

Stroke in pregnancy: A rare case of protein C and protein S deficiency

Snigdha Kumari, Ashok Kumar Biswas, Sukanta Misra

ABSTRACT

Introduction: Stroke in pregnancy is a rare occurrence, its reported incidence being 11–26 deliveries per 100,000. Ischemic strokes are slightly more common than hemorrhages. Subarachnoid hemorrhage, embolism and cerebral venous thrombosis can also occur, albeit with a lesser incidence. Peak incidence of stroke is in the peripartum and postpartum period. Mortality rate of pregnancy associated stroke is 10.13%. Therefore, rapid recognition and management of these patients are imperative to ensure successful outcome. We report a rare case of stroke during pregnancy which has the protein C and protein S deficiency as its precipitating cause.

Case Report: We report a case of a 24-year-old female, with previous cesarean section in her third trimester, suddenly developed weakness of left side of the body and deviation of face to the right. There was no history of head trauma, fever, unconsciousness or seizure. She did not have any skin rash, arthritis or signs of deep vein thrombosis. There was no history of prior fetal loss. Protein C and protein S levels were low.

Conclusion: We report a rare case of stroke during pregnancy which has the protein C and protein S deficiency as its precipitating cause. Stroke is a recognized complication of pregnancy, is uncommon but feared. Stroke in young adults aged 15–35 years is more common in women with poorer outcome in terms of disability and dependence. Most pregnancy related stroke occurs in peripartum and puerperium.

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Introduction: Stroke in pregnancy is a rare occurrence, its reported incidence being 11–26 deliveries per 100,000. Ischemic strokes are slightly more common than hemorrhages. Subarachnoid hemorrhage, embolism and cerebral venous thrombosis can also occur, albeit with a lesser incidence. Peak incidence of stroke is in the peripartum and postpartum period. Mortality rate of pregnancy associated stroke is 10.13%. Therefore, rapid recognition and management of these patients are imperative to ensure successful outcome. We report a rare case of stroke during pregnancy which has the protein C and protein S deficiency as its precipitating cause. **Case Report:** We report a case of a 24-year-old female, with previous cesarean section in her third trimester, suddenly developed weakness of left side of the body and deviation of face to the right. There was no

history of head trauma, fever, unconsciousness or seizure. She did not have any skin rash, arthritis or signs of deep vein thrombosis. There was no history of prior fetal loss. Protein C and protein S levels were low. **Conclusion:** We report a rare case of stroke during pregnancy which has the protein C and protein S deficiency as its precipitating cause. Stroke is a recognized complication of pregnancy, is uncommon but feared. Stroke in young adults aged 15–35 years is more common in women with poorer outcome in terms of disability and dependence. Most pregnancy related stroke occurs in peripartum and puerperium.

Keywords: Stroke, Pregnancy, Protein C deficiency, Protein S deficiency

How to cite this article

Kumari S, Biswas AK, Misra S. Stroke in pregnancy: A rare case of protein C and protein S deficiency. International Journal of Case Reports and Images 2014;5(3):226–229.

doi:10.5348/ijcricri-2014-03-479-CR-10

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Received: 19 August 2013
Accepted: 03 September 2013
Published: 01 March 2014

INTRODUCTION

Stroke in pregnancy is a rare presentation. The highest incidence of stroke is mainly in the peripartum and postpartum period. Ischemic strokes are slightly more common than hemorrhages. Subarachnoid hemorrhage, embolism and cerebral venous thrombosis can also occur, albeit with a lesser incidence. Cerebral venous thrombosis is a rare complication during pregnancy or the puerperium. The cerebrovascular system may be primarily involved in young adults suffering from anticoagulants deficiency. Women with thrombophilia are also more prone to venous thromboembolism in pregnancy and

puerperium. Acquired or hereditary thrombophilia occur in almost two-thirds of women presenting with recurrent miscarriages, preeclampsia, intrauterine growth restriction, abruptio placentae, or stillbirth, which are associated with microvascular thrombosis in placental blood vessels. Women with venous thromboembolism during pregnancy and especially those with thrombophilia need proper management, based on the type of defect, the detailed family history, and the presence of any additional risk factors. These factors are important for antithrombotic drug therapy during pregnancy and the puerperium, and the thrombo prophylactic strategy for future pregnancies. Low molecular weight heparin is effective in improving the outcome of pregnancy in women with previous obstetric complications. Mortality rate of pregnancy associated stroke is 10.13%. Therefore, rapid recognition and management of these patients are imperative to ensure successful outcome.

CASE REPORT

A 24-year-old female, with previous cesarean section in her third trimester, suddenly developed weakness of left side of the body and deviation of face to the right. She was brought to the emergency, examination revealed stable vitals (pulse 90/min, regular, blood pressure 130/84 mmHg, supine), altered level of consciousness (Glasgow coma scale 10), and left sided hemiparesis (power 2/5 in both upper and lower limbs). Deep tendon reflexes were brisk and Babinski sign was present, associated with upper motor neuron palsy of the left facial nerve. Other systemic examination was normal. There was no history of head trauma, fever, unconsciousness or seizure. She did not have any skin rash, arthritis or signs of deep vein thrombosis. There was a history of vertigo, unconsciousness, lower limb weakness, with recovery within an hour, 4 years back for which she received no treatment. There was no history of prior fetal loss.

