Haemophagocytosis in Bone Marrow Aspirates

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In the paper entitled 'Haemophagocytosis in Bone Marrow Aspirate – A Review of the Clinical Course of 10 Cases, Linn et al. [1] analysed 10 cases of haemophagocytosis seen in marrow. The authors observed that 8 patients had a classical haemophagocytic syndrome and an unusually high number (6 cases) had confirmed lymphoma.

Haemophagocytosis occurs in a wide variety of clinical conditions. The diagnosis of the syndrome, however, requires the fulfilment of some additional clinical and laboratory criteria [2, 3]. Since 1993, as a result of our collective work at two large referral hospitals (Maternity and Children Hospital at

Jeddah, Saudi Arabia and Maulana Azad Medical College at New Delhi, India), prominent haemophagocytosis was seen in 14 cases whereas the classical syndrome was diagnosed in 3 (table 1). Ten of these cases were observed in association with a variety of infections, some of which are commonly diagnosed at these hospitals, namely malaria, enteric fever and tuberculosis.

Linn et al. have not submitted data to show that all the criteria advocated by the Histiocyte Society for the diagnosis of a haemophagocytic syndrome were met. Hyper-triglyceridaemia occurs consistently in familial haemophagocytic lymphohistiocytosis

Table 1. Diseases associated with haemophagocytosis with or without haemophagocytic syndrome

Diagnosis
Haemophago-
Haemophagocytic

cytosis

syndrome

Plasmodium falciparum

4

1

Plasmodium vivax

2

-

Enteric fever

2

-

Miliary tuberculosis

-

EBV infection

1

FHL

1

Ulcerative colitis

-

Malignant histiocytosis

-

Gastric adenocarcinoma

NI

Total cases

14

3

Criteria according to Favara [3]. Figures represent the number of cases. EBV = Epstein-Barr virus; FHL = familial haemophagocytic lymphohistiocytosis; NI = not investigated for all criteria.

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[3] and was thought to be a useful marker to distinguish this entity from the virus-associated haemophagocytic syndrome [4]. However, Kikuta et al. [5] observed hypertriglyceridaemia in all 6 of their patients with Epstein-Barr virus infection as was seen in one of our patients. It would be interesting to know whether hyperlipidaemia and/or hypofibrinogenaemia were also a feature in the cases reported by Linn et al.

Finally, we observed that in one patient who had *Plasmodium falciparum* infection, all features of a 'classical' syndrome were present. This suggests that the list of conditions in which a secondary haemophagocytic syndrome may be seen is likely to be larger than suggested previously [3].

References

Linn YC, Tein SL, Lim LC, Lee LH, Teoh G, Goh YT, Tan P: Haemophagocytosis in bone marrow aspirate – A review of the clinical courses of 10 cases. *Acta Haematol* 1995;94: 182-191.

Henter JH, Elinder G, Ost A, FHL Study Group of the Histiocyte Society: Diagnostic guidelines for haemophagocytic lymphohistiocytosis. *Semin Oncol* 1991;18:29-33.

Favara BE: Haemophagocytic lymphohistiocytosis: A haemophagocytic syndrome. *Semin Diagn Pathol* 1992;9:63-74.

Ansbacher LE, Singsen BH, Hosier MW, Grim-minger H, Herbert PN: Familial erythrophagocytic lymphohistiocytosis: An association with serum lipid abnormalities. *J Pediatr* 1983; 102: 270.

Kikuta H, Sakiyama Y, Matsumoto S, Oh-Ishi T, Nakano T, Nagashima T, Oka T, Hironaka T, Hirai K: Fatal Epstein-Barr virus-associated haemophagocytic syndrome. *Blood* 1993;82: 3259-3264.

Announcement

A Syllabus of Thalassemia Mutations (1997)

In collaboration with Dr. Erol Baysal, Dubai, United Arab Emirates, we have prepared an extensive compendium, titled A Syllabus of Thalassemia Mutations (1997), that lists data for each of nearly 200 β -thalassemia alleles (including 17 deletions), the 17 δ -thalassemia and 1 γ -thalassemia alleles, 14 nondeletional and 7 deletional hereditary persistence of fetal hemoglobin determinants, the more than 30 $\delta\beta$ -thalassemia (including $A\gamma\delta\beta$ - and $\epsilon\gamma\delta\beta$ -thalassemia) determinants, as well as the 32 nondeletional and 8 deletional α -thalassemia-2 and 25 deletional α -thalassemia-1 alleles. Frequency data are reviewed in numerous tables and special sections on the Swiss type of hereditary persistence of fetal hemoglobin and Hb H disease are provided. The Syllabus will be available for distribution in the summer of 1997 at a cost of US\$ 50.00/copy plus US\$ 5.00/copy shipping and handling for domestic first class mail, and US\$ 15.00/copy shipping and handling for foreign air mail. Checks, money orders, or purchase orders should be made payable to the Sickle Cell Anemia Foundation.

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