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CASE REPORT

Clinical Features and Prognosis Analysis of Acute Myeloid Leukemia in Children with DEK-CAN-Positive

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SUMMARY

Background: **The goal of the study is to investigate the clinical characteristics and prognostic analysis of acute myeloid leukemia in children with positive DEK-CAN fusion gene.**
Methods: **The clinical characteristics and prognostic analysis methods of a case of acute myeloid leukemia in children with positive DEK-CAN fusion gene were retrospectively analyzed, and the domestic and international literature was reviewed.**
Results: **The patient is a girl, 11 years old. The clinical diagnosis was acute myeloid leukemia M2a. Liver full, spleen large, a few small lymph nodes in bilateral axilla and retroperitoneum. Blood routine: WBC: 34.49 x 10^9/L, RBC: 0.91 x 10^{12}/L, Hb: 30g/L, Plt: 26 x 10^9/L, the proportion of leukocyte classification granulocyte was significantly increased, and the primitive naive granulocyte accounted for 56%. Bone marrow smear: primitive naive myelodysplasia, 74% of myeloid original cells, large cell body, moderate plasma volume, round or irregular nuclei, fine nuclear chromatin, visible nucleoli. Peroxidase (MPO) staining: positive. Immunophenotypic expression of antigens CD117, HLA-DR, CD13, CD33, CD123, partial expression of antigens CD34, CD38, abnormal myeloid original cells. Patients tested by fusion gene were positive for DEK-CAN with FLT3-ITD mutation. The clinical diagnosis was acute myeloid leukemia M2a. Chromosome karyotype analysis showed no split phase. The IA regimen, FLAG regimen, and HIA regimen were given successively, and no remission was achieved.**
Conclusions: **Patients with DEK-CAN fusion gene positive AML have a very poor prognosis, low primary induced remission rate, and high mortality. For confirmed cases, patients in remission with chemotherapy should undergo allogeneic hematopoietic stem cell transplantation as soon as possible to have a chance of long-term survival.**
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KEYWORDS

acute myeloid leukemia, DEK-CAN fusion gene positive, children, FLT3-ITD mutation

INTRODUCTION

Acute myeloid leukemia (AML) is a malignant proliferative disease of the hematopoietic system, which is related to fusion gene formation. Its molecular biological characteristics are of great significance for the diagnosis, treatment and prognosis evaluation of the disease. Among them, the DEK-CAN fusion gene produced by chromosome t (6;9)(p23;q34) translocation occurs in approximately 1% - 2% of children with AML [1]. This type of AML has early recurrence and poor prognosis

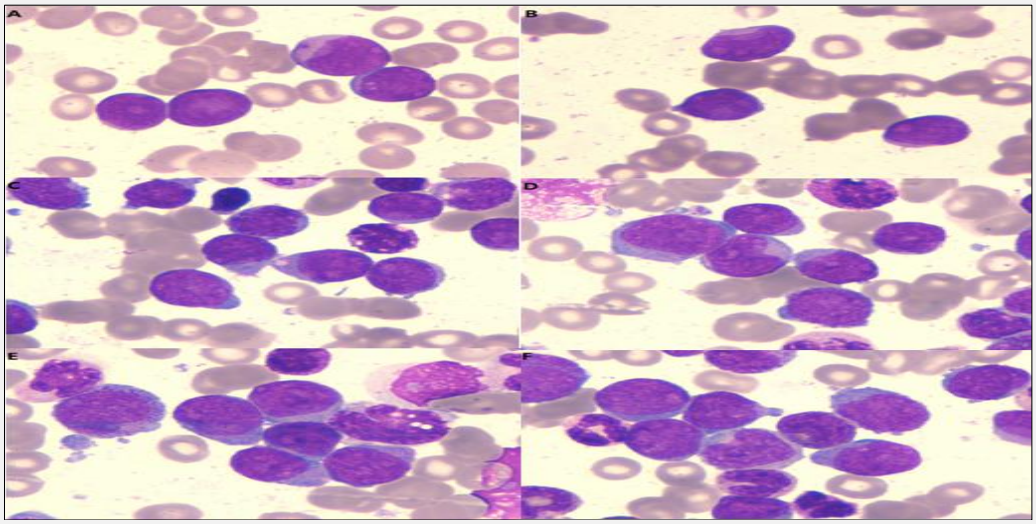

Figure1. (A - B) Blood smears (Giemsa stain 10*100). (C - F) Bone marrow images (Giemsa stain 10*100).

