How to improve our ability to predict adverse events in major surgery of SLE patients?

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Tiziana Bertero[a], Francesco Parisi[b], Giovanni De Rosa[c], Savino Sciascia[a, d,]

[a] Clinical Immunology Department, AO Mauriziano, Umberto I, Via Ferdinando Magellano 1, 10128, University of Turin, Italy
[b] UO Cardiochirurgia, AO Mauriziano Via Ferdinando Magellano 1, 10128, Torino, Italy
[c] UO Anatomia Patologica, AO Mauriziano, Via Ferdinando Magellano 1, 10128, Torino, Italy
[d] CMID, Coordinamento Malattie Rare Piemonte e Valle d'Aosta, Ospedale Torino Nord Emergenza San G. Bosco ed Università di Torino, Piazza del Donatore di Sangue 3, 10154, Torino, Italy

Abstract

Monitoring organ damage in systemic lupus erythematosus (SLE) patients addresses the aspect of the disease which is irreversible, independently of its cause: SLE , drugs and/or co-morbidities. Damage accrual correlates with morbidity, mortality and impaired quality of life. Once damage has occurred further deterioration is to be suspected. Cardiovascular (CV) events are considered the first single cause of death in SLE-patients, partly attributable to accelerated atherosclerosis; thus, monitoring traditional and modifiable CV risks in SLE patients is recommended. Damage assessment could be useful in evaluating cardiovascular surgery risks in SLE patients. In this report the clinical course after an ascending aorta and aortic valve replacement in a 32 year-old caucasian woman is described. Perhaps, time has come for a worldwide challenge to create an updated score to quantify damage in SLE patients.

Keywords

SLE; Disease activity; Organ damage

Sir,

In response to the interesting article “Comparative assessment of vascular function in autoimmune rheumatic diseases: Considerations of prevention and treatment”, recently in Autoimmunity Reviews [1], we would like to offer our experiences in such a complex field.

Monitoring organ damage in SLE patients addresses the aspect of the disease which is irreversible, independently of its cause: SLE, drugs and/or co-morbidities. Damage accrual correlates with morbidity, mortality and impaired quality of life. Once damage has occurred further deterioration is to be suspected [2]. Cardiovascular events are considered the first single cause of death in SLE patients, partly attributable to accelerated atherosclerosis-AS; thus, monitoring traditional and modifiable CV risks in SLE patients is recommended[3] and [4]. Damage assessment could be useful in evaluating cardiovascular surgery risks in SLE patients.

In this report the clinical course after an ascending aorta and aortic valve replacement in a 32 year-old Caucasian woman is described.

She was diagnosed with lupus nephritis (LN) at the age of 8. Her SLE was marked by LN, neuropsychiatric manifestations (NPS), arthritis and mucocutaneous involvement, ANA and antiDNA. In 2006, following an unexplained fetal death at 19 weeks gestation, associated to lupus anticoagulant, a diagnosis of Antiphospholipid Syndrome was made. She was treated with steroids, cyclophosphamide (cumulative dose more than 14 g), azathioprine, cyclosporine and mycophenolate, as well as cardioaspirin and antihypertensive drugs. In 2008, when admitted to our centre,

hydroxychloroquine therapy was started [5]. In 2010, 6 months prior to cardiosurgery, she was hospitalized for a pulmonary life-threatening infection followed by autoimmune haemolytic anemia and thrombocytopenia. The clinical picture resolved in an end stage renal disease-ESRD requiring dialysis or renal transplantation. Due to a 50 mm diameter aorta aneurysm associated to moderate regurgitation of the aortic valve, an ascending aorta replacement was performed; after that, because of the regurgitation of the aortic valve a mechanical prosthesis was implanted.
The consistency of the aortic wall was described by the cardiac surgeon as "supple as jam" (Fig. 1).
The post-operative course was complicated by late recovery of consciousness, cardiac ischemia needing stenting, spinal subdural hematoma, pancreatitis, *Candida* sepsis and iatrogenic thrombocytopenia. The clinical setting was further marked by enteric ischemia and she died two months after the cardiosurgery.
The concomitant presence of juvenile-onset of SLE, great overall disease activity, NPS and renal involvement, cumulative high doses of steroids (which she had taken continuously since 1984) and cyclophosphamide, APS and conventional CV were all poor prognostic factors in our patient.

It is known that in ESRD patients metabolic factors such as uremic toxins not completely removed by dialysis, hormone deficiency, inflammatory mediators and endothelial dysfunction are responsible for increased cardiovascular risk [6] and [7]. SLE seems to be an adjunctive, independent risk factor for death in ESRD [8].
To date, we still wonder whether this surgical intervention should have been avoided. How can we improve our ability to predict adverse events in these patients? Are the tools we commonly use proper to quantify the risk and to predict outcomes of major surgery? Could arterial distensibility assessment be useful [9]? Is the cardiovascular involvement comparable in every vascular district [10]?
Obviously, we do not have answers to these questions. Some suggestions may arise from this experience, as the idea of including dialysis in predictors of CV damage progression in SLE. Moreover, a more extensive use of antimalarials, especially in nephrologic surrounding, is largely desirable [11].
In conclusion, a multidisciplinary assessment in evaluating critical SLE patients is mandatory when surgical treatment is planned, particularly if it is a matter of major surgery[12]. Perhaps, time has come for a worldwide challenge to create an updated score to quantify damage in SLE patients [13].

Take-home messages

•Cardiovascular events are considered the first single cause of death in SLE-patients.
•A multidisciplinary assessment in evaluating critical SLE patients is mandatory when surgical treatment is planned.
•Time has come for a worldwide challenge to create an updated score to quantify damage in SLE patients.

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Fig. 1.
Histological findings revealed irregular thickness and ulcerations of intimal surface and a degeneration of the tunica media. No sign of vasculitis was observed.