ejpmr, 2019,6(3), 374-376

SJIF Impact Factor 4.897

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 2394-3211

EJPMR

FRONTAL MASS AS AN UNUSUAL PRESENTING SYMPTOM IN ACUTE LYMPHOBLASTIC LEUKEMIA

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Article Received on 25/12/2018 **Article Revised on 15/01/2019** **Article Accepted on 04/01/2019**

ABSTRACT

We report a case of a child presented with swelling in the right frontal region, which was diagnosed as small round cell tumour on cytology. Further work up demonstrate presence of lymphoblasts on peripheral blood film & bone marrow. Immunophenotyping was done by flowcytometry, and that confirm a diagnosis of B-ALL. Blasts were CD34+ Tdt+ HLADR+ CD79+ CD 19+ on immunophenotyping. So the final diagnosis precursor B-ALL with extra medullary involvement of right frontal region was entertained.

KEYWORDS: Lymphoblast, frontal mass, flowcytometry, cytology, leukemia.

INTRODUCTION

Acute lymphoblastic leukemia (ALL) is the most common malignancy diagnosed in pediatric age group, representing 25% of all pediatric cancer. B-lymphoblastic leukemia/lymphoblastic lymphoma is a neoplasm of precursor cells that are committeed to the B-cell lineage. Common symptoms and signs are a consequence of bone marrow failure or the involvement of medullary or extramedullary sites by leukemic cells. The primay diagnosis of ALL based on the demonstration of lymphoblasts in pheriphral blood film and bone marrow. For further, categorization of leukemia into B or T lineage, immuophenotyping is mandatory.

CASE REPORT

A 7 yr old male child presented to surgery department with a frontal mass which his parents noticed after trivial head injury. The mass was painless but gradually increasing in size. Local examination revealed a well-defined 2×1 cm mass in right frontal region, firm in consistency, non pulsatile, non tender. The skin over the swelling was stretched but was otherwise normal. On blood counts and haemoglobin were within normal limits. Chest x-ray was normal. CT head was done &reveal chronic extradural haemorrhage along right frontal curvature with fracture of right side of frontal bone. Clinical diagnosis of head injury followed by chronic extradural haematoma was made & patient was treated conservatively. But the frontal swelling progressive in nature & reached upto size of 4×3×2 cm in one month duration. FNAC was done from the forehead mass in surgery OPD. Meanwhile patient also developed paraplegia. MRI brain was advised & reveal large lobulated hyperdense soft tissue attenuation mass lesion in right frontal region with extracranial & intracranial extraaxial component compressing underlying neuroparenchyma with left sided midline shift of 2.5 mm. Erosion and partial destruction of inner/outer table of involved frontal bone. Radiological features were suggestive of neoplastic etiology.

FNAC smears prepared from frontal swelling were stained with Leishman & H/E stain. Smears were cellular comprising of predominant population of dispersed medium to large atypical cells having scant amount of cytoplasm with high N/C ratio, irregular hyperchromatic nuclei & no discernible nucleoli in a haemmorhagic background. Cytological features are suggestive of malignant round cell tumour favouring lymphoma/leukemia over others.

Simultaneously, peripheral blood film was prepared which showed total leucocyte count 70,000/cumm with differentiated count showing >90% blasts, platelet count was 40,000 with dimorphic picture. A diagnosis of acute leukemia favouring acute lymphoblastic leukemia was entertained. Patient was started on chemotherapy & subsequently sample was submitted for bone marrow examination & flow cytometry, that confirmed the diagnosis of B-ALL. Blasts were CD34+ Tdt+ HLADR+

CD79+ CD 19+ on immunophenotyping. So the final diagnosis was precursor B-ALL with extra medullary involvement of rightfrontal region.

Figure 1: Seven year male child presents with a swelling in frontal region.

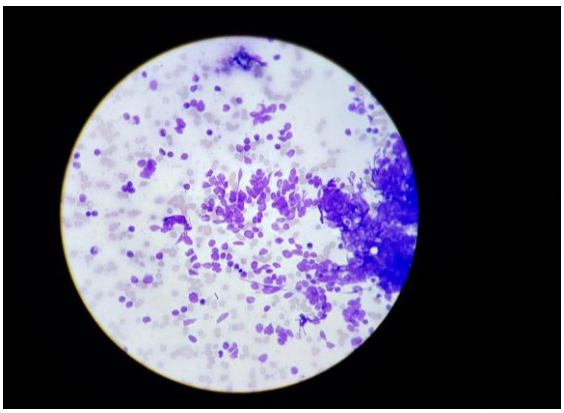

Figure 2: FNAC smears prepared from frontal swelling were cellular comprised of medium sized atypical cells having scant amount of cytoplasm, high n/c ratio, round to oval nuclei with homogenous chromatin pattern.(lesihman stain 400x).

