Letters to the Editors

Adult-onset Still's disease: not always so good

Sirs,

Over the last decade, more attention has been paid to adult-onset Still's disease (AOSD). The search of the key words via MEDLINE (PubMed) ["AOSD" or "Adult Onset Still's Disease" or "Adult Still's Disease"] retrieved 1113 articles published from 1971 to June 2011. Since the beginning of 2011, 23 articles have been published including Riera *et al*. (1). This work described the clinical features and outcome of 41 Spanish patients with AOSD. Even if in seven cases (17%) the use of a biologic agent was required to control disease activity, an overall good outcome was described and no life-threatening complications were reported. This finding is in line with the general belief that AOSD is a rare disorder with a good prognosis. However, we would like to highlight that it may represent a troubling condition needing prompt intervention to prevent a fatal outcome. The cohort from Spain is quite similar to ours (2): both included 41 patients with no significant difference in sex, age (at onset and diagnosis), clinical and laboratory findings. Fever followed by rash and arthralgia/arthritis were the most common symptoms. In our series four patients (9.7%) had a disease resistant to traditional approach and were treated with biological drugs: etanercept in two patients and anakinra in the other two. Another woman was successfully treated with anakinra but she was not included in the description because she was of a different ethnic origin (3). Two out of our 41 patients (4.8%) experienced life-threatening complications such as disseminated intravascular coagulation (DIC), acute respiratory distress syndrome (ARDS) and heart involvement; both required intensive care, and one died. In this case, the autopsy demonstrated myocarditis, serofibrinous pericarditis, pleuritis, and pneumonic infiltrates. Also the other patient experienced DIC accompanied by a myocarditis which resulted in hypokinesia of the myocardium and high frequency atrial fibrillation. The patient was treated with fresh frozen plasma, antithrombin III, and high doses iv corticosteroids (CCS) with benefit. In another large series from Italy, 5 out of 69 (7.2%) patients with AOSD presented a life-threatening complication leading to death in one case (4). As far as we know, 26 fatalities have been reported among the 932 patients with AOSD in the 18 cohorts published over the last 25 years, including at least 14 patients (mortality 2.75%, range 0–9.8%) (Table I). The most frequent conditions potentially leading to a devastating outcome are: DIC and reactive haematophagocytic syndrome (RHS), alone or in association, ARDS, myocarditis (sometimes complicated by cardiac tamponade) and liver failure, more rarely thrombotic thrombocytopenic purpura, haemolitic uraemic syndrome, aseptic meningoencephalitis, neuropathy and status epilepticus [reviewed in 2]. Although all these manifestations are rather rare, clinicians should be aware that AOSD can present with an acute life-threatening onset which may rapidly progress to death; such fearful evolution can be halted by the prompt use of high dose CCS and biologic agents.

Table I. Mortality in 18 studies including at least 14 patients published from 1986.

References	n. patients	M/F	n. deaths	% deaths
Wouters JM (1986) (cited in 2)	45	18/27	0	0%
Reginato (1987) (cited in 2)	23	12/11	1	4.3%
Ohata (1990) (cited in 2)	90	23/67	4	4.4%
Pouchot (1991) (cited in 2)	62	34/28	2	3.22%
Masson (1995) (cited in 2)	65	31/34	1	1.5%
Mok (1998) (cited in 1)	16	5/11	0	0%
Appenzeller (2005) (cited in 1)	16	9/7	0	0%
Singh (2008) (5)	14	9/5	1	7.14%
Mehrpoor G. (2008) (6)	28	7/21	0	0%
Cagatay (2009) (cited in 2)	84	25/59	0	0%
Zeng (2009) (cited in 2)	61	16/45	6	9.8%
Zhu (2009) (7)	77	23/54	1	1.2%
Franchini (2010) (4)	69	29/40	1	1.4%
Priori (2010) (2)	41	18/23	1	2.4%
Kim (2010) (8)	54	15/39	5	9.3%
Lindi Jiang (2011) (9)	70	26/44	3	4.9%
Colina (2011) (10)	76	32/44	0	0%
Riera (2011) (1)	41	16/25	0	0%

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