Large Vestibular Schwannomas Presenting during Pregnancy: Management Strategies

Kushal J. Shah¹ Roukoz B. Chamoun¹

¹Department of Neurosurgery, University of Kansas Medical Center, Kansas City, Kansas, United States

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Address for correspondence Roukoz B. Chamoun, MD, Department of Neurosurgery, University of Kansas Medical Center, Stop 3021, 3901 Rainbow Blvd., Kansas City, KS 66160, United States (e-mail: rchamoun@kumc.edu).

Abstract

Objective Large vestibular schwannomas rarely present in pregnant women. Diagnosis and management of these tumors during pregnancy present a therapeutic challenge.

Methods A 20-year-old primigravida woman at 26 weeks' gestation was transferred to our facility with gait imbalance, left facial weakness, left ear hearing loss, and recent nausea and vomiting. Magnetic resonance imaging revealed a large left cerebellopontine angle mass with extension into the left internal auditory canal and compression of the fourth ventricle resulting in mild hydrocephalus. The patient was admitted with a plan for early delivery at 32 weeks followed by tumor resection. One week later, the patient's headache and neurologic symptoms worsened due to increased hydrocephalus; a ventriculoperitoneal shunt was placed. The next day, an emergent cesarean delivery was performed due to worsening respiratory status. Four days later, a tracheostomy and percutaneous endoscopic gastrostomy tube were placed due to dysphagia. Eight days after the delivery, the mass was resected with a left retrosigmoid approach without complications. Immunohistochemistry confirmed vestibular cellular schwannoma on cranial nerve VIII showing unusually high mitotic activity.

Results The patient was discharged to inpatient rehabilitation on postoperative day 12 without new neurologic deficit. At 1 month, the patient was swallowing without aspiration. Her facial sensation had returned, her facial weakness remained stable, and her gait was significantly improved.

Conclusion If the patient is neurologically stable, the best option is to delay resection until after delivery. If resection is necessary during pregnancy, the optimal time is during the second trimester.

Keywords

- ▶ neuroma
- ▶ acoustic
- ▶ pregnancy
- ▶ schwannoma
- ▶ vestibular
- ▶ surgery

Introduction

Intracranial neoplasms rarely present in pregnant women, and diagnosis and management of these tumors during this time poses a significant challenge.^{1,2} We describe a patient who presented with a large vestibular schwannoma during her second trimester. We review the literature on similar reported cases, summarize the management strategies according to the stage of pregnancy, and present a treatment

algorithm that considers the patient's stage of pregnancy and neurologic status.

Case Report

A 20-year-old primigravida woman at 26 weeks' gestation was transferred to our facility with several weeks' duration of gait imbalance, left facial droop, left facial numbness, hearing loss in the left ear, some slurred speech, and recent worsening

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Fig. 1 Axial T1 noncontrast magnetic resonance imaging showing left vestibular schwannoma measuring 4.7×3.4 cm with compression on fourth ventricle.

of nausea and vomiting. Past medical history was significant for mild developmental delay since early childhood. Neurologic examination showed left facial weakness (House-Brackmann grade III), decreased facial sensation on the left side, and an ataxic gait. The condition of the fetus appeared stable on ultrasound. Magnetic resonance imaging (MRI) of the head revealed a large left cerebellopontine angle (CPA) mass, measuring 4.7×3.4 cm, with extension into the left internal auditory canal and compression of the fourth ventricle resulting in mild hydrocephalus (**Fig. 1**). The patient was admitted to the neurologic intensive care unit and started on intravenous steroids, with a plan for early delivery at 32 weeks followed by tumor resection.

One week after admission, the patient's headache and neurologic symptoms worsened. Computed tomography of the head showed a significant increase in the size of the ventricles as well as mild transependymal edema (**Fig. 2**); thus a right frontal ventriculoperitoneal (VP) shunt was placed. The next day, due to worsening respiratory status and signs of lower cranial nerve dysfunction, the patient underwent emergent cesarean delivery, with delivery of a viable female infant. Four days later, the patient underwent a tracheostomy and percutaneous endoscopic gastrostomy (PEG) tube placement for airway protection and feeding route due to dysphagia.

