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Cutaneous Lymphoblastic Lymphoma Presenting as a Solitary Traumatic Soft Tissue Facial Mass: A Case Report

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Abstract

This case report illustrates the presentation of a six-year-old female with a four month history of increasing redness and swelling of her forehead. At the time of presentation, the clinical history and physical examination did not suggest an oncologic diagnosis. However, due to the persistence and enlargement of the facial lesion, a biopsy was performed and revealed precursor B-cell lymphoblastic lymphoma. This case illustrates an unusual initial presentation of lymphoblastic lymphoma and the effectiveness of routine skin biopsy for skin masses in the early diagnosis of cutaneous pre-B-cell lymphoblastic lymphoma.

Keywords: Lymphoma; Mass; Pediatrics; B-Cell

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Case Report

A six-year-old Caucasian girl presented for admission to the hospital following a concerning workup for a soft tissue mass on her right forehead. This mass initially began as a flesh-colored pinpoint lesion that the parents first noticed following a fall on her forehead four months prior to presentation. Since that time it continued to enlarge and change in color from flesh to bluish-purple in coloration. The parents were not overly concerned at first, but become more concerned when they noticed the lesion continuing to increase in size over four months. Despite this, the parents reported that she did not develop any other concerning symptoms such as fever, weight loss, night sweats, anorexia, nausea, or vomiting. At one point during this four month course, one physician suspected a soft tissue infection (due to associated pain and redness) and treated her with a ten day course of sulfamethoxazole and trimethoprim (TMP) (10 mg TMP/kg/day) with no response. After four months of observation with no improvement despite antibiotic therapy, a skull x-ray and ultrasound were obtained. The skull films were normal but the ultrasound demonstrated a solitary soft tissue mass that was not attached to the skull. Since an oncologic process could not be excluded, a biopsy was obtained to better characterize the mass. The biopsy was concerning for pre-B lymphoblastic lymphoma so the patient was admitted for further evaluation.

Hosiptal Course

On initial hospital examination, she was found to have normal vital signs for her age. She appeared well-nourished and well-appearing. Her physical exam was significant only for a homogeneous bluish-purple 3.5 × 2.5 cm raised circular, edematous mass on her right supraorbital ridge that was tender to palpation. The remainder of her physical exam was unremarkable. She did not have any organomegaly, lymphadenopathy, and cranial nerve dysfunction, joint swelling, muscle weakness, or bone pain. Her hospital work-up included computer tomography (CT) scans of neck, chest, abdomen and pelvis, cerebrospinal fluid (CSF) analysis and bilateral bone marrow aspirates and biopsies to stage a suspected lymphoma. After reviewing the original biopsy slides, the lesion was re-biopsied by fine needle aspiration. Histology of the lesion revealed an atypical lymphoid infiltrate with flow cytometry revealing a population of cells that were positive for B-cell antigens CD19, CD10, CD34, and TdT. Bone marrow aspirates and biopsies revealed no morphologic evidence of

lymphoma cells however flow cytometeric analysis revealed a small population of cells similar to the immunophenotype of her lymphoma cells in her bone marrow. These findings confirmed precursor B-cell lymphoblastic lymphoma (PBLL). Fortunately her cerebral spinal fluid studies were negative for leukemic infiltration.

After diagnosis of PBLL was confirmed, she had surgery for a port-a-catheter placement and started on systemic chemotherapy protocoled through Children's Oncology Group (COG). She started induction therapy with intrathecal cytarabine, vincristine, decadron, doxorubicin, and pegylated asparaginase. She received supportive care with intravenous fluid hydration and allopurinol during her induction chemotherapy regimen. She responded immediately to her first induction cycle with the forehead lesion starting to appear smaller by subsequent days of treatment. Other than an admission for febrile neutropenia, she continued to respond to treatment with no signs of cerebrospinal or further bone marrow infiltration to date, nor recurrence of the forehead lesion.

Discussion

Lymphoblastic lymphoma is one of the most common pediatric non-Hodgkin's lymphomas, however pre-B-cell lymphoblastic lymphoma accounts for less than 10% of all pediatric lymphoblastic lymphoma cases [1]. PBLL is defined as a neoplasm of immature lymphoid cells expressing TdT and B-cell antigens (CD10, CD19, CD22, and CD79a) [2,4,6]. PBLL in children frequently presents at the median age of 14-20 years of age with a female predominance [1,2,4,5,7]. It is usually diagnosed as a bulky mass in solid organs or as painless swelling in the armpit, neck, or groin from enlarged lymph nodes. These lymphomas may present with or without bone marrow or CNS involvement [3,5,7]. In this disorder it is common to see symptoms such as pallor, fatigue, bleeding, and infection. It is also not unusual to have extra nodal involvement including hepatosplenomegaly or central nervous system abnormalities [2]. Laboratory studies may demonstrate pancytopenia, hypercalcemia, hyponatremia, hypokalemia, and elevated lactate dehydrogenase.

