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Case Report

Acute Promyelocytic Leukaemia(APML) in an Adult Patient Presenting with Multiple Lytic Bony Lesions – The First of its Kind – A Case Report with Review of Literature

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ABSTRACT
Acute promyelocytic leukaemia presenting with bony lesions alongside medullary involvement is rare with anecdotal reports having noted this manifestation in a newly diagnosed case. We here report an infrequent presentation of a young adult male presenting solely with multiple axial and pelvic bone lesions initially, with the initial impression being a lymphoma /infiltrative lesion. This is a case report that reiterates the need for prompt diagnosis and a thorough evaluation for early treatment, especially since this is a curable malignancy.

Key words: APML, Isolated bony lesions, Young adult, Rare, Remission.

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INTRODUCTION

Acute promyelocytic leukaemia constitutes 10-15% of all adult acute myeloid leukemia with complete remission rates of 80-90% with ATRA based therapies.[1] Extramedullary disease in association with/without medullary disease is usually seen at relapse with an incidence of 3-5%. The most common sites noted are the central nervous system followed by skin, bones and lymph nodes.[2] We here report a rare case of acute promyelocytic leukaemia in a young male who presented with multiple lytic bony lesions as the presenting extramedullary site.

CASE HISTORY

A 36 year old male patient with no comorbidities presented with localized lower backache for a duration of 1 month in January 2016 to a local hospital. There were no associated neurological deficits. He had no complaints of fever, easy fatiguability, bleeding or headache. His haemogram done initially revealed a Hb- 14.5 g/dl, WBC – 13,100/cumm and platelet count of 2.29 lakhs with a normal differential count and his peripheral smear showed no abnormal cells. He was referred to us with a local part MRI which revealed multifocal enhancing lesions in vertebral bodies and sacrolilitis. A PET/CT scan done at the local hospital prior to his admission at our hospital revealed multiple lytic lesions in the axial skeleton (lower lumbar and sacral bodies) and pelvis (bilateral posterior ilium), along with presacral soft tissue extending to the sacral foramina – opined as lymphoma versus infiltrative disorder. (Figure 1). Our complete blood count (done in April 2016) revealed low platelets of 8,000/cumm and leucopenia–count being 1,300. The bone marrow examination done showed a hyper cellular marrow with atypical cells with moderate amount of cytoplasm, large irregular nuclei and prominent nucleoli. The immunochemistry showed CD99, MPO, CD68 and CD117 positivity. Cytogenetics evaluation revealed (15;17)-suggestive of acute promyelocytic leukemia. The PML - RARα done was 12.26% prior to starting treatment. He received induction therapy with ATRA and ATO as per the GIMEMA[3] protocol. He developed early differentiation syndrome during the first week, however was non-life threatening. There was an initial rise in the total count to 35,000/cumm with a maximum of 23% promyelocytes in the peripheral smear. He had no fever, dyspnoea, or development of pleural/pericardial effusions. He attained complete hematological remission 35 days after initiation of treatment. His bone marrow aspiration /biopsy done post induction revealed complete morphological remission. PML-RAR α is negative and the bone imaging done by PET-CT reveals interval resolution of presacral soft tissue stranding with stable osteolysis in the sacrum and iliac bones (Figure 2).

DISCUSSION

Bony lesions in haematological malignancies are rare, but well known. They are most commonly seen in multiple myeloma, also in acute lymphoblastic leukemia, non-Hodgkin's lymphoma, Waldenstrom's disease, hairy - cell leukemia, myelodysplastic disorders, chronic lymphocytic leukemia, adult T cell lymphoma/leukaemia, chronic phase and blast crisis of chronic myeloid leukemia.[4] Bony le-

