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Case Report

Cerebellar hemangioblastoma during pregnancy: Management options and review of literature

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ABSTRACT

Background: Symptomatic cerebellar hemangioblastomas are extremely rare in pregnant women and the ideal management is not well established. In the present article, we aimed to report a case of large cerebellar hemangioblastoma complicated by pregnancy and managed successfully by surgical resection. In addition, we also discuss management options and review of the current literature pertaining to this pathology.

Case Description: A 22-year-old female presented with a history of headache and vomiting for 4 weeks. She was carrying 28 weeks of pregnancy. She had left cerebellar signs, gait ataxia, and bilateral six nerve paresis. Fundus examination revealed bilateral papilledema. She was diagnosed to have large cerebellar hemangioblastoma with mass effect and obstructive hydrocephalus. She underwent suboccipital craniotomy and excision of lesion in lateral position. She recovered well postoperatively and delivered a healthy baby in the full term. Imaging at10-month follow-up demonstrates no residual lesion or another hemangioblastoma.

Conclusion: Early diagnosis and direct surgery for excision of hemangioblastoma is a good option during pregnancy while avoiding CSF diversion procedures. The symptomatic hemangioblastoma during pregnancy can be safely operated during early pregnancy.

Keywords: Brain tumor, Cerebellar, Hemangioblastoma, Pregnancy

INTRODUCTION

Cerebellar hemangioblastoma is a benign, relatively rare vascular lesion and can be sporadic or associated with von Hippel-Lindau disease. Symptomatic cerebellar hemangioblastomas are extremely rare in pregnant women and the ideal management is not well established.[8] It has been reported that in pregnancy, these lesions may enlarge in size and become increasingly symptomatic, resulting in need for surgical resection.[3,11] Cerebellar hemangioblastomas can cause symptoms due to raised intracranial pressure because of obstructive hydrocephalous and/or edema or due to direct brain stem compression.[3] The detection of hemangioblastoma in pregnancy is often delayed due to the confused presentations between pregnancy and tumor. Management of hemangioblastomas associated with pregnancy is further complicated as it is difficult to make a decision whether a tumor resection should be done early in the pregnancy or not until full-term labor is reached. During pregnancy, rapid increase in the size of these lesions remains poorly understood.[7] It has been suggested that involvement of placental growth factor (PlGF) and its receptor vascular endothelial growth factor receptor 1 (VEGFR-1) in

various pathologic processes can lead to the development and enlargement of peritumoral edema and cysts, which are the principal causes for the development of any symptoms in hemangioblastoma.[7] In the present article, we aimed to report a case of large cerebellar hemangioblastoma complicated by pregnancy and managed successfully by surgical resection. The management of this uncommon neurosurgical condition in pregnancy is presented along with pathophysiological basis, management options, and a review of the current literature.

CASE DESCRIPTION

A 22-year-old female presented with a history of headache and vomiting for 4 weeks. She was carrying 28 weeks of pregnancy (gravida 2, para 1). Her medical history was unremarkable. Neurologically, she was conscious and alert. She had left cerebellar signs, ataxic gait, and bilateral six nerve paresis. Fundus examination revealed bilateral Frisen Grade 4 papilledema and no retinal angioma was observed. The rest of the neurological examination was essentially within normal limits. Per abdominal examination showed a uterine size consistent with 28 weeks of gestation, normal fetal heart rate. Biochemical and hematological parameters were within standard limits. Magnetic resonance imaging revealed an intra-axial mass lesion of 5.4 × 4.5 × 3 cm dimension in the left cerebellar hemisphere, which was hypointense on T1-weighted image and hyperintense in T2-weighted image causing marked distortion of cerebellar hemisphere, with brain stem compression, pressure over the fourth ventricle with enlargement of both lateral ventricles, ballooning of third ventricles, and periventricular lucency [Figure 1]. On administration of gadolinium contrast, administration revealed a brilliantly enhancing mural nodule [Figure 1a]. A diagnosis of cerebellar hemangioblastoma with obstructive hydrocephalus was made. Abdominal ultrasound revealed a single viable fetus with fetal growth appropriate for age. However, maternal ultrasound examination revealed normal liver, pancreas, and kidney.

