EDITORIAL

CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME – CASE REPORT

Tarig Hashim [1], Ahmed Omer [2], Gubara Abdelateef [3]

(1) Ass. Professor, Department of Obstetrics & Gynecology, University of Bahr Elgazal.
(2) M.Sc. Medical Laboratory Technology, Department of Pathology, University of Khartoum.
(3) Assoc. Professor, Department of Obstetrics & Gynecology, University of Science & Technology.

Correspondence:
Email: tarigelhashimi@yahoo.com
Tel.: 0912134451

Abstract:

A 26 year old lady, her first pregnancy complicated by abruptio placentae at term without obvious cause, outcome was fresh stillbirth. In her second pregnancy, she developed severe headache at seven weeks gestation, confusion, apathy, dementia, fits and left lower limb swelling within a period less than 10 days. Magnetic resonance imaging (MRI) revealed bilateral sinus thrombosis. Low molecular weight heparin (Innohep) was started immediately and gradually increased adjusted by (PTT). The patient gradually improved and discharged on Innohep. In spite of, injectable anticoagulant therapy, she developed intrauterine fetal death at seven month.

She delivered spontaneously. The first result of screening for antiphospholipid antibodies was positive with low protein S. She was diagnosed as probable catastorophic antiphospholipid syndrome (APS). She was discharged on oral anticoagulant (warfarin) with adjusted dose according to PT & INR. After a year of warfarin therapy, with laboratory follow-up, the second result of screening antiphospholipid antibodies remained positive with lower protein S., compared with the first result and; drop in protein C, which increases the incidence of thrombosis and makes further pregnancy unadvisable.

Key words:

Antiphospholipid, autoimmune, innohep, abruptio placentae, heparin, surrogation, miscarriage, pregnancy loss, thrombosis, thrombocytopenia, Sudan.

Introduction:

Antiphospholipid syndrome was known as autoimmune condition in the mid-fifties in the last century with its strong bond with specific autoimmune antibodies (Lupus anticoagulant and anticardiolipin) and classical clinical picture (thrombosis, thrombocytopenia and pregnancy loss) (1)

Doors were opened widely for researches and studies to find out the etiological factors, pathological mechanism, genetic inheritance, epidemiology, and effects on other body systems, optimal means of diagnosis, management, and prognosis.

EDITORIAL

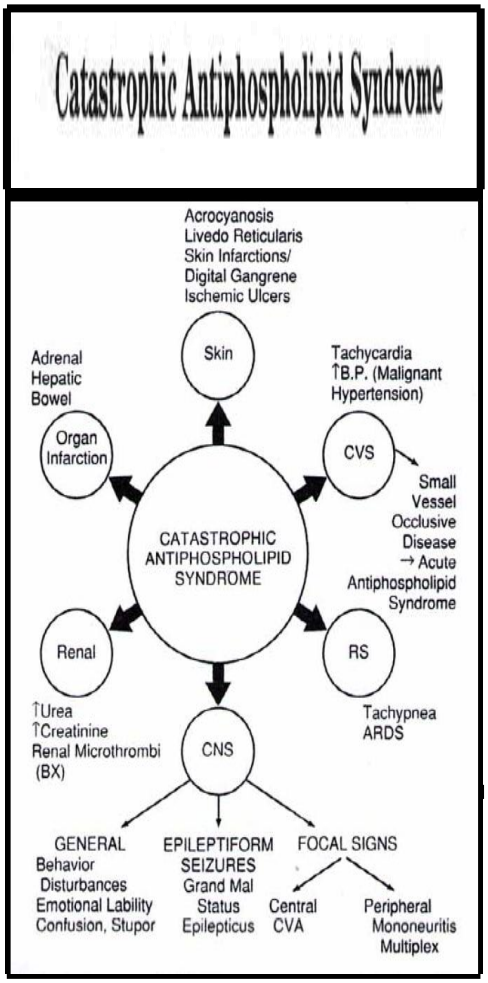

Fig (1)

