Differentiating Vaso-Occlusive Crisis and Infection in Sickle Cell Anemia

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Since leukocytosis is common in patients with sickle cell anemia, it is generally assumed that the white blood cell count is of little diagnostic value. However, few investigators have used them in differentiating infections with vaso-occlusive crisis.[1-3] Recently studies in newborns have stressed the importance of bands/segmented neutrophils ratio in diagnosis of bacterial infections.[4] Importance of bands/segmented ratio has not been described in patients with sickle cell anemia. In this study we evaluated the value of leukocyte parameters, namely, total white blood cell count, absolute segmented neutrophil count, absolute band forms count, and bands-to-segmented ratio in patients with sickle cell anemia during baseline or steady state vaso-occlusive crisis and episodes of bacterial infection.

Methods

Charts of 52 patients (16 female, 36 male) with homozygous sickle cell disease treated at Saint Luke's Hospital during the last four years were reviewed. Their ages ranged from three months to 38 years. Total leukocytic and differential counts were recorded in all 52 patients during baseline or steady state studies, in all 52 during 69 episodes of vaso-occlusive crisis, and in 20 patients during the episodes of culture proven bacterial infections. The infections were two meningitis cases (one, Streptococcus pneumoniae, the other, Hemophilus influenzae type B), three pneumonias (two Streptococcus pneumoniae and one Staphylococcus aureus), three abscesses (two Staphylococcus aureus and one Escherichia coli), one Salmonella typhimurium bacteremia, four Escherichia coli cystitis, one Klebsiella pneumoniae pyelonephritis, and six episodes of suppurative tonsillitis caused by group A beta-hemolytic streptococci. Severity of illness at the time of presentation warranted admission in all cases during crisis and infection.

Results

Bands/segmented ratio was calculated for each episode by dividing absolute band count (per cu mm) with absolute segmental polymorphonuclear leukocytes count (per cu mm). Means and standard deviation of various leukocyte parameters are given in Table 1. The statistical analysis by Student's t test was significant for each of these parameters in different groups. None of the patients during the baseline or steady state had band counts greater than 500/cu mm as compared to 17 percent of patients during crisis and 90 percent of patients with bacterial infections. Distribution of bands/segmented ratio in different groups are shown in Table 2. Seventy-seven percent of patients during baseline studies had a bands/segmented ratio of less than .01 as compared to 36 percent during vaso-occlusive crisis; all patients during the episode of bacterial infection had a bands/segmented ratio of greater than .01. Bands/segmented ratios of $\geq$ .051 were not seen during baseline or steady state studies; however, they were observed in 10 of 69 episodes of vaso-occlusive crisis and 14 of 20 (70 percent) episodes of bacterial infections.

Comment

Patients with sickle cell disease have high total white cell counts related to either decreased margination or increased myeloid activity. As leuko-

Table 1. Mean Leukocyte Parameters in Patients with Sickle Cell Disease (±SD)

Clinical Status of Patients	White Blood Cell Count (per cu mm)	Absolute Segmented Polymorphonuclear Leukocyte Count (per cu mm)	Absolute Bands (per cu mm)	Bands/ Segmented Ratio
Baseline (B)	12,903	7,666	34.6	0.005
(n=52)	±5,566	±4,435	±80.1	±.01
Crisis (C)	17,041	11,123	252.1	0.023
(n=69)	±6,559	±5,232	±330.9	±.03
Infections (I)	26,169	18,613	1,909.2	0.147
(n=20)	±9,599	±9,152	±1,294.1	±.18
P Values (χ^2)				
B vs C	.0004	<.0001	<.0001	<.0001
B vs I	.0008	<.0001	<.0001	<.0001
C vs I	<.0001	<.0001	<.0001	<.0001

Table 2. Distribution of Bands/Segmented Ratio in Patients with Sickle Cell Disease

Clinical Status of Patients	≤.01 No. (%)	.011 to 0.05 No. (%)	≥.051 No. (%)
Baseline (B)			
(n=52)	40 (77)	12 (23)	0 (0)
Crisis (C)			
(n=69)	25 (36)	34 (49.5)	10 (14.5)
Infections (I)			
(n=20)	0 (0)	6 (30)	14 (70)
P values (χ^2)			
B vs C	<.0001 (19.7)		<.002 (8.2)
B vs I	<.0001 (34.6)		<.0001 (45.2)
C vs I	<.001 (10.1)		<.0001 (24.2)

cytosis is usually associated with sickle cell anemia, it is generally believed that leukocyte parameters may be of limited help in evaluating these patients. Data in this study indicate that striking changes in leukocyte count, particularly marked elevation of band forms and high bands/segmented neutrophils ratios are seen during bacterial infections of variable severity and etiologies. This information is of significant value in identifying the sickle cell patients with bacterial infections and vaso-occlusive crisis. These leukocyte parameters are of immense value to clinicians in early identification of children and infants with sickle cell disease who frequently have life threatening bacterial infections with encapsulated bacteria.

References

1. Buchanan GR, Glader BE: Leukocyte counts in children with sickle cell disease. Am J Dis Child 132:396, 1978
2. Pegelow C, Powars D, Overturf GD: Sickle cell disease. Am J Dis Child 133:448, 1979
3. McIntosh S, Rooks R, Ritchey AK, et al: Fever in young children with sickle cell disease. J Pediatr 96:199, 1980
4. Boyle RJ, Chandler BD, Stonestreet BS, et al: Early identification of sepsis in infants with respiratory distress. Pediatrics 62:744, 1978