Acta Paediatrica Hungarica 29 (1–2), pp. 209–211 (1988–89)

Clinical manifestations of various phagocytic defects

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One hundred and three cases of primary immunodeficiencies, aged from 0 to 16 years, who were referred to the Children's Memorial Hospital from the whole country by regional hospitals or individual pediatricians were investigated. Primary immunodeficiencies were diagnosed and classified according to the principles elaborated by a group of experts of the World Health Organization [3].

Twelve children with various disorders of phagocytosis were recognized with the aid of such laboratory tests as the number of granulocytes in the peripheral blood, evaluation of myelogram, NBT reduction test, and test of neutrophil chemotaxis. Disorders of phagocytosis constituted 11.6% of the total number of cases.

Chronic Granulomatous Disease (5 children)

The age at the onset of clinical symptoms and the most frequently observed diseases in these children are summarized in Table I. The infections were always caused by Staphylococcus aureus; with periods without infections lasting up to several years. Pneumonia and bronchitis were not observed.

The main clinical problem in 4 children was liver antibiotic-resistant abscesses. Each of them was operated 1–2 times for liver abscesses. We observed that very good clinical effects were obtained by making aspirations of abscesses under the control of USG and local injection of antibiotics. This enabled us to avoid surgical operations twice in patient F. T., and 3 times in patient B. G. The child F. T. had an abscess in the vicinity of the liver big vessels and the treatment consisted of antibiotics (3 months) and transfusions of leukocytes 3 times per week; the treatment was effective.

All the children received Bactrim as a prophylactic drug.

Chronic Congenital Neutropenia (4 children)

In four children with chronic neutropenia the onset of clinical symptoms appeared in the first year of life (Table II). The course of infections was rather mild and responsive to an-

Acta Paediatrica Hungarica 29, 1988–89
Akadémiai Kiadó, Budapest

TABLE I

Clinical symptoms in children with chronic granulomatous disease

	Age (y)	Age at the onset	Lymphadenitis	Liver abscesses	Skin abscesses	Periodontitis
B. Pi.*	11	5	+	+	—	+
B. P.*	5	2	—	—	+	+
B. G.	16	8/12	+	+	+	+
F. T.	15	7	—	+	+	+
S. P.	24	4	+	+	—	+

* brothers

TABLE II

Clinical symptoms in children with chronic neutropenia

	Age (y)	Age of the onset of symptoms	Pneumoniae	Bronchitis	Otitis media	Skin abscesses	Periodontitis	Lympha denitis
Z. W.	9.5	1	+	+	++	++	+++	—
D. M.	9	11/12	+	+	—	++	+++	+
K. D.	3	1/12	+	+	—	++	+++	—
Z. K.	2	3/12	—	+	—	++	+++	—

\+ sporiadic
++ relatively frequent
+++ chronic infections

tibiotic therapy. The frequency of infections was lower than in antibody deficient children. Pneumonia or bronchitis episodes were rather sporadic as was described [3]. The only chronic infection was periodontitis, caused mainly by Gram-negative anaerobic bacteria (Fusobacterium). This infection in the child D. M. caused premature loss of milk-teeth. Currently the odontum of the permanent teeth of this child are undergoing process of decay. In the other children periodontitis was milder. The range of neutrophils number in peripheral blood was from 0 to 10332/mm^3 and the end stage maturation of granulopoiesis was determined at the level of promyelocyte, myelocyte or metamyelocyte.

In differential diagnosis we excluded neutropenia caused by autoantibodies or induced by drugs. The course of neutropenia did not have cyclic changes. The life-prognosis in chronic congenital neutropenia of unknown heredity was better than in syndromes of autosomal recessive trait described by Kostman [2].

CHEMOTAXIS DISORDERS (3 CHILDREN)

In one family an impaired chemotaxis (below 40% of normal) as a genetic defect transmitted in autosomal dominant trait was documented. The most combersome and common clinical features of mother and her three sons (2, 4 and 6 years old) were

premature exfoliations of teeth due to loss of a periodontal tissue. It was clinically manifested as a slowly progressive periodontal decay due to infection with Fusobacterium and Bacteroides with acute form of gingival inflammation and hypertrophy which was refractory to treatment. The other clinical manifestations observed in this family were: partial hypopigmentation of hair, erythemas and very delicate keratotic spots of the palmar and plantar surfaces. Dilatations of surface skin vessels within cheeks were also observed.

All the family members had predisposition to recurrent bacterial infections of the upper respiratory tract as well as recurrent herpes virus infections, but the course of diseases observed was rather light.

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