A case of Budd-Chiari Syndrome presenting in a lady with newly diagnosed Churg-Strauss Syndrome

Sir, We present a case of Budd-Chiari Syndrome presenting in a lady with Churg-Strauss Syndrome (CSS). A 39-year-old asthmatic lady presented in 2001 with a 6 week history of rash on her lower limbs. She was pyrexial and her peripheral blood film showed marked eosinophilia (56.9% of total white cell count). She was nauseated and had evidence of a lower limb peripheral neuropathy. On admission, ESR was 4 mm/h; coagulation screen, ANA and ANCA were normal. C3 was normal and C4 was low at 0.04 (normal range 0.19–0.45 g/l). Liver function tests were abnormal with raised transaminases. Skin biopsy confirmed an acute leukocytoclastic vasculitis within the dermis and a bone marrow aspirate showed increased eosinophil production with discreet areas of bone marrow necrosis. A diagnosis of CSS was made; she was commenced on high dose oral corticosteroids and intravenous immunoglobulin to reverse the thrombocytopenia. Her clinical condition improved rapidly and her peripheral eosinophil count returned to normal within 2 days.

Four days following admission, she developed abdominal discomfort and swelling, confirmed by ultrasound to be ascites. A diagnostic paracentesis revealed a low eosinophil count in the ascitic fluid and no organisms. A Doppler ultrasound showed absent flow in several hepatic veins due to thrombosis and a diagnosis of Budd-Chiari syndrome was made. She was commenced on heparin and warfarin resulting in a dramatic improvement in symptoms and repeated ultrasound showed resolution of both thrombus and ascites. Anticoagulation was continued for 6 months after which there was no recurrence of thrombus or ascites. Following discharge she received two intravenous pulses of cyclophosphamide (stopped after development of a macular-papular rash), and subsequently oral azathioprine. Prednisolone dose was gradually reduced and she was discharged from clinic after 1 year. She was re-referred in 2004 after developing joint pains, tiredness and further vasculitic rash. ESR was 81 mm/h and eosinophil count 10.5%. Her symptoms responded to 10 mg of prednisolone, which was reduced and stopped over a period of 4 months. She is currently well and has had no further relapses.

Hepatic vein obstruction (Budd-Chiari syndrome) has been reported in association with idiopathic hypereosinophilic syndrome [1], but this is the first reported case in the context of CSS. This association may have been a coincidence, however thrombosis has been documented in CSS previously [2, 3]. Reported cases have been associated with either normal or raised eosinophil counts at the time of thrombus detection. Ames et al. [2] reported three patients with CSS who developed thrombosis; subclavian vein thrombosis, deep vein thrombosis and middle cerebral artery territory ischaemic stroke. In all three cases, eosinophilia was present at time of thrombosis and the patients all had raised levels of fibrinogen. Garcia et al. [3] reported three cases of venous thromboembolism (deep vein thrombosis and pulmonary embolus). Two of these patients had had normal eosinophil counts at presentation, and the third had a blood count with 19% eosinophils.

Thrombosis has also previously been reported in patients with hypereosinophilic syndrome [1], and in a patient with venulitis [4]. It has been hypothesized that there may be a role for eosinophil granule proteins, such as Major Basic Protein (MBP) in the pathogenesis of thrombosis in conditions in which eosinophilia is a feature [5]. Wang et al. [6] hypothesize that the HOSCN, produced by eosinophils, may lead to a pro-thrombotic and pro-inflammatory state occurring in cases of thrombosis in conditions associated with raised eosinophil counts. These authors found that HOSCN induces tissue factor activity in human umbilical vein endothelial cells and the pro-inflammatory pathway p65/p50 NF-κB [6]. Young et al. [4] report a case of Budd-Chiari syndrome in which liver biopsy revealed multiple non-caseating granulomas, and a necrotizing granulomatous venulitis involving the hepatic vein, and a venogram showed hepatic venous obstruction. No systemic cause for thrombosis was found, and the patient had normal eosinophil count and coagulation. This patient improved with oral prednisolone 40 mg/day. Ames et al. [2] state that increased release of von Willebrand factor (vWF) from endothelial cells in vasculitis may contribute to thrombosis. Although not measured in our patient, raised levels of vWF may have been a contributing factor.

In conclusion, as far as we are aware, this is the first reported case of Budd-Chiari Syndrome presenting in association with CSS. Thrombosis has been previously documented, and the underlying mechanism may be related to raised eosinophil count or underlying vasculitis.

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The rehabilitation model rules in RA until biomedicine transforms tomorrow’s rheumatologist into a real thaumaturgus

Sir, In their leading article, Bertinotti et al. [1] discuss the new assessment tools for clinical trials and individual patient evaluation in rheumatoid arthritis (RA). They observe that the doctors’ and patients’ points of view often do not match, with doctors focussing on measuring disease activity, which is of little interest to their patients who only want to improve their state of health. They discuss the problems associated with trying to unify the efficacy/outcome data with patient’s need and, in particular, his/her quality of life.

