Hemolytic-Uremic Syndrome in Asir Region

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Introduction: Hemolytic-Uremic Syndrome (HUS) is a worldwide disease and a common cause of acute renal failure in childhood. Recently the number of cases in our area seems to be increasing.

Aim: The objective of this study was to look at epidemiology, clinical course and complications of hemolytic-uremic syndrome in the Southern region of Saudi Arabia.

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Method: The medical records of 24 patients with typical post diarrhea HUS who were seen in Asir Central Hospital (ACH) between January 1989 and December 1994 were retrospectively reviewed.

Results: Of the 24 patients 15 were boys and 9 were girls with sex ratio 1.7:1. Ages ranged from 7 months to 11 years with a mean of 38 months. Clustering of cases were during spring and autumn time. Shigella dysenteriae was the most commonly isolated micro-organism, found in the stools of 5 patients. Complications occurred in 8 patients (33%) in the acute phase, 5 (21%) had generalized tonic-clonic seizures, while 2 (8%) had depressed level of consciousness. Three of the patients who had generalized seizures developed permanent cortical blindness. One patient developed chronic renal failure. One patient (4%) died in the acute phase after prolonged generalised seizure and coma. The mean age of the group with complications was 33±35 months while it was 41±33 months in the uncomplicated group and mean platelets count was 46±40 x 10^9/L and 58 ± 35 x 10^9/L, respectively.

Conclusion: Males seem to be more affected than females and Shigella was the most commonly isolated organism. Cortical blindness was relatively more common than in other studies and patients with complications were generally younger and had lower platelet count than those without complications.

Key Words: Hemolytic-Uremic Syndrome, Shigella, Children, Saudi Arabia

INTRODUCTION

Hemolytic-uremic syndrome was first described by Gasser et al in 1955. It is recognized as one of the most common causes of acute renal failure in childhood. It is a worldwide disease which has been reported from several countries. There is a scarcity of literature reports on HUS from Saudi Arabia in that only single case reports have appeared recently.

In the past few years, however, an increase in the number of patients with hemolytic-uremic syndrome has been observed in the southern region of Saudi Arabia, and because of that and lack of previous reports from this region a review of patients was initiated. Our study addresses the magnitude of HUS with the emphasis on epidemiology, clinical course of the disease and complications.

PATIENTS AND METHODS

The Asir Central Hospital (ACH) is the main referral hospital for the southern region of Saudi Arabia that serves a child population of one million. The medical records of all children admitted to pediatric ward in ACH with the diagnosis of HUS between January 1989 and December 1994 were reviewed. The diagnosis was based on typical diagnostic triad of microangiopathic hemolytic anemia, thrombocytopenia and acute renal failure. All patients had acute diarrheal disease before they showed the full blown picture of HUS.

The following patient data were obtained from the patients medical records: age, sex, nationality, date of admission, duration of anuria, duration of peritoneal dialysis, complications and family history of the disease. The following laboratory data were analysed: serum creatinine on discharge, lowest platelets count, highest W.B.C., lowest hemoglobin level and stool culture results.

RESULTS

During the 6 year period, 24 children were admitted with HUS. The ages ranged from 7 months to 11 years with a mean age of 38 months. 20 patients (83%) were under 4 years of age. There were 15 boys and 9 girls representing a male to female ratio of 1.7:1. There were 4 Yemani, while the rest were Saudis. In one of the families, 3 children developed HUS following bloody diarrhea while 4 adults developed bloody diarrhea at the same time without progressing to HUS. Most of the
cases were seen during spring and autumn (Fig 1). Eight (33%) were anuric for an average of 10 days. Twelve (50%) required peritoneal dialysis, six of them were anuric before the start of dialysis. Central nervous system (CNS) complications occurred in 7 patients (29%), 5 had generalized seizure and 2 had depressed level of consciousness. Permanent morbidity occurred in 4 patients (17%), three of those who had generalized seizures developed cortical blindness and one child developed chronic renal failure. One patient (4%) died after prolonged seizure and coma.