Source: J. Thromb.Haemostas, 2006; 4:295-306

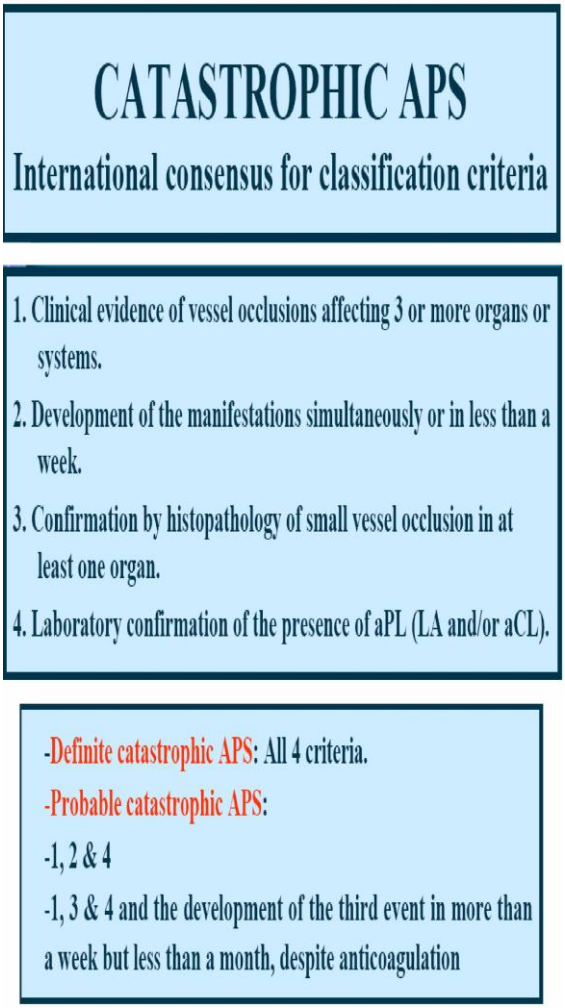

Fig (2)

Source 11th international Congress on Antibodies Sydney, Australia Nov. 2004.

EDITORIAL

The European forums worked out new methods of investigation and clarified classical, unusual and silent manifestations, including the catastrophic form of the syndrome (Fig. 1)**.** The international congress on antiphosophliplid antibodies in Sydney, Australia 2004 updated the classification of the syndrome, and revised the criteria to diagnose the syndrome (Fig. 2) including its catastrophic form, and highlighted the issues, that remained obscure.

Case Report:

The patient was 26 year old Sudanese lady. She has two stillbirths, miscarriage. Her weight was 63Kg, height is 167cm, and blood group is (A+). Regarding gynaecological, past medical history, chronic diseases and congenital are not detected in the family. No consanguinity with her husband. The first pregnancy passed without complications, till term. She was on regular antenatal care visits. She developed spontaneous uterine contractions at term, she was hospitalized, routine investigations and examinations were performed. Labour was accelerated with oxytotcin, when she was at the end of the first stage of labour, the fetal heart sound couldn't be heard. However, acceleration was continued, the outcome was fresh stillbirth. The diagnosis was placental separation.

Fourth months later, the patient developed headache on and off mainly on the right side, two days more the headache became severe and persistent, she received treatment for malaria, enteric fever and antiviral (acyclovir).

The condition day by day deteriorated by developing confusion, apathy, dementia, loss of concentration, and disorientation in place, time, and surroundings, eventually, she developed epileptic seizures. There was swelling in the left calf muscle indicating evidence of (DVT). Accidentally the urine for pregnancy test was discovered to be positive.

The patient was transferred to Khartoum (capital of the Sudan) for further investigations, and management. (MRI), revealed intracranial sinus thrombosis. Basic investigations were requested, including coagulation profile. Low Molecular Heparin (Innohep) was started immediately. The dose increased gradually according to INR to reach 16 thousands per day. She also received dexamethasazone, carbomazipine and beclomethazone inhaler. The patient started to improve gradually. Ultrasound scanning showed 8 week gestation.