A biopsy is needed to confirm the diagnosis of PBLL. The biopsy criteria for the diagnosis of PBLL includes the presence of lymphoblastic histology, FAB-L1 or L2 cytomorphology (French, American and British leukemia subtypes one and two), at least one b-lineage antigen, extramedullary primary, ≤ 5% blasts in the bone marrow, and the absence of surface immunoglobulin and t-lineage antigens [1-7]. Staging for this tumor takes into consideration physical examination, peripheral blood studies, bone marrow analysis, cerebrospinal analysis, ultrasonography, chest X-ray, chest computed topography and magnetic resonance imaging, and skeletal scintigraphy (**Table 1**) [7]. It is important to recognize and differentiate between PBLL and B-cell Non-Hodgkin's (BCNHL) as their treatment protocols differ. PBLL patients are treated with an acute lymphoblastic leukemia- type therapy which requires a longer maintenance phase than other BCNHL treatments.

Unfortunately, primary cutaneous PBLL has a poorer prognosis as compared to Precursor-T-cell Lymphoblastic Lymphoma. However, if primary cutaneous lymphoblastic lymphoma is caught in the early stages (I or II) and treated aggressively with appropriate systemic chemotherapy, the overall survival rate at 5 years post diagnosis is up to 90% [8]. Since cutaneous PBLL is considered a high-grade neoplasm, it is considered a diagnostic and therapeutic emergency. With early diagnosis and prompt initiation of treatment, the prognosis for these patients can be very good. For this reason, it is important for dermatologists and primary care physicians to have an early suspicion and recognize cutaneous lesions or masses that could possibly represent a lymphoma.

Conclusion

Our case report illustrates an unexpected diagnosis of PBLL in a child presenting with a solitary enlarging head mass initially appreciated following head trauma. This case is also unusual due to a delayed diagnostic workup secondary to her young age and absence of nodal and/or extra nodal findings. This case illustrates the importance of early recognition and suspicion of benign appearing masses as possible oncologic processes in children. This case also demonstrates the effectiveness of a skin biopsy for suspicious skin lesions in children with unusual or unexpected presentations.

Table 1 Stages of lymphoma.

Stage	Diagnosis
I	One lymph node site
II	Multiple sites of lymphoma on one side of the diaphragm
III	Lymphoma on both sides of the diaphragm, may present as a mediastinal mass and/or involve the lungs
IV	Lymphoma spread into the CNS, spleen and/or bone marrow

References

1 Belgaumi AF, Al-Kofide A, Sabbah R, Shalaby L (2005) Precursor B-cell lymphoblastic lymphoma (PBLL) in children: pattern of presentation and outcome. J Egypt Natl Canc Inst 17: 15-19.

2 Kawakami T, Kimura S, Kinoshita A, Kondo K, Soma Y (2009) Precursor B-cell lymphoblastic lymphoma with only cutaneous involvement. Acta Derm Venereol 89: 540-541.

3 Lin P, Jones D, Dorfman DM, Medeiros LJ (2000) Precursor B-cell lymphoblastic lymphoma: a predominantly extranodal tumor with low propensity for leukemic involvement. Am J Surg Pathol 24: 1480-1490.

4 Maitra A, McKenna RW, Weinberg AG, Schneider NR, Kroft SH (2001) Precursor B-cell lymphoblastic lymphoma. A study of nine cases lacking blood and bone marrow involvement and review of the literature. Am J Clin Pathol 115: 868-875.

5 Choi J, Ahn HH, Kye YC, Kim SN (2007) Precursor B-cell Lymphoblastic Lymphoma Presenting as Solitary Plaque in a Child. Acta Dermato Venereologica.

6 Bandyopadhyay R, Bandyopadhyay SK, Dhua D, Roy S (2011) Primary cutaneous precursor B-cell lymphoblastic lymphoma with late dissemination. Singapore Med 52: 258-261.

7 Boccara O, Laloum-Grynberg E, Jeudy G, Aubriot-Lorton MH, Vabres P, et al. (2012) Cutaneous B-cell lymphoblastic lymphoma in children: a rare diagnosis. J Am Acad Dermatol 66: 51-57.

8 Reiter A (2007) Diagnosis and treatment of childhood Non-Hodgkin Lymphoma. Am Soc of Hemat Educ Progr 1: 285-297.