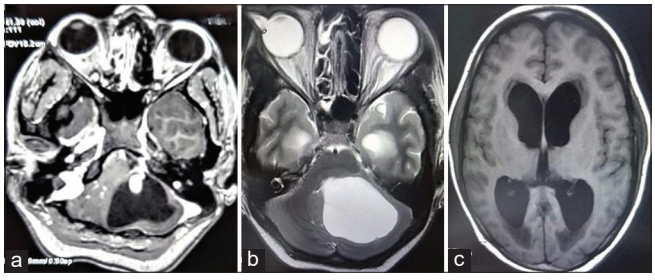

Figure 1: (a) Contrast-enhancing magnetic resonance imaging revealed an intra-axial mass lesion of 5.4 × 4.5 × 3 cm dimension in the left cerebellar hemisphere, enhancing nodule with marked distortion of cerebellar hemisphere, with brain stem compression. (b) T2 axial image showing hyperintense lesion with perifocal edema around the tumor. (c) T1-weighted image showed enlargement of both lateral ventricles and ballooning of the third ventricles with periventricular lucency suggestive of hydrocephalus.

After obtaining the patient's consent, a multidisciplinary team involving obstetricians, anesthesiologists, and neurosurgeons decided to perform surgical excision in view of raised intracranial pressure symptoms. She was considered for emergency surgery and underwent suboccipital craniotomy and resection of lesion in lateral position. Surgery revealed a tense dura and significant herniation of the left cerebellar hemisphere. Exploration of the cystic cavity revealed a vascular cherry red mural nodule that was resected from its insertion into the wall of the cystic capsule. At the end of the surgery, cerebellum was lax, pulsatile and the procedure was uneventful. The histopathological examination of the specimen confirmed hemangioblastoma. She recovered well postoperatively and had marked improvement in neurological status. The ultrasound examination of the fetus on the 10th postoperative day normal viable fetus. She delivered a healthy baby in the full term. She was doing well, at last follow-up at 10 months following surgery. A cranial contrast-enhanced MRI scan, at the last follow-up, revealed no residual lesion or another hemangioblastoma [Figure 2].

DISCUSSION

During pregnancy, cerebellar hemangioblastoma is rare and a patient may need urgent surgical intervention due to a rapid increase in size.[11] When the fetus is still immature, definitive surgical treatment is suggested, especially in the presence of neurological symptoms.[2,11] Symptoms are due to obstructive hydrocephalous, edema, or direct brainstem compression. Only a few cases have been reported in the literature which illustrates direct surgical intervention for hemangioblastomas during pregnancy. We reviewed previously reported cases during pregnancy and also discuss various management options.

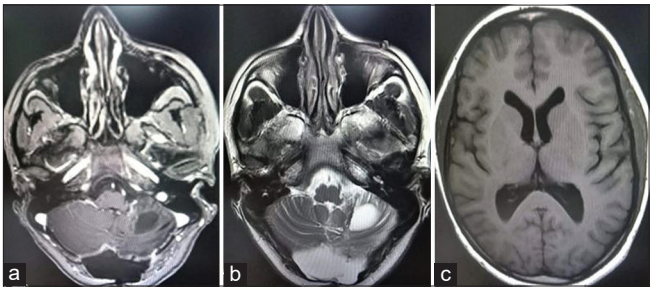

Figure 2: Postoperative magnetic resonance imaging (MRI) at 10 months follow-up. (a) T1-weighted contrast MRI showed postoperative changes with no enhancing nodule and no other hemangioblastoma. (b) T2 axial images showing postoperative changes and disappearance of edema. (c) T1 axial image showed resolution of hydrocephalus.

Mechanism of increase in the size of the lesion during pregnancy

The effect of pregnancy on the natural history of hemangioblastomas remains unclear, though various theories have been proposed.[8] Kasarskis *et al*. proposed that during pregnancy increase in blood volume engorgement of vascular bed may be the cause of the rapid increase in the size of the lesion.[5] Brown *et al*. thought that these lesions had hormonal receptor and expansion of the tumor under hormonal influences during pregnancy.[1] Several metabolic and hemodynamic changes associated with pregnancy such as increased circulating blood volume and cardiac output and decreased plasma osmolarity may also collectively contribute to increased vascularity, the rapid development of peritumoral edema, and associated neurological symptoms.[11] Extracellular and intracellular fluid retention during pregnancy may be another factor associated with an increase in the size of the lesion.[8] Some authors have questioned this hypothesis when it was found that the rate of hemangioblastomas that happen to be symptomatic during pregnancy did not increase when compared with age-matched, nonpregnant female patients.[8]

Laviv *et al*. suggested the involvement of placental growth factor (PlGF) and its receptor vascular endothelial growth factor receptor 1 (VEGFR-1) can lead to the growth of peritumoral edema and cysts, which are the principal causes for the development of any symptoms in hemangioblastoma.[7] Frantzen *et al*. observed in their study progression of cerebellar hemangioblastomas were significantly increased after pregnancy in about 40% of cases.[4] During pregnancy, increase in the size of cerebellar hemangioblastoma and causes a high pregnancy complication rate, hence, recommend close monitoring of such patients during the reproductive phase of women, especially during pregnancy and preconception period.

