Conservative management of cavernous sinus cavernous hemangioma in pregnancy

Case report

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Cavernous sinus cavernous hemangiomas in pregnancy are extremely rare lesions. The precise management of these lesions remains unknown. The authors present a case of a cavernous hemangioma in pregnancy, centered within the cavernous sinus that underwent postpartum involution without surgical intervention.

A 34-year-old pregnant patient (gravida 1, para 0) presented to an otolaryngologist with persistent headache and left-sided facial pain and numbness in the V1 distribution. While being treated for sinusitis, her symptoms progressed to include a left-sided oculomotor palsy and abducens palsy. Magnetic resonance imaging without contrast revealed an expansile mass within the left cavernous sinus consistent with a cavernous hemangioma. The patient was evaluated by a neurosurgeon who recommended close follow-up and postpartum imaging without surgical intervention. Although the lesion enlarged during pregnancy, the patient was able to undergo an uncomplicated cesarean section at 37 weeks. All facial and ocular symptoms resolved by 9 months postpartum, and MRI showed a decrease in lesion size and reduced mass effect. The authors conclude that nonsurgical management may be a viable approach in patients who have an onset or exacerbation of symptoms associated with cavernous sinus cavernous hemangiomas during pregnancy because postpartum involution may negate the need for surgical intervention.

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Key Words • cavernous sinus • cavernous hemangioma • pregnancy • oncology

Cavernous hemangiomas (CHs) are benign, slow-growing vascular anomalies. While most CHs are intracranial, rarely they are found in extraaxial locations, including the orbit18 and the cavernous sinus (CS). Cavernous sinus cavernous hemangiomas (CSCHs) typically result in neurological morbidity related to compression of intracavernous cranial nerves.9,12,13,17

Surgery is considered the preferred method of treatment when lesion location allows safe resection. When anatomical location compromises safe resection,5,14 radiosurgery is an option. Radiosurgery has been shown to reduce the size of lesions while avoiding excessive bleeding and cranial nerve damage.5,14,21 More recently, stereotactic fractionated radiotherapy has also been effective in the treatment of orbital CHs.14

In general, there is a female predominance of CHs, with the peak age of incidence around the age of 40 years.13 Several case reports show that both intracerebral CHs15 and extracerebral CHs within the optic disc,20 retina,19 or optic nerve20 are at an increased risk for hemorrhage during pregnancy, requiring intervention. However, there are no established treatment options for pregnant patients with these vascular lesions. We report a case of extracerebral CSCH that presented during pregnancy and was successfully managed without resection.

Case Report

Presentation. A 34-year-old pregnant patient (gravida 1, para 0) developed persistent headache and left-sided facial pain and numbness at 21 weeks of gestation. She consulted an otolaryngologist who treated her with antibiotics and methylprednisolone for presumed sinusitis. When she developed left eye ptosis at 25 weeks of gesta-
tion, she was referred to an outside neurologist who found ophthalmoplegia of the left eye and sensory deficit in the left V1 distribution.

Clinical Course. The patient was then referred to the emergency department of the NewYork-Presbyterian Hospital where her examination was notable for 2 mm of ptosis in the left eye, impaired adduction, elevation, and depression of the left eye consistent with a partial left oculomotor nerve palsy and an inability to abduct the left eye, consistent with a complete left abducens palsy. Magnetic resonance imaging showed an expansile mass in the left CS (Fig. 1A) presumed to be a schwannoma.

Several weeks later, the patient's left periorbital pain and numbness as well as the vertical motility of the left eye improved. However, repeat MRI showed a slight increase in lesion size and multiple serpentine T2 hyperintense structures, which together were suggestive of a CSCH (Fig. 1B). A cesarean section was recommended to reduce the risk of enlargement and hemorrhage during delivery. The neurosurgery team recommended no acute surgical treatment given the extent of the lesion within the CS, close follow-up, and postpartum imaging to assess for possible spontaneous involution.

At 30 weeks of gestation the patient presented with progressive left orbital pressure, lid edema, worsened ptosis to 5 mm of the left eye and a new 2-mm anisocoria with mydriasis of the left eye. Repeat MRI revealed a significant enlargement of the lesion with extension to the orbital apex (Fig. 1C). There was severe compression of the left internal carotid artery and increased impingement of the left optic nerve. The patient was offered the option of immediate delivery, but she declined. A cesarean section was scheduled for 37 weeks.