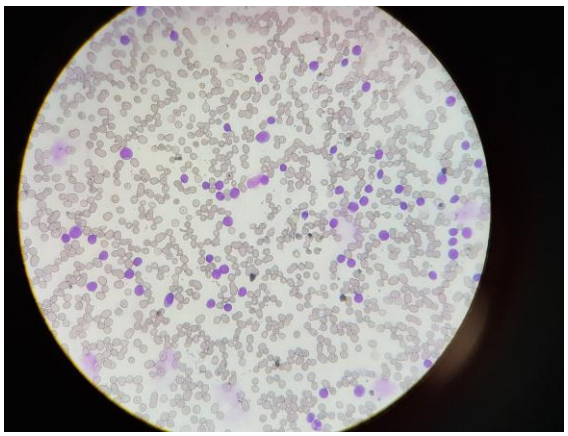

Figure 3: PBF showed predominant population of blast (>90%) conforming to morphology of lymphoblasts (lesihman stained 400x).

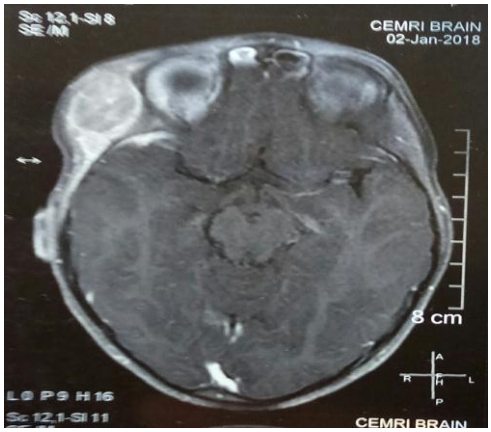

Figure 4: MRI brain showed large lobulated hyperdense soft tissue mass lesion in right frontal region with extracranial and intracranial component compressing underlying neuroparenchyma with left sided midline shift.

DISCUSSION

Precursor B-ALL is the most common of the immunophenotypic categories, that accounts for about 85% of cases in children & 75% in adults. Onset may be insidious and slowly progressive over weeks to months, or acute and explosive. As in this case, intially patient symptoms were gradual in onset but subsequently turns into acute phase. The common presenting signs and symptoms are related to decreased blood counts resulting from bone marrow failure. Generally, patient complaints include fatigue, lethargy, persistant fever, bruising or bleeding, bone and joint pain. Physical findings may include pallor, ecchymoses or petechiae, lymphadenopathy & organomegaly. Only minority of patients present with clinical symptoms which are caused by extra-medullary leukemic infiltrates. Lymph nodes, CNS, skin, gonadal, renal, bone & joint are major sites of extramedullary involvement. In CNS, leukemic blasts can involve the arachnoid, cranial & peripheral nerve roots & brain parenchyma. Intraparenchymal & meningeal mass lesions & haemorrhages can occur in the setting of a blast crisis of more than 3,00,000/cumm leucocyte count. Also, skin involvement is rare, when it occurs, it is associated with a pre-B cell phenotype.

Laboratory features reveal raised white blood cell (WBC) count in 60% of patients with frequent neutropenia. The blood & marrow may be involved at the time of initial diagnosis & the designation of a case as lymphoblastic lymphoma or acute lymphoblastic leukemia is arbitrary.

The diagnosis of ALL is based on the demonstraton of lymphoblasts in the bone marrow. By WHO crieteria, the minimum number of lymphoblasts required for diagnosis is set at 20%. Occasionally, no lymphoblasts can be identified with certainty in blood, despite extensive bone marrow involvement (i.e., aleukemic leukemia). Morphologically, lymphoblasts in ALL are small to medium sized having scanty cytoplasm with high N/C

ratio. Nucleus is generally round to oval & chromatin is quite homogenous. In most cases, nucleoli are small & indistinct or not visualized. Sometimes blood & marrow may contain little or no evidence of involovement, in such cases, diagnosis of precursor B-ALL is preferred.

Occasionally, solid tumors that involve the bone marrow may be confused with lymphoblasts. In pediatric age group, this confusion occurs most frequenty with neuroblastoma, wilms tumor, retinoblastoma. Although tumor cells tend to occur in clumps, they may replace marrow elements diffusely. To differentiate from leukemia immunohistochemistry, immunophenotyping, cytogenetic analysis and electron microscopy required.

Immunophenotyping should be performed in all cases of ALL to differentiate it from AML & to categorize as precursor B & T-ALL. Lymphoblasts of B-ALL express various combinations of CD 19, CD22, CD 79a, CD24, CD10 & CD9 & several lineage-non specific antigens including CD34, TDt, HLADR, CD38 & CD45 as in our case.

More than 95% of cases of precursor B-ALL are Tdt positive. Tdt is useful in differentiating ALL from lymphoproliferative disorders of mature lymphocytes, which are Tdt-negative, but more than 90% of precursor T-ALL & 5% to 10% of AML also express Tdt.

CONCLUSION

Extramedullary presentation of B-ALL is not an infrequent finding & they might present as initial manifestation of the disease. High index of suspicion & awareness is needed in diagnosing this entity. FNAC examination is important because it may help in determining the site of primary tumour in many cases. Also, it limits the number of organs for investigation of the primary tumour& helps tremendously in initiating primary treatment.

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