IJBC 2015; 7(2): 105-109

Face Bones Involvement and Relapse in a Case of Childhood Acute Leukemia

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Submitted: 05-07-2014 , Accepted: 23-11-2014

Abstract

Midface bones are an unusual site for primary presentation and relapse in acute lymphoblastic leukemia. Herein, we describe a case of acute pre B cell lymphoblastic leukemia with leukemic infiltration of maxilla and bone marrow involvement. At the time of relapse, the patient presented again with maxilla involvement and the phenotype changed to biphenotypic lymphoblastic leukemia. Our case suggests that leukemic involvement of the maxilla should be considered in differential diagnosis of patients presenting with acute painful midface swelling. These patients should be evaluated using imaging modalities, blood parameters and histopathology.

Keywords: Face, bone, leukemia, relapse, acute lymphoblastic leukemia.

Introduction

Childhood acute lymphoblastic leukemia (ALL) is the most common type of cancer in children. It has the potential to infiltrate the liver, spleen, lymph nodes and central nervous system. Such extramedullary presentations are important in biology resolution of childhood ALL and predicting the patient's survival. However, facial bones mass is an extremely rare presentation in childhood ALL[1]. Herein, we report a boy presenting with right local maxilla swelling who was diagnosed as an acute lymphoblastic leukemia case. The patient experienced relapse 18 months after the first period of chemotherapy for acute lymphoblastic leukemia, again with a maxilla mass and phenotype switching phenomenon in leukemic blasts.

We present an unusual form of extramedullary organ infiltration in an ALL patient and subsequent relapse with transformed phenotype after starting of the treatment.

Report of the Case

A nine year-old boy presented with approximately one month history of low grade fever, right sided maxilla swelling, and teeth ache unresponsive to analgesic drugs. The patient initially was treated as a case of dental abscess but swelling did not shrink following the antimicrobial therapy. He did not have a history of trauma, weight loss, or facial sensory disturbances.

At the time of hospitalization, the patient was febrile and pale, but lymphadenopathy or hepatosplenomegaly were not detected.

Laboratory assessment revealed normal hemoglobin level for the age, leukocytosis (WBC= 33000/mm^3) with 15% lymphoblast and thrombocytopenia (platelet count = 110000/mm^3). High erythrocyte sedimentation rate (ESR) and lactate dehydrogenase (LDH) levels were recorded at 112 mm/hr and 1220 IU/dl respectively.

Chest radiography and abdominopelvic sonography were normal. Axial and coronal computed tomography showed an expansible lesion within the right part of maxilla (Figure 1).

Bone marrow aspiration and trephine biopsy under light microscopy revealed infiltration of a homogeneous population of blasts constituting of 60% nucleated cells with suspicion of acute lymphoblastic leukemia (Figure 2). Also diffuse infiltration of leukemic cells in maxilla was observed in H&E staining (X100) in the first episode of disease (Figure 3). Flow cytometry of bone marrow aspirate showed positive immunoreactivity of malignant

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cells for TdT, CD22, CD34, and negativity for CD 10, CD13, CD20, HLA-DR and CD3 compatible with diagnosis of precursor B cell ALL (Figure 4).

The patient was treated according to pre B-cell type ALL chemotherapy protocol using vincristine 1.5 mg/m^2, L- asparaginase 6000 IU/m^2, dexamethasone 40 mg/m^2, methotrexate, and intrathecal cytarabine for 18 months.

After 18 mounts since the beginning of the chemotherapy, while the patient was receiving maintenance chemotherapy, he presented again with maxilla re-swelling at the same site of the initial presentation. In physical examination, patient had a bulging over the right maxilla, with mild to moderate local tenderness. Complete blood count (CBC) demonstrated normal limits but ESR was high (35 mm/hr.).

Chest radiography and abdominopelvic sonography were normal. Whole body bone scan showed an area of hyper absorption in right maxilla indicative of an infiltrative phenomenon.

Bone marrow and CSF analysis showed no evidence of leukemic relapse. Right face bone histopathology study demonstrated diffuse infiltration of discohesive, small, relatively uniform cells, with high nuclear-to-cytoplasmic ratio, irregular nuclear borders, and scanty cytoplasm with high mitotic activity. Immunohistochemical study showed biphenotype T-cell and B-cell lymphoblastic leukemia; CD3+, CD5+ (weakly), CD19+ (weakly), CD7+, CD20-, CD68-, CD10-, TdT (polyclonal)+, PAX-5 +, CD 79 α+.

