Recurrent pericarditis in hyper-IgD syndrome

Sirs,

We observed a 12-year-old boy affected by hyper-IgD syndrome (HIDS) who was successfully treated with etanercept, but developed a recurrent pericarditis. The boy suffered, since the age of three months, from recurrent attacks of fever, abdominal pain and arthritis involving small and large joints. Symptoms resolved within a few days with steroid administration but recurred every two weeks. Laboratory findings showed: IgD: 1550 mg/dl; IgA: 1.010 mg/dl; ESR: 101 mm/h; CRP: 5.00 mg/dl. The boy was diagnosed as having HIDS by demonstration of an homozygous missense mutation (C367S) on exon 11 of the mevalonate kinase (MVK) gene. No mutations in MEFV and TNFRSF1A genes were detected. Because of symptomatic therapy inefficacy, etanercept, a TNF-α antagonist, 0.4 mg/kg twice weekly was started. A dramatic recurrence of pericarditis in this patient as no trauma or infection was reported. The hypothesis that etanercept could predispose our patient to pericardial inflammation, presumably of viral origin, is questionable for at least two reasons: firstly, pericarditis occurred after two years since etanercept was started; secondly, the interruption of anti-TNF-α treatment did not avoid pericarditis recurrence. Surely etanercept was effective in aborting attacks of HIDS in our patient. A complete resolution of symptoms was achieved with etanercept and new recurrences were observed after treatment interruption. Other recent studies suggest that ank2 (5), an IL-1 α antagonist, and etanercept (6, 7) are effective in preventing IgD-related febrile attacks; but data about etanercept are contradictory (8, 9, 10).

In conclusion, recurrent pericarditis is a possible clinical finding in patients with HIDS. Etanercept, an anti-TNF-α drug, seems to be highly effective in the treatment of febrile attacks of HIDS. However, further studies are necessary to better understand the long-term efficacy and safety of anti-TNF-α treatment in these patients.

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References


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Fig. 1. Two-dimensional echocardiogram showing large pericardial effusion (arrow).