

Bilateral retinoblastoma presenting as metastases to forearm bones four years after the initial treatment

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ABSTRACT

Osseous metastases from retinoblastoma, the most common ocular malignant neoplasm of childhood, are reported most commonly in the skull and long bones. However, metastases to forearm bones are very rare. Here we present a case of bilateral retinoblastoma with metastases to right forearm bones four years after the initial treatment.

Keywords: Bone scan, metastasis, retinoblastoma

INTRODUCTION

Retinoblastoma is the most common ocular malignant neoplasm of childhood.^[1] It is bilateral in approximately 35% of the cases.^[2] Metastatic disease is rare and usually occurs within the first two years after initial diagnosis, and is extremely rare after that time period.^[3] Osseous metastases are reported most commonly in the skull and long bones.^[2] However, metastases to forearm bones are very rare with only very few reports published till date. Here we present a case of bilateral retinoblastoma with metastases to right forearm bones four years after the initial treatment.

CASE REPORT

An 8-year-old boy treated four years back for bilateral retinoblastoma with enucleation of left eye along with adjuvant chemotherapy and radiotherapy presented with a right forearm swelling. There was no history of trauma. An X-ray revealed a mass lesion in right forearm with destruction of lower one third of right ulna. Later, patient was subjected to bone scan for skeletal metastatic work up. Bone scan was performed three hours after intravenous injection of 5 mCi of ^{99m}Tc MDP. Whole body images showed increased tracer uptake in right forearm bones [Figure 1]. Static images revealed increased tracer uptake

in middle and lower one-third of right ulna consistent with X-ray findings. In addition, bone scan showed involvement of lower one-third of left radius. Later patient underwent biopsy which revealed round blue cell tumour. However, special stains excluded Ewing's sarcoma and other primitive neuroectodermal tumors (PNETs). Involvement of radius picked up by bone scan had important implication in deciding the nature of disease as Ewing's tumor with multi-focal disease as second primary is very unlikely.

Osseous metastases from retinoblastoma are reported most commonly in the skull and long bones.^[2] The prognosis for most children diagnosed with retinoblastoma is good, with a 5-year survival of rate of more than 90%.^[4] Complications include local recurrence, metastases, and the development of a second primary tumor. Focal and diffuse involvement of the bones and

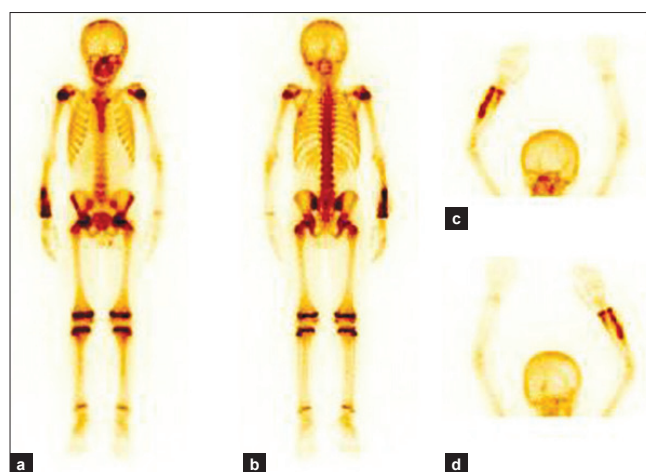


Figure 1: Tc99m-MDP bone scan; (a) whole body anterior view; (b) whole body posterior view showing increased tracer uptake in right forearm bones. Rest of the skeleton shows physiological tracer uptake. Static images of bilateral forearms; (c) anterior; (d) posterior revealing involvement of right ulna and radius

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bone marrow is known to occur, but metastases are usually seen within two years of initial diagnosis and later metastases are said to be extremely rare.^[2,5] These patients are also at risk of increased second primaries like osteosarcoma, leiomyosarcoma, spindle cell sarcoma, malignant fibrous histiocytoma, rhabdomyosarcoma, angiosarcoma, Ewing sarcoma, and PNET. These typically develop after a latent period of 8–15 years.^[6-8] Involvement of multiple bones would point the diagnosis towards metastases rather than multifocal second primary.^[9] Imaging modalities available for skeletal metastatic work up include X-ray, CT, MRI and functional imaging modalities like bone scan. X-rays are in general less sensitive to pick up the metastases, whereas whole body MRI is not widely available, so conventional WB bone scan is essential in the metastatic work up of these patients. In our case, X-ray picked up ulnar involvement but radius involvement was missed. However, bone scan revealed radius involvement suggesting a metastatic disease rather than second primary. Later, histopathology revealed round blue cell tumour involving both radius and ulna and IHC was negative for CD99 excluding diagnosis of Ewings sarcoma.

CONCLUSION

Retinoblastoma can present with skeletal metastases even later than two years of initial treatment. Skeletal scintigraphy remains an essential diagnostic procedure in evaluation of these patients and can have important implications in management.

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