Letter to Editor

Iran J Pediatr
Sep 2010; Vol 20 (No 3), Pp: 369-373

A Case of Congenital Leukemia with Nasal Hemorrhage and Jaundice

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Received: *Mar 01, 2009*; Accepted: *Mar 15, 2010*

Congenital leukemia (CL) is a very rare malignancy which occurs at estimated rate of 1 per 5 million births, and accounts for less than 1% of all childhood leukemias[1,2]. Only approximately 200 patients with congenital leukemia have been reported in the literature[3].

CL has no pathognomonic findings. The proposed diagnostic criteria for CL include: I) presentation in the first 4 weeks of life; II) proliferation of immature myeloid, lymphoid or erythroid cells; III) infiltration of these cells into nonhaematopoietict issues; IV) absence of other diseases which may explain this proliferation[5].

Most CL cases have been reported as Acute Myeloid Leukemia (AML). The acute myelomonocytic (FAB M4) and acute monocytic (FAB M5) subtypes are by far the most frequent AML respectively[4].

Herein we report a case of CL with skin lesions on his abdomen who was hospitalized because of jaundice and nasal hemorrhage.

After an uneventful full-term pregnancy, a boy was born with a birth weight of 3600 gr. No maternal ante partum illness was reported. At the age of 13 days he was readmitted to the hospital because of jaundice, nasal hemorrhage and respiratory distress. Physical examination at this time revealed hepatosplenomegaly with the liver and spleen palpable 5 and 3 cm below the costal margin respectively. Multiple bluish macropapules (blue berry muffin lesion) had been scattered over his abdomen since his birth. Except for the stated signs and tachypnea, other physical examinations were normal.

Laboratory tests showed: white blood cell (WBC) count 59,000 cell/mm^3, Prothrombin Time (PT) >50 seconds, Partial Thromboplastin Time (PTT) >120 seconds, and total serum bilirubin 17 mg/dL with direct bilirubin not exceeding more than 0.5mg/dL. AlT and AST levels were 44 and 51 IU respectively. Blood cultures were negative and serology testing was not indicative of congenital infection. Lumbar puncture did not reveal meningeal involvement. Chest X-ray showed diffuse infiltration. Cerebral ultrasound was normal.

The patient was hospitalized with a primary diagnosis of sepsis, and antibiotics were initiated besides supportive therapy. Gradually all blood cells decreased: WBC count 1400 cell/mm^3, hemoglobin 7 g/dl and platelet 8000/mm^3. Therefore packed cells, platelet, fresh frozen plasma (FFP) and granulocyte colony-stimulating factor (G-CSF) transfusions were given. Despite treatment, his general condition did not improve and echymotic plaques began to expand over all the body.

Bone marrow aspiration was occupied with >80% monoblast cells. A diagnosis of AML, French-American-British (FAB) type M5, was made based on monoblast cells (Fig. 1).

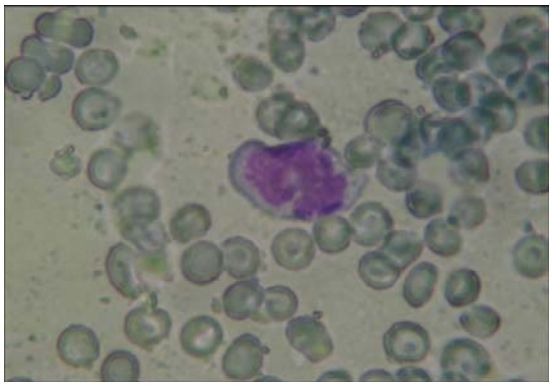

Fig. 1: Monoblast in bone marrow aspiration suggestive of AML type 5

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Immunophenotyping showed CD33: 76%, CD64: 85% and CD45: 91% in the cell population. We recommended chemotherapy. Notwithstanding, boy's parents refused to accept it and the patient received only supportive care (by his parents' consent). Unfortunately, the boy died after 2 months due to severe and diffuse hemorrhage.

