mortality in our series was due to misdiagnosis and associated complications.

All our cases were benign histologically. Malignant changes have not been reported in literature either.

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Kostmann Syndrome

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Kostmann syndrome, which was first described in 1956, is a group of closely related disorders characterized by congenital or infantile neutropenia resulting in frequent episodes of fever, skin infection, boils, aphthous stomatitis and other infections presenting in early life(1,2). Most of the cases described were autosomal recessive(1) and hence consanguinity was common. About 50 cases have been described in the literature so far with no case report in Indian literature. We report a young girl with Kostmann syndrome seen by us at the age of 7 years.

Case Report

A girl aged 7 years, belonging to Christian family from Goa, was referred to us for recurrent infections. She was a product of consanguinous marriage and born full-term by Cesarean section. At the age of 3 months, she developed first episode of fever with otitis externa. Since then, she developed frequent infections affecting skin, ear, throat, mouth, gums and urinary tract at regular intervals. She had spontaneous abscesses at three occasions needing

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surgical drainage. The episode of infections invariably responded to antibiotic therapy within a period of 5 to 10 days. She had measles at the age of 6 months and chicken pox at the age of 2 years with uneventful recovery. She received BCG vaccination at birth and three doses of oral polio and DPT during the first year of life without any adverse reactions. There were no episodes of pneumonia, meningitis or septicemia. Of late, she had developed severe gingivitis needing frequent dental attention.

She had two siblings (brothers) aged 12 years and 10 years who were normal without any history of frequent infections. Parents were apparently healthy.

Her examination revealed an asthenic girl who was dwarf. Her hair had a reddish hue. There were scars of healed or incised abscesses. Examination of the oral cavity revealed multiple aphthous ulcers, poor dental hygiene and gingivitis. Abdominal examination showed mild hepatosplenomegaly.

A review of hemograms carried out at multiple occasions right from infancy showed presence of severe neutropenia with absolute neutrophil count being less than 400/cumm throughout. The total WBC count, however, was invariably normal and this was due to relative lymphocytosis, eosinophilia and monocytosis. Her hemoglobin, platelet count and ESR showed no abnormalities. The marrow examination revealed normocellular marrow, moderate myeloid hyperplasia with maturation arrest at the level of myelocytes. The promyelocyte and myelocyte present at the marrow were large and they showed some evidence of dysmyelopoiesis. Erythroid series and megakaryocytes were normal.

The investigations for assessment of B-lymphocyte, T-lymphocyte functions and complement level were normal (*Table*). Neutrophil function studies could not be carried out due to their severe quantitative deficiency. The child was put on prophylactic antibiotic (oral penicillin) which she has

TABLE–*Immunological Investigations*

	S. Proteins			S. immunoglobulins		
	Electrophoresis			Test	Pt. (mg/dl)	Normal range (mg/dl)
I.	(T) protein	:	6.6 g/dl	IgG	1500	560 - 1500
	Albumin	:	3.1 g/dl	IgA	230	55 - 230
	Globulin	:	3.5 g/dl	IgM	110	27 - 118
	A:G ratio	:	1.0: 1.1			
	α 1-g	:	0.4 g/dl			
	α 2-g	:	0.7 g/dl			
	β-g	:	0.8 g/dl			
	γ-g	:	1.6 g/dl			
II.	Peripheral blood lymphocyte sub-population					
	T-Lymphocytes	:	65% (Normal)			
	B-Lymphocytes	:	17% (Normal)			
III.	S. complement (T)	:	120 mg/dl (Normal)			

been continuing for the last 2 years. She had two episodes of infection during this period–one episode of chest infection and other of acute cervical lymphadenitis. Both responded well to antibiotics.

Results

Infantile agranulocytosis, described by Kostmann is 'probably' a genetic disease of autosomal recessive inheritance. It is clinically characterized by recurrent pyogenic infections which start early in life.

The neutrophil count is extremely low and it is often associated with compensatory eosinophilia, mucocytosis, lymphocytosis and hypergammaglobulinemia(3). The usual sites of infection are skin, ear and oral cavity. The patients may develop bacterial pneumonias and meningitis and other serious infections. Most of the cases described in the literature have died at early age due to fatal infections, but relatively mild course with prolonged survival as noted by us have also been reported(4). Possibly the cases reported under Kostmann's syndrome are a group of closely related disorders. The underlying defect could be of variable nature and hence the outcome may differ.

Chromosomal abnormalities have been reported in the marrow granulocytic cells of children with Kostmann syndrome(5). The bulk of evidence suggests that the marrow maturation abnormalities are related to defective marrow micro environment(6) or possibly to intrinsic cell maturation defect(7). Neutrophil survival is normal.

The treatment is usually with antibiotics which can prolong life for several years(1,2). The only alternative treatment reported in literature is marrow transplantation resulting in partial or complete cure(8). An increased risk of leukemia has been reported in these children(3,4). Splenectomy, corticosteroids and androgens have been of no use(3,4) and splenectomy has often resulted in fatal sepsis. Newer modalities of treatment which can be tried in these children are WBC differentiating agents like 1-25-Dihydroxychole-calciferol and cis-retinoic acid(9). 1-25-Dihydroxychole-calciferol is a potent inducer of cell differentiation *in-vitro* in low concentration, while retinoids especially 13-cisretinoic acid are potent agents causing terminal differentiation with a loss of self-replication capacity(9). Phase I-II trials utilizing these agents in treatment of myelodysplasia have been reported(9). We postulate that these agents could be of use in disorders like Kostmann syndrome and hence are worth a trial.

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NOTES AND NEWS

NATIONAL HEMATOLOGY UPDATE

A Hematology-Oncology Update is being organized by the Kanpur Study Circle and the Hematology-Oncology Chapter of the IAP, on *23rd February, 1992* (Sunday) at Geet Hotel, The Mall, Kanpur. Eminent speakers from all over the country are expected to attend. Those interested in attending the same may send a DD/Cheque for Rs. 150/- in favour of 'NATIONAL HEMATOLOGY UPDATE' payable at Kanpur latest by 15th February, 1992.

Outstation delegates are requested to arrange for their own accommodation and return reservation.

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