

A Disseminated Myeloid Sarcoma Case Transformed into Leukemia

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Abstract

Myeloid sarcoma (MS) is a tumoral mass which is derived from immature myeloid precursor cells. A 28 year old man without a medical history was diagnosed as isolated MS of testis and orchiectomy was performed. After a watch and wait approach, one year later relapse occurred in the other testis without bone marrow involvement. Orchiectomy to the other testis was repeated. One year later, the patient presented to our hospital with masses at inguinal and right popliteal regions. Body 18F-FDG PET/CT showed increased FDG uptake in lymph nodes of aortocaval, paraaortic, paracaval, bilateral common iliac, right external iliac, bilateral inguinal regions with a diameter of maximum 3.7 cm and a SUVmax of 11.9; and also a heterogenous FDG uptake was observed in the muscles of posterior leg region. We performed bone marrow biopsy and aspiration resulting in no pathological infiltration. The patient was treated with induction treatment of AML, followed by consolidation with one cycle of high dose ARA-C. After the first cycle of high dose ARA-C, leucocytosis developed. Peripheral smear revealed blastoid cells. Response could not be achieved with salvage therapies. In conclusion, MS might show a complicated disease course and patients with isolated MS should be treated with systemic chemotherapy at first diagnosis.

Index terms— myeloid sarcoma, acute myeloid leukemia, testis, muscle, lymph node.

1 A Disseminated Myeloid Sarcoma Case

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In conclusion, MS might show a complicated disease course and patients with isolated MS should be treated with systemic chemotherapy at first diagnosis.

2 I. Introduction

Myeloid sarcoma (MS) is a tumoral mass which is derived from immature myeloid precursor cells.

Although MS most commonly develops as an extramedullary presentation of acute myeloid leukemia (AML), it can also accompany myelodysplastic syndrome (MDS) or myeloproliferative neoplasms (1-3). While MS is seen

in % 2-14 of AML patients (1,3), the incidence rate of isolated MS without bone marrow infiltration is only 2 per million (2). Myeloid sarcoma can occur at any time during the course of disease. It is often seen in bone, soft tissue, lymph nodes, periton and gastrointestinal system and rarely seen in genitourinary system and central nervous system (1)(2)(3).

Here, we present a case presenting with isolated testicular MS, relapsing with testis, lymph node and soft tissue involvements, and ultimately experiencing bone marrow infiltration.

3 II. Case

A 28 year-old-man without a remarkable medical history presented to his primary care physician with pain at inguinal region. A testicular mass was found at physical examination. Laboratory results including complete blood count and biochemistry were normal. Body 18F-Fluorodeoxyglucosepositron emission tomography/computed tomography (18F-FDG PET/CT) showed increased FDG uptake in testicular mass, solely. He underwent orchiectomy and biopsy revealed MS. Examinations including bone marrow aspiration and biopsy, and bone marrow conventional cytogenetics were normal without blast infiltration. After a watch and wait approach, relapse occurred in the other testis one year later. Simultaneous bone marrow examination was again normal. Orchiectomy to the other testis was repeated. One year after the second orchiectomy, the patient presented to our hospital with gait disturbance. His physical examination was notable for a mass at inguinal region with a diameter of 3 cm and a mass at right popliteal region with a diameter of 7 cm. Body 18F-FDG PET/CT showed increased FDG uptake in lymph nodes of aortocaval, paraaortic, paracaval, bilateral common iliac, right external iliac, bilateral inguinal regions with a diameter of maximum 3.7 cm and a SUVmax of 11.9; and also a heterogeneous FDG uptake was observed in the muscles of posterior leg region (figure ??). Complete blood cell count was normal. Pathological examination of the excisional biopsy of the lymph node was reported as MS. We performed bone marrow biopsy and aspiration resulting in no pathological infiltration. The patient was treated with induction treatment of AML (3/7: Idarubicin + Cytosine arabinoside (ARA-C)), followed by consolidation with one cycle of high dose ARA-C. His gait disturbance resolved. For monitoring response to therapy we performed 18F-FDG PET/CT which showed increased FDG uptake in lymph nodes of abdomen and iliac region and mass in the right popliteal region with a diameter of 3.5x2.9x7.4 cm (SUV max: 9.2). After the first cycle of high dose ARA-C leucocytosis developed. Peripheral smear revealed blastic cells. Bone marrow involvement was confirmed by bone marrow examination. Salvage therapy with fludarabine + ARA-C + granulocyte colony stimulating factor + idarubicin (FLAG-DA) and later etoposide + mitoxantrone + ARA-C (EMA) were applied to the patient sequentially. However response could not be achieved and patient died.

4 III. Discussion

Although lymph node involvement of MS is often encountered (1,2), testicular (4-6) and muscle involvements (7,8) are rare entities in these patients. Nevertheless, our patient comprised both testicular and muscle involvements consecutively, and also the lymph nodes were affected which was predictable in such a spread of the tumor.

An other distinctive feature of this case is the development of bone marrow infiltration nearly 2.5 years after the diagnosis with the propagation of MS. Whereas leukemic transformation of MS usually occurs after a nearly median 7 months (1). Further more leukemic transformation occurred during treatment after atypical extramedullary relapses. Mostly, MS with chromosome 8 abnormality transforms to AML rapidly with high incidence (2). However cytogenetic analysis of our patient was normal, which may be the cause of late leukemic transformation, although he did not receive systemic treatment at first diagnosis.

To our knowledge, this MS case differs with its disease course, as it presented with isolated testicular MS, relapsed repeatedly with testicular and later with lymph node and muscle involvements, and ultimately experienced leukemic transformation. Patients with isolated MS should be treated at first diagnosis.

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.1 Conflict of Interest

The authors declare that they have no conflict of interest

[Bassichis et al. ()] ‘Chloroma of the masseteric muscle’. B Bassichis , J Mcclay , B Wiatrak . *Int J Pediatr Otorhinolaryngol* 2000. 53 p. .

[Eggenger et al. ()] ‘Granulocytic sarcoma of the testis’. S E Eggenger , A Abrahams , T C Keeler . *Urology* 2004. 63 p. .

[Manabe et al. ()] ‘Granulocytic sarcoma with orbit, cauda equina, muscle and peripheral nerve extension but without bone marrow involvement’. Y Manabe , Y Hamakawa , K Sunami , Y Ohta , N Omori , K Abe . *Intern Med* 2007. 46 p. .

[Yilmaz et al. ()] ‘Granulocytic sarcoma: a systematic review’. A F Yilmaz , G Saydam , F Sahin , Y Baran . *Am J Blood Res* 2013. 3 p. .

[Bakst et al. ()] ‘How I treat extramedullary acute myeloid leukemia’. R L Bakst , M S Tallman , D Douer , J Yahalom . *Blood* 2011. 118 p. .

[Valbuena et al. ()] ‘Myeloid sarcoma involving the testis’. J R Valbuena , J H Admirand , P Lin , L J Medeiros . *Am J Clin Pathol* 2005. 124 p. .

[Avni and Koren-Michowitz ()] ‘Myeloid sarcoma: current approach and therapeutic options’. B Avni , M Koren-Michowitz . *Ther Adv Hematol* 2011. 2 p. .

[Constantinou et al. ()] ‘Testicular granulocytic sarcoma, a source of diagnostic confusion’. J Constantinou , T Nitkunan , M Al-Izzi , T A McNicholas . *Urology* 2004. 64 p. .