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Case Report

Favorable management of symptomatic cerebellar hemangioblastoma presenting with obstructive hydrocephalus during pregnancy: A case report and literature review

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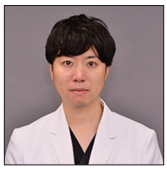

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ABSTRACT

Background: Cerebellar hemangioblastoma is a highly vascular benign tumor and the growth rate of hemangioblastomas is believed to often accelerate during pregnancy; however, the reason for this rapid increase in size remains poorly understood. There are several case reports of symptomatic hemangioblastoma during pregnancy; however, the favorable management strategy has not been well established.

Case Description: A 35-year-old woman, gravida 2 para 1, with no significant medical history presented with vertigo and difficulty walking at around 11 weeks of pregnancy and was referred to our institute at 30 weeks of gestation because of worsening symptoms. Brain magnetic resonance imaging revealed a 5.6 cm cystic lesion with a mural nodule in the right cerebellar hemisphere and the lesion blocked cerebrospinal fluid drainage from the fourth ventricle and brainstem, resulting in obstructive hydrocephalus. After obtaining the patient's consent, a multidisciplinary team consisting of obstetricians and neurosurgeons decided to perform resection of the intracranial lesion following delivery of the fetus by emergency cesarean section in view of the symptoms of increased intracranial pressure. The patient's general condition was confirmed to be stable postoperatively and she was discharged on the 16th day of her hospitalization without any neurological deficits or fetal complications.

Conclusion: Urgent tumor resection combined with cesarean section can be planned once fetal lung maturity is confirmed. Most cases of symptomatic hemangioblastoma during pregnancy have an uneventful gestational course and a favorable outcome for both mother and child.

Keywords: Hemangioblastoma, Neuro-oncology, Neurosurgery, Obstructive Hydrocephalus, Pregnancy

INTRODUCTION

Cerebellar hemangioblastoma is a highly vascular benign tumor, accounting for approximately 3% of all central nervous system tumors.[7,20] The growth rate of hemangioblastomas is believed to accelerate during pregnancy;[4,8] however, the reason for the rapid increase in the size of these lesions remains poorly understood.[10] Cerebellar hemangioblastoma can cause several

symptoms as a consequence of increasing intracranial pressure due to obstructive hydrocephalus or direct brain stem compression.[5] The diagnosis of hemangioblastoma in pregnancy is often delayed either because of the symptoms of pregnancy itself or because of delays in diagnostic imaging due to pregnancy. For these reasons, the favorable management strategy for symptomatic hemangioblastoma during pregnancy has not been well established.[12]

In the present article, we report a case of symptomatic cerebellar hemangioblastoma in a 35-year-old woman at 30 weeks of gestation managed successfully with emergency surgical resection following cesarean section.

CASE PRESENTATION

A 35-year-old woman, gravida 2 para 1, with no significant medical history presented with vertigo at around 11 weeks of pregnancy and gradually became unable to walk. At 15 weeks of gestation, she was diagnosed with vestibular disorder by a local otolaryngologist. She was hospitalized and treated with steroids, but her symptoms did not improve. When she was 30 weeks and 4 days pregnant, her symptoms worsened and she could not walk. She was then referred to our institute for further evaluation and treatment.

Brain magnetic resonance imaging (MRI) revealed a 5.6 cm cystic lesion with a mural nodule in the right cerebellar hemisphere and the lesion blocked cerebrospinal fluid drainage from the fourth ventricle and brainstem, resulting in obstructive hydrocephalus [Figures 1a-c].

After obtaining the patient's consent, a multidisciplinary team consisting of obstetricians and neurosurgeons decided to perform resection of the intracranial lesion following delivery of the fetus by emergency cesarean section in view of the symptoms of increased intracranial pressure. The patient underwent tumor resection through a suboccipital small craniotomy in the three-quarter position following external ventricular drainage. Indocyanine green video angiography showed the location of a vascular nodule thorough the dura and the mural nodule was identified directly underneath. Following cyst decompression and tumor resection without resecting cystic capsule, it was confirmed that the cerebellum had become slack and pulsation was observed [Figures 2a-c]. The surgery was completed without any problems. Histopathological examination of the specimen confirmed hemangioblastoma [Figure 2d].

