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Cerebellar Hemangioblastoma Symptomatic During Pregnancy: A Short Review

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Abstract

Background: Symptomatic hemangioblastoma of the posterior cranial fossa presenting during the pregnancy is very rare. Hemangioblastoma may presents with persistent nausea and vomiting during pregnancy, which may be misinterpreted to be caused by hyperemesis gravidarum, which can further delay the diagnosis of raised intracranial pressure. Previously very few isolated cases of neurosurgical intervention with good outcome is reported in literature, *Aim of study:* To analyze the effect of VP shunt in case of symptomatic posterior fossa hemanagioblastoma in a pregnant women. *Result:* A 28-year- old female with 22-weeks pregnanc, presented with a history of progressive headache, vomiting and diplopia of 4weks duration. She developed ataxia and minimal weakness one week prior to admission. Neuroimaging revealed hemangioblastoma. She underwent ventriculoperitoneal shunt procedure in view of poor neurological status. However, she showed progressive worsening of the neurological symptoms despite ventriculoperitoneal shunt surgery. Finally she was taken- up for emergency midline suboccipital craniectomy and complete excision of right cerebellar hemangioblastoma. After removal of cerebellar hemangioblastoma, she made unremarkable recovery. She delivered a healthy male baby at 37 week of gestation by normal vaginal delivery. *Summary:* Cerebellar hemangioblastomas becoming symptomatic in second trimester pregnancy may warrant CSF diversion surgery and few cases even need definitive excision surgery for haemangioblastoma can be associated with good maternal and foetal outcome. So treating obstetric surgeon and neurosurgeon should consider surgical intervention can yield good outcome.

Keywords

Cerebellar Hemangioblastoma, Pregnancy, Hydrocephalus, Surgical Therapy

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1. Introduction

Cerebellar hemangioblastoma are unusual benign intracranial lesion may remain asymptomatic for prolonged period. However, rarely, some hemangioblastoma may show rapid increase in size during pregnancy causing brain stem compression and obstruction to CSF pathway producing obstructive hydrocephalus. Intracranial neoplasms as a overall presenting during pregnancy is uncommon. About twenty five cases of posterior fossa haemangiobalstoma, which became symptomatic during pregnancy and requiring surgery has been reported. Authors are reporting a case of mangioblastoma, who presented in during second trimester of pregnancy with rapid deterioration of vision.

It represents a therapeutic challenge of decompression of hemangioblastoma without producing any further stress and problem for pregnant women and fetus. In addition to pregnancy induced aggravation of symptoms, the increased vascularity of these tumors, which in turn may initiate new or exacerbate pre-existing symptoms and making them more prone to cause compression and distortion of cerebellar hemisphere and brain stem with obstruction along the CSF pathway. Patient presents with feature of raised intracranial pressure caused by obstructive hydrocephalus in addition to cerebellar and brainstem

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compressive features. Bulent et al [1] reported a case of hemangioblastoma, who noticed worsening of symptoms in the third trimester of pregnancy, which is contributed to delay in neuroimaging diagnosis and management.

2. Case Illustration

A 28-year- old female presented to our neurological services with a history of progressive headache, vomiting and diplopia of 4weks duration. She developed ataxia and minimal weakness one week prior to admission. She was carrying 22-weeks pregnancy (gravida 1, para 0). Her past medical history was not significant. Her general examination was normal. Per abdominal examination showed a uterine size consistent with 22- weeks of gestation, normal fetal heart rate. Neck stiffness was present. Neurologically she was conscious, and alert. Fundi revealed bilateral secondary optic atrophy, however, no retinal angioma was observed. Bilateral six nerve paresis was present, left sided pyramidal minimal hemiparesis was present. Left plantar response was extensor. She had bilateral cerebellar signs, being more prominent on right side. Marked ataxia prevented her to walk. The rest of the neurological examination was essentially within normal limit.

Biochemical and hematological parameters were within normal limits. Magnetic resonance imaging revealed an intra-axial mass lesion of 6x 5x 4 cm dimension in the right cerebellar hemisphere, which was hypointense on T1 weighted image causing marked distortion of cerebellar hemisphere, (fig-1) with brain stem compression, Pressure over fourth ventricle produced obstruction of CSF outflow, with enlargement of both lateral ventricles and ballooning of third ventricles. On T2 weighted image showing hyperintense signal. (fig-2, 3) On administration of gadolinium contrast administration revealed a brilliantly enhancing subpial mural nodule. (fig-4) A diagnosis of hemangioblastoma was made with gross obstructive hydrocephalus. Abdominal ultrasound revealed single viable fetus with fetal growth appropriate for an age. However, maternal ultrasound examination showed normal liver, pancreas, kidney.

