CASE REPORT

Congenital Leukemia
Report of 2 Cases

by

S. UNTARIO, ERWIN SARWONO, E.K. KARIYADI, SYLVIATY M.D. and NETTY RHT.

(From the Department of Child Health, Med School Faculty, Airlangga University/Dr. Soetomo Hospital Surabaya)

Abstract

Two cases of congenital leukemia are reported, one of which was associated with Down's syndrome. Both cases were lymphoblastic as observed morphologically and by the negative Sudanophilia. To the best knowledge of the authors these cases are the first two cases of congenital leukemia reported in the Indonesian medical literature.

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CONGENITAL LEUKEMIA

Introduction

Congenital leukemia is rare, only just over 100 cases have been reported to date [1]. The majority of patients had acute myelogenous leukemia, but newborns with acute lymphoblastic leukemia have been observed.

To fulfill the criteria of congenital leukemia the infant must show clinical and hematological signs of leukemia at birth or shortly thereafter in the newborn period [2]. Some authors include cases up to the 6th week of life when the history suggests that hematological abnormalities date back to the first weeks of life [3]. Pathognomonic criteria would be proliferation of certain precursor cells of the blood leukocytes, and the infiltrations of these immature cells into extramedullary hematopoiesis compartment.

Leukemia in the newborn has been associated with Down's syndrome and other specific congenital anomalies. Patients with Down's syndrome have been observed who were diagnosed as to suffer from leukemia but who recovered completely without specific anti-leukemic therapy. This phenomenon was later ascribed to ineffective regulation of granulopoiesis [4]. Underneath is a report of two cases of congenital leukemia with symptoms and signs present at birth, one of which was associated with Down's syndrome.

Report of Case

Case 1

A baby boy, born at term on March 17, 1990 at the Dr. Soetomo Hospital in Surabaya showed the presence of petechiae and ecchymoses. The delivery was uneventful. The mother was healthy and had a normal pregnancy. She had not attended a prenatal clinic, denied having suffered from a bleeding tendency, but she had consumed traditional herbs regularly for rheumatic fever. When she was one month "late" she once took some traditional drug to abort, but the effort was unsuccessful. The baby was the product of a third pregnancy, he had an older sister of 5 years and a brother of two who were both healthy. After the birth of the second child the mother underwent some procedures for sterilization apparently with no success.

On physical examination the baby did not look sick. The birth weight was 2670 g, the body length 45 cm and the Apgar score was 7 and 8. The heart and lungs were normal, the liver was palpable 1 cm below the costal margin but the spleen was not. No enlarged lymph glands were found. Petechiae and ecchymoses were seen around the mouth and nose, and on the back and extremities. Skin infiltrates were noticed on the right arm (see Fig. 1, 2).

The baby was assessed as suffering from septicemia with hemorrhagic diathesis, and was given ampicillin, gentamycin and Vit-K. A blood culture was done which gave negative results. The chest X-ray picture was normal. Laboratory examination gave the following results: Hb: 23 g/dl, RBC: 4,600,000/cumm, Hct: 72 % and WBC: 83,600/cumm, Platelets: 23,000/cumm. The blood film showed numerous blast cells with a few normoblasts. Bone marrow aspiration on the 2nd day of life revealed the predominant presence of blast cells with scanty cytoplasm containing no granules and a nucleus with rather coarse chromatin with 0 - 1 nucleolus. The blast appeared to be Sudan Black and Peroxidase negative (Fig. 3, 4, 5). The diagnosis of congenital acute lymphoblastic leukemia was made. On day 5 biopsy of the skin infiltrate revealed the presence of lymphocyte sized cells in the dermis and subcutaneous tissue as can be seen in leukemia (pathologist).

The baby developed jaundice on the 4th day of life, and the hemorrhagic spots on the skin increased in number. On day 8 anti-leukemic therapy was started with vincristine 0.35 mg/weekly and dexamethasone orally 3 x 0.5 mg daily. On day 10 the blood picture was as follows: Hb: 17.2 g/dl, RBC: 4,250,000/cumm, WBC: 4,500/cumm, Platelets: 25,000/cumm. The baby's condition deteriorated, he became dyspneic and meteoristic and refused his feeding. A repeat blood exam gave Hb: 11.4 g/dl, WBC: 900/cumm and Platelets: 12,000/cumm.

The patient was put on intravenous fluid drip and oxygen, and cefotaxime-gentamycin plus dexamethasone intravenously. However, the baby's condition worsened, he bled from his nose, developed convulsions and became comatose. On day 13 the baby expired.

The autopsy findings revealed a baby of 43 cm in length, weighing 2650 gman presented with subcutaneous hemorrhages on the head, face, breast and abdomen. The brain was 362.5 grams and showed no bleedings. The heart and lungs were normal. The liver was 189 grams, red brown in color and firm. The spleen was 15 grams, dark red and firm. The pancreas was red-yellowish, both kidneys looked normal. Microscopic examination: The alveoli of the lungs were filled with exudate and erythrocytes, the alveolar septa were thickened with edema of the stroma showing dilated blood vessels. The pancreas consisted of serous glands with the stroma densely infiltrated with lymphocytes of various grades of maturity. No islets of Langerhans or Pancreatic corpuscles were seen. The kidneys showed hemorrhages between the tubules. Other organs showed no abnormal findings.

