

Central Nervous System Tumour in Pregnancy, a Diagnostic Challenge or Management Quandary

Abd Malik NR^a, Ismail H^a, Ali Yeon AA^a, Mohd Rashid NN^b, Wahab MG^c, Awang MS^d

^aDepartment of Obstetrics and Gynaecology, Kulliyah of Medicine, International Islamic University Malaysia

^bDepartment of Obstetrics and Gynaecology, Sultan Ahmad Shah Medical Centre, International Islamic University Malaysia

^cDepartment of Neurosurgery, Kulliyah of Medicine, International Islamic University Malaysia

^dDepartment of Neurosurgery, Sultan Ahmad Shah Medical Centre, International Islamic University Malaysia

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Corresponding Author

Asst. Prof. Dr. Nur Rashidah Abd Malik
Department of Obstetrics and Gynaecology,
Kulliyah of Medicine, International
Islamic University Malaysia,
Kuantan, Pahang
E-mail: rashidahmalik@iiu.edu.my

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ABSTRACT

Central nervous system (CNS) tumour, being rare in pregnancy, posed different challenges. We report three challenging cases managed at our centre. The first case was a diagnostic puzzle, bringing a myriad of differentials since the eighth week of gestation. Diagnosis made after magnetic resonance imaging (MRI) post-delivery at 35 weeks of gestation led to cervical spine meningioma excision surgery and subsequent progressive, remarkable neurologic recovery. Two additional cases were not a diagnostic mystery, yet they posed management challenges. The second case was diagnosed with a cerebellar tumour complicated with hydrocephalus at a pre-viable gestation. Pregnancy was able to be prolonged up to viability at extreme prematurity at 26 weeks when she needed delivery with subsequent cerebellar haemangioma excision and recovery. However, the third case became pregnant during follow-up after radiation therapy for brain glioma, with persistent symptoms necessitating termination of pregnancy.

INTRODUCTION

CNS tumour has a low prevalence of one in 1000 to 2000 pregnancies^{1,2}, with a tertiary neurosurgical centre in the United Kingdom reporting only 33 cases in pregnancy over 10 years.³ We report three cases of pregnancy with CNS tumour, one posed a diagnostic challenge, and the other two cases posed a management predicament.

CASE REPORT

CASE 1

A 40 year old, Gravida 4 Para 3, presented to orthopaedics at 8 weeks' gestation with a one-year history of hand and foot numbness. Cervical myelopathy was diagnosed, and physiotherapy partially alleviated her symptoms. At 20 weeks, the numbness worsened, and she had right limb weakness necessitating ambulation with a wheelchair. Neuromedical assessment showed upper motor neuron lesion with bilateral C5 to T1 hypertonia and hyperreflexia, right-sided reduced muscle power, and hemianaesthesia. Multiple sclerosis was suspected,

but unfortunately, MRI was withheld due to an incompatible right knee implant, which she denied for removal.

She was seen by Obstetrics and Gynaecology at 28 weeks' gestation and followed up fortnightly with neuromedical consults on each visit. There was no progressive neurological impairment, and the foetus was growing well. She was on thromboprophylaxis due to her limited mobility. Further deliberation with orthopaedic and radiological teams at 33 weeks of gestation concluded that, due to long-term placement of the implant, MRI is safe without migration risk but with heating effect. She was planned for an MRI at 36 weeks under the same general anaesthesia (GA) for caesarean section (CS).

Unfortunately, she had progressive weakness involving the left limbs at 35 weeks' gestation, necessitating earlier delivery. The multidisciplinary team decided to combine the delivery with intubation and MRI in the same setting,

prioritising maternal safety and avoiding further delay in diagnosis or intervention. Post-caesarean MRI revealed a 1.5cm X 1.7cm X 2.6cm intradural, extramedullary right dorsolateral cervical spine mass from the cervicomedullary junction to the C3 level (figure 1). This caused compression and left posterolateral displacement of the C1 and C2 spinal cord, with C2 and C3 right neural foramina exiting nerve roots obliteration.

Seven days later, she underwent suboccipital craniotomy with C1/C2 and partial C3 laminectomy and tumour excision (figure 1), after which she had progressive neurological improvement with rehabilitation physiotherapy.

