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Neurofibromatosis In Pregnancy Leading To Eclampsia: A Case Report

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Abstract:
Introduction:
Neurofibromatosis 1[nf1] is one of the most common tumor following autosomal dominant inheritance pattern.patient with nf1 develops plexiform neurofibroma and cutaneous neurofibromas.these tumours are major cause of patient morbidity.an influence of estrogen and progesterone on tumor growth has been suggested but reports on growth or malignant transformation of tumor during pregnancy remain anecdotal.the majority of women with nf1 have healthy pregnancy but need careful monitoring as early diagnosis and treatment result in better outcome.reported incidence of nf1 in pregnancies varies from 1:5000 to 1:8500.fetal complications in female affected includes spontenous abortion,preterm deliveries,iugr,stillbirths.maternal complications include cerebrovascular disorders and hypertensive disorders.
Case summary:
A 28 year female g4p3l1 @35week period of gestation with nf1 with moderate anemia with hypothyroidism with iugr presented to nmch with complaints of breathlessness on exertion.on day 2 of her admission she developed seizures there was no prior history of seizure her bp was 150/100,she was given loading dose of mgso4 following which seizure subsided and patient went in early labour.she underwent lscs for fetal distress and delivered a live female baby of 1.5 kg ,liquor was meconium stained.there were no nf1lession in baby.baby was admitted in nicu in view of respiratory distress.her bp was normal during post-operative period.
Conclusion: the latest literature agrees that pregnancy in patient with nf1 should be considered as at increased risk for obstetric complication.these patient need to be at close antenatal monitoring at tertiary centers for sign of hypertension/pre-eclampsia/eclampsia and iugr that are considered to be responsible for stillbirth,preterm labour and high rate of ceaserean section.
Further more,close observation for sign of disease aggrevation by clinician,expert on nf1 is also needed in order to guarantee the best possible outcome.
Keywords: *neurofibromatosis,iugr,tumor*

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I. Background:

Neurofibromatosis type-1(NF1) is an autosomal dominant tumor predisposition genetic disease,with diverse expression that can affect almost any organ system.It is caused by mutation of the homonymous gene located on chromosome 17.Mutations of the NF1gene creates a syndrome characterized mainly by the development of multiple neurofibromas,café-au-lait spots,lisch nodule{irish hemmartomas},freckling of the axillar or inguinal regions and optic gliomas.incidence of the disorder is 1:2500 -1:3500 regardless of ethnicity or race ,with over two million cases globally.

Due to the hormonal changes pregnancy might cause an increase in the size of all ready existing neurofibromas and appearance of new ones.The majority of women with NF1 have healthy pregnancies,but need careful monitoring as early diagnosis and treatment results in better outcome.The reported incidence of NF1in prgnency varies from 1:5000-1:18500.Fetal complication in affected women includes preterm delivery,spontenous miscarriage,IUGR and still birth.Maternal complications are hypertensive cerebrovascular disease.

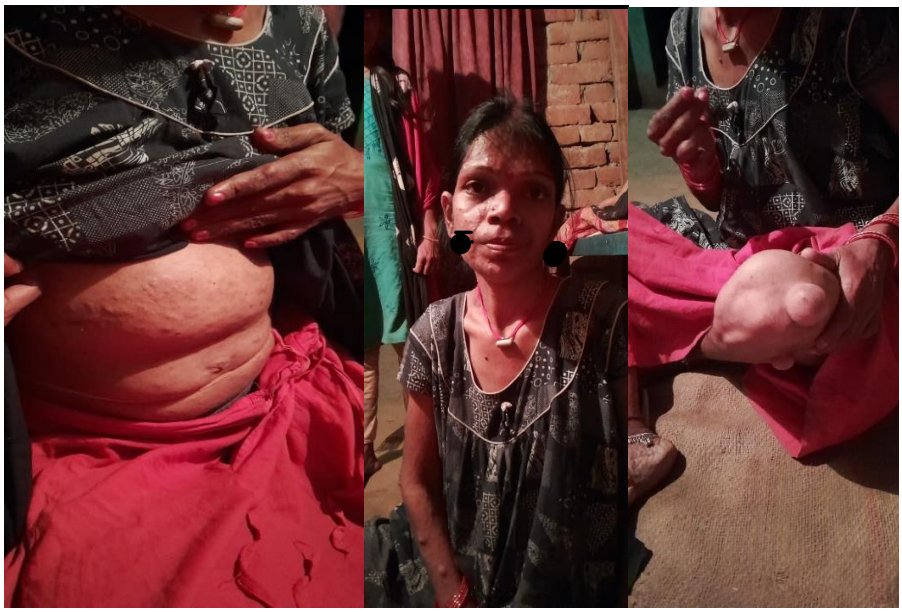
The NF1gene is responsible for production of large protein,called Neurofibromin,which act as atumor suppressor protein due to its function as negative regulator of Ras cellular pathways.Mutations of NF1 gene causes abnormal growth and division in multiple body system .For example, loss of heterozygosity{LOH}in the melanocyte lineage results in café-au-lait macules, hyperpigmented patches of skin and LOH in the schwann cell lineage leads to development of neurofibroma.The clinical expression of this disease is highly variable.Apart from neurofibromas there can be freckling of axillar or inguinal regions,bone deformities, learning disabilities, attention deficit hyper active disorder,gradual hearing loss,ringing in the ears,poor balance,headaches.Phenotypic expression of the NF1gene mutation is extremely heterogenous, therefore molecular diagnosis cannot predict clinical gravity of the disease.

