Shoulder Myeloid Sarcoma: An Initial Presentation of CML Blast Crisis

Ankur Jain¹ · Kamal Kant Sahu¹ · Saniya Sharma¹ · Arvind Rajwanshi¹ · Vikas Suri¹ · Pankaj Malhotra¹

Received: 10 September 2015 / Accepted: 3 February 2016 / Published online: 9 February 2016
© Indian Society of Haematology & Transfusion Medicine 2016

Abstract Myeloid sarcoma (MS) represents extra medullary accumulation of the immature cells of granulocytic series and occurs most commonly in the setting of acute myelogenous leukemia. Its occurrence in chronic myeloid leukemia (CML), myelodysplastic syndrome and other myeloproliferative neoplasm is uncommon. We here in report a 35-year old lady who was diagnosed as CML-chronic phase (CP) in the year 2004 and was on imatinib (400 mg OD) since then with regular follow up and good compliance. She had progression to accelerated phase in April 2014 which was managed by increasing the dose of imatinib to 600 mg OD. In August 2015, she presented with complaints of pain and swelling of the left shoulder suggestive of septic arthritis. Investigations revealed an ill-defined lesion involving muscles around the shoulder and clavicle. Absence of response to antibiotics and negative work up for infectious etiology raised the suspicion for MS which was later confirmed by immunohistochemistry of the aspirate from the lesion. Bone marrow examination was consistent with CML-CP. Hence, the diagnosis of CML with extra medullary blast crises was made. Patient was treated with a combination of high dose imatinib, hydroxyurea, cytarabine and local radiotherapy. Rarity of MS involving the shoulder and its resemblance to septic arthritis has been highlighted in the present case. We emphasize the importance of immunohistochemistry of the aspirate for the timely and correct diagnosis of the cases who do not respond to an initial antibiotic trial.

Keywords Granulocytic sarcoma · Shoulder · Radiotherapy · Chronic myeloid leukemia

We here in report a 35-year old lady who was diagnosed as CML-chronic phase (CP) in the year 2004 and was started on imatinib (400 mg daily). She achieved complete hematological response (CHR) at 3 months and a complete cytogenetic response (CCyR) at 12 months of starting therapy and was in CHR and CCyR till April 2014 when she had progression to accelerated phase (AP) which was managed by increasing the dose of imatinib to 600 mg daily. In August 2015, she presented with complaints of pain and swelling of the left shoulder suggestive of septic arthritis. Local examination revealed tender swelling of the left shoulder joint with redness and induration of the overlying skin and restriction of both active and passive movements (Fig. 1). Spleen was palpable per abdomen (8 cm). Peripheral smear and bone marrow findings were consistent with CML-CP. MRI revealed an ill-defined lesion involving muscles around the shoulder and clavicle suggestive of an abscess (Fig. 2). She was started on broad spectrum antibiotics initially with a suspicion of pyomyositis. Microbiological evaluation of the fine needle aspirate from the lesion was negative for bacteria, fungus and *Mycobacterium tuberculosis*. Absence of response to antibiotics and negative work up for infectious etiology raised the suspicion for MS which was later confirmed by immunohistochemistry of the aspirate from the lesion (Fig. 3). Hence, the diagnosis of CML with extramedullary blast crises was made. Patient was treated with a combination of high dose imatinib (as Kinase domain mutation analysis from the peripheral blood revealed H396R mutation), hydroxyurea, low dose cytarabine and local radiotherapy (RT) which led to a significant reduction in size of

✉ Pankaj Malhotra
hematpgi@gmail.com

¹ Department of Internal Medicine, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India

Fig. 1 **a** Clinical image showing the involvement of left shoulder joint by the myeloid sarcoma. **b** Shoulder is swollen, red and skin is indurated on palpation

Fig. 2 **a** X-ray of the left shoulder showing erosion of the lateral end of left clavicle. **b** MRI of the same region showing an ill-defined heterogeneously enhancing lesion involving the muscles around the shoulder and infiltrating into clavicle

the lesion. MS is reported in about 2–8 % of the cases of AML and can occur after (50 %), concurrent with (15–35 %) or prior to the diagnosis of AML (25 %), after HSCT (<1 %) or rarely as the first sign of relapse after treatment of AML. MS has rarely been described in CML, MDS and other myeloproliferative neoplasm [1]. Occurrence of MS in a patient with CML categorizes the case as extra medullary BC even without evidence of systemic disease. Most common sites of MS reported in the literature includes skin, bone and lymph nodes and the involvement of the CNS, gastrointestinal tract, genitourinary tract, chest

wall, pleura, breast, retro peritoneum, testis, spinal cord and oral mucosa are uncommon ones [2]. Involvement of joints with MS is extremely rare and limited to isolated case reports only [3]. Isolated MS or MS concurrent with AML/CML should be treated with AML based induction chemotherapy regimens [4]. Role of radiation is palliation of pain and the addition of local RT to systemic chemotherapy has been shown to prolong failure-free survival without an improvement in overall survival [5]. Rarity of MS involving the shoulder and its resemblance to septic arthritis has been highlighted in the present case.

Fig. 3 **a** Low power microphotograph (Giemsa $\times 40$) shows hypercellular FNA smears with dispersed population of cells comprising predominantly of blasts with scattered myeloid precursors. **b**, **c** High power microphotographs (Giemsa $\times 1000$) show blasts with dispersed chromatin, round to irregular nuclear membrane, prominent nucleoli and scanty to moderate amount of pale basophilic to granular cytoplasm (*arrows*). **d** Myeloperoxidase (MPO $\times 1000$) staining shows strong cytoplasmic granular positivity in the blasts (*arrows*)

The importance of entertaining the possibility of myeloid sarcoma in cases of septic arthritis which do not respond to antibiotic trial has been emphasized here.

Compliance with Ethical Standards

Conflict of interest None.

Ethical standards On behalf of all the co-authors (listed above) I would like to state that this work has not been submitted elsewhere in any other journal. No involvement or participation of human/animals requiring ethical clearance.

References

1. Avni B, Koren-Michowitz M (2011) Myeloid sarcoma: current approach and therapeutic options. *Ther Adv Hematol* 2(5):309–316

2. Sahu KK, Tyagi R, Law AD, Khadwal A, Prakash G, Rajwanshi A et al (2015) Myeloid sarcoma: an unusual case of mediastinal mass and malignant pleural effusion with review of literature. *Indian J Hematol Blood Transfus* 31(4):466–471
3. Upadhyay S, Rawat SJ, Gupta G, Saxena U (2014) Chloroma (Granulocytic sarcoma): an unusual cause of shoulder pain in chronic myeloid leukemia; a diagnostic dilemma. *J Orthop Allied Sci* 2:20–23
4. Tsimberidou AM, Kantarjian HM, Wen S, Keating MJ, O'Brien S, Brandt M et al (2008) Myeloid sarcoma is associated with superior event-free survival and overall survival compared with acute myeloid leukemia. *Cancer* 113(6):1370–1378
5. Tsimberidou AM, Kantarjian HM, Estey E, Cortes JE, Verstovsek S, Faderl S et al (2003) Outcome in patients with nonleukemic granulocytic sarcoma treated with chemotherapy with or without radiotherapy. *Leukemia* 17(6):1100–1103