

C

1

**A Case of Antiphospholipid Syndrome
Associated with Protein C Deficiency**

Y. S. Nam, S. Y. Han, D. H. Choi, T. K. Yoon and K. Y. Cha

Department of Obstetrics and Gynecology, College of Medicine,
Pocheon CHA University, Pocheon, Korea

= Abstract =

A successful outcome of pregnancy requires an efficient uteroplacental vascular system. Since this system may be compromised by disorders of haemostasis associated with a prothrombotic state, maternal thrombophilia might be a risk factor for fetal loss.

Hereditary deficiencies of the naturally occurring anticoagulants are well recognized conditions predisposing to recurrent venous thromboembolism. Since thrombotic phenomena have been implied as a cause of abortion and stillbirth, these deficiencies might increase the risk of fetal demise.

We have experienced a case of antiphospholipid syndrome associated with protein C deficiency in patient with recurrent spontaneous abortion. So we report this case with a brief review of literatures.

Key Words : Protein C deficiency, Antiphospholipid syndrome, Recurrent spontaneous abortion

C (thrombophilia)
(allele)
heterozygous) 가
S
arginine S glutamine
(cofactor) S
C S
가 가
(homozygous) (compound
가
50%
6 506 C
가
C

29

12

2

36.5°C, 80 / 157cm, 71kg, 가 120 / 80 mmHg,

가

estradiol : < 13 pg/mL, FSH : 6.3 mIU/mL, LH : 1.6 mIU/mL, prolactin : 20.5 ng/mL, TSH : 0.53 μU/mL, G : , M : , : 98 mg/dl, PT : 10.5 sec, PTT : 27.4 sec, : , : , 3 : 118 mg/dl, 4 : 42.3 mg/dl, : , G : , : , : , III : 85.4 %, C : 10 % (: 70 - 140 %), S : 60 %

C heparin 가

, Antithrombin, C , S . Antithrombin serine protease thrombin X VIII V plasminogen C (fibronolysis) (van Hinsberg et al., 1985). S C (Dahlback, 1995). 가 (thromboembolism) 가 (venous stasis), thromboplastin

antithrombin, C, S
가 (Conard et al., 1990 ; Tharakan et al., 1993).
(decidua) (Carp et al., 1989).
C
thrombomodulin 가 (Healy et al., 1995).
10 - 15% antithrombin, C, S
가
5 - 10% antithrombin, C,
S C (activated protein C :
APC) 가 20%
(Koster et al., 1993). C
(V Leiden) 1691
(nucleotide) V
(Bertina et al., 1994) .
V Leiden 3%
가 1%
(Rees et al., 1995) .
(Koeleman et al., 1994) . 가
가 (Infante -
Rievard et al., 1991) . C S
(Malia et al.,
1990). C S
(Healy et al., 1995).
Antithrombin III, C, S

가 (uteroplacental insufficiency) 가
가
가
가 V Leiden (homozygosity)
(Koeleman et al., 1994 ;
Rosendal et al., 1995).

가
가
가
C S
thrombin C K (zymogen)
thrombomodulin
S C Va
VIIIa thrombin (Esmon,
1992).

C Va 506
arginine glutamine
Va 506
arginine (biphasic) arginine
306
C 562 arginine VIIIa
336 arginine
C 4 mg/ L
8 S
40% S C4b
C S
가 가 C
가 C
10 C 4%가 가
(Miletich, 1990).

C
 가 C 가
 가 C (penetrance) 가
 C 160 가 가
 (missense mutation)가 B IX
 C 가
 가 C
 가 가
 1 C 2
 C S C V
 C S 3 가
 C C
 가
 10% 가 C
 S 가 C S
 V 506 arginine
 (heterozygote)
 가 가 C S 가
 (clinical penetrance)

REFERENCES

- Bertina RM, Koeleman RPC, Koster T et al : Mutation in blood coagulation factor V associated with resistance to activated protein C. 1994, 369, 64 –67.
- Carp HJA, Frenkel Y, Many A et al : Fetal demise associated with lupus anticoagulant : clinical features and results of treatment. Gynecol Obstet Invest 1989, 28, 178 –84.
- Conard J, Horellou MH, van Dreden P et al : Thrombosis and pregnancy in congenital deficiencies in AT III, protein C or protein S : A study of 78 women. Thromb Haemost 1990 . 63, 319 –20.
- Dahlback B, Carlsson M, Svensson PJ : Familial thrombophilia due to a previously unrecognized mechanism characterized by poor anticoagulant response to activated protein C : prediction of a cofactor to activated protein C. Proc Natl Acad Sci USA 1993, 90, 1004 –8.
- Dahlback B : The protein C anticoagulant system : inherited defects as basis for thrombosis. Thromb Res 1995, 77, 1 –43.
- Esmon CT : Protein S and protein C. Biochemistry, physiology, and clinical manifestation of deficiencies. Trends Cardiovasc Med 1992, 2, 214 –219.
- Healy AM, Rayburn HB, Rosenberg RD et al : Absence of the blood clotting regulator thrombomodulin causes embryonic lethality in mice before development of a functional cardiovascular system. Proc Natl Acad Sci USA 1995, 92, 850 –4.

- Infante – Rievard C, David M, Gauthier R et al : Lupus anticoagulants, anticardiolipin antibodies and fetal loss. *N Engl J Med* 1991, 325, 1063 –66.
- Koeleman RPC, Reitsma PH, Allaart CF et al : Activated protein C resistance as an additional risk for thrombosis in protein C – deficient families. *Blood* 1994, 84, 1031 –35.
- Koster T, Rosendaal FR, de Ronde H et al : Venous thrombosis due to poor anticoagulant response to activated protein C : Leiden Thrombophilia Study. *Lancet* 1993, 342, 1503 - 6.
- Malia RG, Kitchen S, Greaves M et al : Inhibition of activated protein C and its cofactor protein S by antiphospholipid antibodies. *Br J Haematol* 1990, 76, 101 –7.
- Miletich JP : Laboratory diagnosis of protein C deficiency. *Semin Thromb Hemost* 1990, 16, 169 –176.
- Rees DC, Cox M, Clegg JB : World distribution of factor V Leiden. *Lancet* 1995, 346, 1133 –34.
- Rosendaal FR, Koster T, Vandenbrouke JP et al : High risk of thrombosis for factor V Leiden(activated protein C resistance). *Blood* 1995, 85, 1504 –8.
- Tharakan T, Baxi LM, Diuguid D : Protein S deficiency in pregnancy : A case report. *Am J Obstet Gynecol* 1993, 168, 141 –2.
- van Hinsberg V, Bertina R, van Wijngaarden A et al : Activated protein C decreases plasminogen activator – inhibitor activity in endothelial cell - conditioned medium. *Blood* 1985, 65, 444 – 51.