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Case Report

Pregnancy with neurofibromatosis type II: the enigma continues

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ABSTRACT

Neurofibromatosis type II in pregnancy has variable presentations. The recurrence rate and bilaterality of CP angle tumour is very high. We present a case of bilateral schwanomma diagnosed in midpregnancy, for which patient was operated in second trimester and postpartum radiotherapy will now be given.

Keywords: Neurofibromatosis, Schwanomma, Autosomal dominant trait, Spinal anaesthesia

INTRODUCTION

Neurofibromatosis type II affects about 1 in every 40000 people without regard to sex or race. They are at high risk of developing tumours (Schwanomma) on both nerves to the ears. This nerve has two portions. Acoustic concerned with hearing and vestibular concerned with balance inputs to the brain.[1] The early symptoms of Neurofibromatosis type II are dysfunction of these nerves- hearing loss, tinnitus (Acoustic) and problems of balance (Vestibular). These tumours are also called schwanomma as they arise from Schwann cells covering the nerve. However, they can arise from other cells as well like ependymoma and meningioma. Some patients of Neurofibromatosis type II can also present with special type of cataract called juvenile posterior sublenticular opacity.[2,3]

CASE REPORT

Mrs. ABC 26 years of age, married since 4 years, $G_2P_1IUFD_1$, previous caesarean section, at present 38.5 weeks period of gestation, known case of Neurofibromatosis type II, with bilateral cerebellopontine angle schwanomma operated in 2014.

Patient is a known case of neurofibromatosis type II and was diagnosed to have bilateral cerebellopontine angle schwanomma in April 2014. She underwent right retrosigmoid craniotomy with excision of right CP angle in 9th October 2014. She then complained of swelling and pain on operated site on 30th October 2014. Diagnosed to have pseudomeningocoele, which had settled subsequently following lumbar puncture done 3 times.

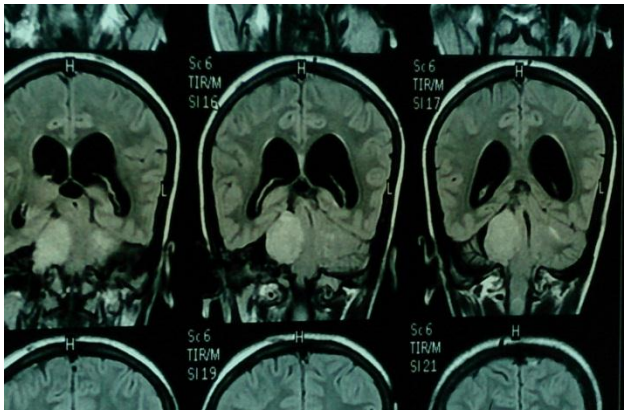

Figure 1: MRI brain in coronal view showing tumour at cerebello-pontine angle).

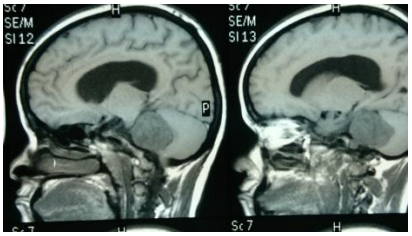

Figure 2: Tumour at cerebello-pontine angle displacing midbrain structures anteriorly in sagittal section of MRI brain.

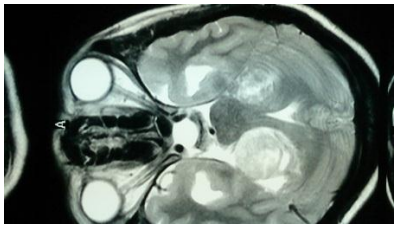

Figure 3: Transverse section of MRI brain showing tumour at right CP angle.

Patient was also diagnosed to have deranged blood sugars in October 2014. Started on inj. insulin 8 units thrice a day. Presently she is also taking tab. levipril (500 mg) OD. Patient had significant family history as her mother had expired of brain tumour (details not known). Patient was vitally stable when came to the hospital and was taken for emergency caesarean section in view of scar tenderness in labour. Perinatal outcome was healthy female child of 3.5 kg. Postoperative course was uneventful.

DISCUSSION

Two major treatment options available for neurofibromatosis type II are surgery and radiotherapy. Though they do require surgery once in the lifetime, it is not without risk as the tumour lies very close to nerves, it is likely to damage the delicate nerve. There are additional risks associated with anaesthesia as well. Radiation therapy also is associated with risks and benefits which must be outweighed vis a vis.[4] Neurofibromatosis type II is a genetic disease that is passed from parent to child during conception. All persons with Neurofibromatosis type II have 50-50 risk of affecting the children. Disorders such as neurofibromatosis type II which affect both sexes equally and are transmitted to 50% of their children are termed as autosomal dominant. Exact segment of DNA that causes neurofibromatosis type II has been identified on chromosome 22 and can be identified and isolated with FISH technique or hybridization.[5-7]

CONCLUSION

The outcome of baby will be evident only in due course of time. As the tumours, even in genetically predisposed patients appear at a later age, in late teens. The only way to confirm if child would be affected in future is to subject the baby to chromosomal evaluation for segmental DNA analysis during antenatal period. However, in the present case scenario, it was not possible as the woman had presented for antenatal registration in almost 7th month of gestation when time limit for terminating pregnancy was exceeded well in advance. In the case presented here, only way to find out if child will be affected is either subject them to chromosomal study or subject the child to audiometric evaluation.

Hence, the enigma still continues as whether the baby will be affected or not, as there is no conclusive way to diagnose whether the baby has the defective gene.

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