

“CASE SERIES OF RARE NEUROLOGICAL DISORDERS IN PREGNANCY”

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ABSTRACT

A variety of neurological disorders may be encountered during pregnancy and perperium. These disorders may be unrelated to the pregnant state (i.e. meningitis) or peculiar to or more prevalent during pregnancy i.e. eclampsia, pelvic neural compression, cortical various thrombosis etc. Pregnancy may affect the course of pre-existing neurological disorders such as epilepsy A secondary neurological disorder (i.e. encephalopathy) can affect a pregnant patient with non-neurological medical disease. A through neurological assessment can distinguish between neurological symptoms that are common in pregnancy (such as dizziness, pain and urinary frequency) and those that might indicate a most significant disorder. All neurological conditions may present with both medical and obstetric challenges to clinicians who are unfamiliar with the management within the context of pregnancy.

KEYWORDS: Pregnancy, Neurological Disorders, Maternal & Fetal Outcome

INTRODUCTION

Aims of the Study

Establishing early diagnosis and early intervention can prevent maternal and fetal outcome in neurological disorders associated with pregnancy.

Material and Methods

From Jan-2014 to May-2014 we received five interesting cases of neurological disorder associated with pregnancy at Bowring & Lady Curzon Hospital, Bengaluru. The management of these five cases is discussed below.

CASE NO 1 PRES (Posterior Reversible Encephalopathy Syndrome) SYNDROME

Twenty year old primi para presented on post OP day 4 after cesarean section with headache followed by 4 episodes of generalized tonic clonic seizures and bilateral visual loss.

On Examination

BP-150/90mmHg, patient was in altered sensorium. CT scan-hypodensity in right parietal region. Patient was treated with valproate, nifedipine and mannitol. During the stay in the hospital, patient's vision, sensorium improved and seizures controlled. Hence discharged after adequate control of blood pressure and advised to continue antihypertensive and antiepileptic. MRI done after 4 weeks was normal.

DISCUSSIONS

Symptoms of PRES

Seizures (74%), Altered sensorium or encephalopathy, Headache, Visual changes (3), Others: aphasia and sensory changes.

Etiology of PRES

Hypertension (61.0%), cytotoxic medications (19.0%), Preeclampsia or eclampsia (6.0%) autoimmune Systemic conditions, including sepsis.

Management of PRES

High degree of suspicion is required to make the diagnosis, Management depends on the etiology and should be initiated in a timely manner. The treatment of the underlying cause is typically sufficient to reverse this condition.

Word of Caution

This condition can lead to irreversible brain insult if treatment is delayed or if there is prolonged brain insult, in which case a brain insult then becomes irreversible brain infarctions.

Conclusions

Urgent diagnosis and early management of this condition are imperative as it directly impacts the neurological outcome. Brain CT perfusion can play an important role in the diagnosis.

CASE NO 2: NON SYSTEMIC VASCULITIC NEUROPATHY

Twenty four years old primi gravid with 22 weeks gestation presented in tertiary care hospital with scan report showing “**INTRAUTERINE DEMISE**”. The Patient was admitted previously in neurology department with bilateral lower limb weakness 20 days back. Superficial peroneal nerve biopsy was done, showed axonopathy of vasculitic etiology. On examination, patient had hypotonic, reduced power and sluggish reflexes in all 4 limbs sparing ocular and facial muscles. Pregnancy was terminated by inducing with mifepristone and misoprostol. ANA and RA factor were negative. After getting discharged, patient lost follow up. This patient needed detailed workup to avoid further pregnancy losses.

Discussions

Vacuities of exclusively the PNS is not infrequent, although less common than systemic vasculitis. This condition is also known as non-systemic vasculitis. Patients typically present with subacute, painful, multifocal, asymmetric, distal-predominant neuropathy.

