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Management of Giant Vestibular Schwannoma in Second Trimester Pregnancy – Review of Literature

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Abstract

Vestibular schwannoma is considered as the one of the most common primary brain tumor in middle age group especially in females, however incidence is extremely uncommon. Although surgical management of giant vestibular schwannoma (GVS) in non-pregnant state is associated with high morbidity and mortality. However, in the recent years more and more surgeons prefer surgical management with relatively better outcome. Pregnancy is associated with hormonal changes, systemic changes and increased circulatory volume and associated hypertension, latent diabetes mellitus, pre-eclampsia further complicate the surgical management in the pregnancy. As anaesthetic medication and surgical procedure should not put stress on either mother as well as developing foetus and ideally pregnancy should continue till expected date of delivery and normal delivery of foetus at term should be aim. There is no definite line of management due to paucity of literature as most of authors suggested initial CSF diversion surgery in view of considering added risk as surgical management of GVS itself is associated with very high morbidity and complications rate, and pregnancy further aggravates the risk concerning not only to the pregnant women but also live foetus, and considering these, primary surgical management were usually deferred till few weeks following delivery of the baby or rarely Caeserian section for foetus delivery consecutively followed by resection of GVS was reported. Authors report an extremely rare case of giant vestibular schwannoma in pregnancy, who underwent retromastoid craniectomy in second trimester, she tolerated surgery very well and pregnancy continued with single live foetus, baby was delivered normally at term. Foetus had no congenital malformation. To the best of knowledge of authors, current case is first case in literature, which underwent successful giant Vestibular schwannoma excision with continuation of pregnancy with healthy live foetus. Authors briefly discuss natural history, operative procedure, timing of surgery, management of vestibular schwannoma in pregnancy with aim to monitor foetal well being and outcome and pertinent review of literature.

Keywords

Giant Vestibular Schwannoma, Pregnancy, Second Trimester, Sensorineural Hearing Loss

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1. Introduction

Even though vestibular schwannomas rarely present during pregnancy, symptoms may appear or worsen particularly in this period. The clinical picture may include tinnitus, hearing abnormalities, and in large tumors, brain-stem and cerebellar compression with involvement of additional lower cranial nerves. Large vestibular schwannomas present a great challenge in peripartum management of both the mother and the foetus. [1-5] Classically having a benign clinical course and characteristic radiological appearance, intraoperatively acoustic schwannoma usually contain defined plane of separation with normal brain tissue. [6-9] Author present a female carrying 20 weeks pregnant middle aged lady with giant vestibular schwannoma located in the left cerebello-pontine angle.

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2. Case Illustration

This 37-years-old female with 4 months of amenorrhea presented in outpatient services with left sided hearing impairment of insidious onset, gradually progressive, painless without any earache or ear discharge with tinnitus for four months. A mass in the cerebello-pontine angle identified with four months of pregnancy, referred to our institute. There was no associated headache, vomiting, seizures, hypertension or diabetes mellitus. On examination she was carrying a- 18 weeks of pregnancy with single foetus. On ultrasonography abdomen showed foetus movement with adequate liquor with grade -1 placenta with normal foetal parameters. On admission, she had stable vitals, neurological examination showed left sided sensorineural hearing loss**,** however, the trigeminal nerve, facial nerve and lower cranial nerve were intact, with positive cerebellar signs, and rest of examination were essentially within normal limit. Magnetic resonance imaging revealed (Figure 1, 2, 3) large heterogeneous extra axial, well circumscribed, lobulated, encapsulated ,mass of size 4x3.9x3.6 cm showing iso to hypo-intense signal on T1 image with areas of necrosis & haemorrhage, and showing hyper intense signal on T2 image and heterogeneous enhancing on gadolinium administration. (Figure 4) The lesion was causing expansion left cerebello-pontine angle cistern with clear rim of CSF at periphery with ice- cream cone appearance of extension of tumour entering internal acoustic meatus without any intra canalicular extension. It was not associated with obstructive hydrocephalus with significant mass effect compressing left brain stem, cerebellum, stretching of the 7th nerve - 8 th nerve complex. There was no evidence of any dural attachments differentiating from meningioma.