Eight days after delivery, the patient underwent resection of the CPA mass with a left retrosigmoid approach (**Fig. 3**). There were no complications. Immunohistochemistry confirmed vestibular cellular schwannoma, although the mitotic activity in the tumor exceeded the usual range of four or fewer mitoses per 10 high-power fields typically noted in cellular schwannoma. In addition, although cellular schwannoma has been reported in the posterior fossa, cranial nerve VIII is a somewhat infrequent location for this histologic variant.

Fig. 2 Axial noncontrast computed tomography scan showing hydrocephalus with evidence of transependymal flow.

The patient recovered well and was discharged to inpatient rehabilitation on postoperative day 12. There was no new neurologic deficit. The infant did well without a ventilator or artificial nutrition, and she was discharged to home in stable condition. At 1-month follow-up, the patient's tracheostomy and PEG tube were removed and she was safely swallowing without evidence of aspiration. Her facial sensation had returned, her gait was significantly improved, and her facial weakness remained stable at House-Brackmann grade III.

Discussion

Vestibular schwannomas (also called acoustic neuromas, acoustic neurinomas, or acoustic neurilemmomas) are rare,

Fig. 3 Axial T1 with contrast magnetic resonance imaging showing resection of vestibular schwannoma with no evidence of residual tumor.

generally slow-growing tumors of Schwann cell origin, usually arising from the vestibular portion of the eighth cranial nerve. These tumors are essentially benign and found more frequently in women.³ In addition, intensely vascular vestibular schwannomas are more common in women, which correlates with the larger size being more common in women as well.³

Vestibular schwannomas can appear for the first time during pregnancy, or symptoms may worsen during the last trimester.^{4,5} In 1917, Harvey Cushing was the first to report that the growth rate of acoustic neuromas accelerates during pregnancy.⁶ Allen et al described seven patients from 1912 to 1972 in whom acoustic neuroma was undiagnosed until after delivery. The authors attributed the acceleration of tumor signs and symptoms in late pregnancy to hormonal stimulation of tumor growth or to hormonally induced vascular dilation within the tumor.⁷ In contrast, Beatty et al reported that pregnancy does not significantly accelerate the cellular growth of acoustic schwannomas.⁸ The hormonal influence of estrogen and progesterone on vestibular schwannomas has been studied by multiple groups since 1981, and it remains unclear what role these hormones play in the accelerated tumor growth rate.⁵ Although results are conflicting regarding the frequency of estrogen receptor expression in these tumors, there is consensus that a small subset of vestibular schwannomas do exhibit estrogen binding.⁹

The mechanisms of accelerated tumor growth during pregnancy remain unclear; however, two leading theories exist. First, increased blood volume during pregnancy leads to engorgement of the vascular bed, which in turn leads to increased tumor size. Second, direct hormonal effects from progesterone and estrogen receptors mediate tumor growth.⁵ In their study of 16 vestibular schwannoma specimens, Brown et al did not find any obvious correlations between estrogen receptor expression levels and patient age, gender, or tumor size; however, the authors concluded that antiestrogen therapy might prove an effective modality because decreased proliferation and increased apoptosis have been reported with its use.¹⁰ Studies are ongoing regarding the use of tamoxifen and raloxifene to impede the growth of vestibular schwannomas.¹⁰

Only 31 cases of vestibular schwannomas diagnosed or becoming symptomatic during pregnancy have been reported in the literature (► **Table 1**) including our case. Among these cases, the most frequent presenting symptom was hearing loss. Based on the information available, all cases except two were diagnosed in the second or third trimester of pregnancy. Only six cases required surgical resection of the vestibular schwannoma prior to delivery. All the infants were delivered at 36 weeks to term, except for two cases, one at 27 weeks (this case) and one at 33 weeks.

