Case report

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Neurofibromatosis Type 2 in Pregnancy: A Rare Case

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Abstract

Neurofibromatosis type 2 in pregnancy is an extremely rare condition. Central nervous system involvement with shwannomas and spinal tumours is very common. This condition is very challenging for obstetricians especially when the neurological complications starts manifesting. Close obstetric care, timely interventions and Multidisciplinary approach is the key to successful outcome in cases of Neurofibromatosis 2 in pregnancy.

Keywords: Neurofibromatosis Type 2; Pregnancy; Schwannomas; *Paraparesis*.

Introduction

The birth incidence of Neurofibromatosis type 2 lies between 1 in 33 000–40 000 [1]. Patients typically present with a gradual, progressive and often asymmetrical hearing loss. The mean age of onset of symptoms is 24 years whilst non-neurofibromatosis patients with sporadic tumors present in their midforties.

In this case 21 year old female patient, diagnosed case of neurofibromatosis type 2, presented in an emergency ward as primigravida with 31weeks of pregnancy. Chief complaints were breathlessness and lower limbs weakness. On admission she had right facial paresis and aspiration pneumonitis. She had multiple café au lait spots and subcutaneous neurofibromata. Patient was admitted in intensive care unit. At 20 weeks of gestation she had observed weakness in her lower limbs which was gradually progressed during next few months.

On inquiry patient gave history of surgery at the age of 12 years for a painful neck mass. Histopathological examination of the mass was suggestive of neurofibroma. In 2012 spastic paraparesis progressively increased along with sensory impairment below D12 level. Laminectomy D6 to D9 and D12 to L1 with excision of intraspinal schwanoma was done. Patient was fine after surgery. She got married in 2014 and completed her education upto Master of Arts. She conceived in Feb 2015.

Antenatal period was uneventful till fifth month of pregnancy when lower limb weakness was observed. Patient presented to us at 31 weeks of gestation with severe deterioration of neurological status, raised intracranial tension with pressure effect on the brainstem and paraparesis. She had aspiration pneumonitis due to lower cranial nerve involvement. She received dose of steroids and higher antibiotics and was under observation in intensive care unit.MRI suggestive of bilateral acoustic neuroma, supratentorial meningioma, left intraventricular meningioma, obstructive hydrocephalus and scalp neurofibroma suggestive of neurofibromatosis type-2. Chest radiograph shown pneumonitis and prominent bronchovascular markings in both the lung fieids. Ultrasonography at 32 weeks of gestation showed live fetus in the uterus with mild reduction in liquor. In view of acceleration in the tumor growth leading

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to further risk to mother and fetus, planned cesarean section decided after anaesthesia & medicine evaluation. Elective cesarean section with extraventricular drainage done under general anaesthesia. Patient had undergone definitive neurological surgery later and discharged after three weeks with complete recovery of lower limb weakness.

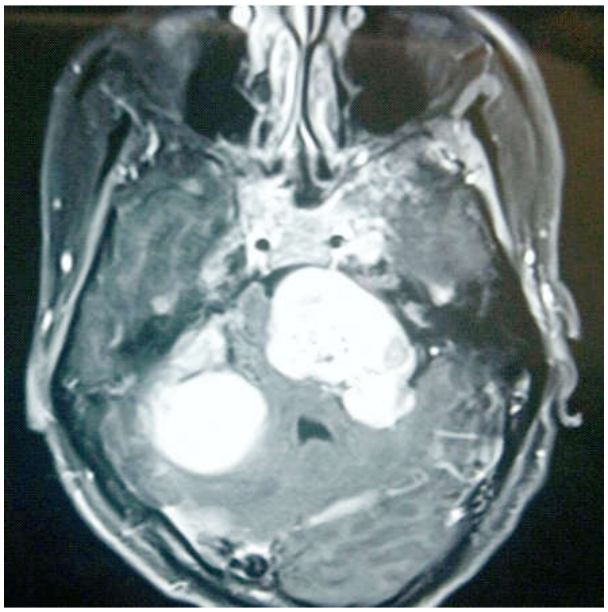

Fig. 1: Post contrast T1W MRI image in axial plane shows well defined lobulated, homogenously enhancing discrete lesion in bilateral CP angle causing mass effect and distortion of brainstem. These lesions are suggestive of bilateral acoustic schwannomas.(Pre-operative)

