A Case of Zieve's Syndrome Presenting with Myalgia: Not to be Confused with Polymyalgia Rheumatica

Sir—We report a case of Zieve’s syndrome initially diagnosed as polymyalgia rheumatica (PMR).

A 59-yr-old woman was referred by her general practitioner with myalgia and a raised plasma viscosity (PV) of 2.25 [normal range (NR) 1.57–1.75], as a case of PMR unresponsive to prednisolone therapy (15 mg o.d.) given for a 2 month period. At the initial hospital consultation, PMR was considered unlikely as her myalgia was generalized, so prednisolone was withdrawn. Although the patient was tearful, examination was otherwise unremarkable. Over the subsequent 8 weeks, her aching worsened, then nausea, vomiting, abdominal pain and dark urine with mild jaundice developed. Her haemoglobin had fallen within 2 weeks from 14.9 to 8.8 g/dl with no evidence of haemorrhage. A reticulocytosis was noted (5.8%; NR < 2), and fragmented erythrocytes were seen on the blood film. Other investigations at this time were: PV 2.13 mPa, bilirubin 41 μmol/l (NR 5–17), alanine aminotransferase 68 IU/l (NR 10–60), alkaline phosphatase 216 IU/l (NR 40–140), γGT 1049 IU/l (NR 7–64), cholesterol 31.1 mmol/l (NR < 5.2), triglyceride 19.2 mmol/l (NR < 2.3), sodium 129 mmol/l (NR 130–155), ferritin 1529 μg/l (NR 15–300). Other tests, including Coomb’s test, viral serology and hepatic autoantibodies were normal. A CT scan of the abdomen showed fatty infiltration of the liver and a normal pancreas.

Zieve’s syndrome consists of jaundice, hepatic dysfunction, hyperlipidaemia and transient haemolytic anaemia associated with alcohol abuse [1], although the latter may not be suspected [2, 3]. Red blood corpuscle metabolism has been found to be abnormal [2, 4–9], predisposing to haemolysis, possibly through a circulating haemolysin, such as lysolecithin [1] which is elevated in such patients [10]. Hyperlipidaemia often precedes the episode of haemolysis and in our case lipaemic serum was noted in several samples during the 4 month period prior to the full-blown presentation. Unfortunately, retrospective measurement of lipids was not possible. Haemolysis is thought to be precipitated by a rapid fall in lipids [2, 10], which is supported in part by our case as the reticulocytosis peaked after the fall in lipids. Cholesterol has been shown to have an inhibitory effect on the action of lysolecithin [11] and it is postulated that lysolecithin acts only when lipid levels fall.

A raised PV has not been reported previously. This led to an initial diagnosis of PMR in our patient,

![Graph](https://example.com/graph.png)

**Fig. 1.**—Changes in cholesterol (Chol: mmol/l), triglyceride (TG: mmol/l), reticulocyte count (Retics: %), haemoglobin (Hb: g/dl), alanine transaminase (ALT: IU/l), alkaline phosphatase (Alk Phos: IU/l) and bilirubin (Bil: μmol/l) during a 20 week period following admission with Zieve’s syndrome.
although the generalized nature of myalgia and the lack of response to prednisolone suggested otherwise. It is worth speculating that extreme hyperlipidaemia, as in this case, caused elevation of the PV. This would explain the elevated PV at the onset of her symptoms when lipaemic serum was also noted. Similarly, myalgia has not been reported. Although alcohol abuse causes an acute or more chronic myopathy which may be painful [12], the myalgia may have been, at least in part, psychosomatic in origin due to her stressful domestic situation. This case illustrates the importance of a careful clinical assessment when presented with the combination of an elevated PV and myalgia.

J. C. Martin, A. Ross,*, D. Watson,† M. M. O'Sullivan

Departments of Rheumatology, *Medicine and †Haematology, Maelor Hospital, Wrexham, Clwyd, LL13 7TD

Accepted 13 November 1995

Correspondence to: J. C. Martin, Department of Rheumatology, Maelor Hospital, Wrexham, LL13 7TD.

Polymyalgia Rheumatica as the Rheumatological Manifestation of Myelodysplastic Syndrome in a Chinese Patient

Sir—Myelodysplastic syndrome (MDS) is a heterogeneous group of clonal stem cell disorders character-ized by ineffective haemopoiesis resulting in cytopenias with normal or increased marrow cellularity. Rheumatological features were found in 10% of patients with MDS [1]. These included cutaneous vasculitis, peripheral neuropathy, lupus-like syndrome, Achilles tendinitis and seronegative polyarthropathy which might precede the diagnosis of MDS [2, 3]. However, it is not until recently that Kohli and Bennett [4] described the possible association of polymyalgia rheumatica (PMR) and MDS. As PMR is fairly common in Orientals, the presence of another disorder like a haemic malignancy may well be a chance co-existence of two separate diseases. We report a Chinese patient with MDS who presented concurrently with PMR symptoms which responded to low-dose steroid therapy. The relative rarity of PMR in our locality suggests that the two diseases may be causally related.

A 59-yr-old Chinese woman presented in January 1993 with malaise, weight loss, anorexia, polyarthralgia and limb girdle stiffness. She was anaemic and leucopenic (Hgb 7.0 g/dl, MCV 92.4 fl, WBC 2500/ mm3 with 75% neutrophils, 5% lymphocytes, 1.5% blasts). A marrow biopsy revealed mild hypercellularity and dysplastic features of the red cell and granulocyte precursors. There were 8% blast cells. Cytogenetic study of the marrow cells was normal. She required monthly blood transfusion, but no chemotherapy was instituted in view of her age and the small proportion of blast cells. She was initially treated conservatively for her joint and girdle symptoms. In March 1993, her polyarthralgia worsened and there was increasing stiffness and pain over the shoulder and pelvic girdles. She was depressed. Physical examination revealed marked wasting of the shoulder girdle and hip muscles, but there was no active synovitis. She was pale and had 2 cm splenomegaly. Repeat blood tests confirmed anaemia, leucopenia and elevation of alkaline phosphatase to 230 U/l. Her ESR was 140 mm/h and ultrasonography of the liver did not reveal any obstructive lesions. HBsAg, anti-HCV, rheumatoid factor, ANA and anti-dsDNA antibody were all negative. Her serum IgG was mildly elevated, but there was no monoclonal gammapathy. X-Rays of the affected parts were unremarkable. A temporal artery biopsy was negative. The rheumatological diagnosis was PMR and she responded dramatically to low-dose steroid therapy. The relative rarity of PMR in our locality suggests that the two diseases may be causally related.

The annual incidence of PMR has been estimated at around 50/100 000 for persons over the age of 50 yr [5]. The great majority of patients are Caucasians and PMR is an uncommon disease in Orientals. The reason for this ethnic difference is unclear and a genetic factor probably contributes because certain HLA class II antigens, like DR4, have been found to be more prevalent in Europeans with PMR [6].