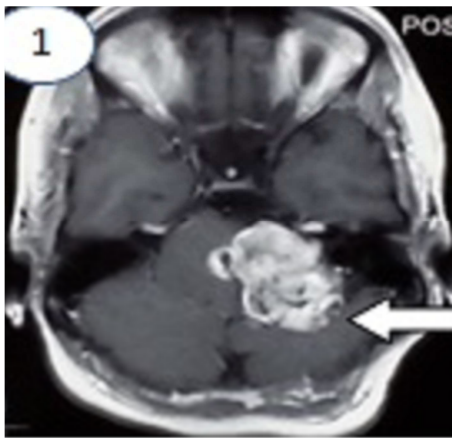

Figure. 1. Contrast enhanced MRI brain, axial section showing large heterogeneous enhancing mass lesion in left cerebello pontine angle causing displacement and distortion of cerebellum and brainstem.

Pure tone audiometry was also done suggestive of left sided sensorineural moderate hearing loss. Brain stem evoked potential – inter peak latencies increased on left side suggestive of left impaired conduction in auditory pathways. A diagnosis of left sided giant vestibular schwannoma with second trimester pregnancy with vial foetus was made and planned for surgical intervention under general anaesthesia.

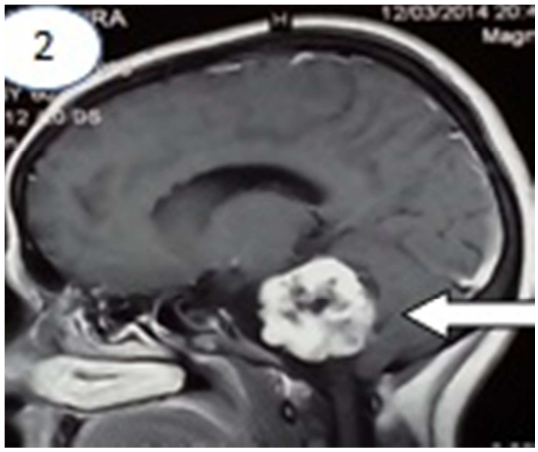

Figure. 2. Contrast enhanced MRI brain, sagittal section showing large heterogeneous enhancing mass lesion in left cerebello pontine angle causing displacement of cerebellum and obliteration of fourth ventricle.

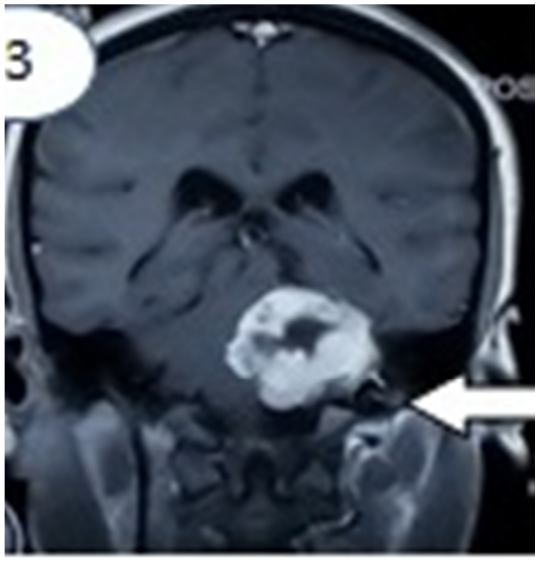

Figure. 3. Contrast enhanced MRI brain, coronal section showing large heterogeneous enhancing mass lesion in left cerebello pontine angle and extending into the left internal acoustic meatus.

She underwent left retromastoid suboccipital craniectomy in the lateral position with special precaution to avoid pressure on abdomen with foetal heart monitoring, with trans-oesophageal echocardiography. Gross total resection of tumour was done. Lazy retromastoid 'S" shaped incision was given. Galeal, muscle layers separated, a 4x6 cm size suboccipital craniectomy with opening of foramen magnum rim was made, leading to visualisation of transverse and sigmoid sinus at its boundary of bony craniectomy. After dural opening, cisternal magna was opened to assist further

draining of CSF. Tumour was visualized after retracting cerebellum, arachnoid was dissected and internal decompression started. Lesion was soft, variegated consistency, CUSA suckable, moderately vascular, lobulated mass with poorly defined plane was present. Facial nerve was identified and anatomically preserved. A gross total resection of was done. Operative duration was five hours only with about 800 ml blood loss. Histopathology of specimen was consistent with schwannoma.

