CASE REPORT

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A 9-Year Old Boy with Leukemia and Oral Manifestation: A Case Report and Review of Literature

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Abstract

Background: Childhood leukemia is not a common disease. However, the most common malignant neoplasms in children are the leukemias, and 77% of these cases are acute lymphoblastic leukemia (ALL). **Case Report:** We report a case of precursor B-cell ALL, initially presented by unusual manifestations such as; teeth ache, loose teeth and gum swelling, knee pain and a normal cell blood count (CBC). It was first considered to be Histiocytosis. Several laboratory workups have been performed to rule out the immunologic, infectious and rheumatologic diseases. A bone marrow aspiration revealed the diagnosis of leukemia. However, acute myeloid leukemia is more famous for being accompanied by oral presentation; our patient was afflicted by ALL. The patient was first referred to a dentist for the oral manifestations. **Conclusion:** This case reminds the importance of systemic evaluation of localized signs and symptoms in children and detailed history taking, follow-ups and physical examination.**[GMJ.2016;5(4):215-18]**

Keywords: Leukemia; Acute Lymphoblastic; Oral Manifestations; Histiocytosis; Langerhans-Cell

Introduction

The most common malignant neoplasms in children are the leukemias, and 77% of these cases are acute lymphoblastic leukemia (ALL). In leukemia, the genetic abnormalities in a hematopoietic cell lead to uncontrolled proliferation of cells. The ALL is more prevalent in boys, and its peak incidence is at 2-3 year of age. The initial manifestations of ALL are often nonspecific, such as anorexia, malaise, irritability, fatigue, and low-grade fever. Bone and joint pain especially in the lower limbs and joint swelling may be present [1-3].

Case Presentation

A 9-year old boy of Kurdish origin was brought to our hospital with teeth and jaw ache, gingival swelling, loose teeth and left knee pain. The problems had started from about 40 days ago with pain and swelling in the left knee. He could not bear weight on his left lower limb. The pain of the left knee got worse by activity. The swelling of gum and pain of the chin had started from 3 weeks earlier, and he had a toothache since the 2 weeks ago. The jaw and toothache radiated toward the head.

He felt that all of his teeth had become loose;

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therefore, he could just eat watery foods. On physical examination, both sides of the left knee were found to be tender. The range of motion of the left knee had reduced, and it was painful on both active and passive motions, but it was not warm or erythematous. The other joints were not painful and were normal on examination. The patient did not have weight loss, loss of appetite or skin rashes. The vital signs were stable, and he did not have a fever, nausea or vomiting. On examination of the left eye, he had lateral gaze palsy, and it deviated to the right (which is suggestive of the involvement of the left sixth cranial nerve). He reported having double vision. Funduscopic retinal exam of both eyes and other neurologic examinations were also unremarkable. On oral exam, he had poor oral hygiene, several teeth decays, and swollen gum without bleeding, swollen upper lip and chin (more prominent on the left side) and a subcutaneous mass were palpated on the left maxilla beneath the eye. At both sides of the neck, several small size, mobile, and non-tender lymph nodes were palpated. The 1st and 2nd teeth of upper and lower jaws were examined gently and were found to be loose. The rest of the teeth were also felt to be loose, and these were the reasons for which he was, in the first place, referred to a dentist for the oral complaints. Abdominal exam was not remarkable, and no hepatosplenomegaly was detected. His past medical history had a history of varicella infection about 4 months ago which was cured without complications. Twenty days before coming to our clinic, he had travelled to mountainous rural areas and contracted Herpes Labialis on the lower lip that caused numbness and tingling on the lip and chin, which lasted about 3 days and then chin scaling, pain of the gum and teeth, inflammation of the gum, and loosing of the teeth started in a few days afterwards. The complete blood count (CBC), and inflammatory markers (ESR and CRP) and laboratory work-ups (calcium, AST, ALT) were within nnormal limit.

Considering an unremarkable cell blood count (CBC), and the oral signs and symptoms, the clinicians considered Histiocytosis as the first diagnosis, then malignancies and immunologic disorders as the second possibilities. A radiography of the left knee revealed lytic lesion with periosteal reaction (Figure-1). A three-phase bone scintigraphy showed fusiform uptake in the lateral aspect of the distal physis of the left femur which could be tumoral involvement.

The bone marrow aspiration exam was performed and its result demonstrated hypocellular marrow, with decreased myelopoiesis, erythropoiesis, and megakaryopoiesis, with more than 90% large lymphoblasts with cytoplasmic vacuole that suggested of ALL (Figure-2).

Flow cytometry analysis and morphology of the bone marrow aspiration showed that about 60% of the bone marrow cells were the blast population which were positive for CD45/CD10/CD19/CD20/CD22 and HLA-DR that were consistent with the diagnosis of common precursor B-cell ALL (common ALL).

Chemotherapy for high-risk ALL was performed. The oral presentation (gingival hy-

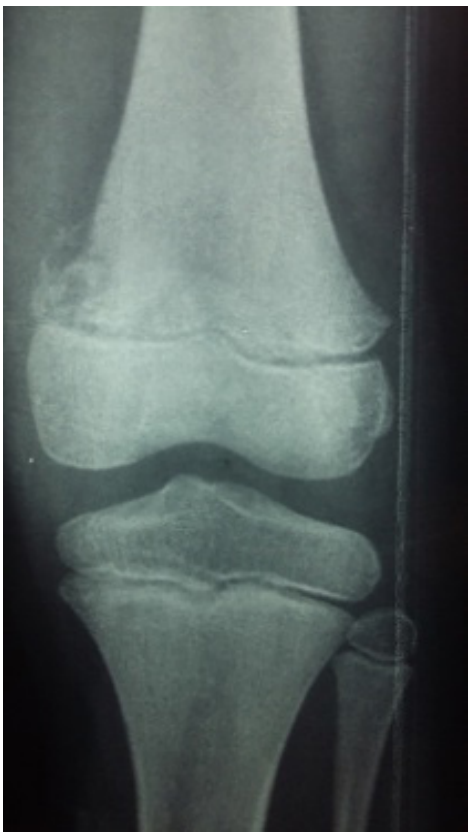

Figure 1. Radiography of the left femur. A lytic lesion with the periosteal reaction on the distal epiphysis of the left femur.

