153. Clinical - Haematological Profile of Paroxysmal Nocturnal Haemoglobinuria (PNH) : Ten Years of Experience

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Object of the Study : To study the clinical - haematological profile of paroxysmal nocturnal haemoglobinuria.

Material and Methods : Thirty-five patients of PNH over a period of 10 years from Jan. 1994 were analysed retrospectively. All the patients were diagnosed on the basis of positive Ham’s and positive sucrose lysis test.

Summary of results : Eleven cases were females and 24 were males (M:F - 2:2). The age varied from 12 to 50 years, however, 94% of the cases were below 38 years of age. 31 of the 35 cases (88.6%) had symptoms of anaemia at presentation and 26 of them had been transfused. Jaundice was present in 12 cases at presentation (34%), while 14 (40%) gave a history of dark coloured urine. 10 cases had bleeding manifestations. Only one patient developed DVT on treatment while another patient presented due to severe sepsis with aspergillosis fungemia. One patient had post traumatic intracranial bleed, which recovered with platelet transfusions. Other complications were: herpes zoster - 1, pneumonia - 1 and neutropenic fever in 1.

In 17 cases followed up for 3 months or more: there was complete response in 4; partial response in 8 and no response in 5. The earliest response was after 1 month and median after 3 months. The follow up after other courses is less than 3 months: there is partial response in 2 and no response in 8 (including 1 death). The number of cases and follow up is insufficient to compare the response rates of the different preparations, but there was no difference in toxicity profile.

Conclusions : This study shows that administration of ATG has acceptable toxicity, provided facilities are available in terms of blood and platelet transfusions along with management of infective and immunological complications. After ATG therapy and with cyclosporin maintenance, a response may be expected after 3 months.
plasma haemoglobin with a range of 3.5 mg% to 19.0 mg%. 26 of
the cases had undergone a bone marrow examination of which 10
marrows (28.6%) were hypocellular. All the patients were
managed with supportive care wylsolone and/or danazol. Menabol
and Cyclosporine were given to the patients with aplastic anaemia.
Danazol seems to be an effective drug for management as a good
number of patients in our series showed improvement achieving
transfusion independence.

Conclusion: PNH is a relatively uncommon disease but not
rare as we found 35 cases over a 10 year period. As ours was a
hospital based study we could not make any estimates of
prevalence/incidence in the Indian population. Commonest
manifestations are those of anaemia and about 40% gave a history
of coca coloured urine at some point of time. Thrombotic episodes
are rare.

* Adjudged Best Papers and got an award of Rs. 1000/- each from Chairman Scientific Committee, Diamond APICON 2005.