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Hemangioblastoma and Pregnancy: A Case Report and Review of Literature

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Abstract

Objective: There is no increase in the frequency of hemangioblastoma in pregnancy compared to the same age group, but it may become symptomatic and the symptoms may be confused with the symptoms of pregnancy. As the symptoms such as severe nausea, vomiting and headache rarely continues with treatment, it is essential to consider brain tumors in differential diagnosis in cases with persistent nausea and vomiting. In addition, pre-diagnosis of this type of patients will enable the team to be prepared for emergency surgical intervention that may develop at any time during pregnancy. In this article we aimed to present a case of pregnant with brain tumor that had followed until term.

Case: Following an uneventful first pregnancy, in the second pregnancy of 37-year-old G2P1 woman, the first two trimesters of pregnancy follow-up revealed normal pregnancy features except for nausea and vomiting. The patient was referred to the neurology outpatient clinic when the patient's nausea and vomiting did not respond to medical treatment and the complaint of frequent headache was added. The patient's neurological examination did not reveal any problem. The patient again presented with nausea, vomiting and severe headache at the 35th week of pregnancy follow-up. Complete blood count, biochemistry, bleeding profile and blood pressure evaluation revealed normal findings and no neurological deficit was determined during her physical examination. The patient was asked to undergo an emergency MR (Magnetic resonance) examination, MR report showed a cystic mass lesion with a size of approximately 48 x 42 mm in the right cerebellar hemisphere, which had compression onto the 4th ventricle and moving the cerebellar hemisphere to the left. Elective Caesarean section was performed at 36th week of gestation. Craniotomy was performed four days after the caesarean section, the brain tumor was resected and the histopathological examination revealed hemangioblastoma.

Conclusion: The diagnosis was delayed due to the symptoms mimicking pregnancy symptoms, therefore intracranial masses should be kept in mind in common

pregnancy symptoms such as nausea, vomiting and headache that do not respond to treatment.

Keywords: Hemangioblastoma; Pregnancy; Magnetic resonance; Craniotomy

Introduction

Hemangioblastoma is one of the commonly encountered brain tumors which are commonly originated from cerebellum, medulla, spinal cord, cerebral hemisphere, meninges and the retina. These tumors constitute 7.3% of the posterior fossa tumors and 1-2% of all intracranial tumors [1,2]. One fourth of these tumors are accompanied with autosomal inherited von Hippel-Lindau syndrome and 75-80% is sporadic [3]. These tumors are slowly growing tumors and commonly present with the findings related to intracranial pressure increase. Magnetic resonance imaging shows a contrasting cystic or solid mass. The most common site of origin is cerebellar paramedian site as a well-defined mural nodule, which can be separated from the surrounding tissue. These tumors are seen in both sexes with equal ratio and most frequently observed at 35-45 years of age. It can also be seen in children and the elderly. Cerebellar hemangioblastoma also takes the name Lindau. They are called as Lindau tumor when it is accompanied with Von Hippel-Lindau syndrome (VHL). In addition, kidney, liver, pancreas and adrenal cysts, renal cell carcinoma (RCC), pheochromocytoma can be seen cases with these kind of tumors, however sporadic hemangioblastoma are usually encountered at a younger age [4-5].

In some cases with these tumors, secondary polycythemia may be found in the construction of tumoral erythropoietin. Tumors have microscopically benign appearance, forming stromal cells and capillary vessels between endothelial cells [6]. They may show focal micro invasion to the neighboring neural parenchyma, despite having well-borders. Recurrence may occur after incomplete excision. Although there is no sign of significant cytological malignancy, distant metastases are rarely seen [1].

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In this article we aimed to present a case of pregnant with brain tumor that had followed until term.