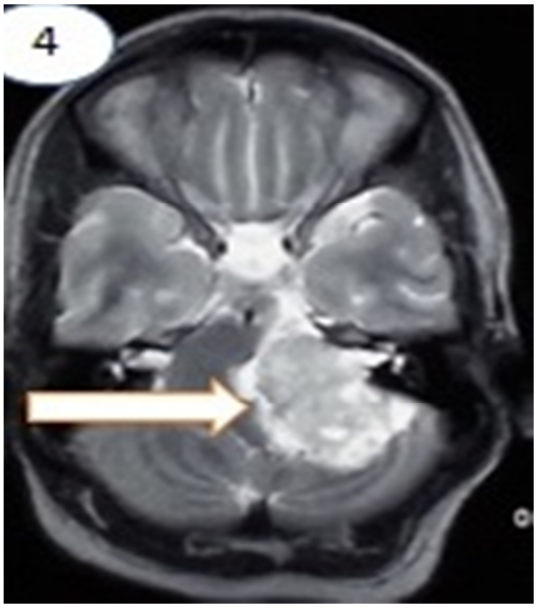

Figure. 4. MRI brain, axial section T2W image showing enlargement of left cerebello pontine angle cistern with well circumscribed extraxial mass lesion.

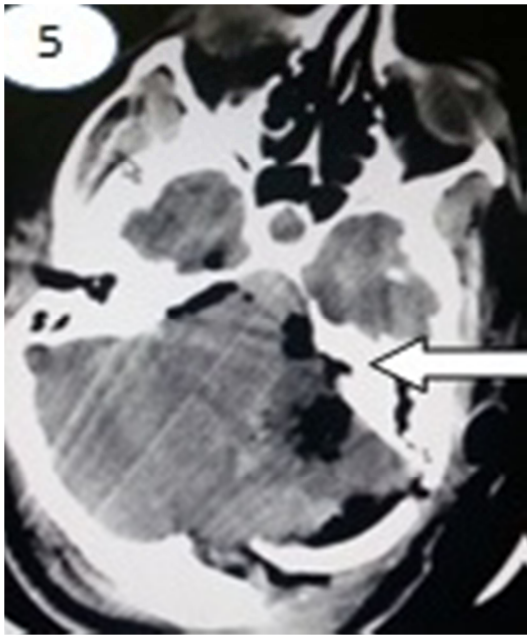

Figure. 5. Postoperative non-contrast computed tomography scan of brain showing complete removal of vestibular schwannoma with opening of fourth ventricle in a 37-year-old-female.

Post operatively, she was extubated and kept in the neurosurgical intensive care unit for two days for monitoring of mother as well as foetus. She developed left sided grade 3 facial paresis but gag and cough reflex was intact. Post operative non-contrast computed tomography with lead apron on abdomen for foetal shielding suggestive of no residual tumour, clear operative cavity and reduction in size of ventriculomegaly (Figure 5) without any operative site haematoma. She was discharged with a GCS of 15/15 score, accepting oral feeds well with persistent 7 th nerve paresis'. Post-operatively, ultrasound abdomen for foetal well being was done suggestive of good foetal movement with adequate liquor and regular foetal cardiac activity.

3. Discussion

Vestibular schwannoma is a benign intracranial tumour of the vestibular nerve myelin sheath. Even though, it presents infrequently in pregnant women. Patient may become symptomatic during the pregnancy, as pregnancy exacerbates the clinical course of intracranial tumours. It has been shown to expand rapidly during pregnancy, possibly due to hormonal changes. [1-5] The other hypotheses are that there is a direct hormonal effect on tumor growth rate, mediated by hormonal receptors. [1-5]

The increase in size and vascularity of the lesion can lead to acute bleeding, which causes exacerbation of symptoms. [3] Non-specific symptoms of headache, nausea and tinnitus, commonly attributed to pregnancy itself, may delay the diagnosis of acoustic neuroma in pregnant women. [1, 4]

Diagnosis is based on clinical history, examination and MR imaging of brain, shows lesion typically located in the cerebello-pontine angle with pressure effect on the brainstem and displacing or occluding fourth ventricle causing obstructive hydrocephalus. However, computerized tomography san of brain avoided as associated radiation hazard to foetus. For foetal wellbeing ultrasound can detect details of cardiac status of foetus, attachment of placenta, and adequacy of amnion and position and lie of foetus.