We are reporting a case of neurofibromatosis which was complicated due to development of eclampsia in third trimester.

II. Case Report:

28 year old G4P3L1 @35weeks period of gestation with neurofibromatosis1 with moderate anemia with hypothyroidism with IUGR presented to NMCH with complaints of breathlessness on exertion.On examination at the time of admission her general condition was normal and she was afebrile with pallor present,icterus absent.There were multiple big and small neurofibromas all over the body, she also gave history of increase in the size of neurofibromas during pregnancy.Her BP was 130/90mmHg and urine albumin was 1+ ,Pulse rate 98bpm.On per abdomen examination uterus was 30 weeks in size {IUGR},relaxed,cephalic with regular fetal heart rate 130bpm.

On ultrasound fetal growth parameters corresponding to 30 weeks period of gestation.Her labs value Hb%7.7gm%,BT:2.11 min,CT:4.15min, Blood Group: B positive, LFT- wnl, KFT and fundoscopy was with in normal limit.On day two of admission she developed seizures and her BP was 150/100mmHg. She was given loading dose of Inj. MgSO4.Her pregnancy was terminated by cesarean section in view of fetal distress. Her post-operative period went un eventful and patient was discharged with healthy baby.



III. Discussion:

There is limited information on pregnant women with NF1,but many authors have suggested increased risk of maternal as well as fetal complications.Maternal complications as reported, include increase of tumor burden as a rise in number and size of tumors such as neurofibromas café-au-lait spots,optic gliomas and malignant transformation of tumors.Hypertensive complications like gestational hypertension and pre-eclampsia as well as cerebrovascular complications are also of significant importance.NF1 predisposes to pheochromocytoma and renal artery stenosis, both of which cause secondary hypertension.Fetal complications consist of spontenous abortion of first trimester,still birth,IUGR, preterm cesaren delivery and oligohydromnios.Fetal distress,neurofibromatosis lessions on the newborn,malpresentations and cephalopelvic disproportion due to undiagnosed pelvic neurofibrommas and pelvic bone contractures as well as severe pre eclampsia,abruptio placenta is reported to increase risk of cesarien section.In most studies,an important

percentage of patients,usually more than 50% affected by NF1,during pregnancy,have reported an increase in terms of number and size of preexisting neurofibromas.

Since 1906 Brickner has described nodular lesions, that appear during pregnancy and gradually disappear after delivery. Later Sharpe and Young(1937)and Mortiz and Snider(1962) stated that pregnancy may provide a growth stimulus on neurofibromatosis skin lesions and that way promote diagnosis of disease.Swapp and Main in (1973) released an intresting study of 10 NF patients and their 24 pregnancies.In five out of 10 patients,the lesions of neurofibromatosis appeared for the 1st time during pregnancy in other five patients lesion increase in size and number.The lessions regressed considerably after delivery in seven out of ten patients.They also stated that hypertension during these pregnancies is more than a chance association possibly due to neurofibromatosis vasculopathy.All ten patients to the end of their pregnancies have shown significant rise of mean blood pressure,while five of them were already hypertensive in first visit.Several case reports and studies of more patients suggest that pregnancy might worsen NF1 diseases tumor lesions or stimulate the rise of new ones or even provoke malignant transformation and emergence of pheochromocytoma.

Recently Well et al.(2020) published a retrospective study that investigated the effect of pregnancy on tumor burden in 13 patients with NF1,matched with 13 nonpregnant patients as control group.In this study although some NF1 patients experienced a subjective increase of NF1 related clinical symptoms and tumor growth during pregnancy,growth of plexiform and cutaneous neurofibromas in pregnant patients,with MRI observation,was not significantly different compared to non-pregnant patients.Furthermore,no patient developed new plexiform neurofibroma(PNF) and no PNF underwent malignant transformation,which was expected.

Many authors in the past have reported cases of women with NF1 that presented complications in one or more pregnancies .Subsequently most of the studies on larger patient samples as well as retrospective register based analysis have confirmed that pregnancy complications are significantly increased among women with NF1.

Segal et al.(1999) showed up with the evaluation on outcomes of 13 pregnancies on 8 patients with NF-1,matched 1 to 5 with a control group.The incidence of hypertension in the study group was 12.5% versus 4.6% in the control group,preterm delivery 30.8% vs 6.1%,IUGR 46.2% vs 8.9%,still-birth 23%vs 1.5% and caesarean section 38.5% vs 7.7% respectively.

In our case also patient became hypertensive and developed seizures for which loading dose of Mgso4 was given and cesarien section was done due to fetal distress.

IV. Conclusion:

As we have seen that pregnancy in patients with NF1 should be considered at increased risk for obstetrics complications. These patients need to be at close antenatal monitoring at tertiary centers for signs of hypertension/pre-eclampsia and intrauterine growth restriction that are considered to be responsible for still births,preterm labour and higher rates of cesarean sections.There should be close observation for signs of disease aggravation by clinicians,eperts on neurofibromatosis inorder to have healthy mother and baby.

Abbreviations:
IUGR:Intrauterine growth restriction
LOH:Loss of heterozygosity
NF:Neurofibromatosis

Conflicts of interest:
The authors declare that they have no conflict of interest.

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