

Case report of a parturient with cerebellar hemangioblastoma and acute hydrocephalus in relation to Von Hippel-Lindau disease

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INTRODUCTION

Von Hippel-Lindau disease (VHL) is a rare autosomal dominant condition predisposing to neoplasia affecting several organs, including central nervous system haemangioblastomas, retinal hemangioblastomas, clear cell renal cell carcinomas, pheochromocytomas, neuroendocrine tumours of the pancreas and endolymphatic sac tumours . The natural history of VHL in pregnancy is unclear[1] We present the management of a parturient with VHL at our tertiary centre.



CASE REPORT

A 25 year old primigravida with a history of previous cerebellar hemangioblastoma resection, known stable cerebellar haemangioblastomas and bilateral adrenalectomies for phaeochromocytoma, was reviewed by the multidisciplinary team throughout pregnancy. Prior to pregnancy, regular surveillance with two yearly stable magnetic resonance imaging (MRI) brain was undertaken and MRI lumbar spine at 30 weeks gestation showed no hemangioblastoma. Plasma metanephrines in pregnancy were normal. However, the parturient was admitted at 35+5 weeks with a three week history of poor balance and falls.

New imaging showed progressive hemangioblastoma in the left cerebellar tonsil which was compressing the fourth ventricle and causing acute hydrocephalus. After multi disciplinary team discussion, the patient underwent elective caesarean section at 36 weeks, with a second surgery for posterior fossa haemangioblastoma resection on the following day. Neuraxial anaesthesia was contraindicated due to the presence of hydrocephalus. Caesarean section was performed uneventfully under general anaesthesia using propofol/remifentanyl infusions and rocuronium, with attention to neuroprotection. Postoperatively, the patient was monitored in a level two setting on both occasions with good recovery, though subsequently required further surgery for ventriculoperitoneal shunt insertion.

DISCUSSION

An individualised approach should be undertaken for the antenatal and peripartum management of complex parturients. Our multidisciplinary team decision was to perform caesarean section and neurosurgery on two separate occasions rather than during one general anaesthetic. Maternal and fetal considerations included those related to urgency of neurosurgery, risk of postpartum haemorrhage and prone surgical positioning for posterior fossa surgery.

This case prompted literature review of VHL in pregnancy. We highlight that the progression of VHL in pregnancy appears unclear [1].

We found international consensus guidelines for surveillance of organ systems in VHL with reference to pregnancy [2].

CONCLUSION

Although the evidence is limited, the current recommendation [1,2] for women with quiescent Von Hippel-Lindau disease is two yearly surveillance MRI of the craniospinal axis. When considering pregnancy, it is recommended that surveillance MRI with contrast be performed prior to a planned pregnancy to characterise hemangioblastomas and/or associated cysts before conception.

If a patient becomes symptomatic while pregnant, MRI of the neuroaxis can be obtained without contrast if the clinician suspects a developing hemangioblastoma and/or hemangioblastoma associated cyst. Furthermore, if a CNS hemangioblastoma becomes symptomatic during pregnancy, tumor resection can be performed safely with careful maternal and fetal monitoring.

REFERENCES

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