Case Study

The 37-year-old G2P1Y1 woman gave her first birth by a Cesarean section four years ago. There was no specific feature in the patient's history and family history. In the first two trimesters of second pregnancy follow-up; except for nausea and vomiting, there were no additional problems. The patient was referred to the neurology outpatient clinic when the patient's nausea and vomiting did not respond to treatment and the complaint of frequent headache was added. The patient's neurological examination did not reveal any problems. The patient also expressed that she had the same complaints in the first pregnancy. At her last visit, the patient again presented with nausea, vomiting and severe headache at the 35th week of pregnancy. Hemogram, biochemistry, bleeding profile and blood pressure evaluation revealed normal findings and no neurological deficit was determined during her physical examination. The patient was asked to undergo an emergency MR examination, MR report showed a cystic mass lesion with a size of approximately 48 x 42 mm in the right cerebellar hemisphere, which had compression onto the 4th ventricle and moving the cerebellar hemisphere to the left (Figure 1). Neurosurgery and neurology consultation was requested. The patient was admitted to the perinatology clinic for follow-up and after determination of fetal well-being and the general condition of the patient became stable, the patient was referred to the 3rd stage health institution with the suggestion of neurosurgical and neurological consultation. Elective Caesarean section was performed at 36th week of gestation. Craniotomy was performed four days after the caesarean section, the brain tumor was resected and the pathology report was determined as hemangioblastoma. Routine neonatal evaluation revealed a healthy newborn.



Figure 1: Showing a cystic mass lesion.

Discussion

Diagnosis, follow-up and treatment of brain tumors encountered during pregnancy require a multidisciplinary approach [7]. In our case, there was no other clinical symptom except nausea, vomiting and headache, which increased occasionally until the 35th week of pregnancy. In this article, we aimed to report a case of cerebellar hemangioblastoma complicated by pregnancy and managed successfully.

Malignant brain tumors are fortunately a rare disease and incidences of these tumors shown to be similar between pregnant and non-pregnant women. Previous report indicated the importance of early and effective multidisciplinary management of these disorders according to condition of patient and age of pregnancy [8].

In a previous report of case with hemangioblastoma during pregnancy, pregnant woman was reported to have disturbed consciousness with poor general condition. In their case, a large tumor in the cerebellar vermis along with an obstructive hydrocephalus was detected following imaging modalities. Patient was scheduled to surgical excision of the tumor and extraction of the fetus. Due to the rapid neurological deterioration secondary to tumor progression, authors reported to plan an emergency cesarean section. Surprisingly following fetal extraction, the level of consciousness was reported to be improved. Tumor resection was postponed until the patient's general condition improved. However, a second neurological deterioration was observed secondary to the worsening hydrocephalus. Worsening in hydrocephalus was claimed to be due to the increased cerebral blood flow following uterine contraction. As a result patient underwent an emergency surgery for the brain tumor two days after delivery. Hemangioblastoma was diagnosed by histopathological examination [9]. In our case, neurosurgical intervention was also postponed for 4 days, hydrocephalus was not detected in our case before delivery and no deterioration observed after delivery.

Von Hippel-Lindau disease is a rare genetic disorder which gives rise to a range of tumors including central nervous system hemangioblastoma. A case of caesarean section in a patient with symptomatic cerebellar hemangioblastoma associated with von Hippel-Lindau disease was reported. An intracranial pressure monitor was inserted before surgery, which enabled intracranial pressure to be monitored throughout [10].

The importance of performing a neurological examination in all cases of persisting headache and/or dizziness during pregnancy was emphasized. Other symptoms that require further evaluation were reported to be profound dizziness with instability of gait and the presence of isolated or bilateral lateral rectus palsies in any patient with a headache, which should be investigated by urgent imaging to exclude a space-occupying.

In our case, pregnant woman had only persistent nausea and vomiting. The literature on the characteristics of intracranial neoplasms during pregnancy consists largely of case report.

There are some case reports of hemangioblastoma worsening or being diagnosed in pregnancy. It was suggested that

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expansion of the tumour vascular bed, increases in blood volume and/or altered hormonal milieu might have some impact on tumor physiology. Furthermore, cerebellar hemangioblastomas were shown to have progesterone receptor [11]. Although it is expected to see deterioration in the symptoms during pregnancy, our case did not show any deterioration in the symptoms up to term pregnancy.

Hemangioblastoma is one of the rare brain tumors seen in pregnancy. In our case, the diagnosis was delayed due to the clinical symptoms mixed with pregnancy symptoms, therefore intracranial masses should be kept in mind in common pregnancy symptoms such as nausea, vomiting and headache that do not respond to treatment. With the early diagnosis of intracranial masses seen during pregnancy, the risk of newborn and maternal mortality will be minimized by intervening in a multidisciplinary center with obstetrician, neurosurgeon and newborn specialist.

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