



Case Report

Primary Multifocal Osseous Hodgkin Lymphoma: A Case Report

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ABSTRACT

Hodgkin lymphoma (HL) is primarily involves Lymphatic system at one or more sites. Hodgkin Lymphoma restricted to bone only; Also known as Primary osseous Hodgkin lymphoma is exceedingly rare with a reported incidence of 0.25% in HL. We report a case of primary multifocal osseous Hodgkin Lymphoma of spine in a middle aged man, who was treated with multiagent chemotherapy and is disease free more than a year after Diagnosis.

Key Words: Primary osseous Hodgkin Lymphoma, bone only lymphoma, Tumor.

INTRODUCTION

Hodgkin Lymphoma is a systemic disease commonly presenting as lymphadenopathy in cervical and mediastinal region. Lymphomas represent only 8% of primary malignant bone tumors.

⁽¹⁾ At presentation osseous involvement in Hodgkin lymphoma is uncommon and in late stages 10-20% involvement may be seen.^(2,3) Hence it is extremely rare for patients to present with primary Hodgkin lymphoma of the bone. If more than one osseous site is involved with no extra osseous site at presentation it is termed as Primary osseous Hodgkin lymphoma. It is essential to rule out other medical conditions especially osteomyelitis before diagnosis of primary osseous Hodgkin lymphoma may be made. We are presenting a rare case of primary multifocal osseous Hodgkin

lymphoma involving the vertebral column, who is treated with ABVD (Adriamycin, bleomycin, vincristine and dacarbazine) for 6 cycles along with radiation therapy, with complete response to treatment.

CASE REPORT

A 54 year old male presented with low grade intermittent fever, backache and weight loss. On clinical examination he had no peripheral lymphadenopathy, systemic examination was normal with no neurological deficits. He was evaluated and initial investigations showed anemia (Hb: 9g %) and elevated alkaline phosphatase (634U/L). Patient underwent Magnetic resonance imaging of spine which showed (fig: 1) multifocal vertebral involvement extending from T-3 to lumbosacral vertebrae, a clinical suspicion of

tuberculosis was made. He was subjected to Fine needle aspiration from T3 vertebra which showed Reed Sternberg cells (Fig.2), and further confirmed by immunohistochemistry which was positive for CD15, CD30 and negative for T cell antigens. Hence patient underwent staging work up with CT scan of thorax and abdomen, bone marrow aspiration and biopsy which did not reveal evidence disease. Bone scintigraphy (image not available) was done which corresponded with involvement as seen on MRI spine. Patient was staged as stage 4 B and was started on ABVD for 6 cycles (Adriamycin, bleomycin, vincristine and dacarbazine). He was reassessed after 3 cycles with MRI of spine which showed good response to therapy. Upon completion of 6 cycles patient is asymptomatic and post treatment MRI spine has normalized. Patient did not develop any lymphatic or extra osseous disease during course of treatment. He is on follow up since last one year with no evidence of disease.

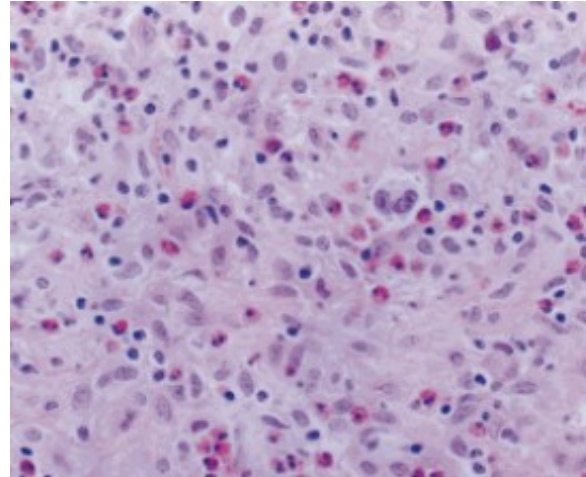


Fig. 2: Shows Reed sternberg cell on biopsy specimen from Dorsal (D3) spine on H&E stain.

DISCUSSION

Hodgkin's disease usually presents with painless peripheral lymphadenopathy most commonly involving cervical lymph nodes. It has bimodal age incidence curve with the first peak occurring in second decade and third decade and second peak in sixth decade. The historical incidence of skeletal involvement in Hodgkin lymphoma varies from 10-20% mostly seen in advanced stages.⁽²⁾ Primary bone involvement is seen in 0.25% of cases. Skeletal involvement may present in four different ways: POHL (either solitary or multifocal); simultaneously in osseous and non-osseous sites; or recurrence of disease at osseous sites. In order to make a diagnosis of primary osseous Hodgkin lymphoma there should not be nodal or extra osseous disease at presentation. In osseous Hodgkin lymphoma axial and proximal appendicular skeleton is most commonly involved with spine, proximal humerus and femur being most common. In one of the study radiologic appearance of lesion varies from osteolytic to osteosclerotic to mixed forms. One study showed that 75% were lytic, 13.6% sclerotic and 11.4% mixed.⁽³⁾ The radiological features in general are not characteristic, differential diagnoses include



