

Case Report

Hemangiopericytoma of the foramen magnum in a pregnant patient: A case report and literature review

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Abstract

Background: The presentation of intracranial hemangiopericytomas is very rare, and only one case of a hemangiopericytoma during pregnancy has been reported in the literature. The management of these lesions poses a great challenge to the neurosurgeon, since the physiological and hormonal changes of pregnancy can exacerbate the symptoms of this highly vascularized neoplasm and pose different risks to both the mother and the fetus. We report the case of a patient who had sudden onset of intracranial hypertension at the ninth week of gestation due to a hemangiopericytoma of the foramen magnum and review the literature in this regard.

Case Description: A 23-year-old female who presented with signs and symptoms of intracranial hypertension at the ninth week of gestation was initially thought to have hyperemesis gravidarum. Because her symptoms persisted, she was found to have intracranial hypertension due to a tumor in the foramen magnum. She was treated by means of derivative surgery to allow for her pregnancy to progress beyond the first trimester, and at the 22nd week of gestation she underwent a sub-occipital craniotomy with partial tumor removal. Pathology was consistent with hemangiopericytoma. Both the mother and the fetus had positive outcomes.

Conclusions: To our knowledge, this is the second intracranial hemangiopericytoma presenting during pregnancy to be reported in the literature, and it is the first one of its kind to be located in the foramen magnum and causing severe intracranial hypertension.

Key Words: Hemangiopericytoma, hyperemesis gravidarum, intracranial hypertension, intracranial neoplasms, pregnancy complications

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INTRODUCTION

The presentation of intracranial tumors during pregnancy is rare, and international data estimate an incidence of approximately 15 in 100,000.^[2] Even though the

etiology of primary central nervous system (CNS) tumors has not been clearly established yet, there is experimental, biological, clinical, and epidemiological evidence that suggests the influence of sexual hormones in their development.^[3] However, this subject is still very

controversial, for the literature provides evidence both in favor and against the influence of female hormonal factors in the development of intracranial neoplasms, and many of these data are inconsistent.^[3,4,7,16] Intracranial hemangiopericytomas, in contrast, are a very rare entity that represents 1% of all intracranial tumors, and only one case presenting during pregnancy has been reported in the literature.^[1] Physiological changes during pregnancy can exacerbate the symptoms related to the intracranial neoplasm, as well as affect the intracranial pressure.^[5,12,14] The management of these lesions is therefore a very complex challenge for the neurosurgeon, because the location and natural history of the tumor, as well as the risks and benefits for both the mother and the fetus, must be taken into consideration. We present the case of a patient with symptoms of intracranial hypertension during the first trimester of her pregnancy due to an intracranial hemangiopericytoma.

CASE PRESENTATION

A 23-year-old female patient, with no known past medical history, was referred to our centre at the 9th week of her first pregnancy because of incoercible vomiting and mild dehydration. She was admitted to the Obstetrics department, where she received symptomatic treatment with antiemetics and prokinetics. She was discharged on the third day with the diagnosis of hyperemesis gravidarum. Nevertheless, 2 weeks later, the patient was readmitted for presenting incoercible vomiting, dizziness, and headaches. On this occasion, vitamin B6 was added to her treatment, and she was discharged on the fifth day.

On the 16th week of gestation, the patient was readmitted because she persisted with the symptoms. At this time, the patient was obtunded and disoriented, and she also complained of an oppressive occipital headache.

On physical examination, left VI cranial nerve palsy, papilledema, and right ankle clonus were found. Brain magnetic resonance imaging (MRI) was performed, suspecting a dural sinus thrombosis. However, the study revealed triventricular hydrocephalus with transependymal edema due to a tumor in the posterior fossa [Figures 1 and 2].

The patient was assessed by the authors, and she underwent an urgent ventriculoperitoneal shunt with a high pressure valve without any complications. In spite of a favorable course in the immediate postoperative period, the patient experienced neurological decline with blurry vision, vomiting and headaches 10 days later. These symptoms did not respond to medical treatment with dexamethasone. In view of the latter, an endoscopic third ventriculostomy was performed, and her derivative system was removed. At the same time, we discussed with the Obstetrics department the need to operate on the mass, since the patient showed progressive involvement of the brainstem.

At 22 weeks of gestation, the patient underwent a right sub-occipital craniotomy with partial resection of a highly vascularized mass in the posterior margin of the foramen magnum. Its intraoperative biopsy reported a low-grade stromal tumor, suggestive of hemangioblastoma. The surgery was performed without any obstetric or neurologic complications. In the postoperative period, the patient only presented mild right hemiparesis, which remitted within a few days. The definitive biopsy showed the presence of a vascular neoplasm with irregular “stag-horn” cells, and positive immunohistochemistry for CD34 and vimentin. All of the latter was compatible with hemangiopericytoma. The patient continued to be followed at the Neurosurgery and Obstetric outpatient clinics after the 26th week of gestation. She underwent a cesarean section at the 36th week because of intrauterine growth restriction, and it was performed without any



Figure 1: T1-weighted sagittal view showing a hypointense lesion in the posterior fossa, with displacement of the brainstem and the cerebellum

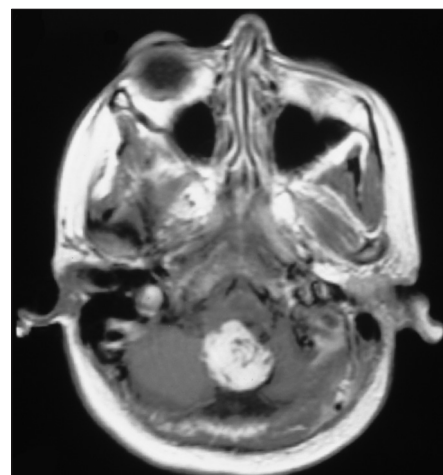


Figure 2: T1-weighted axial view showing mildly enlarged ventricles and a contrast-enhancing lesion in the posterior fossa, exerting mass effect on the brainstem

complications. Currently, both the mother and the baby are in good health.