General Principles of Management

Management of a vestibular schwannoma diagnosed or exacerbated during pregnancy may have conflicting risks and benefits for the mother and the fetus. The optimal treatment strategy for both mother and fetus is close observation, followed by tumor resection after delivery.^{1,4,5,10,11} The second-best strategy is delivery of the infant (if late enough

in the third trimester), immediately followed by tumor resection.⁵ Cesarean delivery is the preferred method due to the risk of increased intracranial pressure during phase 2 of vaginal delivery.^{5,12} Some authors have reported cases of spontaneous vaginal delivery prior to tumor resection, but the risks remain uncertain.⁴

Vestibular Schwannomas in the First Trimester

Surgery during the first trimester imposes the highest risk to the fetus due to the effects of general anesthesia.⁹ Therefore, if surgery can be delayed until after the first trimester, the risk of spontaneous abortion is greatly diminished.^{9,12} If obstructive hydrocephalus is present due to mass effect, then a VP shunt could be placed to delay primary surgery until later in the pregnancy. Anesthesia and operative time for VP shunt placement is much shorter than that required for tumor resection, and a VP shunt assures continuous cerebrospinal fluid (CSF) drainage.¹² Despite all best efforts, surgical resection during pregnancy may sometimes still be required.¹³

Vestibular Schwannomas in the Second Trimester

If tumor resection is necessary during pregnancy, the second trimester is the most favorable time for both mother and fetus^{5,12} because surgery with general anesthesia during this stage presents the lowest surgical and anesthetic risk to the mother and fetus.⁹ If hydrocephalus is present, a VP shunt could be placed to delay tumor resection until after delivery. However, if it is early in the second trimester and there is concern that the patient may require tumor resection during the pregnancy, strong consideration should be given to primary resection during this time.

Vestibular Schwannomas in the Third Trimester

Surgery during the final trimester of pregnancy poses the highest risk to the mother. This risk is likely due to hemodilution, decreased functional residual respiratory capacity, increased oxygen consumption predisposing to hypoxemia, and capillary venous engorgement of the airway.⁹ If obstructive hydrocephalus is present, a VP shunt could be placed to delay tumor resection until after delivery.² If late enough in the pregnancy, delivery of the infant followed by surgical resection of the tumor is an option that poses less risk to both mother and fetus.⁵ When there is a large symptomatic tumor along with raised intracranial pressure and hydrocephalus, proper management is critical.¹² Thacker et al reported successful resection of a giant acoustic neuroma during the third trimester without detriment to either mother or fetus.¹⁴ The optimal strategy is drainage of CSF prior to emergency cesarean delivery, immediately followed by tumor resection.^{2,12} Resection may be delayed several days to a week to enable restoration of hemodynamic stability.¹² However, Kacchara et al describe a case in which cesarean delivery was followed immediately by surgical resection, and both mother and infant recovered uneventfully.⁵

► **Fig. 4** represents a general treatment algorithm for vestibular schwannomas diagnosed or becoming symptomatic during pregnancy.

Table 1 Patients with vestibular schwannomas diagnosed or becoming symptomatic during pregnancy^a