**Figure 1**
Seasonal occurrence of Hemolytic-Uremic syndrome and sex distribution in Asir Central Hospital, Abha, Saudi Arabia.

The stool cultures were positive in 6 patients (25%) with 5 cases of Shigella and one case of Campylobacter infections. All patients had thrombocytopenia but only two patients developed purpura. Age, white blood cells count, platelets count, hemoglobin level in complicated and uncomplicated group are shown in Table 1.

**DISCUSSION**

In the last 6 years, we have admitted 24 patients with typical post diarrheal HUS to Asir Central Hospital. In the first 3 years (1989 - 1991) we admitted only 3 cases. The incidence seems to be increasing as we have admitted 21 cases within the last 3 years without any appreciable increase in the number of cases of diarrhea. This increase in incidence has been reported in other countries and could be due to increase in virulence of the causative organisms.

We believe that the incidence of HUS in this region is much higher because some mild cases might have been missed, or not referred to us as the response to the usual conservative management is quite reassuring in such mild cases. Gastroenteritis is still the leading cause of acute renal failure in this region. Of the 134 children with acute renal failure admitted in the last 6 years, only 18% had HUS, while severe dehydration from gastroenteritis was the cause in 48%. Three (12%) of our patients with HUS came from one family, four adult members of the same family had bloody diarrhea at the same time without progressing to HUS. This may indicate that these family members had the same source of infection and that the pathogenesis of HUS may be affected by host factors such as age.

All our patients were between 7 months and 11 years of age at onset (mean age 38 months). Those who were under 4 years of age constituted 83%. The mean age at onset seems to be similar to what has been reported in the United States but older than those reported in Argentina and South Africa. In contrast to previous reports, there were more males (M:F ratio 1.7:1). Although cases were admitted throughout the year, there was notable clustering in autumn and spring as has been noted by others (Fig. 1). The dialysis rate in our study (50%) is similar to other reports (51-57%). Our criteria for initiating dialysis were not different from other centres, since early initiation of peritoneal dialysis might help in preventing complications. CNS complications ranging from...
depressed level of consciousness, to seizures and blindness were observed in 7 (29%) patients. Our results are comparable to previous reports on CNS complications (range 25 - 40%). 10,11 These complications carry the potential risk of death and long term disabling consequences.11 One girl, 4-years old, died during the acute phase of the disease as a result of worsening coma and seizure. Of the 7 who had CNS complications, 2 had depressed level of consciousness and 5 had seizures, complicated by cortical blindness in 3 (60%). The latter complication was relatively higher than in other studies. Hahn et al reported cortical blindness in 2 patients (12%) out of 16 with CNS complications following hemolytic uremic syndrome.12 It was not possible to assess the long term complications due to failure of attendance for follow up. Robson et al reported a poor prognosis in younger patients with high white cell count and thrombocytopenia on admission.10 In our series, it also seemed that the younger patients and patients with severe thrombocytopenia were more likely to develop CNS complications and have poor prognosis; however, the difference was statistically insignificant. The mean age of the group with CNS complications was 33 ± 35 months while it was 41 ± 33 months in the uncomplicated group. There was no difference between their hemoglobin level and white blood cell count.

All patients except one made full recovery of renal function. This one, a 9-year-old boy continued to have raised creatinine level on follow up. This is in agreement with the previous observation that the older patients are more likely to develop end stage renal failure.15

Although all our cases of HUS were post-diarrheal, we were only able to isolate organisms from stools in 25% of cases. E.coli 0157. H7 has been reported to be the commonest isolated micro-organism from patients with HUS from North America. Our yield could probably have been higher if E. coli 0157. H7 test was available. However, our results compare well with reports from India and Pakistan where organisms were isolated from stool in 27%-35%.14,15 and Shigella was the commonest organism isolated.

In conclusion, this first major report of HUS from Southern part of Saudi Arabia tends to support the increase in incidence of HUS globally.2-6 In contrast to other studies, males seem to be more affected than females and Shigella was the most commonly isolated organism. Cortical blindness is commoner than in other studies. Further studies from this area will be necessary to confirm or explain these differences in the clinical presentation of HUS.

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