CASE NO 3: SLE with Recurrent TRANSIENT ISCHEMIC ATTACKS

Twenty two years old gravid 2, abortion 1, with 28 weeks + 5days gestation, presented with pain abdomen and bleeding PV Patient had history of first abortion at two –and half, patient had developed recurrent episodes of headache and giddiness. She was treated in NIMHANS-tertiary care hospital and she was advised not to conceive and started on treatment with Wysolone and ecospirin. Provisional diagnosis of Systemic vasculitis was made. Patient has conceived again after one month. She was asked to continue pregnancy under coverage of heparin. Patient also gives history of photosensitivity, joint pains, hair loss and ulcers since 8 months. Patient developed recurrent attacks of TIA with left hemiparesis and twisting of tongue initially once in 15-20 days, but since one month, once in 2-3 days and resolves on its own without any deficits. Patient expelled a dead macerated baby weighing 1.2 kgs. On post natal day1, Patient developed left sided weakness. On examination, patient has telogen effluvium, oral ulcers in hard palate with pseudo membrane, deviation of tongue to left, deviation of angle of mouth to right, weakness of all 4 limbs, lower limbs & upper limbs. CT scan was normal. ANA reports were not available. Patient’s attenders were not willing to get evaluated further.

She was treated with Wysolone, warfarin and antibiotics. Symptoms resolved with treatment and patient got discharged.

Discussions

Systemic lupus erythematosus (SLE) is an autoimmune disease that predominantly affects women of reproductive age. Pregnancy and its outcome is a major concern to most SLE patients. Queries regarding the risk of disease flares during pregnancy, chance of fetal loss, and the safety of various drugs are often raised. With the improvement in the understanding of the pathogenesis of SLE and the judicious use of immunosuppressive drugs, better disease control can now be achieved and SLE patients should not be deprived of the opportunity for bearing children. Prepregnancy counseling and close collaboration with other specialists such as the obstetricians and the perinatologists is essential in optimizing the maternal and fetal outcome in lupus pregnancies. It is certain that lupus can be life threatening to both the mother and her fetus-infant. During pregnancy, lupus improves in a third of women, remains unchanged in a third, and worsens in the remaining third. Thus, in any given pregnancy, the clinical condition can worsen or flare without warning. In general, women with lupus and chronic vascular or renal disease should limit family size because of morbidity associated with the disease as well as increased adverse perinatal outcomes. The fetus should be closely observed for adverse effects. Fetal growth is monitored, and careful attention is given to the development of hypertension. Unless hypertension develops or there is evidence of fetal compromise or growth restriction, pregnancy is allowed to progress to term. Women with renal disease have a high incidence of gestational hypertension and preeclampsia, but if their disease remains in remission, they usually have good pregnancy outcomes.

CASE NO 4: CEREBRO VASCULAR ACCIDENT (CARDIO EMBOLIC STROKE)

Introduction

Pregnancy is a physiological state associated with increased blood volume, enhanced cardiac output, increased preload, reduced after load and increased contractility of heart. It is also a procoagulant state due to markedly increased blood levels of procoagulant factor I, VII, VIII, IX and X. Pregnancy is associated with decreased protein S levels and reduced activated protein C levels. Venous stasis and alteration in platelet function also occurs during pregnancy. All these changes predispose a patient to both ischemic (arterial and venous) and hemorrhagic stroke during pregnancy and immediate post partum period. We hereby present a patient of rheumatic heart disease with prosthetic heart valve on oral anticoagulants who developed an ischemic stroke in her immediate post partum period. The epidemiology, clinical features and causes of various types of stroke during pregnancy and postpartum period is also reviewed.

Twenty five years old gravida -III, para-II with previous two LSCS, known case of Rheumatic heart disease, Severe mitral regurgitation, mild stenosis, Grade -2 on Inj. Penidure once in twenty one days and Metoprolol 10mg, presented with pain abdomen. Emergency LSCS was done. On post op day 4 Patient developed one episode of generalized tonic clonic seizures. Sudden onset of weakness of Left upper and lower limbs and slurring of speech, deviation of angle of mouth to right. On examination, patient had Left UMN type seventh nerve palsy and UMN type Left upper and lower limb weakness. CT scan showed hypodensity in temporo frontal region in cortical grey white matter of MCA territory. Patient was treated with antiplatelets, anticoagulants, antiepileptic, antibiotics and Mannitol. Patient improved within 24 hours. Early diagnosis of embolic stroke and treatment with antiplatelets is vital in reducing maternal morbidity. Patient has to continue these antiplatelets and Penidure to prevent stroke in future. It is widely believed that pregnancy and the period shortly after pregnancy are associated with an increased risk of stroke. In a study by Kitner SJ et al relative risk for cerebral infarction during pregnancy, adjusted for race and age is 0.7 (95% confidence interval, 0.3 to 1.6) (2). It increases