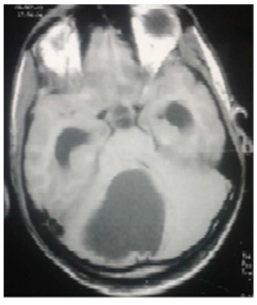

Fig. 1. MRI brain, axial section T1WI showing an intra-axial mass lesion, sized 6x 5x 4 cm in the right cerebellar hemisphere, with hypointense signal causing marked distortion of cerebellar hemisphere and brain stem compression.

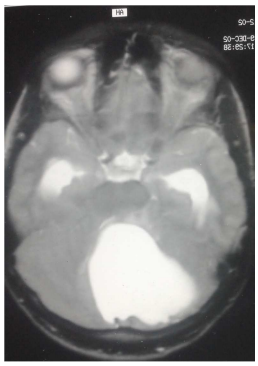

Fig. 2. MRI brain, T2WI, axial section showing hyperintense intra-axial mass lesion in the right cerebellar hemisphere causing marked distortion of cerebellar hemisphere and obstructive hydrocephalus.

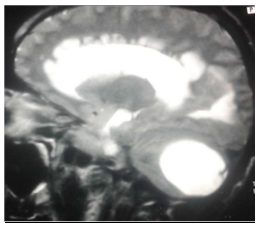

Fig. 3. MRI brain, T2WI, sagittal axial section showing hyperintense intra-axial mass lesion in the right cerebellar hemisphere causing marked distortion of cerebellar hemisphere.

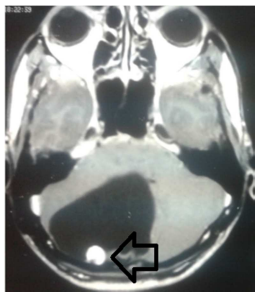

Fig. 4. MRI brain, Gadolinium enhanced image showing brilliantly enhancing mural nodule (arrow head within large no n-enhancing cyst.

She was taken up for surgery with consultation of obstetrician. She underwent CSF diversion, left sided ventriculoperitoneal shunt procedure. She had no improvement in the postoperative period. In fact, she showed deterioration in the neurological status after 48 hours, an emergency cranial CT scan revealed well-decompressed lateral ventricle with mild increase in size of right cerebellar hemangioblastoma. She underwent emergency right paramedian suboccipital craniectomy and excision of vascular cherry red mural nodule with straw colored cyst content. At the end of surgery, cerebellum was lax and pulsating. The histopathological examination of specimen revealed confirmed hemangioblastoma. In the postoperative period, she had marked improvement in the neurological status. The ultrasound examination of fetus on seventh postoperative day following second surgery revealed healthy child. She delivered a healthy female baby at the full term. She was doing well, at last follow-up one year following

surgery. A cranial contrast enhanced computed tomography scan, at the last follow-up revealed no residual lesion or other hemangioblastoma.

3. Discussion

Cerebellar hemangioblastoma presenting during pregnancy is very rare. [1] Only isolated few case- reports of surgical intervention during pregnancy has been reported. These can present for the first time during pregnancy or symptoms may worsen during pregnancy. As surgical procedure for excision of cerbellar, hemangioblastoma should not produce any further stress to pregnant women and foetus. The patient may require urgent surgical therapy due to rapid development of obstructive hydrocephalus and brainstem compression. Our patient also required urgent surgery as reported by Bulent et al. [1]

Patient may become symptomatic during the pregnancy. Previous reports noted that pregnancy seems to exacerbate the clinical course of intracranial tumours including hemangioblastoma of posterior cranial fossa. Ogasawara et al reported a case of spinal hemangioblastoma showed deterioration during the pregnancy in a case of Von Hippel – Liandau disease. [14]

Various hypothesis were proposed to explain the rapid neurological deterioration of hemangioblastoma patient during pregnancy. According to first hypothesis, rapid expansion or engorgement of vascular bed, which is presumably the result of generalized increase in blood volume that occurs during pregnancy. Karskis et al supported the vascular engorgement of hemangioblastoma probably accounts for some of patients during the pregnancy. [5] The other hypotheses are direct hormonal effect on tumor growth rate, mediated by hormonal receptors. [13, 15] During pregnancy the maternal plasma volume increases from the six week of gestation to the peak volume of 3500 ml by the 32-34 weeks. [12] Several metabolic and hemodynamic changes associated with pregnancy may in fact also may be collectively responsible for enlargement and increased vascularity hemangioblastoma. Arterial hypertension or pre-eclampsia and tendency to retain extra cellular and intracellular fluid during pregnancy are considered to additional predisposing factors for development of increase in size of adenoma and associated increase intracranial pressure. These changes may accentuate the symptoms associated with adenoma during pregnancy. The cardiac output rises by about 20% during the first trimester, and continue to persist until end of pregnancy. These changes are seconadry to increased trophoblastic activity leading to increased production of oestrogen and progesterone. [12, 13, 15, 16]

