CASE REPORT

Parotid Gland Enlargement as a Presenting Manifestation of Acute Lymphoblastic Leukemia

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Abstract

A young female presented with symmetric polyarthritis, generalized lymphadenopathy, hepato-splenomegaly and bilateral parotid gland enlargement without sicca symptoms. A second case of 15 months old child presented with short duration pyrexia with generalized lymphadenopathy, hepato-splenomegaly and bilateral parotid gland enlargement. Both the patients had out of proportion anemia on examination. Investigations confirmed CD 10+ B-cell acute lymphoblastic leukemia (ALL) in both the cases. Fine needle aspiration cytology of parotid glands in both the cases showed infiltration by lymphoblasts. We propose that ALL should be included in the differential diagnosis of bilateral parotid gland enlargement especially if associated with lymphadenopathy and hepato-splenomegaly.

Key Words

Acute lymphoblastic leukemia, Parotid gland, Arthritis, Mumps

Introduction

Parotid gland involvement in acute myeloid leukemia has been reported rarely as an uncommon presenting manifestation (1), or relapse of disease (2,3). However parotid gland enlargement as a presenting manifestation of acute lymphoblastic leukemia (ALL) is very rare (4). Herein two cases of ALL with parotid gland enlargement as the presenting manifestation are reported.

Case-1

A 23-year-old female presented with bilateral symmetrical inflammatory arthritis involving wrist, metacarpo-phalangeal, elbow, knee and ankle joints for 3 months, bilateral parotid gland enlargement, menorrhagia and symptomatic anemia for 1 month. Examination revealed severe pallor, cervical lymphadenopathy, hepato-splenomegaly and bilateral parotid gland enlargement. Musculo-skeletal examination revealed synovitis of both the wrists and knee joints. Bone tenderness was negative. Investigations revealed hemoglobin (10.3 gm/dl), total leukocyte count (13,000/mm^3) with 85% lymphoblasts and platelet count (65,000/mm^3). Renal and liver function tests were normal with a mildly raised lactate dehydrogenase-(LDH) of 512 U/L (normal up to 450 U/L). Bone marrow examination confirmed ALL. Immunophenotyping of the blasts revealed CD 10+ precursor B-cell ALL. Cerebro-spinal fluid cytology for malignant cells was negative. Fine needle aspiration cytology of both the parotid glands revealed infiltration by lymphoblasts.

Case-2

A fifteen month boy presented with moderate grade fever and bilateral parotid gland enlargement of 10 days duration. Examination revealed severe pallor, generalized lymphadenopathy, hepato-splenomegaly and bilateral parotid gland enlargement. Investigations revealed hemoglobin 4.6 gm/dl, total leukocyte counts of 42,500/mm^3 with 67% lymphoblasts, platelet count of 1,10000/mm^3, normal renal and liver function tests and markedly

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elevated LDH (2179 U/L). Bone marrow examination confirmed ALL. Immunophenotyping of the lymphoblasts revealed CD 10+ precursor B-cell ALL. Cerebro-spinal fluid cytology for malignant cells was negative. Fine needle aspiration of both the parotid glands revealed infiltration by lymphoblasts (Fig . 1).

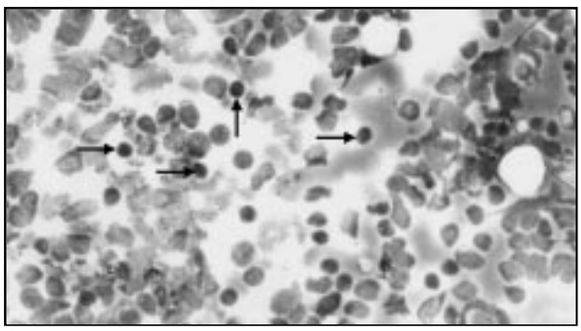

Fig. 1 Fine needle aspiration smear of parotid gland showing diffuse infiltration by lymphoblasts (Black Arrows). (H&E X 400)

Discussion

Our first patient was misdiagnosed as rheumatoid arthritis and had received NSAIDs, prednisolone and methotrexate for it. The other differential diagnosis in such a case could be primary Sjogren's syndrome, but the absence of sicca symptoms ruled out that possibility. Fukuzawa et al (4), had described a 74 years old male with dryness of oral cavity and parotid gland enlargement as the presenting manifestation of adult T-cell leukemia and proposed that adult T-cell leukemia should be included in the differential diagnosis of such a presentation. Our second patient was misdiagnosed as mumps. However, the presence of leukocytosis and out of proportion anemia was suggestive of acute leukemia.

Parotid gland involvement in ALL has been reported as post-chemotherapy second malignancy (5). However, infiltration of parotid gland by lymphoblasts is distinctly uncommon at presentation. In both the cases, there was infiltration of the parotid gland by CD10+B-cell lymphoblasts, which (to the best of our knowledge) is not reported in literature. Parotid gland has been reported to be a sanctuary site for relapse of acute myeloid leukemia either in isolation (3), or in association with central nervous system (2).

The significance of parotid infiltration by lymphoblasts is not clear. It also remains unanswered whether parotid gland involvement in ALL warrants aggressive local or systemic therapy or signifies a risk for second malignancy or may later on act as a sanctuary site. Unfortunately, both of our patients did not opt for therapy due to financial constraints and a chance to evaluate the course of ALL with parotid gland involvement was lost.

Both of these cases highlight a unique clinical presentation of ALL. We conclude that ALL should be included in the differential diagnoses of parotid gland enlargement in both children and adults.

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