TWO CASES OF FATAL CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME

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Introduction

Catastrophic antiphospholipid syndrome (CAPS) is a rare and fatal condition secondary to antiphospholipid syndrome (APS) characterised by venous and/or arterial thromboembolism within a short period of time in the presence of positive AP antibodies. Thrombocytopenia is seen in >60% of cases. Here we report 2 cases of women with fatal CAPS.

Case reports 1.

A 55-old female affected by APS (excluded overlap SLE), treated initially with warfarin, due to previous DVT, than apixaban after a new EP episode, was admitted to our Unit for dyspnea: we found a mitral valve injury. After few days she developed a neurologic impairment with evidence of post-ischaemic alterations, likely embolics, and a sensory–motor neuropathy. We concluded for probable CAPS and started antibiotics plus high dose corticosteroids, unfractionated heparin and plasma exchange. However, we observed further worsening. The patient quickly succumbed to her illness. Case report 2.

A 59-old female affected by SLE with CSN impairment and APS with thrombocytopenia and recurring DVT – EP episodes treated with low dose methylprednisolone, iloprost and heparin (warfarin suspension for severe thrombocytopenia impairment) was admitted to our Unit for appearance of diarrhoea with abdominal pain. Blood tests showed further worsening of platelet count. We started antibiotics with clinical improvement. Taking into account the patient history CT was performed, with evidence of massive arterial and aortic arterial disease severe thrombotic obstruction. Two days later she developed acute pain on the left foot with massive venous thrombosis of the left femoropopliteo axis. We started unfractionated heparin, despite low platelet value, as rescue therapy. The patient progressively worsened with further progression of the arterial occlusions, until death, 3 days later.

Conclusion

CAPS can lead to acute multiorgan failure and can be associated with infections or SLE. Early recognition is essential for effective life-saving treatment. Management of these patients is complex, especially in those with thrombocytopenia. Anticoagulation plus steroids plus plasma exchange should are the first line therapy but our two cases show that they are not always applicable and effective. New therapies, such as rituximab and eculizumab, may be options, but time is a fundamental variable.

WOMEN WITH SYSTEMIC LUPUS ERYTHEMATOSUS MORE OFTEN ACHIEVE PREGNANCY, AND HAVE A SHORTER TIME TO PREGNANCY COMPARED TO WOMEN WITH RHEUMATOID ARTHRITIS

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Conclusion

Women with SLE more often achieve pregnancy, and have a shorter time to pregnancy compared to women with RA.
Objectives To examine possible differences in the ability to get pregnant and time to pregnancy (TTP) in women with systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA).

Methods Data from RevNatus, a Norwegian nationwide observational register of women with rheumatic diseases planning pregnancy was used. We compared the number of women with SLE and RA achieving pregnancy, TTP and pregnancy outcome (live birth or pregnancy loss). Fifty-three women with SLE and 180 women with RA with a pregnancy wish had follow-up until pregnancy and known pregnancy outcome or at least one year if not achieving pregnancy. With cox regression we adjusted for maternal age, parity and use of disease-modifying antirheumatic drugs (DMARDs).

Results The two groups were similar concerning maternal age, parity, smoking, BMI, educational level, prior pregnancy loss and prior preeclampsia and/or preterm birth. Women with SLE had longer disease duration (p=0.001), more often active disease (p=0.002) and more often used a DMARD (p<0.001). Live birth occurred in 38 (71.7%) women with SLE and 104 (57.5%) women with RA, while pregnancy loss was experienced in 9 (17.0%) and 26 (14.4%), respectively. Six (11.3%) SLE-women and 50 (27.8%) RA-women did not achieve pregnancy during follow-up (Figure 1a). Women with SLE had almost doubled pregnancy rate compared to women with RA (pregnancy ratio 1.91, CI 1.27 to 2.88, p=0.002), indicating subfertility.

Conclusions Women with SLE more often achieve pregnancy and have shorter time to pregnancy compared to women with RA.

LONG-TERM FOLLOW-UP OF 320 CHILDREN BORN TO MOTHERS WITH SYSTEMIC AUTOIMMUNE DISEASES: A MULTICENTRE SURVEY FROM 24 RHEUMATOLOGY CENTRES IN ITALY

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Background Rheumatic Diseases (RD) frequently affect women during reproductive age, therefore counselling on family planning is crucial for their quality of life. Children’s outcome is a major topic, but no large studies are available. This study aimed at assessing the long-term health conditions of children born to women with RD.

Methods 24 Italian Rheumatology Centres distributed the questionnaire (65 multiple-choice and 12 open-answer