There is an *arbitrary* difference between congenital leukemia (manifest itself within the first days of life) and neonatal leukemia (diagnosed in the first 6 weeks of life)[5].

Hypothesized factors involved in the etiology include: congenital anomalies and genetic predisposition [t (4;11) (q21;q23) is the most frequent karyotypic abnormality)][6], besides parental exposure to environmental toxins, alcohol and tobacco[5].

Skin lesions may be the first manifestation of CL. Leukemia cutis refers to the direct infiltration of cutaneous tissues by leukemic cells and occurs in approximately 25% to 30% of CL cases. They are violaceous nodules approximately 1 to 2.5 cm in diameter, accompanied by purpura, petechiae, and ecchymoses[7]. Hepatosplenomegaly, spleno-megaly, manifestations of anemia and thrombocytopenia are common. Sepsis and pneumonia owing to neutropenia and respiratory distress may occur[5]. In accordance with literature, our patient presented many of these signs as well.

Most common Immunophenotypes in CL are CD33, CD13, CD14, and CD15.[5,8]. The infant in this study also had a high CD33 (76%).

Hematologic parameters may be normal or present profound leukocytosis (50,000 to >100,000), anemia, and thrombocytopenia as in our case. Leukemic blasts are usually found in the peripheral blood. Lactic dehydrogenase and liver function test may be abnormal.

Examination of the cerebrospinal fluid frequently reveals the presence of leukemic blasts[5], although in this study it was negative.

Some differential diagnosis of CL include sepsis, birth-related hypoxia, congenital infections due to TORCH, neuroblastoma, Transient Myeloproliferative Disorder and hemolytic disease secondary to Rh incompatibility[5]. In our report, the primary diagnosis was sepsis.

The survival rate of congenital AML is reported to be as low as 26%[2], and to our knowledge, there are approximately 20 reported cases of congenital leukemia in which spontaneous remission has occurred[9,10]. Our patient also expired, although chemotherapy was not performed.

Key words: Leukemia; Neonatal jaundice; Congenital; Bleeding

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Experience with a New Technique for Laparoscopic Hernia Repair in Small Children and Infants

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Received: *May 15, 2009*; Accepted: *Mar 10, 2010*

During recent years, the trend toward laparoscopic approach for hernia repair in children has been increasingly justified [1,2]. The ability to detect and repair the contralateral patencies simultaneously, along with safety of the procedure are the cornerstones of the selection of the laparoscopic approach as a reliable alternative to the conventional open techniques[3].

Although many authors believe that the laparoscopic inguinal hernia repair is superior to the traditional approach in the view of improved cosmesis and fewer recurrences, there are still some issues about its popularity, especially regarding the acceptable cosmetic results, along with the short operative time and brief hospital stay and the high success rate of open conventional technique.

We performed 50 inguinal hernia repairs by laparoscopic technique during 2.5 years from April 2006 to October 2008 in our hospital.

Forty-one children including 34 males and 7 females underwent operation by this technique. Hernia in 25 cases was right-sided and in 7 left-sided. Two patients had recurrence following previous hernia repair through groin incision. 32 cases presented with unilateral hernia and 9 patients had bilateral inguinal hernia.

The age of the patients ranged from 4 months to 5 years. The median age was 11 months. The mean operative time for unilateral repairs was 20 minutes and for bilateral ones 35 minutes. The scars on the abdominal wall were small and minute (one 5mm incision for umbilical port and a 3mm stab incision ipsilateral to the hernia for working cannula) and the cosmesis was excellent. There were no intra-operative complications and we had no conversion. The follow-up rate at six months was 100% and we had no recurrences or any other complications such as testicular atrophy.

Inguinal hernia in pediatric age group is a common problem and all the pediatric surgeons are fully familiar with the various aspects of its traditional surgical repair through the groin incision which has a high success rate and acceptable cosmetic results with few complications[4,5].

By far one of the drawbacks of this conventional technique is its inability to rule out the contralateral patent processus vaginalis and synchronous hernia.

