**ABSTRACT**

The more common causes of parotid enlargement in children are infections and inflammatory conditions. Primary neoplasms of the parotid glands are rare in pediatric age group; however, secondary malignancies have been reported in survivors of childhood leukemia. The parotid glands have been the sites of relapses in acute lymphoblastic leukemia (ALL) and acute myeloid leukemia. However, bilateral parotid involvement as an initial presentation of ALL is rarely reported. We present a case of an 8-year-old boy who presented with bilateral parotid enlargement as an initial manifestation of ALL. General physicians should be aware of this extramedullary presentation of acute leukemia when they see a child presenting with organomegaly or abnormal blood counts.

**Keywords:** Children, leukemia, parotid enlargement

**INTRODUCTION**

Parotitis is a common symptom seen among pediatric population, with infection and inflammation being the commoner etiologies. Pallor, bone pain, bleeding manifestations, lymphadenopathy and hepatosplenomegaly are the well known presenting features of leukemia, that physicians are usually familiar with. Parotidomegaly as the presenting feature of Acute leukemia is rarely reported. Hence, we need the physicians to be aware of this unusual presentation of a common pediatric malignancy, when they encounter an abnormal blood counts picture. Diagnostic delays can be avoided with the knowledge of identifying an unusual marker of leukemias.

**CASE REPORT**

An 8-year-old boy presented with a 2-week history of abdominal distention and bilateral swelling of the parotids for 1 week. The parotid swelling was not painful and was not associated with difficulty in opening the mouth or mastication. The child had no complaints of dryness of mouth or eyes before the development of swelling nor previous history of similar swellings in the past.

There was no history of fever, gum bleeds, skin bleeds, bone pains, or joint pains. He was fully immunized for his age. On examination, he was found to be pale. No lymph nodes were palpable, bilateral diffuse; nontender swelling was present in the pre- and infra-auricular region, lifting the lower lobule of the ear. Liver was palpable 6 cm below the right costal margin and spleen was palpable 3 cm below the left costal margin. Testis was normal. Central nervous system (CNS) examination

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did not reveal any abnormality. Hemogram revealed hemoglobin of 6.9 g/dl, white blood cell count of 14.2 × 10^9/L, and platelet of 30 × 10^9/L. Mumps serology was negative. Peripheral blood showed 10% neutrophils, 75% lymphocytes, 20% lymphoblasts, 7% monocytes, and 2% eosinophil. In view of bicytopenia and peripheral blood showing atypical lymphocytes with hepatosplenomegaly, bone marrow aspiration was performed which revealed 68% lymphoblasts, and immunohistochemistry was conclusive of common acute lymphoblastic leukemia antigen (CALLA)-positive pre-B-cell acute lymphoblastic leukemia (ALL).

Karyotyping from bone marrow and peripheral blood fluorescence in situ hybridization for BCR ABL, MLL, TEL acute myeloid leukemia (AML), and t(1:19) was normal. Cerebrospinal fluid analysis showed no malignant cells. The child was started on the Children’s Oncology Group ALL protocol and parotid swelling subsided within 72 h of starting the steroids. The bone marrow study for minimal residual disease done at the end of induction period was negative. The child is currently in consolidation phase of chemotherapy and doing well.

**DISCUSSION**

The common causes of bilateral parotid swelling in children are of infective etiology, followed by inflammatory conditions. Mumps being most common, followed by *Staphylococcus aureus*, *Cytomegalovirus*, Epstein–Barr virus, coxsackievirus, tuberculosis, and human immunodeficiency virus. The other less common systemic causes are Sjogren’s syndrome, Wegener’s granulomatosis, and sarcoidosis. Chronic sialadenitis, Juvenile recurrent parotitis, and Kimura’s disease should also be considered in recurrent and chronic cases of parotid enlargement. Salivary gland involvement in children with HIV is well recognized and the gland is firm, nontender, and is usually asymptomatic. Parotid neoplasms are rare in pediatric population.

Bilateral parotid swelling as an initial manifestation of acute leukemia is very rarely reported. Although the occurrence of secondary malignancy in the parotid or salivary glands after chemotherapy and radiotherapy treatment for ALL or AML is well reported, parotidomegaly as an initial manifestation in ALL is rare.[1,2] Biswas et al. in their study about the uncommon clinical features of acute leukemia in West Bengal identified only one child with parotidomegaly among the 75 cases.[7] Ünal et al. have reported a case of a 3-year-old girl who had presented with bilateral swelling of parotids and dactylitis, was serologically positive for mumps IgM, and later diagnosed to have ALL with CNS involvement, in view of persistent symptoms.[8]

In the largest case series of four children who presented with parotid enlargement as initial presentation of acute leukemia reported by Kulkarni and Marwaha, only one had bilateral parotidomegaly. All the four children had massive hepatosplenomegaly and lymphadenopathy at diagnosis. None of them had CNS involvement though one of the children who took treatment had a combined bone marrow and CNS relapse after 18 months of treatment. It has been proposed that parotid involvement could be the result of bulky extramedullary disease. Diagnostic delays of more than 1 month have been reported when children present with parotid swelling as the initial manifestation of acute leukemia.[9]

Chloromas, gum hypertrophy, and granulocytic sarcoma are recognized in 5% of acute myelomonocytic leukemia and monoblastic leukemia, but exocrine gland involvement in the form of parotid involvement has been reported only in two pediatric cases as of now.[10] The recent reporting of bilateral parotid enlargement at ALL presentation is in a 6-month-old girl with infant ALL who had bilateral parotid swelling and bilateral nephromegaly at presentation.[11]

Till date, all the children who presented with parotidomegaly in ALL had CALLA-positive B-cell ALL.

Although Naithani and Mahapatra and Saha et al. had reported magnetic resonance imaging/biopsy of the parotid gland to confirm tumor infiltration, in our case, as the peripheral smear showed 20% blasts and bone marrow aspirate was conclusive of ALL, no further studies were done to confirm the same. The complete resolution of the parotid enlargement within 72 h of starting the steroids also was confirmative of the leukemic infiltration of the glands.

Only Naithani and Mahapatra have reported facial nerve involvement with parotid enlargement as presenting features of acute leukemia.[10] As parotid could be a sanctuary site, larger studies and longer follow-up are needed to evaluate its prognostic significance.
CONCLUSION

Unusually prolonged swelling of the parotid glands with subtle changes in the blood parameters should raise a high index of suspicion among the physicians and should rule out the noninfectious cause for the same. Awareness among general practitioners about such an unusual presentation would help in avoiding the diagnostic delays.

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Conflicts of interest
There are no conflicts of interest.

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