Successful outcome in a twin pregnancy with hereditary motor and sensory neuropathy type –II complicated with heart disease and preclampsia superimposed on chronic hypertension.

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Abstract: Heridetary motor and sensory neuropathy (HMSN TYPE-2) reflects reduction in the number of primary motor and sensory neurons. The occurrence of this disease is rare in pregnancy but may be exaggerated in pregnancy leading to preeclampsia / eclampsia. Here is a 28 years old 2nd gravidae with twin pregnancy at 31 weeks hospitalized with HMSN TYPE-2 disease and was managed successfully with good feto maternal outcome.

Introduction

Hereditary motor and sensory neuropathy (HMSN) Type –II is otherwise known as neuronal form of Characot Marie-Tooth disease (CMT) [1]. It reflects reduction in the number of primary motor and sensory neuones1.Inheritency is usually autosomal dominant. So the occurrence of HMSN Type –II in pregnancy is very rare, but may be exaggerated during pregnancy [2-3]. Pedigree is of paramount importance in the diagnosis of Charcot –Marie tooth disease, HMSN Type II has later age of onset, whereas in Type –I the onset disease in most of the cases is from childhood [4]. Patient most commonly present with difficulty in walking due to distal leg muscle weakness and wasting. Here we are reporting a twin pregnancy with HMSN Type –II complicated with heart disease, preeclampsia superimposed on chronic hypertension and its outcome.

Case history

A 28 YEAR OLD 2ND GRAVIDA ($P_{O+O+1+O}$) with twin pregnancy at 31 weeks of gestation referred to our outpatient clinic complaining of progressive thinning of bilateral upper and lower limbs and tendency to fall during walking. She also complained of tingling sensation of hands and feet, slipping of shoes and difficulty in holding objects for last 4 years but all these symptoms aggravated during this pregnancy. Her previous records revealed that she was known a case of CMT disease as per nerve biopsy report done in 2002 showing demyelination with axonopathy having possibility of hereditary motor and sensory neuropathy Type-II. She was having a raised blood pressure. For which she was not treated. Though the woman had irregular antenatal check ups. She was immunized against tetanus and her 1st trimester was uneventful. From 2nd trimester onwards she experienced aggravation of

the neurological symptoms and swelling of lower limbs with raised blood pressure. On admission she was 56 kg and found to be conscious cooperative. Anemia with bilateral pedal oedema were present. Her blood pressure record was 160/100 mm Hg. Respiratory system was clinically normal. Systemic examination of cardiovascular system showed cardiac enlargement with LVS_3. There was no murmur or pericardial rub. Examination of nervous system showed the woman to be conscious and oriented without any cranial nerve deficit. Bilateral both upper limb and lower limb motor power was grade 4/5 with preservation of tendon reflexes. Spine and cranium were normal. Sensory examination was normal. Abdominal examination revealed uterine height corresponding to 34 weeks of gestation with two fetal heartbeats on Rt and Lt spino umbilical line. Her haemoglobin was 7.8 gm/dl packed cell volume was 26% urine showed moderate poroteinuria blood sugar level renal and liver function tests were within normal limit. Ultrasonography revealed twin pregnancy one at breech presentation. Other at cephalic presentation without any congenital abnormalities. ECG showed left ventricular hypertgrophy (voltage criteria) and echocardiograpy showed borderline systolic and diastolic dysfunction EMG revealed denevation of all four limbs, motor and sensory neuropathy with axonal demyelination.In our antenatal ward we advised antihypertensive medications with (methyldopa, Nifedipine, Labetolol) proper nutrition with regular iron and calcium supplementation. Maternal and fetal care were taken properly by regular blood pressure checkup by availing cardiotocography, ultrasonography facilities. At 35 weeks of gestation she developed abnormally raised blood pressure, severe proteinuria in 24 hour urine output, mild arteriolar attenuation in opthalmoscopic examination. Giving prophylactic magnesium sulfate we had to go for delivery of babies by lower uterine segment cesarean section under general anesthesia. Post operative period went of well without any complication. By end of 10^{th} post operative day she gradually got relief from neurological symptoms. We discharged healthy mother and her two healthy daughter two weeks after delivery. On 6^{th} week and 3^{rd} month follow up the women was found to be physically fit and now under treatment of cardiology and neurology department.

Discussion

Charcot –Marie-tooth Disease is a hereditary motor and sensory demyelinating polyneuropathy with potentially debilitating peripheral symptoms. The Pregnant patient with this disease has increased rate of presentation anomalies, postpartum hemorrhage, so the rate of operative delivery on higher side [5].Respiratory and vertebral anatomy can also be affected, we should take precaution on the part of anesthesia if any operative intervention is needed [6]. Low socio economic status of the patient and inhabitant of a remote village bard us to analyze the pedigree. Our patient was having pregnancy with one of the twin in the breech presentation. She developed superimposed preeclampsia which was managed successfully by timely decision of operative delivery and proper execution of anticonvulsant, anti-hypertensive therapy. For this complicated pregnancy multi disciplinary approach plays the pivotal role. Successful pregnancy outcome is not unusual if managed with

combined collaboration. Pre –pregnancy counseling should be encouraged for better prospects.

Reference

1. Donaghy M. et al Disorders of peripoheral nerves, Walton. J. Brain's Diseases of the Nervous System 10th edition, New Delhi, Oxford University Press, 1998 : P 590-2.
2. Pullock M. Nukuda H. Kritchevsky M. Exacerbation of charcot Marie tooth Disease in pregnancy neurology (NY) 1982 : 32 : 1311-4.
3. Gastacet JL, Bendm J Livet MO Philip N et al Charcot marie, tooth disease exacerbation in pregnancy Rev Neurol (Paris) 2003 oct 156(10) 890-1.
4. Rudrik Schonehorn S, Rohriq D Nicholson G Zerresket a\ et al Pregnancy and Delivery sin CMT-1 Neurology 1993 Oct 43(10)(: 2001 -6
5. Hoff JM Gilhus NE, Dalvioet AK et al pregnancy and deliveries in patients with chrcot-Marie Tooth Disease Neurology 2005 Feb 8 : 64(3)- 459-62.
6. Greenwood JJ Scott WF et at Charcot – Marie – Tooth Disease peripartum management of two contrasting clinical cases Int. J obstet. Anasth 2007 April 16(2) 149-54.

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