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A pediatric case of acute lymphoblastic Leukemia presenting as a skull mass

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ABSTRACT **Background:** Acute lymphoblastic leukemia (ALL) is a clonal hematologic disorder and comprises a group of lymphoid neoplasms that morphologically and immunophenotypically resemble B-lineage and T-lineage Precursor cells. Although it is not uncommon for acute myelogenous leukemia to manifest initially as an extramedullary myeloblastic tumors, so-called myelosarcomas. Thus the aim of this report is to present for the first time ever a case of precursor B-cell (pre-B-cell) ALL that manifested as skull mass. **Case presentation:** A five year old boy presented with a two months history of swelling in the right neck that's increasing with time and proceeded with skull swelling by almost three months. Physical Examination showed multiple right sided cervical Lymph nodes enlargement, non tender slightly mobile and scalp swelling firm to hard, non-mobile and abdomen examination revealed neither Hepatosplenomegaly nor other masses. **Conclusions:** This is a rare case of pre–B-cell ALL presenting initially as a skull mass, It is extremely rare for ALL to present in this manner, in addition to that this case explores that leukemic infiltrate should be considered when the initial mass is getting larger with time.

KEYWORDS : ALL, Child, skull mass, case report

Background:

Acute lymphoblastic leukemia (ALL) is a clonal hematologic disorder and comprises a group of lymphoid neoplasms that morphologically and immunophenotypically resemble B-lineage and T-lineage Precursor cells [1]. It evolves through uncontrolled proliferation and damaged differentiation of leukemic blasts that compromise the normal hematopoiesis. Thus patients usually present with symptoms resulting from bone marrow failure. The most common symptoms include fever (caused by leukemia or a secondary infection secondary to neutropenia), fatigue and lethargy (as a result of anemia), bone and joint pain, and a bleeding diathesis (related to thrombocytopenia). Patients with precursor T-cell ALL often present with a mediastinal mass with or without associated pleural effusions, which may lead to respiratory distress and other signs of superior vena cava syndrome.[1] Common extra medullary sites of involvement include lymph nodes, liver, spleen, and meninges, whereas less commonly, ALL may infiltrate orbital tissues, testes, tonsils, and adenoids.[6]

The clinical onset of ALL is most often acute, although a small percentage of cases may evolve insidiously over several months. [1] ALLs represent 75% of all acute leukemia (which in turn represent34% of all cancers in this age group), with a peak incidence at 2 to 5 years of age.[2]

A sharp peak in ALL incidence is observed among children aged 2 to 3 years (>90 cases per 1 million per year), with rates decreasing to fewer than 30 cases per 1 million by age 8 years.[3,4]

Prognosis and management of patients with ALL depends on many factors, including cell lineage (i.e., pre-B cell, or T cell), age, presence of genetic mutations, and central nervous system involvement.[8]

Case Report:

This is a case of five year old boy without any significant past history who presented with a two months history of swelling in the right neck that's increasing with time, preceded with skull swelling by almost 3 months. However, parents claim that the skull swelling didn't show significant increase in size and denied any prior history of trauma or any other systemic disease. Physical examination showed multiple rights sided cervical Lymph nodes enlargement, around 3cmx3cm, firm, and non tender slightly mobile and scalp swelling measuring around 5cmX 3cm firm to hard, non-mobile, non tender, abdomen examination revealed neither Hepatospl enomegaly nor other masses.

Imaging:

CT head with contrast, showed 3 cm x1 cm right frontal vertex subcutaneous soft tissue swelling extends to skin without fracture or bone changes and no intracranial extension. Neck Ultrasound showed, bilateral lymph node enlargements. The largest one on the right side measures 1.6cm in transverse diameter and on the left side measures 1cm in transverse diameter. Additionally,

complete blood count and differential was completely normal. Excisional biopsy of the enlarged lymph node and skin biopsy was done and sent for histopathology. Right neck lymph node and scalp showed B-cell lymphoblastic lymphoma/leukaemia.

Section of lymph nodes showed total effacement of the nodal architecture by medium-sized atypical lymphoid cells with convoluted nuclei, condensed nuclear chromatin and indistinct nucleoli. Also the skin section showed the same neoplastic cells diffusely infiltrating the dermis and sparing the epidermis. These atypical cells are positive for terminal deoxynucleotidyl transferase (TdT) , CD-10, CD-19, and CD-34 . They were negative for gamma and kappa light chains, CD-13, and *c-myconcogene*.

Bone Marrow Aspirate Differential Count: Blasts 51%; Pr omy elocytes 3%; Myelocytes 4%; Metamyelocytes 7%; Band Neutrophils 3%; Neutrophils 13%; Monocytes 0%; Lymphocyes 0%, Atypical Lymphocytes 0%, Eosinophils 2%, Basophils less than 14.8:%, Erythroids 17%.

The Flow cytometry findings of the Cervical Lymph Node biopsy and flow cytometry findings of Bone (Marrow Aspirate); and the Bone Marrow Morphology findings are Concordant. The picture, entirely taken together, is consistent with Precursor B-Acute Lymphoblastic Leukaemia.

Lumber puncture was negative for malignant cells :Molecular studies were indicative for the presence of a hyper diploid karyotype in up to 49% of cells .The FISH signal patterns indicate the presence of trisomy 4,tetrasomy 4, trisomy 9, trisomy 17 and trisomy 21, however cytogenetics studies didn't reveal any of the poor prognostic translocations. Based on these data standard -risk treatment plan was initiated by induction phase for 4 weeks which include 3 drugs, dexamethasone (10 mg/m2/day for 4 weeks, vincristine (1.5 mg/m2/dose every week for 4 weeks, peg-aspariginase (2500 units/m2/dose once on day 5in addition to that, during this phase the patient also received 2 doses of intrathecal methotrexate and one dose of intrathecal cytarabine as prophylaxis

for central nervous system. Four weeks after treatment bone marrow aspirate examination demonstrated complete remission of the leukemia.Subsequently the patient continues on treatment as planned for standard risk ALL.

Discussion and Conclusions:

This is a rare case of pre–B-cell ALL presenting initially as a skull mass It is not unusual for patient with Acute Lymphoblastic Leukaemia to present with Lymphadenopathy and hepatosplenomegaly, moreover, in about 5% of patients with B-precursor ALL, bone pain, arthritis, and limping are the only symptoms, leading to delays in diagnosis. [8]Though in Acute myeloid leukemia where extramedullary myeloid cell tumor known as a granulocytic sarcoma or chloroma have been described rarely in literature as the presenting symptoms of AML with no evidence of leukemia, either in the peripheral blood or in bone marrow . [12]In Acute lymphoblastic Leukemia the extramedullary mass seldom described as the presenting symptoms, aside from few case report of orbital mass[9,10].unusual presentation of Acute lymphoblastic leukemia has been reported in the literature as described in one case report of Precursor B-cell acute lymphoblastic leukemia presenting as obstructive jaundice and Aplastic Anemia. [11, 8]The patient responded well to the risk-stratified treatment, this response to treatment seems to be similar to B-cell ALL presentingwith Peripheral or bone marrow infiltration manifestation, which has an overall cure rate approaching 80% with modern chemotherapeutic regimens.[2]

It is extremely rare for ALL to present in this manner, moreover, this case explores that leukemic infiltrate should be considered when the initial mass is getting larger with time.

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