Uncommon pathology of thyroid, myeloid sarcoma: a case report

P. Kumar Mandal[a], Anindya Adhikari[b], Kumar Praveen[c], Samrat Dutta[d]

Department of Pathology [a]Sagar Dutta Medical College, Kolkata, [b]Bankura Sammilani Medical College, Bankura, [c]GB Pant Medical College, New Delhi, [d]North Bengal Medical College, Darjeeling, West Bengal, India

Correspondence to Anindya Adhikari, MD, Basudevpur, Banipur, Sankrail 711304, Howrah, West Bengal, India
Tel: + 91 943 329 2336;
e-mail: anindyaadhikari04@gmail.com

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Myeloid sarcoma (MS) of the thyroid in a patient suffering from chronic myeloid leukemia (CML) is very rare. Herein, we report a case of MS of the thyroid in a 50-year-old woman who was a diagnosed as a case of CML but discontinued treatment for the last 1 year despite satisfactory initial response to chemotherapy. She came with weakness, fever, and painful swelling of the thyroid for the last 1 month. Present hematological examination diagnosed CML in blast crisis phase. Ultrasonography revealed multiple nodules along with calcification in the right lobe of the thyroid. Guided fine needle aspiration from thyroid nodule disclosed MS of the thyroid. Our report is not only rare but also stresses the need for regular chemotherapy, discontinuity or irregularity of which calls upon such complication.

Keywords:
chronic myeloid leukemia, myeloid sarcoma, thyroid

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Introduction

Myeloid sarcoma (MS), also called granulocytic sarcoma, intramedullary myeloblastoma, or chloroma, is a rare solid tumor composed of immature myeloid cells [1]. The tumor may precede or occur concurrently with acute or chronic myeloid leukemia (CML). Known sites for MS are skin, soft tissue, lymph nodes, and bones [2]. The thyroid is an uncommon site for MS.

Case report

A 50-year-old woman presented with a history of weakness, abdominal discomfort, intermittent fever, and pain in the neck for 1 month. History begins 2 years back when she was diagnosed with CML and treated with imatinib. That time, clinically she responded well to medicine and the hematological parameters also showed improvement with fall in total leukocyte count to 22 000 from 110 000/cm^2 and rise in hemoglobin from 6.2 to 9.5 g/dl. However, after 7 months, she started taking medicine first irregularly and then completely discontinued drugs and was ultimately lost to follow-up. Now, she has pallor, nodular thyroid enlargement (Fig. 1a), and moderate hepatosplenomegaly. Examination of peripheral blood revealed total leukocyte count of 67 000 and myeloblast of 25%. Bone marrow study showed myeloblast of 52% (Fig. 2a) suggestive of blast crisis phase of CML. Fluorescent in situ hybridization for breakpoint cluster region gene (BCR)/Abelson murine leukemia viral oncogene (ABL) translocation assay showed BCR/ABL fusion signal detected (Fig 2b) in 72% of cells. Whole-abdomen ultrasonography (USG) showed hepatosplenomegaly, whereas USG of the neck (Fig 1b) showed multiple nodules with calcification in both lobes of the thyroid, more on the right side. USG-guided fine needle aspiration from thyroid nodule showed myeloblasts, myelocytes in background of thin colloid, and a final diagnosis of MS of thyroid was rendered. Immediately she was put on chemotherapy (CT) for acute myeloid leukemia (AML). Responding well to treatment her thyroid swelling reduced gradually. Blood and bone marrow picture returned to CML of chronic phase again. Unfortunately, she was again lost to follow-up after 3 months.

Discussion

MS, also known as extramedullary myeloid tumor, is a tumor mass composed of myeloblasts or immature myeloid cells occurring in an extramedullary site or in bone [1]. The clinical presentations of MSs vary and are dependent on the sites of involvement. Commonly involved sites are subperiosteal bone structures of the skull, paranasal sinuses, sternum, vertebrae and pelvis, lymph nodes, and skin [1]. Rare sites reported in the literature include the pancreas, heart, brain, mouth, breast, gastrointestinal tract, and so on [2]. A single tumor or sometimes multiple nodular masses may occur. MS may be found in one of the four settings – (a) in patients with known AML in the active phase of the disease; (b) in patients with chronic myeloproliferative disorder (CMPD) or myelodysplastic syndrome, in

