806  Correspondence


Str., A double-contrast arthrographic technique was used in the first six frozen shoulder patients studied, but no advantage was found over the single-contrast technique, and the latter technique was used in the remaining patients. These details were unfortunately omitted from the final draft of the paper. However, the procedures were performed under x-ray control, and adequate joint outline was obtained with both methods. There was no difference in the rate or extent of recovery of pain or range of passive movement and no difference in the frequency or severity of side effects with either technique. We therefore stand by our observation which is, as Wright and Haq 1 suggested, that arthrographic features are not consistent in patients with painful stiff shoulders.

Department of Rheumatology, A I Binder
Whittington Hospital, London N19 5NF.
Department of Rheumatology, B Hazleman
Addenbrookes Hospital, Hills Road, Cambridge.
Department of Radiology, P Tudor
Addenbrookes Hospital, Cambridge.

Reference

Still’s disease and haemophagocytic syndrome

Str., We were very interested in the reports by Heaton et al. 1 and by Morris et al. 2 on two patients with Still’s disease and virus-associated haemophagocytic syndrome (VAHS). Although no research of haemophagocytosis syndrome had ever been made in previous reports, it is striking to observe that acute episodes with similar clinical and biological features had already been reported and considered either as Rye’s syndrome 3 or as consumption coagulopathy 4 in systemic juvenile chronic arthritis.

For many years we have been intrigued by this life threatening complication and reported it in 1979 5 as a consequence of either virus infection, gold therapy, or other recent modifications in drug administration. We recently published a comprehensive study of seven patients, 6 in whom we observed the association of features of consumption coagulopathy, pancytopenia, liver function alterations of various degree, and metabolic disturbances with changes suggestive of proteolysis. In our view, macrophage or other accessory cells such as Kupffer’s cells or endothelial cells might be the main cells responsible for this syndrome. Indeed, we had observed in histological material from our patients features of macrophage activation with phagocytosed material. The role of these cells is also suggested by a comparison of the VAHS occurring in Still’s disease with the main symptoms observed in two other rare and severe conditions: the accelerated phase of the Chédiak-Higashi syndrome 7 and the familial erythrophagocytic lymphohistiocytosis. 8 In the two latter the clinical manifestations include lethargy, fever, hepato-splenomegaly, pancytopenia, and profuse bleeding with laboratory evidence of liver dysfunction, coagulation anomalies of complex origin, with a fibrinolytic process, and possible intravascular coagulation. These two syndromes are known to be associated with haemophagocytosis, 7, 8 and macrophages show in-vitro evidence of hyper-activation. 7 Thus VAHS or drug induced HS in JCA, and HS in Chédiak-Higashi syndrome and in familial lymphohistiocytosis share common features of a probably systemic macrophage activation.

The question remains why systemic JCA patients are more susceptible to virus or drug induced HS. This underlines the vulnerability of patients with systemic JCA, and great caution must be taken when treating with high dose aspirin, gold salts, when adding another non-steroidal anti-inflammatory drug, or when a virus infection occurs. Most of the patients seem to survive only if they are rapidly treated with high dose steroid. 6

Groupe de Recherches d’Immunologie et de Rhumatologie Pédiatriques, 7
INSERM U 132,
Hôpital Necker-Enfants Malades,
149 Rue de Sèvres, 75 743 Paris, Cédex 15.
France

References
Still's disease and haemophagocytic syndrome.

A M Prieur, A Fischer and C Griscelli

*Ann Rheum Dis* 1985 44: 806
doi: 10.1136/ard.44.11.806

Updated information and services can be found at:

http://ard.bmj.com/content/44/11/806.citation

**Email alerting service**

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

**Notes**

To request permissions go to:

http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:

http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:

http://group.bmj.com/subscribe/