The authors then go on to discuss the OMERACT core set, the ACR20 and the Disease Activity Score (DAS), which are made up of both subjective and objective measures. For example, the DAS is comprised of two objective measures (number of swollen joints and ESR or CRP) for the doctors and two objective measures (number of tender joints and patient’s assessment of global health...
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Combining these two types of measures can cause problems, especially if we are to base our clinical decisions about DMARD management upon them. For example, when the DAS is measured in patients with fibromyalgia (and no RA) they score so highly on the subjective measures that in spite of an absence of joint swelling and a normal ESR, their DAS scores are as high as patients with moderately active RA [4]. About 20% of patients with RA also have fibromyalgia, which will confound the meaning of the outcome measure in these cases [5].

The desire to include the patient’s view in the medical assessment of RA has led to the present day confusion described in the editorial. This comes to no surprise to one mentioned in despatches as a rheumatologist ‘of past decades’ in my ‘long lethargy’ caused ‘by the absence of effective drugs’. In these distant times rheumatology was closely linked to rehabilitation. Indeed my certificate of specialization declares me to be trained in Rheumatology and Rehabilitation. We were all too well aware that we did not have a medical cure for the patient with chronic RA and our multidisciplinary teams managed such patients by means of the bio-psycho-social rehabilitation model [6].

Put simply this postulates that pathology (e.g. active synovitis) gives rise to impairment (e.g. loss of joint movement); which leads to disability (e.g. reduced mobility); which results in handicap (say not being mobile enough to get to the workplace and so not fulfilling the role of the worker). In this model different outcome measures are used to measure its different functions. For example, pathology—ESR and swollen joints; impairment—range of joint movement; disability—HAQ; and handicap—a patient-generated measure like the Patient Generated Index (PGI) [7]. The functions are also used as a focus of the rehabilitation plan. For example, DMARDS are used to minimize the ESR and number of swollen joints; physiotherapy is used to try to maximize the range of joint movement; gait retraining, a walking stick and car modification is used to improve mobility so that the patient can return to whatever activities he/she has chosen in her PGI [6]. The bio-psycho-social approach ensures that factors not directly associated with the pathology such as pain, fatigue, deformity, depression, family factors and coping style, etc. are considered.

This model with its different outcome measures and focuses for a rehabilitation plan is far more likely to meet the need of the patient. Today’s rheumatologists are much closer to the thaumaturgus (miracle worker) quoted by Bertinotti et al. than those of past decades. We have powerful designer drugs that have a dramatic effect on the pathology of RA. Unfortunately, we do not yet have a cure and almost every patient still has a degree of impairment, disability and handicap. Until we are able to eradicate RA we need to apply the biomedical model strictly to the drug treatment and concentrate only on the pathology by using measures such as number of swollen joints, volume of synovial hypertrophy measured by ultrasound or MRI and acute phase reactants. To meet the patient’s needs we should move to the bio-psycho-social rehabilitation approach to facilitate the multidisciplinary team in minimizing the patient’s handicap.

When biological science has developed treatment to obliterate RA, so that the rheumatologist is a real thaumaturgus, the medical model on its own will suffice, until then we need to use separate outcomes measures for the doctor and the patient. Combining them sometimes results in confusing signals that are all too easy for the biomedically based doctor to misinterpret and leave the patient unheard.

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Rehabilitation, an old useful tool in the midst of a new therapeutic era for rheumatoid arthritis

Sir, Dr J. G. Jones raises an important issue about rehabilitation that was neglected in our editorial. The rheumatologist should look at the triad pathology–disability–handicap not only from the point of view of the outcome measures, but also as a ‘world’ where, after the diagnosis is made and the treatment started, an active rehabilitation procedure should be seen as an imperative part of the therapeutic strategy, and started as soon as possible. This attitude may eventually lead the rheumatologist to add another valuable weapon in the fight against disease-related disability. In fact, drugs can switch off joint inflammation, but only a correct rehabilitation programme can promote an optimal recovery of joint functionality and range of motion.

On the other hand, some doubt on the real efficacy of rehabilitation in the early phase of rheumatoid arthritis (RA), where treatments usually guarantee the better response, have been recently addressed [1]: the necessity for controlled clinical trials to determine the exact efficacy of different rehabilitation programmes in rheumatic diseases has been stressed. Thus, the rehabilitative approach is currently under scrutiny because of lack of standardization of disease status [2], as indicated by the lack of uniformity and acceptance of the Patient Generated Index (PGI) [3]. Moreover, only few recent studies have considered the proposed bio-psycho-social approach as potentially valuable but still not conclusive [4, 5].

Finally, International Classification of Functioning, Disability and Health (ICF) is considered by the WHO as a powerful tool to study health and health-related states, outcomes and determinants. ICF permits also the comparison of the data obtained from different countries, because it supplies a coded information for health status. ICF was mainly created to improve communication between different users (health care workers, researchers, policy-makers and the public).

In order to correctly evaluate the contribution of rehabilitation to the therapeutic strategy, two problems should be considered: the first that still today the rheumatologists have scarce experience with the ICF, and the second, that likely only tertiary centres are adequately provided with dedicated personnel (physicians and HPs) that can permit the assessment, during a normal 20-min evaluation of the patients status, of all the variables of the bio-psycho-social approach suggested by ICF. In addition, some authors recognize...