Frantzen et al. analyzed the effect of pregnancy on von Hippel-Lindau in a total of 29 cases and observed progression score of cerebellar hemangioblastomas was significantly after pregnancy in about 40% cases in their study. [17] Further, authors concluded pregnancy accelerates the increase in the size of cerebellar hemangioblastoma progression and causes a high pregnancy complication rate and recommend close monitoring of such patients during reproductive phase of women specially during pregnancy and preconception period. Erdogan et al. noted incidence of posterior fossa hemangioblastoma getting symptomatic during pregnancy is extremely low. [18] However, pregnancy aggravates the clinical course of intracranial tumors. Author cautioned the treating neurosurgeons and obstetricians must be aware and careful about clinical course as surgery may be necessity to relieve the mass effect even during pregnancy, which if ignored can cause deleterious effect on maternal as well as fetal survival. [18]

Contrary to the view that pregnancy causes enlargement of haemangioblastoma and size of the peritumoral cyst, Ye DY et al analyzed a total of thirty-six consecutive females, out of which nine got pregnant and comparing non-pregnant control noticed size of the cyst and nodule progression is almost similar in the pregnant and non-pregnant female. Authors also concluded pregnancy may not be associated with increase in the size of peritumoral cyst development or also enlargement of cerebellar hemangioblastoma specially in patients with Von Hippel – Liandau disease. [19]

Authors feel large size and increased vascularity of hemangioblastoma during pregnancy make them more vascular and large size, which may worsen the symptoms in a pre-existing tumor, this is well illustrated by our case, as after CSF diversion procedure, she had mild improvement as compensatory mechanism and again started deterioration at 48 hours, might explain ongoing increase in the size of hemangioblastoma, which might be causing increasing pressure effects over brain stem. Similar observation was also made by Kasarsis et al where ventriculoperitoneal shunt was carried on 18 year –old -women with second month of pregnancy, showed rapid deterioration after 12 days necessitating emergency surgery for hemangioblastoma surgery. [6] Roman sky et al also reported a 20- year -old female with second month of pregnancy, combined surgery of ventriculoperitoneal shunt, caesarian section for delivery of fetus and suboccipital craniectomy was done for surgical remove of hemangioblastoma. [5] However, in case of Von - Hippel Lindau disease, where symptomatic hemangioblastoma was operated during pregnancy, the other small hemangioblastoma may increase in size may require surgical excision also. Surgery for hydrocephalus and hemangioblastoma in a single setting was carried out in a 7 -

weeks of gestation with good outcome of pregnancy by Naido et al. [2] Nathan et al reported a 28- year female underwent only tumour decompression without CSF diversion procedure during second trimester pregnancy. Resulted in continuation of pregnancy with good outcome. [3] Inoueet al. reported a- 19-year-old woman with cerebellar hemangioblastoma, carrying also 35 weeks pregnancy, developed headache, nausea, and general fatigue with disturbed consciousness, and the patient's general condition was poor on admission. On imaging large haemangioblastoma in the cerebellar vermis causing obstructive hydrocephalus. She underwent an emergency cesarean section and two days later underwent posterior fossa surgery of haemangioblastoma excision. [20]

In contrary, our case was carrying 22- week pregnant and pregnancy continued after intitial VP shunt surgery followed by intracranial surgery for haemangioblastoma excision after 2 days. It is interesting and unique case of surgical excision with good maternal and pregnancy outcome.

4. Conclusion

Cerebellar hemangioblastoma, a rare posterior cranial fossa lesion, should be considered in differential diagnosis of persistent nausea and vomiting, when associated with focal neurological deficit during pregnancy. Early diagnosis and direct surgery for excision of hemangioblastoma alone or in association with VP shunt surgery. As only surgery for CSF, diversion procedure may not be suffice and may require emergency surgical excision of hemangioblastoma as observed in our case. The symptomatic hemangioblastoma during pregnancy can be safely operated during pregnancy, if associated with of failure conservative management or not responding with progressive worsening of neurological deficit.

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