While she was on antenatal care visit, in her seven month' gestation ultrasound revealed Intrauterine Fetal Death (IUFD) in spite of taking Innohep with the same dose 16000 IU/ day since she was discharged. The cause most likely due to placental thrombosis and severe placental insufficiency. Fortunately she delivered spontaneously. She was screened for thormbophilia as well as antiphospholipid antibodies. The last two investigations were done in Germany. All laboratory findings regarding system and organs were normal. She was shifted to 9 mg of Warfarin daily. The result of screening for thromophilia and antiphosphilipid antibodies was received and confirmed the presence of antiphospholipid antibodies, and a decrease in protein S.

After more than one year on warfarin 9mg daily, the patient went to Khartoum for follow-up, carrying in her mind two substantial questions that concerned her for the whole last year. The first question is for how long she will be on medication? The second what is the future of her reproductive life as she now aged 27 years old?

Prognosis and Outcome:

The mortality of the condition is high despite therapy. Mortality is of the order of 50%. In a recent analysis of the CAP Registry focusing on mortality (2), the major cause of death was identified in 81/114(71.1%)

patients. However, once patients with catastrophic APS have recovered they usually have stable course with continued anticoagulation. (3)

Discussion:

Further pregnancy is not without hazards and might seriously endanger the patient's life. The site, the mechanism of thrombosis, and the aetiology still are unknown so that no body should take the risk to advice pregnancy, it should be avoided by contraception that does not contain estrogens. Barrier and intrauterine device are advisable methods of contraception.

The new thrombophilia screening result done on 15-9-2009 could give an answer to these questions. The result shows significant decrease in protein S in comparison with the first result in addition to a protein C drop which was normal. The decrease in protein S can be attributed to the prolonged medication with warfarin (4) but the drop in protein C can be explained by other factors. The net result is increased incidence of thrombosis indicating that further pregnancy is unadvisable.

Both the patient, and her husband were counselled for surrogation, that encountered by moral and religious teaching of Islam, in addition to the culture of the eastern societies, however in some societies this method is acceptable. Adoption is acceptable from an Islamic point of view.

Conclusion

Multidisciplinary approach is important in such conditions. No empirical treatment for pregnant ladies. Clinics, including obstetrician, haematologist, and neurologist and specialized centre are crucial for decision-making, in such cases.

So in conclusion the lady experienced episodes of vessels occlusion (DVT), intracranial thrombosis and the third event resulted in IUFD due to the placental thrombosis, however, we can't be sure when she developed IUFD which could have happened immediately after she went back home, therefore, we think that, it is a probable catastrophic antiphospholipid syndrome with low protein S, although protein S is normally low in pregnancy.

Acknowledge:

The authors would like to thank Dr. Anwar Alkordofani head Department of Pathology Faculty of Medicine U of K and Prof. Elsanwsy (Department of Obs. & Gyna. Gazera University), for their advices and invaluable comments.

References

1. Hughes GR. Thrombosis, abortion, cerebral disease and the lupus Anticoagulant. Br Med J 1983; 287:1088-9.
2. Bucciarelli S, Espionsa G, and Cervera R, et al. Mortality in the catastrophic antiphospholipid syndrome: Cause of death and prognostic factors in a series of 250 patients. Arthritis Rheum 2006; 54:2568-2576
3. . Erkan D, Asherson RA, Espinosa G, et al. The long-term outcome of catastrophic antiphospholipid syndrome survivors. Ann Rheum Dis 2003; 62: 530-533
4. Lane DA Mannucci PM, Bauer KH Bertina RM, Bochkov NP, Boulyenkov EK, Milotich JP, Rosendal FR. Inherited thrombophilia part 1. Thromb Haemost. 1996; 76: 561-662.