An urgent non-contrast magnetic resonance imaging scan of brain was done which showed acute infarct in right centrum semiovale and right gangliocapsular region in the right middle cerebral artery territory (Figure 1). Patient's complete hemogram, liver function tests, serum creatinine and electrolytes were normal. Abdominal ultrasound showed a single, live fetus with normal parameters. Echocardiography was normal. She had normal prothrombin and activated partial thromboplastin time. The patient was seronegative for hepatitis B, C and HIV. Antinuclear factor by indirect immunofluorescence method was negative. Anticardiolipin antibodies, antineutrophil cytoplasmic antibodies (pANCA and cANCA) were negative. Blood homocysteine level, Lupus anticoagulant screen time was normal. Mutation of factor V could not be detected by real time polymerase chain reaction. Protein C (0.9 mg/L, reference range 1.67–3.16 mg/L) and protein S (7 mg/L, reference range 55–123 mg/L) levels were low.

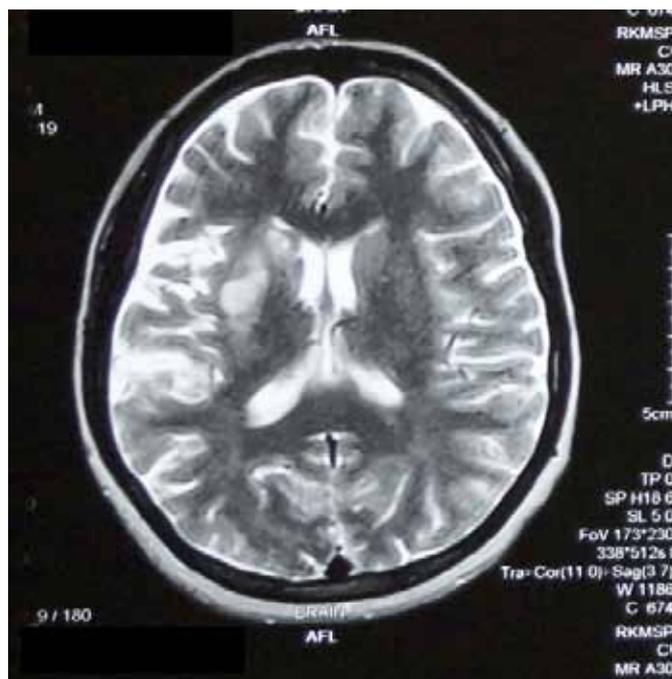


Figure 1: A non-contrast magnetic resonance imaging scan of brain showing an acute infarct in right centrum semiovale and right gangliocapsular region in the right middle cerebral artery territory.

She was diagnosed as a case of protein C and protein S deficiency and was started on low molecular weight heparin (60 mg intravenously twice daily) for 10 days. It was stopped 24 hours prior to elective cesarean section with patient's International normalized ratio 2.0. A three-kilogram healthy male baby was delivered. After 4 hours of cesarean section, low molecular weight heparin (60 mg subcutaneously once daily) was restarted. Subsequently, the patient was maintained with oral anticoagulant (2 mg once daily) with an overlap period of 3 days with a target International normalized ratio of 2.53.

DISCUSSION

Arterial thrombosis is reported in protein C, protein S, antithrombin deficient patients [1]. The incidence of pregnancy related stroke lies between 11–26 deliveries per 100,000 [2]. Prophylactic anticoagulation offers no benefit. Randomized control trials of treatment protocol in women with thrombophilia and pregnancy is needed. Stroke is a recognized complication of pregnancy, has to be taken care of. Stroke in young adults aged 15–35 years is more common in females with poorer outcome in terms of disability and outcome. Most pregnancy related stroke occurs in peripartum and puerperium [3]. Cerebral venous thrombosis is a rare complication during pregnancy or the puerperium [4]. The cerebrovascular system may be primarily involved in young adults suffering from anticoagulants deficiency [5]. Considering

the importance of prothrombotic state, especially caused by deficiency of protein S, any patient presenting with features of cerebrovascular accidents should be thoroughly investigated for any natural anticoagulants deficiency, in whom no other etiologic factors can be determined [6]. Hence, thrombophilia screening might be justified in women with pregnancy loss, and treatment with low molecular weight heparin might be considered for those with pregnancy loss and thrombophilia [7]. Women with thrombophilia are also more prone to venous thrombo embolism in pregnancy and puerperium [8]. Although initial results on use of thrombolysis in pregnancy have been encouraging, further evaluation with regard to maternal and fetal risk is warranted.

CONCLUSION

We report a rare case of stroke during pregnancy which has the protein C and protein S deficiency as its precipitating cause. Stroke in young adults aged 15–35 years is more common in females with poorer outcome in terms of disability and outcome. Hence, women with venous thromboembolism during pregnancy and especially those with thrombophilia need proper management, based on the type of defect, the detailed family history and the presence of any additional risk factors.

Author Contributions

Snigdha Kumari – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ashok Kumar Biswas – Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Sukanta Misra – Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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ACKNOWLEDGEMENTS

No grants or funds were needed for this study. We have no conflict of interest.

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Article citation: Kumari S, Biswas AK, Misra S. Stroke in pregnancy: A rare case of protein C and protein S deficiency. *International Journal of Case Reports and Images* 2014;(3):225–228.



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