Case 2

A baby girl, born on May 22, 1991, showed severe intrauterine growth retardation, having a body weight of 1770 grams after 40 weeks of gestation. The baby was born to a 23 year old para 1 mother who was apparently healthy. The mother was VDRL negative, of group O and denied having had X-ray irradiation for diagnostic or therapeutic purposes during or before her pregnancy. The baby showed epicanthal folds and simian lines and was phenotypically mongoloid, no enlargement of the liver and spleen were noted. She gave a good cry, the Apgar score was 8 and 9 and was apparently healthy until later in the day when she began to vomit brownish fluid. She was given intravenous fluid drip and Vit K 1 mg i.m. The Hb was 19.5 g/dl, the leukocytes 56,000/mm3, the platelets were sufficient in number. The next day the vomit was distinctly bloody, the baby became icteric and the cry was weak. Her serum bilirubin was 9 mg/dl, her blood group was 0. She was given ampicillin and plasma transfusion as a surrogate of intravenous feeding. On her 4th day of life she developed petechiae on her right cheek and a repeated blood exam gave the following results: Hb 23.9 g/dl, platelet count 16,000/cumm, WBC 16,200/cumm, with blast cells in the blood smear. A bone marrow puncture was performed and the aspirate showed a picture that was dominated by lymphoblasts. The blasts had a high nuclear-cytoplasmatic ratio, dense nuclear chromatin with 0 - 1 nucleolus and the cytoplasm was Sudan-Black negative. The baby's condition deteriorated and she expired on the ninth day of life. No autopsy was performed.
Discussion

The cases described above were both diagnosed as congenital leukemia since the signs and symptoms appeared in the first few days of life. Besides showing leukemic blood and bone marrow pictures case 1 had skin infiltrates, an important sign of congenital leukemia [5]. At autopsy leukemic cells infiltrates were found in the pancreas but not in other organs. We wonder whether the treatment could have cleared most parenchymatous organs of leukemic infiltrates.

Nies (1965) has shown that during remission 11 of 15 patients had an evidence of leukemia in other sites of the body [6]. However, the induction treatment of his patients was inadequate by present standards. Case 2 had also typical leukemic blood and bone marrow pictures. This baby was phenotypically mongoloid and as such might have transient or pseudo-leukemia on follow-up [4] but for the fact that she showed proliferation of lymphoblastic instead of granulocytic cells. In the literature transient leukemia cases were predominantly myeloblastic [7], except for one case described by Clark etal. (1981) that was lymphoid [8]. Chromosomes studies were not done on this patient, nor was autopsy permitted by the parents.

In both cases the leukemia was lymphoblastic. This was morphologically observed in the giemsa stained smear and confirmed by the negative Sudan-philia of the blasts [9]. This seemed some what unusual. In the literature congenital leukemia was mostly myeloid or monomyeloid though the lymphoid type was also observed [11,10]. However, it is not clear in how many of the cases cytochemistry was done to establish the diagnosis. Wolk (1974) described a 3 month old boy with leukemia who would have been diagnosed as myeloblastic, but which turned out to be lymphoblastic after cytochemical studies [11].

The combination of mongolism and leukemia is supposed to give a 20 times higher frequency than expected [12]. Of our two cases one was phenotypically mongoloid while the other was apparently normal. This fact reflected the greater frequency of leukemia in the mongoloid since the number of births of normal babies is very much higher than that of mongoloid babies. In Down's syndrome transient leukemia has been reported [8,13] and opinions were divided in the interpretation of this phenomenon. Ross et al. (1963) ascribed it to temporary ineffective regulation of granulopoiesis in connection with the chromosomal anomaly [4]. considered it as spontaneous remission such as may happen with neuroblastoma [7]. Some of these cases showed a relapse [14,15,16] but it was not established whether it was a real relapse or the occurrence of a new (type of) leukemia. Our patients died shortly after diagnosis, thus follow-up on the course of their disease were not possible.

To the best knowledge of the authors these two cases of congenital leukemia are the first cases reported in the Indonesian medical literature.

Figure 1. Baby boy (Case 1) showing petechiae on the face
Figure 2. Skin infiltrate on right fore-arm of case 1

Figure 3. Lymphoblasts in bone marrow of case 1
Figure 4. Sudan Black negative blast in bone marrow of case 1. inset: SB positive granulocyte in peripheral blood of same patient

Figure 5. Peroxidase negative blast in bone marrow of case 1. inset: Peroxidase positive granulocyte in peripheral blood of same patient
REFERENCES


SPECIAL ARTICLE

The Impacts of Genetic Counseling on The Quality of Children

by

SJARIF HIDAJAT EFFENDI
(From the Department of pediatrics, Padjajaran University, Bandung)

Introduction

The success of our National Family Planning Program and awareness of people to the motto of: "The norm of a small, happy and prosperous family" has led our people, especially the eligible couples, to think no more about quantity, but the quality of their children. There is a tendency that they do need assurance of completely healthy children, the present or the coming ones. This fact leads our Health Service Personnel to the awareness that they must be able to solve people's problems related to their desire for good quality children.

Increasing the children's quality of life greatly depends on the efforts within the following stages of their lives: conception, pregnancy, delivery and neonatal conditions. It means that marriage/genetic counseling, antenatal care, and perinatal services play a very important role. Every disturbance within these periods will lead to serious damage to the growth and development of organs, especially the brain. This condition might result in handicapped and malformed children.

As infectious disease are brought under control and decreasing, the relative importance of overcoming disorders that are wholly or partly genetic has been an decrease in the perinatal. Mortality rate, insincerity of infections and malnutrition in hospitals in Indonesia, but cases of congenital hereditary disorders are increase.

Every year thousands of families are affected by the birth of an abnormal child. About 0,5% of newborns have chromosom abnormality with moderate to severe phenotypic effects.