SIR, In the June issue of this journal, Thonhofer et al. reported on treatment of a patient with Schnitzler syndrome [1]. We would like to comment briefly on their findings.

The observation that the patient deteriorated when started on treatment with tumour necrosis factor (TNF)-inhibitors, adalimumab as well as etanercept, is intriguing, also with regard to pathophysiology. However, the use of TNF-inhibitors in Schnitzler syndrome was already described in 2005 by Lin et al. [2], whose patient ameliorated when etanercept was given at a dose of 25 mg twice weekly.


We have recently reviewed the cases of Schnitzler syndrome in the literature and concluded that few if any therapies other than the IL-1 receptor antagonist anakinra are able to induce complete remission in Schnitzler syndrome [7]. There is no need for co-administration of methotrexate to anakinra in Schnitzler syndrome [4], we have seen this in at least eight more patients, either in our clinic, published [5, 6] or through personal communication (V. Akhras, London, UK). We feel that co-administration of methotrexate is not indicated, especially in view of its potential side-effects.

We agree that the use of methotrexate (MTX) in SS is not supported by rational insights into the pathogenesis of this syndrome. Our decision to combine anakinra with MTX was influenced by classical therapy schemata of rheumatoid arthritis. Indeed, we tapered and discontinued MTX in this patient and for the now overseen period of 2 months the patient did not relapse while being treated with anakinra monotherapy.

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Letters to the Editor

1741

Comment on: Adult-onset Still’s disease and myocarditis: successful treatment with intravenous immunoglobulin and maintenance of remission with etanercept

SIR, We read with interest the case of dilated cardiomyopathy complicating adult-onset Still’s disease (AOSD) by Kuek et al. [1]. The firm diagnosis of myocarditis was established on clinical
grounds and was substantiated by the raised cardiac specific muscle enzymes. I, however, think that the cause of recurrent and partially responsive myocarditis in an immunocompromised individual who has a long-standing chronic inflammatory illness, merits further study to exclude other causes for myocarditis. This is especially true when an acclam for a successful therapeutic regimen is being expressed and its wider use is encouraged.

The negative viridae serology does not exclude viral myocarditis (by far the commonest cause of myocarditis) since tests of many of the viruses implicated in the causation of myocarditis are not part of the routine virology screen. Furthermore, the fact the patient suffers with AOSD does not rule out other specific forms of idiopathic or viral myocarditis [2, 3]. Chronic bacterial, fungal or infiltrative cardiomyopathies are possibilities needing also to be explored [4].

All these disorders would have similar initial presentation of dilated cardiomyopathy but would greatly differ in the natural course and prognosis of the disease [5, 6]. Many warrant additional or different modalities of therapy and may require cardiac transplant much earlier than cases with inflammatory myocarditis [7].

The positive response to intravenous immunoglobulin is well reported in various forms of myocarditis, especially with viral myocarditis [8–10]. Giving anti-tumour necrosis factor (TNF) therapy is yet another reason for being more diligent in excluding cardiac infective process before the inception in this particular case [11]. This patient already had three attacks of acute myocarditis with dramatic course.

For all of the above, I think a histological diagnosis using transvenous cardiac biopsy would be the appropriate step at the outset of any future relapse. The procedure is relatively safe and can be done as a day case [12]. The information gained will be invaluable. Polymerase chain reaction (PCR) for specific viruses and bacterial studies for chronic infective myocarditis can be readily tested [13]. The various histological examinations and immunohistochemical studies will exclude other primary forms and infiltrative disorders. The histopathological picture would after all confirm the diagnosis of inflammatory myocarditis. A biopsy directed therapy would present a solid cause and effect evidence. It would also substantiate the clinical decision to use other targeted therapy.

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Comment on: Adult-onset Still’s disease and myocarditis: successful treatment with intravenous immunoglobulin and maintenance of remission with etanercept: reply

Sir, We thank Dr Binymin [1] for his interest and comments regarding our case report. Is it certainly true that an infective or other infiltrative cause for the myopericarditis may have been present in our patient. We feel, however, that the recurrence of the cardiac inflammation at the same time as the flare of Adult-onset Still’s disease (AOSD) makes a secondary phenomenon more likely. The fact that the patient’s cardiac function also returned to normal completely in between paroxysms without residual dysfunction supports this association. Furthermore, despite being immunosupressed, with low-dose corticosteroid and methotrexate, the co-existence of two rare diseases would be less likely than a unifying diagnosis. His improvement both from the AOSD and myopericarditis perspective, with high-dose corticosteroid and intravenous immunoglobulin also lends weight to the connection. We agree whole-heartedly that a transvenous cardiac biopsy would have been enlightening; however, the use of an invasive procedure to make a diagnosis, would only be beneficial if specific therapy at the time of critical illness were effective. A scarcity of studies looking at endomyocardial biopsy in the very acute setting exists and its usefulness in tertiary care is controversial [2]. After consultation with our cardiology colleagues, it was felt that under the circumstances and in the light of the previous history and the empiric treatment he had already received, cardiac biopsy was not necessary. If however the patient does relapse, as highlighted by Dr Binymin [1], cardiac biopsy will be contemplated.

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