Management of giant vestibular schwannoma itself posses a great neurosurgical challenge due to size of lesion, relatively smaller capacity of posterior fossa comparing to supratentorial compartment and poor surgical plane with brain stem, availability of small surgical space for manipulation, bleeding and these factor intraoperative factors, collectively may contribute to neurological worsening and in the rare cases leads to morbidity and occasional mortality. [8] Optimal management must balance long-term functional outcome with tumor control. Most surgeons advocate surgical resection as the first-line

treatment modality for vestibular schwannoma in non-pregnant female. Improved knowledge of the patho-physiology and natural history of vestibular schwannoma led to possible complete surgical resection. Ramina et al. reported complete resection was possible not only in all primary but also in recurrent and large residual cases in single-stage surgery with a relatively lower morbidity rate, but preservation of the facial nerve is difficult due to adherent scar tissue. [9] Total surgical removal is the only treatment option for these giant lesions. Samii et al. evaluated the outcome of radical surgery in giant vestibular schwannoma and concluded total tumor removal can be achieved using a retrosigmoid approach with zero mortality and further hearing preservation is also possible. [10] Satyarthee et al. favoured single stage surgical excision of GVS instead of staged surgery for giant vestibular schwannomas. [8]

Increased risk to mother is attributed to, increase in the maternal plasma volume from the six week of gestation to the peak by the 32-34 weeks of pregnancy. [8, 11] Several hemodynamic and metabolic changes associated with pregnancy may be responsible for enlargement and increased vascularity, 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 and associated increase intracranial pressure during pregnancy. [8]

In the pregnancy, surgical management of giant vestibular schwannoma is associated with excess risk in addition to routine surgical risk, increased maternal risk and foetal risk leads to higher incidence of morbidity and mortality and premature delivery. Wang and Young's [5] advocated multidisciplinary approach is required involving obstetricians, neurosurgeons and anaesthetists.

Management is dependent on the gestational stage of the pregnancy and the neurological status, with severe symptoms necessitating urgent surgical intervention. Surgery performed during the first trimester poses the greatest anaesthetic risk to the foetus, in terms of spontaneous abortion and fetal teratogenicity [1, 2, 5].

The anaesthetic risk to the mother increases during the later stages of pregnancy, due to physiological changes such as reduction in functional residual respiratory capacity and expansion of blood volume causing alteration of anaesthetic drug distribution. [2] Provided there are no impending neurological complications, surgical intervention can be safely delayed until the postpartum period. [5]

If the lesion size is giant and symptoms are severe due to obstructive hydrocephalus, early ventriculoperitoneal shunting alone could be performed, followed by tumour resection after delivery. [6] If the patient is in an advanced stage of pregnancy, the best foetal and maternal outcomes are provided by emergency caesarean section after drainage of cerebrospinal fluid, followed by definitive neurosurgery. [1] In the current study, our case had giant vestibular schwannoma with hearing loss in pregnancy, operated in second trimester with moderate blood loss, having more vascularity of tumour observed intraoperatively as compared to non-pregnant females and males in general population with equivalent plane of dissection and tumour consistency with good post- operative recovery without foetal compromise. However during follow-up, she delivered normal healthy baby followed by no further intracranial recurrent lesion with good prognosis but grade 3 facial nerve paresis was persisting.

Regarding surgical management of very small vestibular schwannoma in pregnancy, patient can be advised to carry on through pregnancy and undergo normal delivery and vestibular schwannoma can be safely excised after delivery with an interval of few weeks to months, to allow time for reversal of physiological changes of the pregnancy.

While schwannoma size being less than 3 cm but minimally symptomatic are also advised to continue pregnancy but advised elective caesarean section delivery to avoid rise in the intracranial pressure during second stage of labour. However, symptomatic patient with giant tumours are advised microsurgery as in our case also had coexistent obstructive hydrocephalus preferably in second trimester. Quick resection is planned for maximum safety of both mother and child.

However, radiosurgery is also considered as alternative to surgical management of vestibular schwannoma. Its advantages are outdoor procedure, no anaesthetic risk and few hours are required for completion of procedure. However, major limitation is size of vestibular schwannoma as our case had GVS and suitability criteria for radiosurgery is size less than three cm in diameter. Further associated hydrocephalus and brain stem rotation are other relative contraindications. Another important issue is radiation exposure risk to foetus during radiosurgical procedure and possible teratogenicity needs also to be considered. The global toxicity of a single-shot or hypo-fractioned radiosurgery would have been greater than the operative anesthesia.

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