Autoimmune type antiphospholipid antibodies in a patient with Q fever

Q fever is a zoonosis caused by Coxiella burnetti which has been associated with a range of haematological manifestations like bone marrow necrosis, haemophagocytosis, haemolytic anaemia, transient mononuclear gammopathy, lymphadenopathy mimicking lymphoma, transient hypoplastic anaemia and spontaneous splenic rupture. Also a number of autoantibodies have been reported in association with acute Q fever including antiphospholipid antibodies –aPA (lupus anticoagulant-LA), anticardiolipin –aCA- and anti-β2 glycoprotein-I-β2gpl-) which are one of the most relevant autoimmune markers reported in this setting.1

Typically (but not always) the aPA transiently found in association to infectious diseases belong to the infectious type (β2gpl-independent) as opposed to the autoimmune type (β2gpl-dependent) frequently reported in patients with autoimmune disorders. This positivity for anti-β2gpl antibodies along with LA are considered the most specific aPA for the development of clinical manifestations of the antiphospholipid syndrome, but still an increased thrombotic risk has been reported for individuals with the so called infectious-type aPA.2,3

We report a case of a patient with acute Q fever who developed a transient strong global positivity for LA, aCA and anti-β2gpl consistent with autoimmune type aPA.

A 39-year-old woman presented with isolated spiking fever, fatigue, myalgias and arthralgias over the last 2 weeks and hepatosplenomegaly as the sole relevant finding; other microbiological test results were irrelevant. A positive serological result revealed an isolated abnormality of liver function tests suggesting lymphoma, transient hypoplastic anaemia and anemia which along with appropriate HLA presentation and structurally homologous peptides shared by many other viral, bacterial or parasitic microorganisms.

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Antiphospholipid antibodies complicating acute Q fever are transient in virtually all cases and they tend to clear before the positivity of specific serological assays do.8 IgM of aPA are mainly produced but IgG elevations are also often found. From our knowledge there are no data available on the prevalence of anti-β2-gpl antibodies in patients infected by this microorganism. Some authors have suggested that given this consistent association between a prolongation of the APTT over 10 seconds the control time and a clinical scenario consistent with Q fever can be useful to reinforce the degree of suspicion of Q fever, especially in cases like ours in which fever is the only presenting feature and an aPA can be demonstrated.7 Anyway these clinical and biological features can be reproduced in a number of other infectious diseases and lack any reasonable degree of specificity to give them such a degree of diagnostic value.

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Antiphospholipid antibodies have been reported to be a common event in the setting of infection by Coxiella burnetti; actually the prevalence of aCA and LA is between 42-54% and 76% respectively.6 IgM isotypes of aCA are mainly produced but IgG elevations are also often found. From our knowledge there are no data available on the prevalence of anti-β2-gpl antibodies in patients infected by this microorganism. Some authors have suggested that given this consistent association between a prolongation of the APTT over 10 seconds the control time and a clinical scenario consistent with Q fever can be useful to reinforce the degree of suspicion of Q fever, especially in cases like ours in which fever is the only presenting feature and an aPA can be demonstrated.7 Anyway these clinical and biological features can be reproduced in a number of other infectious diseases and lack any reasonable degree of specificity to give them such a degree of diagnostic value.

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mune phenomenon; phase II antibodies to Coxiella burnetti and aCA are different antibodies and the aCA activity in patients with Q fever is β2-gp1-independent in over 85% of cases. Occasionally Q fever-associated aPA are of the autoimmune type but no manifestation of the antiphospholipid syndrome has been reported in those cases so far. Our patient is an example of this type of aPA presenting with a strong and asymptomatic positivity for all the aPA routinely tested.

The general lack of thrombotic complications associated to these type of aPA makes any type antithrombotic therapy unnecessary; however the slightly increased risk of thrombosis reported in individuals with infectious-driven aPA makes prophylactic measures reasonable during the positivity of the antibodies should clinical situations susceptible of a significantly increased thrombotic risk arise during that period.

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References