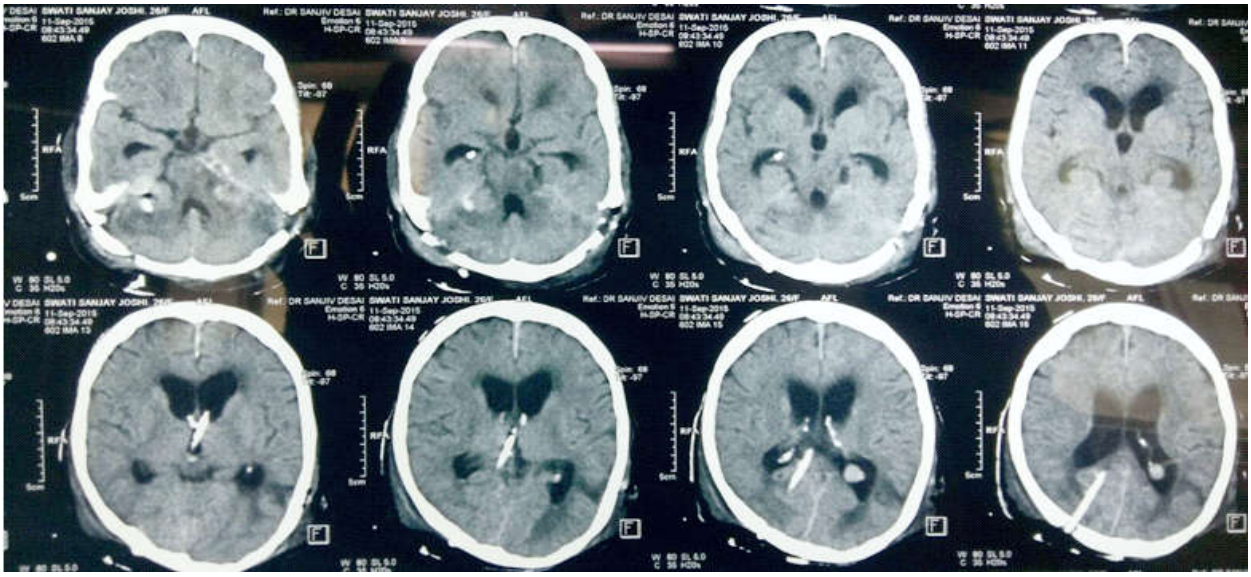

Fig. 2: Axial CT study shows lesions in the bilateral CP angle reduced in size. Mass effect on the brain stem reduced. Tip of the ventricular drainage tube is seen in the frontal horn of left lateral ventricle. (Post-operative)

Discussion

Pregnancy with neurofibromatosis type 2 is very rare and very few cases studied till date [2]. These are mostly slow growing benign tumors. It is an autosomal dominant condition. The types of tumors involved include schwannomas, meningiomas, ependymomas, and neurofibromas. Schwannomas are classically located at the superior vestibular branch of cranial nerve VIII bilaterally. Inferior vestibule and peripheral nerves can be involved. The

Symptoms of vestibular schwannomas include hearing loss, imbalance, tinnitus, headache, mastoid ache, raised intracranial pressure (ICP). These vascular tumors are more frequently seen in women [3].

The course of event in pregnancy is mostly decided by the acceleration in the growth of these tumors and subsequent pressure effects. Vestibular schwannomas can appear for the first time during pregnancy, or symptoms may worsen during the last trimester [4,5].

The theories put forth for the acceleration of the growth of vestibular schawannoma in pregnancy are related to the hormonal and hemodynamic changes occurring in pregnancy. Influence of estrogen and progesterone hormone on the growth of neurogenous tumors like vestibular schwannomas have been studied by multiple groups. Estrogen receptors were identified on the tumor. Hormonally induced vascular dilatation in the tumour was observed under the influence of oestrogen and progesterones, though results are conflicting [6,7].

Several vascular and metabolic changes occur during pregnancy. Pregnancy associated hypoproteinemia, anaemia, gestational hypertension or pre-eclampsia cause retention of extra cellular and intracellular fluid. It increases the size of the tumour and intracranial pressure during pregnancy. Increase in plasma volume starts from 6th week of pregnancy and peaks at 32 -34 weeks. The average increase in the plasma volume upto 40-45 % was observed at 32 weeks of gestation. This mediates the tumor growth and vascularity [8].

Hemodynamic and metabolic changes in pregnancy alters the manifestation of the disease. Presentation of patient varies case to case depending on the neurological status and state of the pregnancy. Abortions, preterm labour, increase rate of *cesarean* section, neonatal and maternal morbidity commonly seen in cases of neurofibromatosis in pregnancy.

The optimal treatment strategy for such a patient is close observation, periodic neurological assessment and obstetric care. Operative interventions advised mostly in the second trimester. Rate of spontaneous abortions and teratogenecity with anaesthetic drugs are higher in the first trimester [9,10]. Neurological surgery advisable if the neurological status deteriorated significantly at any juncture of the pregnancy.

In this case patient had uneventful antenatal course till second trimester. The anaesthetic risk to mother increases in third trimester due to physiological changes such as reduction in functional residual capacity and hypervolemia causing alteration of anaesthetic drug distribution. Patient in the study had complaints of breathlessness and severe lower limb weakness in the third trimester and was kept under close observation in intensive care unit. She had severe deterioration of her neurological status in view of raised intracranial tension. Most of the studies concluded that the best foetal and maternal outcomes in the third trimester are provided by emergency *cesarean* section after drainage of cerebrospinal fluid, followed by definitive neurosurgery [11-13]. Wang & Young's [14] advocated multidisciplinary approach while managing complicated neurofibromatosis type 2 patients in pregnancy.

Our case had giant vestibular schwannoma with parapareis in pregnancy. She was operated at 32 weeks. Extraventricular drainage was performed by neurosurgeon to relieve intracranial tension and simultaneous *cesarean* section was done by Obstetrician. Patient deliverd a live male baby of 1.5 kg. Intra operative course was uneventful. Patient was kept under observation in intensive care unit. The neurological status of the patient was improved partially due to reduction in inrtracranial pressure.

Major neurological surgeries are advocated in the postpartum period. Post delivery physiological changes in pregnancy starts reverting that aids in anaesthesia as well as overall better outcome of the surgery. A week later vestibular schwannomas were resected. Surgery took place with moderate blood loss. Patient had significant improvement in next two weeks.

She started walking without support after 2 weeks. Patient with her baby was discharged with complete recovery of her neurological status though partial ptosis of the right eye persisted for few weeks.

Conclusion

Neurofibromatosis type 2 in pregnancy has wide spectrum of its manifestation. Most of the neurological surgery have a doubtful outcome in pregnancy. As in this case vascular tumour like schwannomas posing significant challenges during pregnancy are to be managed by obstetrician with the help of other expert faculties like neurosurgeon, anaesthesiologist and intesivist. Definitive diagnosis, genetic counselling and information releted to the tumor course in pregnancy is to be emphasised upon the pregnant female and her family. Releiving intracranial pressure prior with cesarean section followed by definitive neural surgery aids in optimum

outcome of the patient.

Conflict of Interest

No

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