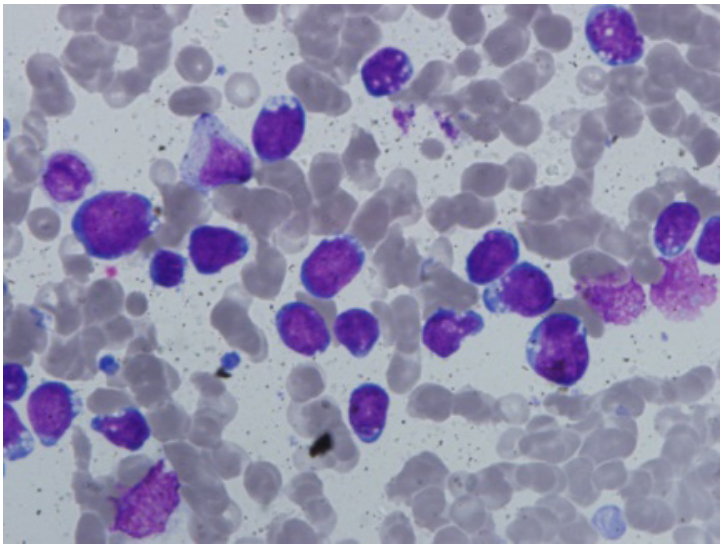

Figure 2. The bone marrow aspiration smear stained by wright giemsa method shows hypocellular marrow, with decreased myelopoiesis, erythropoiesis, megakaryopoiesis, and more than 90% large lymphoblasts with cytoplasmic vacuole that suggested of ALL (original magnification ×100)

perplasia and loose teeth) resolved in a few weeks after the initiation of the treatment.

The chemotherapy regimen with prednisolone, vincristine, daunorubicin, and L-asparaginase started as the induction phase. In about one month (until the end of the induction phase of the chemotherapy), the loose teeth improved, and he began to eat and chew the solid food. After two months, the oral manifestations improved completely. In the seventh day of the induction phase, the conducted bone marrow aspiration indicated less than 10 percent of blast cells; therefore, the patient was considered eligible to be treated on the rapid early response protocol. During the 2-year follow-up, he was completely well and in complete remission.

Discussion

In Langerhans cell histiocytosis (LCH), oral involvement could be the first and sometimes the only affected site. Oral manifestation of LCH may include gingival ulceration and inflammation and floating teeth. Maxilla and mandible are the commonly affected bones, and solitary intraosseous lesions may cause facial swelling [1]. The LCH in all children with jaw pain or swelling and/or loose teeth should be considered [2].

Gingival hyperplasia is commonly seen in acute leukemia which can be localized or generalized and caused by inflammation or leukemic infiltration. It mainly affects the interdental papillae and the marginal gingiva. While it is less frequent in chronic leukemia [3]. Calcium channel blockers, cyclosporine, anticonvulsants, hereditary gingival fibromatosis, chronic inflammation and leukemic infiltration, may lead to gingival hypertrophy [4].

Throughout the course of leukemia, periodontal lesions are common. The most common signs and symptoms of the oral cavity in leukemia included gingival hemorrhage, petechiae, ecchymoses, infections, mucosal ulcers, and gingival enlargement. Poor oral hygiene, neoplasia, and systemic diseases may cause gingival hyperplasia [5]. Gingival enlargement has been reported as an early sign of AML [6]. Gingival hyperplasia is mostly seen in AML, commonly in acute monocytic leukemia (M5) and acute myelocytic leukemia (M1, M2) [7]. In the AML, gingival hyperplasia and leukemic infiltration could be aggravated with pre-existing periodontal disease [8]. Manifestations of bone marrow failure such as pallor, fatigability, bruising, and epistaxis will occur in the course of the illness. Lymphadenopathy, cranial nerve neuropathy, hepa-

tosplenomegaly, headache, seizure, testicular enlargement are caused by organ infiltration. An enlarged mediastinal node may compress the airways and lead to respiratory distress. The AML causes secondary to bone marrow failure and may present with manifestations that are not common in ALL such as distinct masses (chloroma or granulocytic sarcomas), infiltration of gingiva, subcutaneous nodules or disseminated intravascular coagulation [9]. Leukemia treatment in ALL may commonly lead to dental problems which need close follow-up and care [10].

Because of the initial oral presentation in our patient, such as teeth ache, loose teeth and gum erythema and swelling he was first referred to the dentist office. He also had knee pain, which has not initially grabbed the proper attention. Later he was referred to the pediatric department, and hematology assessment performed, and a CBC provoked the doctors to consider Histiocytosis as one of the probable diagnoses. A bone marrow exam proved the diagnosis of leukemia. However, AML is more famous for being accompanied by an oral presentation, our patient was afflicted by ALL, which is an unusual presentation in ALL.

Conclusion

This case reminds the importance of systemic evaluation of localized signs and symptoms in children and detailed history taking, follow-ups and physical examination. The first clue to the diagnosis in this patient was the presence of bone pain accompanied by the oral manifestations which with further investigation proved to be leukemia.

Conflict of Interest

The authors declare no conflicts of interest in preparing this manuscript.

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