With the advent of minimal access surgery, many pediatric surgeons accepted it, as an suitable and reliable alternative to previous techniques, considering its superiority for handling tissues during repair of recurrent inguinal hernias and also for its capabilities in regard to justifying and managing the synchronous subtle contralateral hernia[6,7].

However, there are still some issues about the introduction of laparoscopic inguinal hernia repair as the gold standard method, specially taking the possible longer operative time and the inevitable need for three separate ports which is the case in routine laparoscopic herniotomy techniques into consideration.

The modified and new laparoscopic technique employed by the authors has an acceptable short operative time with only one working trocar located ipsilateral to the hernia, using extra-corporeal tying, that yields excellent cosmesis.

Key words: Laparoscopic surgery; Childhood; Inguinal hernia;

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Asthma Knowledge Level of Primary School Teachers in Babol, Iran, 2008

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Received: Sep *20, 2009*; Accepted: *May 05, 2010*

Regarding the fact that two thirds of children's life passes in school, it is important for teachers to have enough knowledge about asthma and how to encounter the disease. Children become panic in asthma attack and this behavior is normal. These children should be supported by other children in the school. Teachers should identify asthmatic patient. Also it is important that patient's classmates be aware of this disease [1]. So, the aim of this study was to assess the knowledge of teachers about asthma and its treatment in order to design asthma plan action based on our findings. This cross-sectional descriptive analytical study performed in winter 2008 with randomized sampling in Babol. After coordination with department of education, 5 or 6 teachers in every school were randomly chosen in all 80 schools. These teachers were given specifically designed questionnaires including demographic data (age, sex and level of education) and 16 questions about asthma knowledge. Each correct answer was graded 1 and wrong answers 0 (the maximum score of knowledge questionnaire was 16). Comments of many epidemiologists and clinicians (specialist in asthma and allergy of pediatric and adult lung specialist) were considered to enhance content validity. The Cronbach's α coefficient (α=0.822) was used to evaluate questionnaire reliability. Study was approved by ethical committee of Babol University of Medical Sciences and Health Services. Data were analyzed by software SPSS, for statistical tests we used Mann-Whitney and Kruskal-Wallis. The mean age of participants (n=425) was 42.7±7.3 years. The results showed that the mean knowledge score of our teachers was 12±2.3. This means that the mean knowledge score was intermediate, while most of researches have shown that their teachers' knowledge level about asthma was low. Frock and colleagues (2008) found an insufficient knowledge about bronchial asthma among 120 teachers of physical education in Schleswig-Holstein University [2]. Abdel Gawwad and collaborators (2007) reported that most of school staff had poor to fair level of asthma knowledge and management practices. They found it very important that training is directed to all staff as pre-service and in-service programs [3]. The results of Rodehorst (2003) indicated that although teachers had a favorable attitude toward asthmatic students, their knowledge about asthma was low[4]. Our results indicate that mean knowledge level of teachers in our study is higher than in other studies performed in the world but regarding the importance and prevalence of the disease in our region, ongoing efforts to improve asthma management in public schools through teacher education and policy development should be supported.

The mean knowledge score of teachers had no significant difference regarding the age (12.2±2.1 in teachers under 40 years and

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12.1±1.8 in teachers over 40 years, *P*=0.318). Ones and colleagues (2006) also showed there was no difference between age groups [5]. The results of our study showed that there was also no significant difference in knowledge score between sexes (11.8±2.3 in males and 12.1±2.2 in females). Forck[2] (2008) had come to a similar result. Our findings revealed a significant difference in knowledge score regarding educational degree of the teachers. Teachers with bachelor degree had higher knowledge scores (11.7±1.5 in under diploma, 12±2.2 in diploma, 11.6±2.6 in junior college and 12.4±1.8 in bachelor, *P*=0.031), but Forck (2008) found no differences regarding educational level of the teachers [2]. Ones indicated that the knowledge level of the teachers was not related to their educational level [5]. The lack of a standard questionnaire that makes comparison of different studies possible can be regarded as a limitation of the present study.

Key words: Asthma; Knowledge; Teachers; Primary school

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