Based on the diagnosis of biphenotype acute lymphoblastic leukemia, chemotherapy was started with vincristine 1.5 mg/m^2, prednisone 40 mg/m^2/day, doxorubicin 30 mg/m^2, MTX 4mg/m^2, PEG-asparaginase 2500 IU/m^2. For better local control, patient received radiotherapy of the right maxilla according to radiotherapist recommendation. Ten days after initiation of the chemotherapy, swelling, pain and local tenderness resolved. Tertiary relapse occurred 5 months after the second relapse as diffuse aleukemic leukemia cutis, without bone marrow or CNS relapse or other extramedullary involvement (Figure 5). Again he was treated by first course of MRC12 protocol including adriamycin (33.5 mg /m^2 day 1,3,5), cytosar (100 mg /m^2 day 1 to 10) and etoposide (100 mg /m^2 day 1 to 5). Remission was induced by this regimen and he received allogeneic bone marrow transplant from a matched related donor immediately.

Discussion

This case was interesting because of the unusual extramedullary presentation of the leukemia, similar presenting and relapse site, and switching of phenotype from immunophenotypic into biphenotypic leukemia with the same presentation site.

A case of leukemic infiltration of the maxilla, was described by Bakathir and Al-Hamdani in 2009 [2]. They described a clinical case of relapse of precursor B-cell acute lymphoblastic leukemia in maxilla of a 19-year-old female patient who presented with facial swelling, sensory disturbances of the face, and teeth mobility 10 months after a successful allogeneic bone marrow transplantation. The oral and dental presentations were the only features indicating leukemic relapse in this patient similar to our patient. This patient had a maxilla involvement just as a presentation during relapse, but our patient had both primary maxilla involvement, and relapse at the same site as well as a different phenotype at relapse.

In 2011, a 35-year-old male with adult acute lymphoblastic leukemia was reported presenting with an osteolytic lesion of the mandible [3]. There was no definitive involvement in other craniofacial bones. A panoramic radiograph taken 4 months before had indicated no bony involvement. In this case a CBC count showed a slightly decreased red blood cell count, but white blood cell count, differential cell count and platelet counts were normal. Routine chemistry of the patient revealed hypercalcemia with an increased level of parathyroid hormone-related protein. But our patient had normal biochemical and hematological laboratory tests. In this case mandibular lesion was osteolytic but our patient had a non-lytic lesion with opacified pattern.

In 2011, Fallahinejad Ghajari et al. [4], reported a 12-year-old girl with symptoms of pain and swelling in the buccal vestibule and also at the posterior part of the right palate of the maxilla diagnosed with ALL and undergoing complete chemotherapy.

In 2006 Karimi and Eshghi reported a 6-year-old boy with unusual huge enlargement of maxilla, mandible and soft palate as well as gingival hypertrophy which led to secondary respiratory

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Figure 1: Computed tomography showing an expansible lesion within the right part of maxilla.

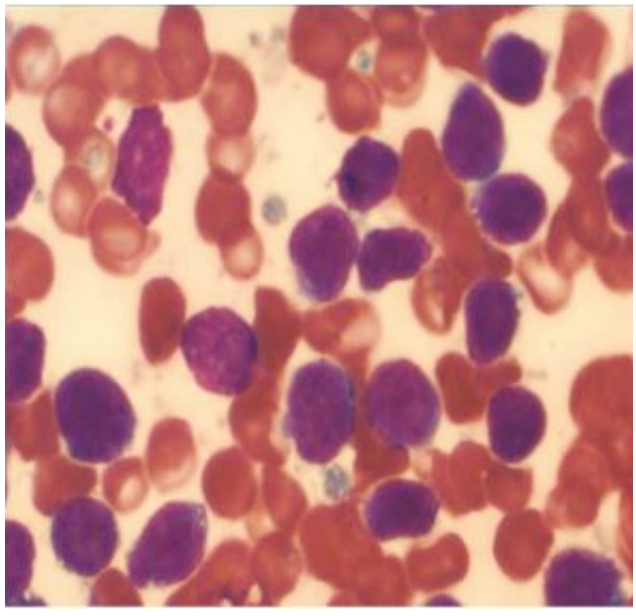

Figure 2: Bone marrow aspiration and trephine biopsy under light microscopy shows infiltration of a homogeneous population of blasts constituting of 60% nucleated cells with suspicion of acute lymphoblastic leukemia.

