkin's lymphoma (mixed cellular pattern) by finding the diagnostic Reed-Sternberg cells.
Standard blood test showed that the WBC (white blood cell) count was 21.23 × 109 L−1 with the neutrophil rate of 84.3% and lymphocyte rate of 11.2%; Hb was 109 g/L and platelet count was 540 × 109 L−1). The biochemical results showed ESR and CRP were also increased.

Based on the complete staging workup the final diagnosis for this 29-year-old patient on November 2021 was confirmed as classical Hodgkin's lymphoma stage IVA, predominately bony disease with very low-grade uptake in supraclavicular lymph nodes. She had a number of high-risk factors including stage IV disease, high white cell count and low albumin which does increase her risk in general. Escalated BEACOPP regimen was started for the patient. Four cycles of chemotherapy were completed in 12 weeks and she had complete response to treatment.

## Discussion

Osseous HL presenting with the bony pain and lesion, is an unusual presentation of this disease. A painless progressive enlargement of the lymph node is the commonly observed clinical presentation of Hodgkin's lymphoma. Primary skeletal involvement is very rare. The occurrence of Hodgkin's lymphoma in skeletal muscle is observed in the middle age to older population and frequently long bones are usually affected. Bone pain is the most common presentation, followed by bone mass and pathological fracture. Neurological abnormalities as a result of nerve compression are also observed in these patients (4).

Radiological investigation for skeletal involvement displays either lytic or sclerotic lesions, however lytic lesions are more frequently encountered compared to sclerotic lesions. The common radiographic differential diagnosis includes osteomyelitis, primary sarcomas of the bone, non- Hodgkin's lymphoma, metastatic lesions, and leukemia.

Because, both radiological and clinical features are non-specific for the primary osseous Hodgkin's lymphoma, histopathological examination remains the mainstay of diagnosis. Balancing the disease control with the occurrence of early and delayed treatment related side-effects is required, when choosing the preferred first-line treatment. Patients with advanced stage of Hodgkin's lymphoma are most likely to relapse and thus need intensive treatment. Combination chemotherapy is the mainstay of treatment for those patients.

An intensified front-line regimen has been developed by the German Hodgkin Lymphoma Study Group (GHSG) consisting of bleomycin, etoposide, doxorubicin, vincristine, cyclophosphamide, prednisone and procarbazine for the treatment of Hodgkin's lymphoma.

The escalated BEACOPP treatment protocol has higher-than-standard doses of doxorubicin, etoposide and cyclophosphamide. Escalated BEACOPP regimen elicits effective tumor control, with an overall increase in 11% at 10 years as compared to COPP-ABVD. GHSG advocates BEACOPP regimen as the standard treatment strategy for patients with high risk factors and advanced-stage Hodgkin's lymphoma (5, 6). Treatment of Hodgkin Lymphoma with eBEACOPP regimen displayed frequent and long-lasting remissions. However, the estimated

increased risk of secondary malignancies is not reported (7). Previous reports comparing eBEACOPP with other regimens showed that eBEACOPP treatment showed good overall survival rate and less relapses with a comparable rate of secondary neoplasia. A meta-analysis study encompassing 10,000 patients from 14 studies showed that eBEACOPP treatment regimen displayed marked survival benefit of 7% in advanced stage of Hodgkin's lymphoma as compared to the ABVD regimen (8). The outcome of patients with osseous Hodgkin's lymphoma has been reported similar to that of patients with lymph node involvement, (9, 10) and this was true for our patient as well.

## Conclusion

Even though it's uncommon, the primary osseous Hodgkin's lymphoma should be considered in the differential diagnosis of ongoing bony pain. Lack of classic type B symptoms and lymphadenopathy can make the diagnosis challenging. Remarkable response was noted in the patient with the current chemotherapeutic regimens.

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