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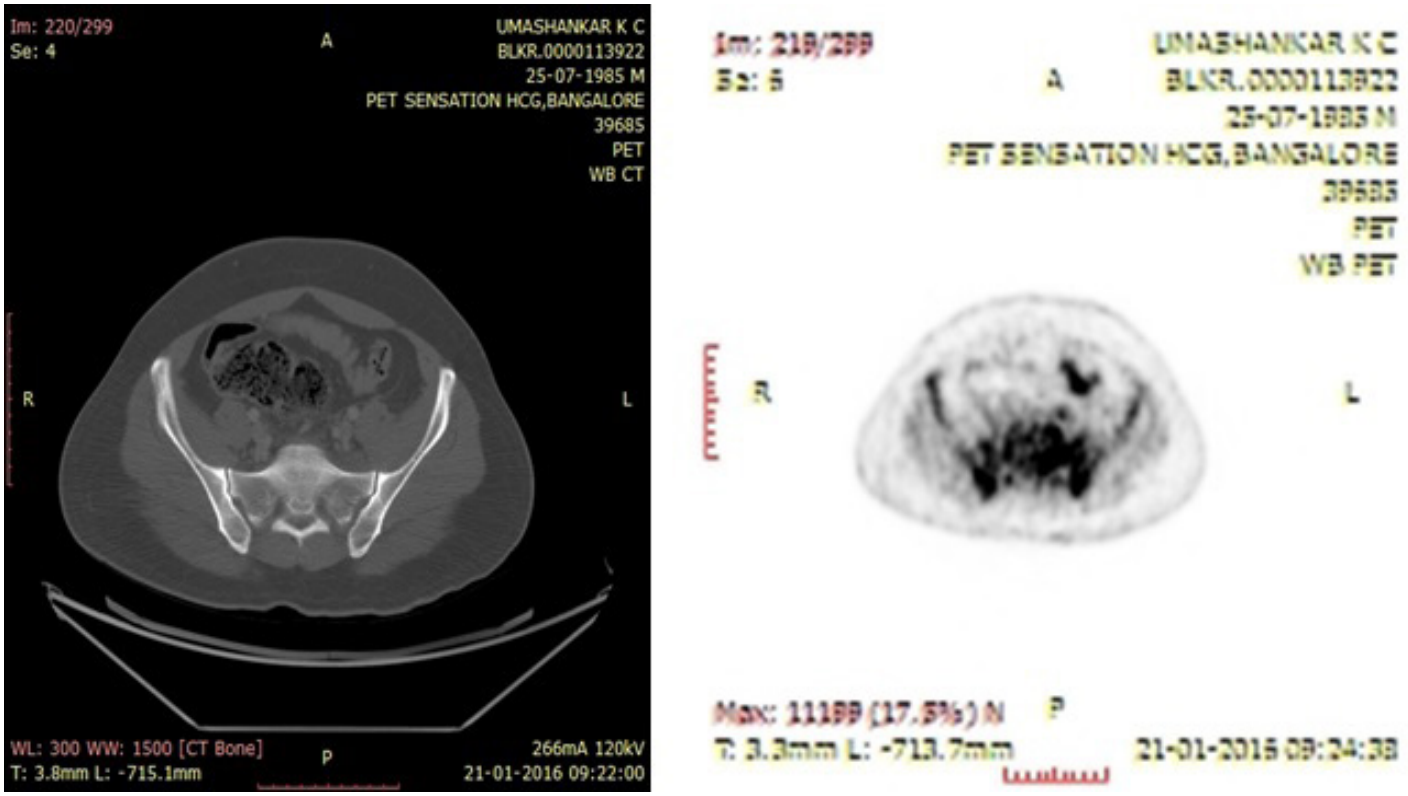

Figure 1: Pretreatment FDG- PET/CT imaging suggestive of vertebral lesion.