Case	No. of weeks gestation at diagnosis	Study	Age, G/P	Right/Left	Size, cm	Symptoms	Tumor resection during pregnancy	Tumor resection after delivery	Weeks at delivery
1	8–9	Doyle and Luxford ⁹	28, G1/P0	Left	2.0	Left ear hearing loss	X		Term
2	11	Doyle and Luxford ⁹	29, G1/P0	Right	4.5	Right ear hearing loss, right facial numbness	X		Term
3	20	Gaughan and Harner ⁴	30, G2/P1	Right	4 × 4	Intractable vomiting	X		Term
4	20–22	Kachhara et al ⁵	27, G2/P1	Right	4.5 × 4	Headache, vomiting, hearing loss, diplopia, gait ataxia, right facial paresis and numbness	X		Term
5	24	Allen et al ⁷	16, G1/P0	Left		Hearing loss, vertigo, facial weakness, eye burning		X	Term
6	24	Magliulo et al ¹	24, G1/P0	Right	4.5 × 4	Slow progressive hearing loss, mild headache		X	Term
31	26	Shah and Chamoun (this report)	20, G1/P0	Left	4.7 × 3.4	Gait ataxia, hearing loss, lip numbness, nausea vomiting, facial droop, dysphagia		X	27
7	28	Allen et al ⁷	21, G1/P0	Right		Hearing loss, dizziness		X	Term
8	28	Allen et al ⁷	G2/P0			Hearing loss, tinnitus, headache		X	Term
9	28	Beatty et al ⁸	33		4.0		X		Term
10	29–40	Allen et al ⁷	24, G3	Right		Hearing loss, facial weakness and diminished facial sensation		X	Term
11	30	Hsiao et al ¹⁵	30, G2/P1	Left	4.5 × 4	Hearing loss, tinnitus, diplopia, slurred speech, unbalanced gait, headache, nausea, vomiting		X	36
12	30	Kurowska-Mroczek et al ¹³	33	Right		Progressing problems with visual acuity		X	33
13	31	Thacker et al ¹⁴	27, G1/P0	Left	Large	Vomiting, unsteady gait, poor balance, visual disturbance, hearing loss	X		38
14	32	Allen et al ⁷	31, G4			Sudden unilateral hearing loss		X	Term
15	34	Cushing ⁶	34	Left		Ataxia, speech changes, drooling, tongue and face numbness, deafness			36–40
16	34	Bédard et al ¹⁶	30, G2/P1	Right	3.8 × 3.8 × 3.8	Right facial numbness, decreasing right auditory function		X	36
17	35	Beni-Adani et al ¹²	24, G4	Right	6	Hearing loss, mild ataxia, hemiparesis, dysphagia, slurred speech, headache		X	37
18	36	Kachhara et al ⁵	30, G2/P1	Left	4	Papilledema, headache, vomiting, imbalance		X	36

(Continued)

Table 1 (Continued)

Case	No. of weeks gestation at diagnosis	Study	Age, G/P	Right/Left	Size, cm	Symptoms	Tumor resection during pregnancy	Tumor resection after delivery	Weeks at delivery
19	38	Sharma et al ²	23, G1/P0	Right	3.9 × 2.4	Headache, eye pain, facial weakness, hearing loss		X	38
20		Allen et al ⁷	24	Bilateral		Bilateral hearing loss		X	Term
21		Beatty et al ⁸	29		4.5			X	Term
22		Beatty et al ⁸	34		2.5			X	Term
23		Beatty et al ⁸	33		3.5			X	Term
24		Beatty et al ⁸	28		3			X	Term
25		Beatty et al ⁸	23		3			X	Term
26		Gaughan and Harner ⁴			5.5	Dysequilibrium, tinnitus, headache, nausea, vomiting, diplopia, dysgeusia		X	Term
27		Gaughan and Harner ⁴			4.5	Dysequilibrium, hearing loss, tinnitus, headache, facial paresthesia, nausea, vomiting, diplopia		X	Term
28		Gaughan and Harner ⁴			4	Dysequilibrium, hearing loss, tinnitus, headache, facial paresthesia, nausea, vomiting, otalgia		X	Term
29		Gaughan and Harner ⁴			5	Hearing loss, headache, diplopia		X	Term
30		Gaughan and Harner ⁴			3	Dysequilibrium, hearing loss		X	Term

Abbreviations: G, gravida; P, parity.

^aIn order of number of weeks' gestation at diagnosis.

Note: Blanks in the table signify that the information was not available.

Fig. 4 Treatment algorithm for vestibular schwannoma diagnosed or becoming symptomatic during pregnancy. VP, ventriculoperitoneal.

Conclusion

Vestibular schwannomas in pregnant patients pose a challenging treatment dilemma. Although these tumors are uncommon, a sound treatment strategy involving a multidisciplinary team of obstetrics, anesthesia, and neurosurgery allows for the best outcome for both mother and fetus.^{2,12} Treatment options depend on the patient's stage of pregnancy and neurologic status.² Every patient should be evaluated on a case-by-case basis to determine the best treatment strategy and minimize the risks to both mother and fetus. If the patient is neurologically stable, the best treatment option is to delay resection until after delivery. If resection is necessary during pregnancy, the second trimester is the optimal time for surgery.

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