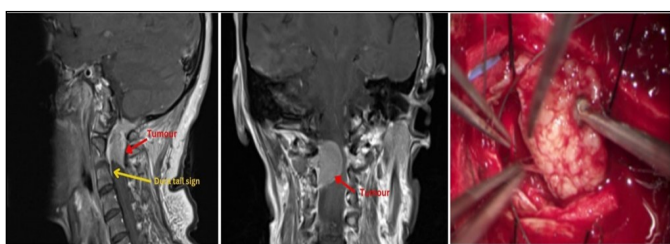


Figure 1: (from left to right): Intradural Extramedullary cervical spine tumour with dura tail sign in sagittal and coronal section MRI and midline durotomy at C1-C3 level with near total tumour removal

Two months later, she needed minimal help with daily activities and ambulating without wheelchair, and was completely independent in daily living activities one year after. Her baby, born at 2.05kg, was also thriving well. Histopathologic examination (HPE) of the tumour showed meningiothelial meningioma (grade 1).

CASE 2

A 36-year-old Gravida 2 Para 1 at 23 weeks of gestation presented with a four-month history of recurrent headache, intermittent vomiting, and new onset of blurred vision. She exhibited positive cerebellar signs with left-sided unbalanced gait. MRI revealed a left cerebellar tumour with hydrocephalus.

An Ommaya catheter insertion enabling daily tapping of cerebrospinal fluid (CSF) drainage relieved her recurring symptoms for 19 days. However, her symptoms worsened despite regular tapping of CSF and administration of intravenous steroids. Intravenous magnesium sulphate was given for fetal neuroprotection, and Caesarean section was performed under GA at 26 weeks, delivering a 1.17kg

baby with a good APGAR score and placed under neonatal intensive care support for extreme prematurity. A repeated MRI showed a 3.1cm X 2.5cm X 2cm left cerebellar mass with perilesional oedema and mass effect onto the left midbrain, pons, and medulla oblongata. There was fourth ventricle compression with dilated bilateral lateral and third ventricles with crowding of cerebellar tonsils and tonsillar herniation (figure 2).

She had a left cerebellar suboccipital craniotomy and tumour excision (Figure 2) three days post-delivery.

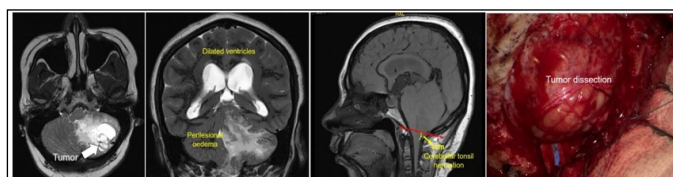


Figure 2: (from left to right): Cerebellar tumour with perilesional oedema in MRI axial section, coronal section, cerebellar tonsil herniation in MRI sagittal section, and left cerebellar suboccipital craniotomy and tumour excision.

She was discharged well three days after surgery. One month later, she remained asymptomatic with no residual tumour on MRI. Her child was discharged at 2.41kg after three months. HPE of the tumour reported as haemangioma.

CASE 3

A 40-year-old lady presented with a generalized tonic-clonic seizure. MRI showed a minimally enhancing tumour in the left frontal region measuring 4cm X 3cm X 2.6cm. She underwent a parietal craniotomy and tumour debulking surgery (Figure 3). A postoperative MRI one month later revealed residual tumour measuring 2.4cm X 2cm X 3cm. HPE reported as diffuse astrocytoma (grade II).

She completed 5 fractions of stereotactic radiotherapy. Her seizure was poorly controlled despite optimum anti-epileptic medication. Three months after the completion of radiotherapy, she was pregnant with twin pregnancy at 9 weeks. An urgent multidisciplinary team discussion was organized. Her conception was calculated to be after completion of her radiotherapy.

In view of poor seizure control on anti-epileptic and risk of contrasted MRI to fetus, which was essential in disease

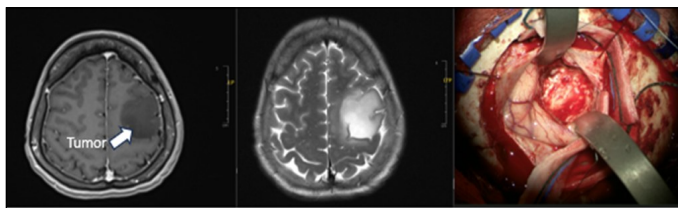


Figure 3: (from left to right): Minimally enhanced left frontal tumour on axial T1-weighted image with gadolinium and hyperintense on T2-weighted image. Intraoperative picture showing tumour debulking.

assessment and management, the managing team has collectively opted for termination of pregnancy (TOP) under general anesthesia. This decision for pregnancy termination was reached following consensus among the treating neurology, neurosurgery, obstetrics, and Shariah teams, in discussion with the patient and her husband.