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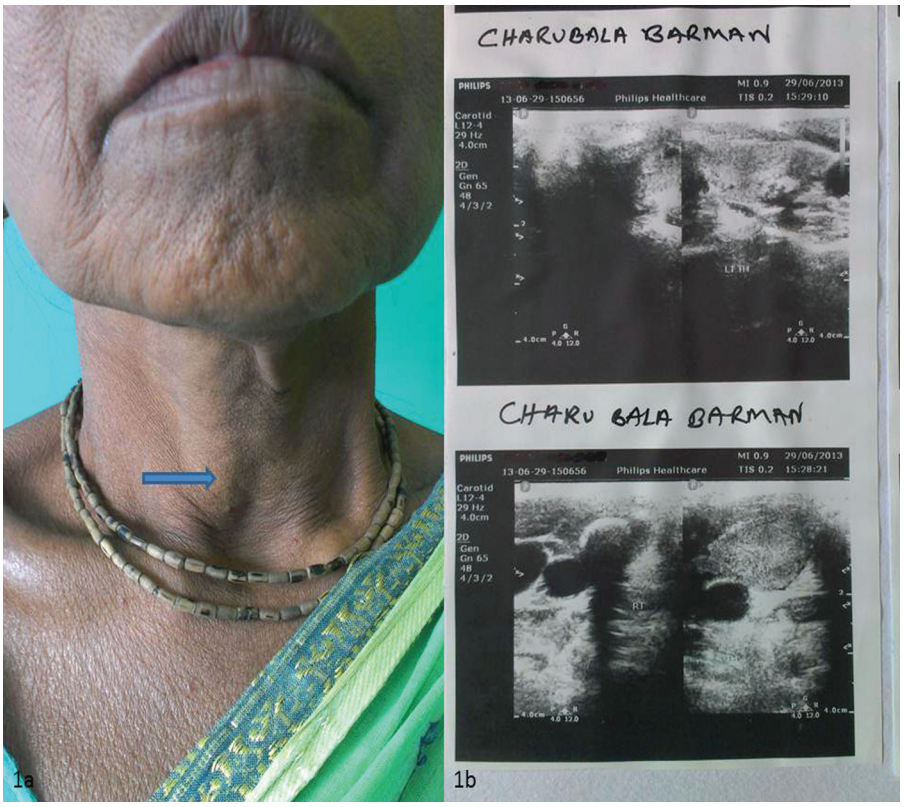
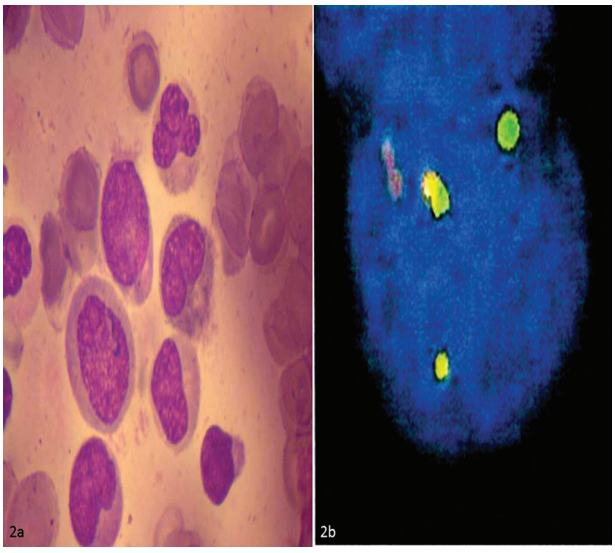
Fig. 1



a: Nodular thyroid enlargement. b: USG of the neck.

Fig. 2



a: Myeloblast of 52%. b: Signal detected.

whom MS may be the first manifestation of blastic transformation; (c) as he first manifestation of relapse in previously treated patients of primary or secondary acute leukemia; (d) *de novo* in healthy individuals [1,3]. They occur in 2.5–9.1% of patients with acute myelogenous leukemia and five times less frequently in patients with CML. MS patients with CMPD have a negative prognosis, because these tumors often occur during acute transformation [4].

It typically occurs in the later decades of life (median age: 56 years; range: 1 month to 89 years) [5,6]. Studies show equal sex predilection for MS [4]. Although MSs are cytologically variable, most often they are composed of medium-sized to large blastic cells with ovoid vesicular nuclei with medium-sized or large centrally located nucleoli and dispersed chromatin. Their cytoplasm is scant to moderate. Cytochemical stains such as Sudan Black, periodic acid–Schiff, myeloperoxidase (MPO), and nonspecific esterase are helpful to identify the lineage. MPO stain was positive on fine needle aspiration smears in our case. Immunohistochemistry with MPO, lysozyme, CD-68, CD-33, CD-34, and others can be performed on tissue sample for further confirmation [1,7]. Although the diagnosis is often thought of in patients with an established history of AML, myelodysplastic syndrome, or CMPD, in others without such histories the diagnosis is often missed. The differential diagnoses are non-Hodgkin lymphoma, small round cell tumors, undifferentiated carcinoma, melanoma, etc. The current recommended treatment regimen in patients presenting with isolated MS or MS presenting concomitantly with AML is conventional AML-type chemotherapeutic protocols and the role of radiotherapy in addition to

systemic CT is not established, although it is often given [8]. Our patient's outcome remained unknown as she was lost to follow-up repeatedly.

Conclusion

Discontinuation or even suboptimal dose of CT in a case of CML may lead to blastic transformation, which may present as MS. As the prognosis deteriorates in MS the correct diagnosis is important and the line of management has to be changed in CML.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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