The patient recovered from general anesthesia and was extubated the next day. Postoperatively, the patient continued to experience dizziness and general malaise, but her symptoms resolved spontaneously with continued symptomatic treatment with oral medication. Postoperative brain MRI revealed that the tumor had been removed with no obvious residual tumor and the hydrocephalus had improved [Figures 3a-c]. The patient and neonate's general condition was confirmed to be stable and she was discharged home on the 16th day of her hospitalization without any neurological deficit and with the newborn in good condition at discharge. As for the mother, postoperative imaging studies confirmed the absence of other cerebrospinal hemangiomas, retinal hemangiomas, renal cell carcinoma, adrenal tumors, and pancreatic tumors, which were negative for Von Hippel-Lindau disease. As for the fetus, the child was not subjected to an aggressive systemic examination because he was only 2 weeks old at the discharge. At her follow-up examination 3 months postoperatively, there was no recurrence of symptoms, hydrocephalus, or tumor regrowth.

DISCUSSION

Mechanism of increase in the size of hemangioblastoma during pregnancy

Cerebellar hemangioblastoma is a benign vascular tumor and accounts for approximately 3% of all central nervous system tumors.[7,20] The combination of hemangioblastoma and pregnancy is rare because of its low incidence. The previous publications have focused on the effect of pregnancy on this tumor and there was a hypothesis that pregnancy could potentially accelerate hemangioblastoma progression through fluid retention, increasing venous pressure, and the occurrence of hormone receptors on tumor cell in patients with Von Hippel-Lindau disease.[1,6,22] However, it was also reported that the progression of symptoms in hemangioblastoma was simply a result of the natural course of the disease rather than the effects of pregnancy because gestational age is consistent with the peak age for the presence of hemangioblastoma symptoms.[21] Although there is currently no direct evidence of a relationship between pregnancy and hemangioblastoma progression, managing these patients to safely control the tumor and protect the fetus has been a major challenge.

Favorable management for hemangioblastoma during pregnancy

A variety of management options for hemangioblastoma occurring during pregnancy has been reported in the relevant literature, including conservative management with close observation, spinal fluid detour, and direct surgery. Conservative management with close observation is not a viable option for symptomatic patients because these conditions can deteriorate during pregnancy.[15]

To date, there are a total of 12 English reports regarding direct surgical treatment of symptomatic hemangioblastomas during pregnancy in 16 cases, primarily in case reports or case series [Table 1]. In these reports, 14 of the 16 patients underwent urgent tumor resection (UTR) during gestation,

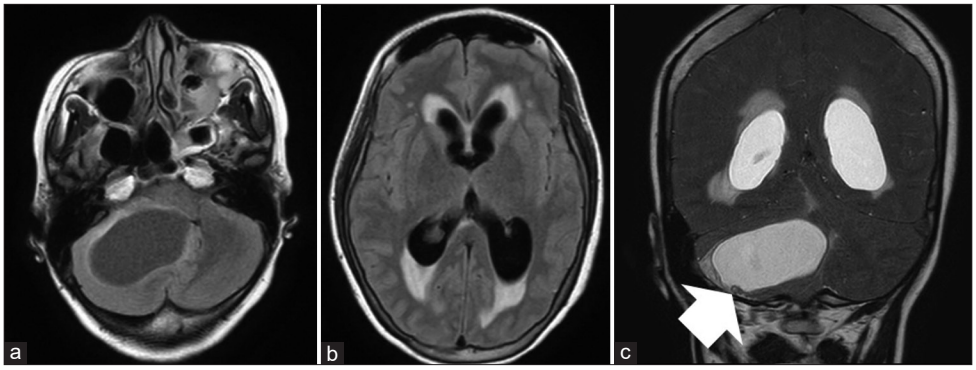

Figure 1: Preoperative FLAIR axial imaging showing a 5.6 cm cystic lesion in the right cerebellar hemisphere, with the lesion draining the fourth ventricle and brainstem, resulting in obstructive hydrocephalus (a and b). T2 coronal image showing a mural nodule in the cyst (white arrow) (c).