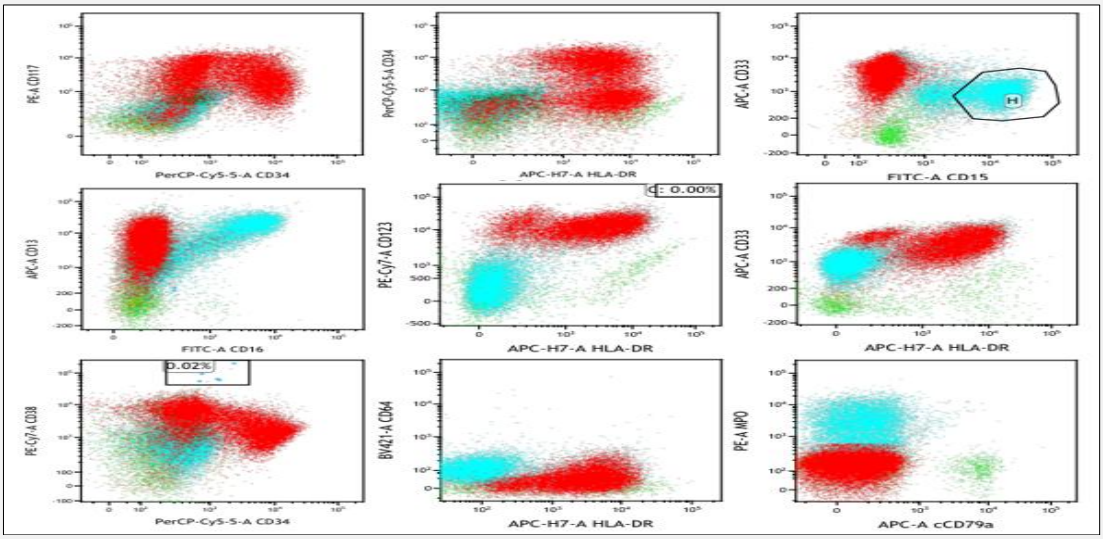

Figure 2. Flow cytometry: Immunophenotypic expression of antigens CD117, HLA-DR, CD13, CD33, CD123, partial expression of antigens CD34, CD38, abnormal myeloid original cells.

and is classified into a separate clinical subtype [2,3].

CASE REPORT

The patient is a girl, 11 years old. Main cause was pallor and fatigue for 7 days and abnormal blood routine for 1 day. Liver full, spleen large, a few small lymph nodes in bilateral axilla and retroperitoneum. Physical examination: Body temperature 37.9℃, pulse 110 beats/minute, breathing 22 times/minute, clear, poor spirit, anemic appearance, old large ecchymosis visible in both lower limbs, stable breathing, pale lips, pharyngeal congestion, no enlargement of tonsils. Laboratory examination: C-reactive protein 36.2 mg/L, D-dimer 1.82 mg/L, prothrombin time 15.70 second, fibrinogen 1.32 g/L. Blood routine: WBC: 34.49 x 10^9/L, RBC: 0.91 x 10^{12}/L, Hb: 30 g/L, Plt: 26 x 10^9/L, the proportion of leukocyte classification granulocyte was significantly increased, and the primitive naive accounted for 56%. Bone marrow smear: primitive naive myelodysplasia, 74% of myeloid original cells, large cell body, moderate plasma volume, round or irregular nuclei, fine nuclear chromatin, visible nucleoli (Figure 1). Peroxidase (MPO) staining: positive. Immunophenotypic expression of antigens CD117, HLA-DR, CD13, CD33, CD123, partial expression of antigens CD34, CD38, not expressing MPO, TDT, cCD3, cCD79a and other myeloid and lymphoid markers, abnormal myeloid original cells (Figure 2). Patients tested by fusion gene were positive for DEK-CAN with FLT3-ITD mutation. Chromosome karyotype analysis showed no split phase. The IA regimen, FLAG regimen and HIA regimen were given successively, and no remission was achieved.