Management

Various management options have been described in the pertinent literature for cerebellar hemangioblastomas encountered during pregnancy which include conservative management with close observation, CSF diversion, and direct surgery. As these symptoms may get worse during pregnancy, so conservative management with close monitoring is not a feasible option in symptomatic patients. Some authors have recommended direct open surgery for a symptomatic hemangioblastoma during pregnancy, however, in high-risk pregnancies, particularly those at risk for preterm labor, symptomatic hemangioblastomas can be treated with more conservative approach-like CSF diversion.[11]

Naidoo *et al*. performed definitive surgery in two patients at 21 weeks and 33 weeks of gestation with good outcome.[9] Similarly, Nathan *et al*. reported a 28-year female underwent tumor resection without CSF diversion procedure during second-trimester pregnancy, and the pregnancy was continued with good outcome.[10] The literature concerning direct surgical treatment of hemangioblastomas in pregnancy consists largely of case reports or case series [Table 1].

The severity of maternal symptoms and gestational age should be taken into account while deciding appropriate treatment. CSF diversion is an option, but the patient can deteriorate after shunt surgery.[5] On the other hand, several shunt-related complications may follow this approach.[11] Some authors have reported neurological deterioration after shunt surgery in symptomatic cerebellar hemangioblastomas which necessitated direct emergency surgery for hemangioblastoma.[5] The possible cause of deterioration

Table 1: Studies illustrating direct surgery (without shunt) for cerebellar hemangioblastoma during pregnancy.

Studies	Age (year)	Gestational age at presentation	Size of lesion (centimeter)	Location	Outcome
Kasarskis *et al*., 1988[5]	18	2nd month	2.5×2	Cerebellum	Good
Nathan *et al*.,1995[10]	28	2nd trimester	4×3.5	Cerebellum	Good
Naidoo *et al*., 1998[9]	26	21 weeks	2.9×2.3	Cerebellum	Good
	26	33 weeks	4.5×4	Cerebellum	Good
Delisle *et al*., 2000[2]	35	30 weeks	5×3	Cerebellum	Good
Erdogan *et al*., 2002[3]	35	24 weeks	3.5	Cerebellum	Good
Kenyon *et al*., 2009[6]	33	28 weeks	5×4	Cerebellum	Good
Xiu-jian *et al*., 2018[8]	31	6 weeks	1.2	Medulla	Abortion
	27	29 weeks	4.2×3.6	Cerebellum	Good
	26	23 weeks	4.2×1.9	Cerebellum	Good
	24	19 weeks		Medulla	Good
Present study	22	28 weeks	5.4×4.5×3	Cerebellum	Good

after shunt surgery may be rapid growth of the tumor due to reexpansion of the tumor intravascular bed and due to loss of cushion effect between CSF and lesion causing direct compression over brain stem.[5]

In view of overall management options, direct surgery is a better option if patients present in early pregnancy. It can avoid repeated exposure of general anesthesia and shunt-related complications (i.e., bleeding during placement of the catheter, infection, shunt malfunction, shunt exposure, and rare bowel perforation).[11] Those patients present after 32–34 weeks during pregnancy can be managed with minimal intervention and the combination of cesarean section and surgical resection of the tumor can be planned once fetal lung maturity has been accelerated.[11,12] In the case of early pregnancy, optimal treatment strategy should be selected on an individual basis considering the risks and benefits for both mother and fetus. As in the present case, surgical resection should be done in lateral position, which secures uteroplacental perfusion, and provide adequate intraoperative exposure for the neurosurgeons.[6,8]

This case highlights the significance of performing a detailed neurological examination in all cases of persisting headache and/or vomiting during pregnancy. As in the present case, the presence of lateral rectus palsies in any patient with a headache necessitates urgent imaging to rule out a space-occupying lesion. The case reported here is important because it suggests a direct surgical approach to the management of symptomatic cerebellar hemangioblastomas in pregnant women.

CONCLUSION

Cerebellar hemangioblastomas are extremely rare during pregnancy and should be included in differential diagnosis for those patients presented with persistent nausea and vomiting associated with focal neurological deficits. Early diagnosis and direct surgery for excision of hemangioblastoma is a good option during pregnancy while avoiding CSF diversion procedures. The symptomatic hemangioblastoma during pregnancy can be safely operated during early pregnancy. Further, case reports and series are required to support this recommendation.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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