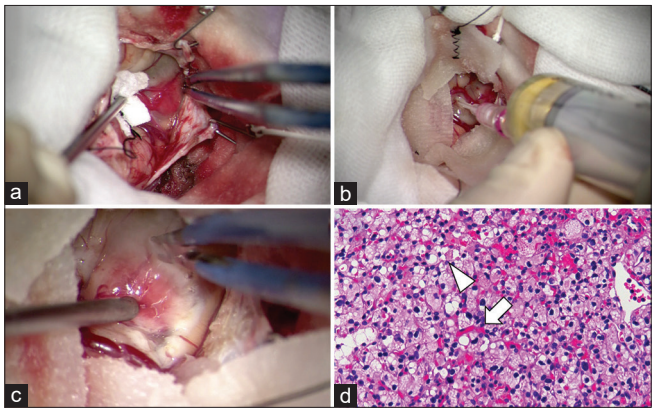

Figure 2: Intraoperative view showed that the dura was incised and a mural nodule was identified directly underneath (a). The tumor was punctured and the pale yellow contents were aspirated; internal decompression was performed (b). The mural nodule was removed as a mass without resecting the cystic capsule and it was confirmed that the cerebellum had become slack and pulsation was observed (c). Histopathological examination of the specimen revealed a rich vascular network (white arrow) and bubbly vacuolated cells with mild nuclear enlargement and clear cytoplasm (arrowhead) (d).

and the pregnancies were continued; in 12 patients, the infant was delivered at full term with no problems.[2,3,5,8,9,12-15,19] Naidoo and Bhigjee performed UTR in two patients at 21 and 33 weeks of gestation, achieving good results.[13] Similarly, Nathan *et al*. reported that a 28-year-old woman underwent UTR without spinal fluid detour during the second trimester of pregnancy with good results and continued pregnancy resulting in a full-term delivery.[14] Spinal fluid detour may also be an option for temporary symptomatic improvement; however, patients may deteriorate after shunting and may experience shunt-related complications.[17] Kasarskis *et al*. reported neurological deterioration after shunting in a patient with symptomatic cerebellar hemangioblastoma and described the need for direct emergency surgery on the hemangioblastoma.[8] The causes of deterioration after shunting include rapid tumor growth due to intravascular re-expansion of the tumor and direct pressure on the brainstem due to loss of the cushioning effect between the spinal fluid and the lesion. Ma *et al*. reported in 24 hemangioblastoma in pregnancy, four patients had external ventricular draining as an initial treatment; two patients had abortion at 6 weeks and 16 weeks, and other two patients had emergent cesarean section.[12] From this result, external ventricular draining has not shown positive results in maternal as well as fetal outcomes and is not recommended as a first choice of treatment. To the best of our knowledge, there are no case reports of endoscopic third ventriculostomy for hemangioblastoma during pregnancy.

Based on the previous literature, six cases were reported with brainstem compression and three cases were reported without brainstem compression [Table 1]. There was no difference in treatment strategy between patients with and without brainstem compression. Further case reports are needed, and at this point, it is not considered to be an indicator of decision-making.

Overall, these reports suggest that direct surgery is a better option, especially in early pregnancy. Repeated general anesthesia and shunt-related complications during pregnancy can both be avoided. Moreover, in patients who present after 30–32 weeks of gestation, as in this case, combined of cesarean section and surgical resection of the tumor can be planned if fetal lung maturity is confirmed.[17,18] Once obstetric criteria are met, an emergency cesarean section with epidural anesthesia should be employed to avoid prolonged labor and increased cerebrospinal fluid pressure.[16] A cesarean delivery should be preferred if there are any signs of increased intracranial pressure.[12] A multidisciplinary discussion between neurosurgeons, anesthetists, and obstetricians is necessary before surgery.[11] In this case, intracranial hypertension due to cerebellar hemangioblastoma in the mother was observed, and the delayed venous return associated with pregnancy contributed to the intracranial

Table 1: Summary of direct surgical treatment of symptomatic hemangioblastomas during pregnancy as reported in the past literature.