DISCUSSION

Intracranial tumors are a relatively rare complication in the setting of pregnancy. Glial neoplasms are the most common, followed by meningiomas and acoustic neuromas, all of which have similar frequencies to nonpregnant controls.^[14] Hemangiopericytomas, in contrast, represent less than 1% of intracranial tumors, and they are more common in men than in women,^[1, 6] which makes their presentation during pregnancy very infrequent.

One of the challenges faced by physicians when diagnosing intracranial tumors during pregnancy is that the symptoms of intracranial hypertension, such as nausea and vomiting, can be mistaken for pregnancy-related issues, such as hyperemesis gravidarum.^[12] In our case, it is not surprising that her symptoms were attributed to hyperemesis, for the nausea and vomiting due to this condition tend to peak over the first trimester. Nausea and vomiting due to intracranial tumors appear later because of the hemodynamic changes of pregnancy, which mediate a common pathway to increase the mass effect of the intracranial tumor.^[13]

To date, only one case of intracranial hemangiopericytoma during pregnancy has been reported in the literature.^[1] Their patient presented at 38 weeks of gestation with diplopia and VI cranial nerve palsy, without any signs of intracranial hypertension. Her MRI showed the presence of a hyper-intense image in the right middle fossa, but surgery was deferred to the second month postpartum, for she did not present major neurological involvement. Her histopathology was compatible with hemangiopericytoma, and both the mother and the child had a good clinical outcome.

Unlike the previous case, our patient presented symptoms at an earlier gestational age, and she suffered progressive neurological decline with intracranial hypertension and involvement of the brainstem. The latter was, to a large extent, due to the location of her tumor in the foramen magnum. Therefore, she required a more aggressive management, including a sub-occipital craniotomy at the 22nd week of gestation. Currently, there are no guidelines for the management of intracranial tumors in pregnant patients, because the available evidence is limited to case reports or small patient series.^[2] Therefore, decision-making in the management of these patients becomes extremely complex.

In 2009, Ng and Kitchen created an algorithm to guide the management of pregnant patients with intracranial tumors.^[10] In case of neurologically stable, early pregnancies, these authors recommend allowing

the pregnancy to progress until the second trimester. Once this point has been reached, they recommend neurosurgery. The reason why they choose the beginning of the second trimester is that the hemodynamic changes have not reached their peak yet, which renders the risk of intraoperative bleeding not significant. Furthermore, the fetus has already completed organogenesis, which accounts for the risks of teratogenicity to be decreased. In contrast, if the patient is unstable, these authors advocate for a neurosurgical approach, regardless of gestational age.

In our case, two surgical interventions, namely the ventriculoperitoneal shunt and the third ventriculostomy, were performed in order to treat her intracranial hypertension and allow for her pregnancy to advance. The goal was to avoid exposing the mother and the fetus to the anesthetic and surgical risks of a craniotomy. These measures allowed for the pregnancy to progress until the second trimester, which is when the mass was surgically treated, without further complications. Thus, greater neurological decline of the mother was prevented, and the pregnancy progressed until the birth of a healthy preterm infant. This course coincides with the one observed by Terry *et al.*^[14] In their retrospective study of an American obstetric population with intracranial tumors, they determined that neurosurgical intervention during pregnancy could contribute to the continuation of the pregnancy to term.

Much has been discussed about the correlation between hormonal factors and the development of intracranial tumors. Specifically, it has been noted that the incidence of meningiomas is twice as high in women as in men, while gliomas are 1.5 times more frequent in men.^[9] In addition, there is evidence in favor of meningioma growth during pregnancy, as well as a strong association between meningioma and breast cancer.^[8] Specifically, European and American studies have found a correlation between parity and the risk of presenting meningiomas,^[3,16] while an early menarche and a late menopause seem to be protective factors against gliomas.^[4] Nevertheless, the latter is still controversial, and many studies have not been able to show a direct correlation.^[7,9]

Hemangiopericytomas have not been studied as comprehensively, for they are a rare histological type of intracranial tumors. However, molecular studies have shown that these neoplasms strongly express receptors for the placenta-induced growth factor (PIGF) and the vascular endothelial growth factor (VEGF) 1.^[11] Furthermore, it is well known that during pregnancy, these factors are involved in the trophoblastic invasion of the uterus and placental development.^[15] Therefore, it can be hypothesized that the increase of these factors during pregnancy contributes to hemangiopericytoma tumor growth and the mass effect on the surrounding tissues. In addition, the changes in the mother's intravascular

volume can cause these highly vascularized tumors to ingurgitate and become edematous, which creates an even larger mass effect.^[13]

In conclusion, our case evinces the complexity of managing intracranial tumors during pregnancy. Protean changes in the hormonal and physiological milieu inherent to the pregnant state can favor the manifestation of these neoplasms, be it through tumor growth itself or through the hemodynamic changes of pregnancy. Therefore, it is very important to provide early care. Since evidence is controversial, and since there are no validated guidelines, it is extremely important for an interdisciplinary team of neurosurgeons and obstetricians to approach each case individually, in order to achieve the best results for the mother and the child.

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