DISCUSSION

DEK-CAN fusion gene positive AML is a relatively rare type in clinical practice. It has the following characteristics: the onset age is younger, the median age is 25 - 30 years old, and the incidence rate is not different between men and women [4,5]. According to FAB classification, chromosomal translocations mainly occurred in AML (M1, M2 or M4), and the incidence was 0.5% to 4.0%. Most patients did not have splenomegaly or lymph node enlargement. The proportion of primitive cells in peripheral blood and bone marrow, white blood cell count and platelet count were not different between children and adults, and the peripheral blood hemoglobin of children was lower than that of adults. Of the patients, 44% had basophilic granulocytosis in the bone marrow, most of which were accompanied by the presence of Auer bodies, and 67% of the patients had pathological hematopoiesis in the bone marrow [4]. Immunotyping often expressed CD13, CD33, CD34, CD38, CD45, HLA-DR, and TdT, and about 70% with (t6;9) (p23;q34) FLT3 mutations are present in patients with abnormal AML [6,7]. In this case, the patient is a girl, 11 years old. The clinical diagnosis was acute myeloid leukemia M2a. with liver full, spleen large, a few small lymph nodes in bilateral axilla and retroperitoneum. Blood routine: WBC: 34.49 x 10^9/L, RBC: 0.91 x 10^{12}/L, Hb: 30 g/L, Plt: 26 x 10^9/L, the proportion of leukocyte classification granulocyte was significantly increased, and the primitive naive accounted for 56%. Bone marrow smear: primitive naive myelodysplasia, 74% of myeloid original cells, large cell body, moderate plasma volume, round or irregular nuclei, fine nuclear chromatin, visible nucleoli. There was no pathological hematopoiesis, no increase in basophils, and no Auer bodies. Immunophenotypic expression of antigens CD-117, HLA-DR, CD13, CD33, CD123, partial expression of antigens CD34, CD38, abnormal myeloid original cells. Patients tested by fusion gene were positive for DEK-CAN with FLT3-ITD mutation. High expression of CD34 and HLA-DR usually suggests a poor prognosis. This is because the high expression of these two antigens means that leukemia cells are closer to the original hematopoietic stem cells, strong proliferation ability, low differentiation, easy to lead to disease recurrence, and relatively insensitive to chemotherapy drugs [8]. The prognosis is poor, the complete response rate of induction therapy is 65%, the 5-year overall survival rate is 28% in children and only 9% in adults, and the mortality rate after relapse is extremely high. The overall survival rate of patients with allogeneic hematopoietic stem cell transplantation was higher than that of patients with autologous hematopoietic stem cell transplantation or chemotherapy alone. This patient failed to reach remission after IA regimen, FLAG regimen and HIA regimen, and is a refractory leukemia. AML patients with DEK/CAN have a short survival period, poor prognosis, and high mortality after recurrence. (t6;9) (p23;q34)/DEK/CAN-positive AML patients should undergo allogeneic hematopoietic stem cell transplantation at an early stage, and arsenical therapy CAN also be a good clinical treatment option.

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Declaration of Interest:
All authors declare: 1. All views and data in this paper are supported by references and data. The manuscript has not been published before and is not being considered for publication elsewhere. 2. All authors have contributed to the creation of this manuscript for important intellectual content and read and approved the final manuscript. We declare there is no conflict of interest. 3. This paper is published with the consent of patients, in line with ethical requirements.

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