Author (year)	Age	Gestational age at presentation	Tumor location	Lesion size (cm)	Brainstem compression	Treatment	Maternal outcome	Fetal outcome
Broager (1949[2]	26	6 months	Cerebellum	N/S	N/S	UTR during gestation	Alive	Delivery at term
Scarcella *et al.* (1961)[19]	27	6 months	Cerebellum	4.0	N/S	UTR during gestation	Alive	Delivery at term
Kasarskis *et al.* (1988)[8]	18	2nd month	Cerebellum	2.5	N/S	UTR during gestation	Alive	N/S
Romansky *et al.* (1992)[18]	20	8 months	Cerebellum	N/S	No	UTR following C/S	Alive	Good
Nathan *et al.* (1995)[14]	28	2nd trimester	Cerebellum	4.0	N/S	UTR during gestation	Alive	Delivery at term
Naidoo and Bhigjee (1998[13]	26	21 weeks	Cerebellum	2.9	Yes	UTR during gestation	Alive	Delivery at term
	26	33 weeks	Cerebellum	4.5	Yes	UTR during gestation	Alive	Delivery at term
Delisle *et al.* (2000)[3]	30	30 weeks	Cerebellum	5.0	No	UTR during gestation	Alive	Delivery at term
Erdogan *et al.* (2002)[5]	35	24 weeks	Cerebellum	5.0	Yes	UTR during gestation	Alive	N/S
Kenyon *et al.* (2009)[9]	33	28 weeks	Cerebellum	5.0	No	UTR during gestation	Alive	Delivery at term
Rehman *et al.* (2009)[17]	27	29 weeks	Cerebellum	4.1	Yes	Cyst drainage during gestation and tumor resection following delivery	Alive	Delivery at term
Ma *et al.* (2018)[12]	31	6 weeks	Medulla	1.2	N/S	UTR during gestation	Alive	Abortion
	27	29 weeks	Cerebellum	4.2	N/S	UTR during gestation	Alive	Delivery at term
	26	23 weeks	Cerebellum	4.2	N/S	UTR during gestation	Alive	Delivery at term
	24	19 weeks	Medulla	3.7	N/S	UTR during gestation	Alive	Delivery at term
Nayak and Kumar (2020[15]	22	28 weeks	Cerebellum	5.4	Yes	UTR during gestation	Alive	Delivery at term
Present case, 2021	35	30 weeks	Cerebellum	5.6	Yes	UTR following C/S	Alive	Good

UTR: Urgent tumor resection, N/S: Not specified, C/S: Cesarean section

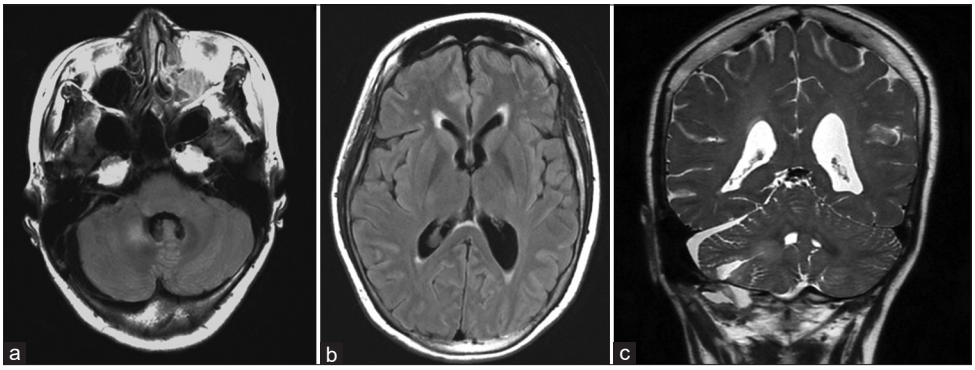

Figure 3: Postoperative FLAIR axial imaging showing that the tumor was successfully removed with no obvious residual tumor, and T2 coronal imaging showing that the hydrocephalus had improved (a-c).

hypertension was considered. Since there was a possibility that the tumor resection would not improve the intracranial pressure quickly, an emergency cesarean section was chosen after confirming fetal lung maturity.

Prognosis

The previous publications reported that most pregnant patients with hemangioblastoma during pregnancy experienced an uneventful gestational course [Table 1]. In our literature review of perinatal case reports, there were no maternal deaths due to neurological deterioration, and both maternal and fetal outcomes were quite favorable; regarding fetal prognoses, only one pregnancy ended in spontaneous miscarriage.[12] Other neonates, including preterm infants, were in good condition with no congenital malformations. Thus, reported maternal and fetal outcomes have been good in the past literature. Unfortunately, there are few case reports on the difference in outcome between early term delivery and full-term delivery in terms of fetal prognosis. Further case accumulation and investigation are needed.

CONCLUSION

We reported a case of symptomatic cerebellar hemangioblastoma during pregnancy and reviewed the literature. UTR combined with cesarean section can be planned once fetal lung maturity can be confirmed. A multidisciplinary discussion among neurosurgeons, anesthetists, and obstetricians is necessary to determine a treatment plan. Most cases of symptomatic hemangioblastoma during pregnancy have shown an uneventful gestational course and a favorable outcome for both mother and child.

Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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