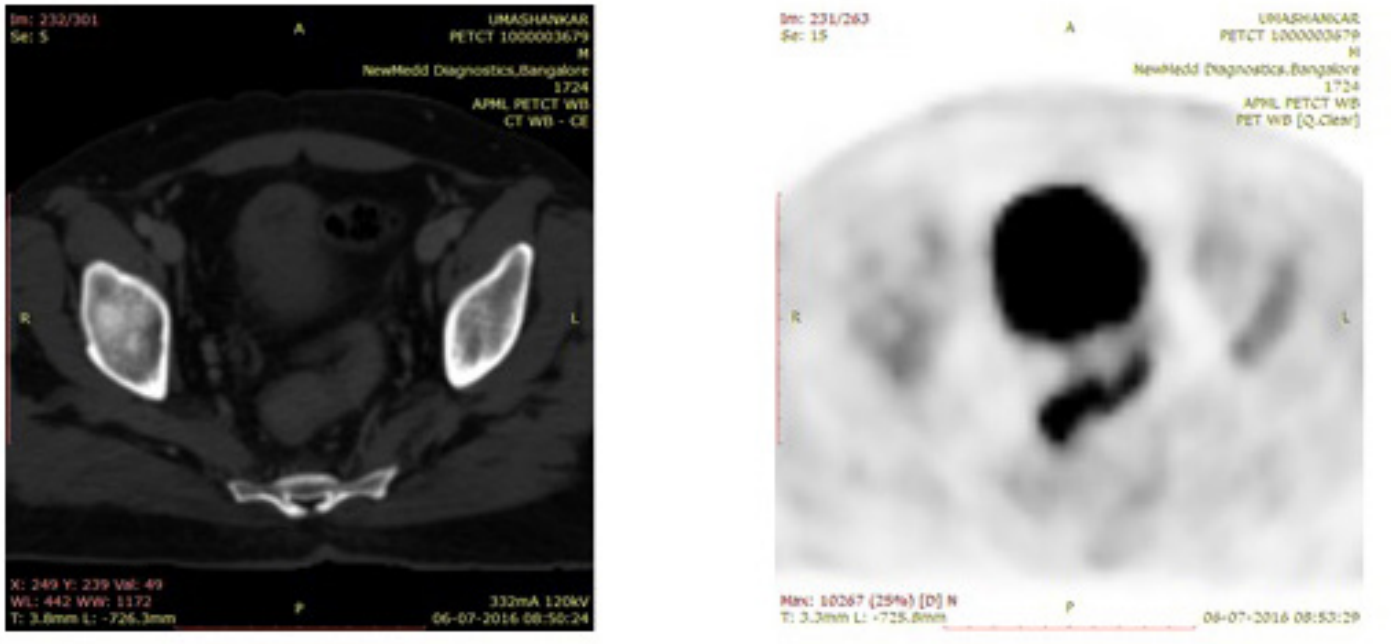

Figure 2: FDG-PET/CT imaging done post induction- with reduction in uptake in the vertebra.

sions, although unusual in acute myeloid leukaemia (AML), have been documented in case reports. The pathogenesis of bone destruction in leukemia remains poorly defined. Abnormal production of parathyroid hormone by malignant cells has been demonstrated.[5] The radiological findings described in leukemias include metaphyseal lucent bands, bone erosions, periosteal reactions, lytic bone lesions, reduced bone density, permeative destruction and vertebral collapse. Due to widespread red bone marrow in childhood, more than 50% of children with leukemia reveal skeletal abnormalities; however, this is seen in less than 10% adults.[6] Bone pain in acute leukemia is due to proliferation of bone marrow, pressure effect, compression fractures and osteoporosis.[5]

The involvement of bone has been reported in few case reports in APML,[7,8] whose patients presented with multiple lytic bony lesions and medullary disease.[9,10] have reported promyelocytic sarcoma of the ulna. The above reports are in the paediatric age group. There are few reports in literature for adult patients who had APML and who presented with single bone lesions.[11-13]

Ours is probably the first case in an adult of promyelocytic leukaemia with multiple bony lytic lesions at presentation. The outcomes of those with bony lesions with concomitant medullary involvement has been noted not to affect survival outcomes in these patients of AML[5,6] however these are seen to be associated with higher initial WBC count, CNS infiltration, and chance of later relapse, which are associated with poor prognosis.[7] Our patient's disease behaved clinically like a low/intermediate risk APML with a rather non stormy course during treatment. There were no complications associated with a rising total count such as acute respiratory distress syndrome or coagulopathy as seen in high risk group APML, which are seen to be fatal in our set up. Whether the presence of concomitant presence of extramedullary disease will be prognostic for overall survival in APML will have to be followed up and studied. This being a very infrequent presentation in APML, drawing any conclusions regarding behaviour of disease, treatment response and outcomes would remain difficult.

CONCLUSION

The main learning points in this case would be that multiple lytic lesions in a young patient should always prompt a thorough evaluation to rule out the presence of a haematological malignancy. Prompt evaluation and diagnosis will be lifesaving before development of complications, which occurs due to lack of consideration of haematological malignancies in the differential diagnosis in young patients presenting with multiple bony lytic lesions.

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CONFLICT OF INTEREST

None

ABBREVIATONS USED

APML: Acute promyelocytic leukaemia; **AML:** Acute myeloid leukaemia; **ATRA:** All-trans retinoic acid; **PML:** Promyelocytic leukaemia; **RARα:** Retinoic acid receptor–α.

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