At 35 weeks of gestation, the patient was referred to a neuro-ophthalmologist for declining left visual acuity. Neuro-ophthalmic examination confirmed a complete left abducens palsy and partial left oculomotor palsy, including a mild adduction defect, mild ptosis and mydriasis, worse in light. However, there was no relative afferent pupillary defect. Visual acuity in the left eye improved with a +2 lens, consistent with blurry vision from a lens accommodation paresis associated with the oculomotor palsy rather than from optic nerve compression (Fig. 2).

Postpartum Course. The patient underwent the scheduled cesarean section without complication. Two weeks postpartum, she developed complete ptosis and decreased supra- and infra-adduction of the left eye, consistent with worsened oculomotor nerve function. Furthermore, in addition to left mydriasis in the light, a relative miosis in dim light had developed, consistent with a new left Horner’s syndrome. Magnetic resonance imaging demonstrated a further increase in size of the lesion (Fig. 1D).

The neurosurgery team recommended a steroid treatment course. Two weeks into this treatment, the patient had improved vision and decreased ptosis, but she continued to have diplopia with leftward gaze and left eye pressure exacerbated by recumbence. Magnetic resonance imaging showed a decrease in lesion size (2.7 × 1.8 × 1.4 cm compared with 3.4 × 2.7 × 2.0 cm) and a reduced mass effect on the left internal carotid artery.
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![Diagram of Pupil Dilation and Sympathetic Chain]

Fig. 2. Accommodation paresis due to left oculomotor palsy with mydriasis in light. CN = cranial nerve.

Compression had resolved (Fig. 1E). By approximately 4 months postpartum, the left oculomotor palsy had resolved and there was 50% left abducens function. Notably, by 9 months postpartum, the patient’s neurological examination had returned to normal, and MRI showed a significant decrease in size of the CH (Fig. 1F).

Discussion

Cavernous hemangiomas are benign vascular anomalies that contain a characteristic honeycomb network of vascular spaces lined with endothelial cells and sparse amounts of loose connective tissue. It is important to note that extracerebral CHs such as those within the CS, optic nerve, optic disc, or retina should be differentiated from intracerebral CHs because of existing differences in clinical behavior, imaging, and response to treatment.

Various studies have noted a concomitance of pregnancy and the onset of symptoms associated with CH, but the number of cases reported during pregnancy remains small. Our patient first experienced symptoms in the 2nd trimester of her pregnancy. While the link between pregnancy and the formation of lesions remains unclear, several hypotheses concerning changes in angiogenic signals and hemodynamic parameters have been proposed.

Pregnant women are known to express increased levels of both vascular endothelial growth factor (VEGF) and progesterone. Cavernous hemangiomas express proangiogenic Flk-1 VEGF receptors and progesterone receptors on both endothelial and smooth muscle cells. Upregulation of these two endocrine signals may be associated with the growth of these lesions. Pregnancy also brings about a number of hemodynamic changes including the expansion of plasma volume and estrogen-induced increase in vascular compliance either through direct interaction with vascular smooth muscle or induction of nitric oxide release by the endothelium. Ohata et al. described cavernous sinus hemangiomas as a “cluster of small balloons” that can change in size according to blood pressure. According to this model, the vasodilation, increased heart rate, and hypervolemia that are part of the physiological state of pregnancy may increase the size of a CSCH as seen in this case.

Surgical treatment of CH has been associated with low risk and effective symptom alleviation. However, lesions that are close to the arteries and nerves of the orbit, as in this case, are difficult to resect safely and can make complete removal complicated. Surgical alternatives such as stereotactic radiosurgery have been used successfully to treat CHs. In our pregnant patient, both surgical and radiosurgical treatment were considered last-line treatment.

The biggest threat to the patient’s safety was hemorrhage from the CSCH. The morbidity from hemorrhage during pregnancy and/or labor is notable in intracerebral CHs as well as those within the optic disc and retina, which require surgical intervention. The rupture of a CH is less likely than rupture of a carotid-cavernous aneurysm, which has an incidence of 1.5%. Therefore, we felt that the best course of action would be to see whether symptoms improved postpartum, as has been reported elsewhere. If the CSCH ruptured, the hemorrhage would likely be contained within the dural sinus.

The patient’s symptoms did not resolve soon after delivery. The CSCH enlarged during the first 2 weeks following delivery before it started to decrease in size around 4 weeks postpartum. The patient’s net gain of approximately 800 ml of fluid during the cesarean section may account for the modest increase in CSCH size seen immediately postpartum.

In conclusion, nonsurgical treatment is a potential option for CSCH in pregnant women. Postpartum involution may negate the need for surgical intervention.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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