Figure 1: MRI spine on T1 showing hypointense lesions in Vertebrae from T3-L4.

primary sarcomas of the bone, non-Hodgkin lymphomas, leukemia, metastasis, and, especially, osteomyelitis. Although there have been many advances in the diagnosis of Hodgkin lymphoma, histopathological diagnosis still depends on the demonstration of Reed Sternberg cell which accounts for about 1% of cell population in most biopsy specimens the rest being lymphocytes, granulocytes, histiocytes, plasma cells and fibroblasts. Availability of immuno-histochemistry has made the diagnosis straightforward. The most commonly observed subtype of Hodgkin lymphoma in primary osseous setting is nodular sclerosis subtype.⁽⁴⁾ On review of literature the most common histopathological differential diagnosis is acute osteomyelitis, this diagnosis was suggested by large areas of necrosis with prominent polymorph nuclear cell infiltration, alternating with areas of fibrosis and chronic inflammation.

The term “Primary osseous Hodgkin lymphoma” should be applied cautiously as review of literature suggests simultaneous involvement of lymph node areas on imaging. Therefore imaging with MRI, CT scan or Positron emission tomography is essential before ruling out other sites of involvement.⁽⁵⁾ Hence without complete imaging the diagnosis of Primary osseous Hodgkin lymphoma remains questionable.

On review of literature there are more than thirty reported cases of osseous Hodgkin lymphoma.⁽⁶⁾ Bony involvement in Hodgkin lymphoma both osseous and non osseous was first described in 1934 by craver et al.⁽⁷⁾ Most of the cases reported earlier than 1950 s did not have diagnostic CT or MRI to demonstrate simultaneous nodal involvement. Ostrowski *et al*⁽⁴⁾ reported two cases of POHL, who were among 25 patients diagnosed with osseous Hodgkin's disease from a group of over 500 patients known to have had Hodgkin's lymphoma, at the Mayo clinic between 1927

and 1996. Five of the twenty five had POHL; three had disease at a single bony site and two had multifocal bony disease. Gross *et al.*⁽⁸⁾ presented two cases in adolescents (12 years and 17 years) who presented in pattern similar to our case. Both presented with back pain and raised inflammatory markers. Investigation revealed widespread osseous involvement. In the case of the 17 year old, treatment was delayed by a misdiagnosis of eosinophilic granuloma. Bone involvement in Hodgkin lymphoma would be staged as stage IV disease according to Ann Arbor staging (Table 1) and traditionally would have poor prognosis as bone involvement is seen in advanced disease. POHL has good prognosis based on review of literature and has responded well to chemotherapy or radiation therapy alone. Primary osseous Hodgkin Lymphoma is a very rare disease and diagnosis can be made only after meticulously ruling out nodal and extra nodal sites. In developing countries tuberculous osteomyelitis still remains the commonest cause of vertebral involvement, POHL though rare is one of the differential diagnosis.

Table 1: Staging of Lymphoma: Ann Arbor classification.

Stage I	Disease in a single lymph node region
Stage II	Disease in two or more regions on the same side of diaphragm
Stage III	Disease in Lymph node regions on both side of Diaphragm
Stage IV	Diffuse or disseminated involvement of one or more extralymphatic organs or tissues with or without associated lymph node enlargement.

CONCLUSION

Primary osseous Hodgkin lymphoma is very rare and most common differential diagnosis being tuberculous osteomyelitis. However it must be considered in the differential diagnoses.

REFERENCES

1. Unni KK. Dahlin's bone tumors: general aspects and data on 11, 087 cases. 5th edition. Philadelphia: Lippincott-Raven, 1996.
2. Horan FT. Bone involvement in Hodgkin's disease: a survey of 201 cases. *Br J Surg* 1969; 56:277-81.
3. Granger W, Whitaker R. Hodgkin's disease in bone, with special reference to periosteal reaction. *Br J Radiol* 1967; 40:939-48.
4. Mary L. Ostrowski, Carrie Y. Inwards. Osseous Hodgkin Disease: *Cancer* 1999; 85:1166-78.
5. McElwain Tj, Selby P: Hodgkin's disease 1st edition. Oxford: Blackwell S.C. Publications; 1987:105-106.141-146.
6. Clare R Langley, Simon JW Garrett. Primary multifocal osseous Hodgkin lymphoma .*world journal of surgical oncology* 2008; 6:34.
7. Craver. F., and Copeland.D, *Archs Surg* , Chicago, 28, 1062.
8. Gross SB, Robertson WW Jr, Lange BJ, Bunin NJ, Drummond DS:Primary Hodgkin's disease of bone. A report of two cases in adolescents and review of the literature. *Clinical Orthopaed Rel Res* 1992, 283:276-280.

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