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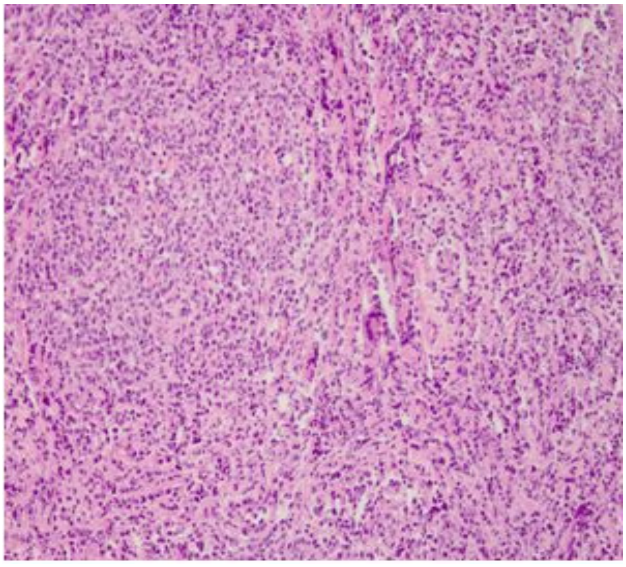

Figure 3: Diffuse infiltration of leukemic cells in maxilla (H&E staining, X100) in the first episode of disease.

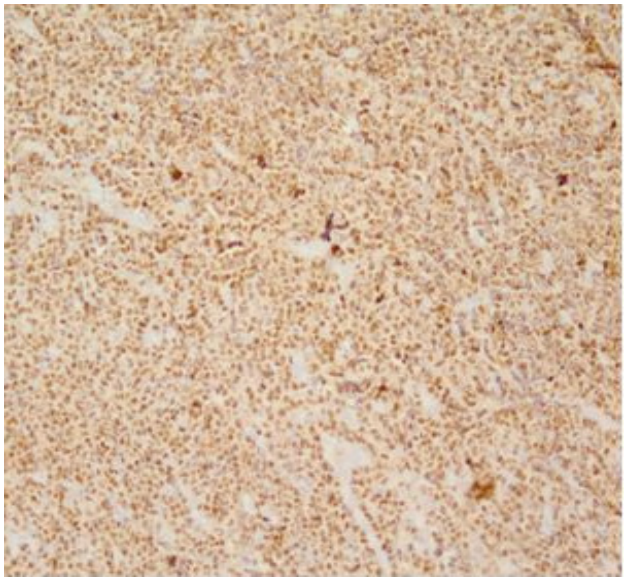

Figure 4: In Immunohistochemistry diffuse nuclear immunostaining with TdT in leukemic cells was observed in the first episode of the disease.

and feeding difficulties. Morphologic and flow cytometric evaluation of the bone marrow aspiration showed T cell type acute leukemia and the patient was considered as a case of T cell lymphoma/leukemia [5].

In our case, lineage switching happened with same presentation. The reason for this phenomenon may be the incapability of pre-B ALL chemotherapy to induce clinically significant antileukemia effect on T cells. This may be not only due to their inability to function as APC (Antigen Processing Cell) but also due to their potential to

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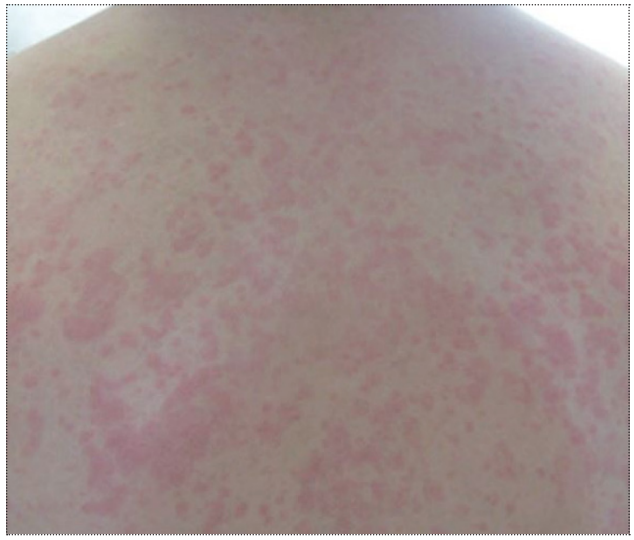

Figure 5: Tertiary relapse as diffuse infiltration of leukemic cells in the skin occurred 5 months after the second relapse.

induce tolerance.

Biphenotypic acute leukemia (BAL) and every transformation in cell lineage or phenotype is a very rare phenomenon possibly arising from hematopoietic pluripotent stem cells with generally poor outcome. Patient can be a candidate for allogeneic bone marrow transplant after remission. The presence of a different cell phenotype at the time of relapse, a so-called phenotype switch (in lymphoid band), is a relatively rare event. Phenotype switch may result from chemotherapeutic eradication of one phenotype and subsequent expansion of a second phenotype of an originally biphenotype leukemia. Alternatively, chemotherapy may induce modulation of antigen expression in a leukemic phenotype that retains the potential for T or B-lymphoid differentiation [6]. Also this phenomenon can happen due to technical failure in immunophenotyping of blast cells.

Conclusion

Our case suggests that leukemic involvement of the maxilla should be considered in differential diagnosis of patients presenting with acute painful midface swelling. These patients should be evaluated using imaging modalities, blood parameters and histopathology.

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