to 8.7 for postpartum period after live or still birth (95% confidence interval, 4.6 to 16.7)(2). For intra cerebral hemorrhage, the adjusted relative risk is 2.5 during pregnancy (95% confidence interval, 1.0 to 6.4) but 28.3 for post partum period (95% confidence interval, 13 to 61.4). Cardio embolic stroke is the most common cause of stroke in young and an important cause of pregnancy associated stroke. Rheumatic heart disease, prosthetic heart valves and post partum cardiomyopathies cause most of the cardioembolic stroke in these patients (7). Prosthetic heart valve especially ball and cage valve are quite thrombogenic, hence, anticoagulation must be continued during pregnancy and post partum period and international normalization ratio(INR) should be maintained between 2 to 3(8). Even on oral anticoagulation stroke recurrence rate is 4% and 2% per year in patients with mitral and aortic valve disease respectively.

Conclusions

Pregnancy and post partum state are associated with increased risk of ischemic and hemorrhagic stroke. Diagnosis of etiology of stroke during pregnancy and post partum period is a challenge and requires detailed history, physical examination and carefully chosen investigations. Preeclampsia and eclampsia related stroke, cerebral venous thrombosis and cardioembolic stroke are important causes of ischemic stroke in these patients. Patients with valvular heart disease are also at risk of hemorrhagic stroke due to anticoagulants and mycotic aneurysms. Management of these patients involves supportive care and careful use of drugs so as to prevent the stroke recurrence, minimize their side effects by regular monitoring of their dose and at the same time taking care of teratogenic effects and fetal well being.

CASE NO 5: VON HIPPEL LINDAU DISEASE

VHL is a heritable, autosomal dominant, neoplastic disorder that predisposes to development of specific types of benign and malignant tumors. The estimated prevalence of VHL disease is 1 in 35,000 to 1 in 40,000

Clinical manifestations: Individuals with VHL syndrome may be at risk for development of CNS hemangioblastomas, retinal angiomas, endolymphatic sac tumors, clear-cell renal carcinomas, pheo chromocytomas, pancreatic neuroendocrine tumors, epididymal cystadenomas in men, and cystadenomas on the broad ligament in women.

Twenty four years old primi gravid with 32 weeks gestation, presented with chronic headache aggravating since 2 months. Neurologist opinion was taken. Neurological examination was normal. CT scan showed hemangioblastoma in the cerebellum. Genetic study in the patient and the siblings showed mutation in VHL gene. She was advised tumour excision by neurosurgeon. Hence early decision was taken and elective LSCS done. Later patient underwent surgery for excision of hemangioblastoma. Patient improved and headache subsided in the postoperative period. There is no current way to reverse the presence of the VHL mutation in patients. Nonetheless, early recognition and treatment of specific manifestations of VHL disease can substantially decrease complications and improve quality of life. For this reason, individuals with VHL disease are usually screened routinely for retinal angiomas, CNS hemangioblastomas, clear-cell renal carcinomas and pheochromocytomas. CNS hemangioblastomas are usually surgically removed if they are symptomatic.

CONCLUSIONS

Timely diagnosis and management of neurological diseases during pregnancy poses major therapeutic challenges to neurologists and other non-neurologist health care providers. Pregnancy is a unique period in life associated with significant hormonal and other physiologic changes in female patients, which may trigger or alter the course of neurological and psychiatric disorders. In addition, many diagnostic procedures that can be performed in non-pregnant

women are prohibited during pregnancy for safety reasons. Therapeutic decisions and management of a pregnant patient with neurological disorders heavily depends on the issue of the reasonable balance between the risks of no treatment versus active treatment for the mother and her fetus.

ACKNOWLEDGEMENTS

We acknowledge the help we received from Department of Neurology, Neurosurgery and Radiology, of Bengaluru Medical College & Research Institute, Bengaluru and NIMHANS, Bengaluru.

Conflict of interest: No Financial help received from any quarters in preparing this manuscript.

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