

Primary Non-Hodgkin's Lymphoma of the Pelvic Bone in a Child

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Summary

We present a rare case of a child with primary Non-Hodgkin's lymphoma of the pelvic bone and recommend a combination of local radiotherapy and systemic chemotherapy for the treatment of this condition.

Key Words: Primary lymphoma of bone, Non-Hodgkin's lymphoma

Introduction

Primary lymphoma of the bone is a rare extranodal lymphoma and accounts for less than 0.2% of all biopsy analysed primary bone tumours¹. Its occurrence in childhood is rare. We report here a 10-year-old Chinese girl with a primary Non-Hodgkin's lymphoma of the pelvic bone.

Case Report

A 10-year-old Chinese girl presented with a one month history of fever and pain in the left hip and inability to walk. Clinical examination revealed a miserable looking girl who was febrile and who had tenderness in the left hip. The range of movement of the left hip was limited in all directions. There was no hepatosplenomegaly or lymphadenopathy. The other systems were normal. Preliminary investigations showed Hb 9.1 g/dl, WBC $12.7 \times 10^9/L$ (N 74%, L 24%, E 2%, no blast cells present), platelet $568 \times 10^9/L$ and ESR 135 mm in the first hour. The X-ray of the pelvis showed destruction

of the left ischium with evidence of new bone formation. The blood cultures were negative. A preliminary diagnosis of osteomyelitis was made and an exploration and biopsy of the left ischium was performed. The histopathological examination showed that the child had high grade Non-Hodgkin's lymphoma of the pelvic bone and the biopsy was negative for acid fast bacilli. Further investigations showed a negative HIV titre, a normal 24 hour urine VMA (vanillyl mandelic acid) and alpha fetoprotein level. The lactate dehydrogenase level was elevated at 712 U/L (normal range 230-460 U/L) and the bone marrow examination did not reveal any lymphomatous infiltration. The chest X-ray was normal and an ultrasound examination showed that the liver and spleen were normal and that there were no para-aortic lymphadenopathy. The technetium bone scan showed slight relative increase in tracer uptake in the left pelvic bone at the region of the tumour. A CT scan of the pelvic bone showed tumour arising from within the ischium with destruction of the cortex of the bone.

CASE REPORT

The patient was commenced on vincristine 1.5 mg/m², cyclophosphamide 1000 mg/m², adriamycin 40 mg/m² and prednisolone 60 mg/m², and was planned for local radiotherapy. The patient however absconded after one course of chemotherapy.

Discussion

Primary Non-Hodgkin's lymphoma of the bone (PNHLB) is an uncommon type of primary bone tumour accounting for less than 0.2% of all biopsy analysed primary bone tumours. In childhood, it is even rarer. There are no exact figures available concerning the incidence of childhood PNHLB but in a series from Japan, Ueda *et al*² reported 34 cases of PNHLB in a period from 1961-1988 and of these, there were only two cases involving children less than 12 years of age.

The criteria for the diagnosis of PNHLB are as follows³:

- (a) A primary focus restricted to a single bone on clinical examination at first admission.
- (b) Unequivocal histological proof from the bony lesion itself.
- (c) Nodal involvement, if at all present, restricted to regional nodes only.
- (d) No evidence of dissemination of disease.



X-ray of pelvis showing destruction of bone at the left ischium

To our knowledge, this patient is the first reported case of childhood PNHLB in Malaysia.

Because of its rarity in childhood, the treatment experience is limited and is based on that in adults. The treatment strategies vary and in some institutions, treatment is limited to local radiotherapy in cases where localised disease can be established by accurate staging. In other institutions, a combination of local radiotherapy and chemotherapy is advocated in view of the potential of the tumour to disseminate. In a series from Brazil by Bacci *et al*⁴, where there were 26 cases of PNHLB who were treated with a combination of local radiotherapy and systemic chemotherapy with vincristine, adriamycin and cyclophosphamide, no local relapses were noted and three out of the 26 cases developed a systemic relapse giving a 88% survival. The mean follow-up period for this study was 87 months (range 30-160 months) with 20 patients being followed-up for more than five years. In the series from the Memorial Sloan-Kettering Cancer Centre, USA,⁵ 16 cases of childhood PNHLB were treated with chemotherapy with the LSA2-L2 protocol⁶ and the survival rate was 81%. In other series where the disease is treated by radiation therapy alone the event free survival is only 40-50%⁷. As such, chemotherapy is probably the most important modality of treatment. It should include agents given in dosages high enough to cause tumour regression, prevent local recurrence as well as bone marrow and CNS dissemination. We would recommend that treatment consist of administration of systemic chemotherapy and local radiotherapy to the lesion in the affected bone.

Conclusion

In conclusion, we present here a rare case of a child with a primary Non-Hodgkin's lymphoma of the bone. The recommended treatment for this condition would be systemic chemotherapy and local radiation therapy.

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