MRI post-termination of pregnancy showed a residual tumour of 3.9cm X 2.7cm X 2.8cm with post radiation necrosis component and worsening local mass effect (Figure 4). She was commenced on long-acting contraception with progesterone implant and subsequently started on a course of corticosteroid therapy with a single anti-epileptic. With ongoing reassessment and monitoring, she has remained seizure-free for up to eight months following the termination of pregnancy.

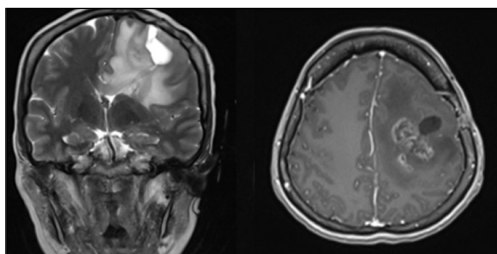


Figure 4: A coronal T2-weighted image showing left frontal cystic encephalomalacia with local mass effect, and residual tumour plus post-radiation necrosis seen on axial T1-weighted image with gadolinium

DISCUSSION

Diagnostic challenges of CNS tumour in pregnancy are contributed by its rarity and mimicry of pregnancy-related symptoms^{1,4,5,6}, such as vomiting, back pain, and numbness of extremities.⁷ Pre-existing tumours may only first manifest symptoms in pregnancy due to rapid growth attributable to pregnancy hemodynamic and hormonal changes^{5,6,7,8} and increased growth and angiogenic factors.^{1,2}

Pregnancy has been associated with tumour-accelerated growth. Meningioma, as illustrated in Case 1, is among the

commonest spinal tumours that is slow growing and may be asymptomatic for years.⁶ Reports have shown meningioma expressing progesterone and oestrogen receptors^{3,4,6}, therefore causing the accelerated growth in pregnancy. Haemangioma has also been shown to only become symptomatic in pregnancy with rapid worsening symptoms in the third trimester, due to oestrogen and progesterone angiogenic growth-promoting effect⁹ alongside increased blood volume¹ and elevated venous pressure, especially in the third trimester^{5,6} as evident in Case 2.

Henceforth, delayed diagnosis compounded by pregnancy-associated accelerated growth would jeopardise maternal safety and increase morbidity and mortality.² Therefore, a high index of suspicion followed by proper investigation is imperative in women with neurologic symptoms in pregnancy.

MRI is best in identifying deep soft tissue structure, is safe in the second and third trimesters, with some reservations in the first trimester.¹ Gadolinium contrast is shown to have no additional risk to pregnant women, but inconclusive in foetal safety, with yet undocumented long-term concerns on kidney and neurodevelopment, but teratogenicity in animal studies has been shown only with high repeated dosing.¹ Nonetheless, when benefit outweighs the risks, the judicial use of MRI in pregnancy is justified.

Despite the unfortunate delay due to an MRI-incompatible implant in Case 1, postpartum diagnosis enabled immediate surgical intervention for the spinal tumour.

In contrast, diagnosis was not a challenge in Cases 2 and 3, but the management quandary in diagnosed CNS tumour in ongoing pregnancy is to have timely intervention for optimal maternal and foetal outcome.^{4,9} Conservative approaches would reduce premature foetal morbidity and mortality, but in extensive and rapid neurological involvement, immediate intervention is needed.^{3,5}

Antepartum surgery has multiple challenges with difficult prone positioning in the third trimester,⁸ increased risk of epidural venous bleeding⁸ and risk of foetal compromise² with difficult monitoring⁸ during surgery. Hence, when pregnancy reaches 32 weeks, early delivery followed by surgery is recommended^{4,5,7}

In Case 2, the diagnosis was at 23 weeks. The initial management goal was to prolong pregnancy to a viable gestational age while maintaining maternal stability. However, ensuing neurological deterioration despite maximal supportive measures necessitated immediate tumour resection. Despite severe prematurity at 26 weeks, the timely multidisciplinary reassessments and subsequent decision for delivery of the foetus with neonatal support, followed by surgical tumour resection, have brought a favourable outcome for both mother and baby.

Case 3 demonstrates maternal morbidity early on in pregnancy; hence, TOP is appropriate to prevent further maternal deterioration and mortality. Although no reported increase in miscarriages was reported from a recent systematic review¹⁰, the patient has conceived within three months' post-radiotherapy for diffuse astrocytoma, before reassessment of treatment response and operability could be performed.

MRI with gadolinium contrast, essential for differentiating post-radiation necrosis from tumour recurrence, was avoided due to foetal safety concerns. Thus, continuation of pregnancy would have hindered appropriate imaging and treatment planning, delaying critical management. Following multidisciplinary deliberation, termination was recommended to permit full diagnostic evaluation and timely oncologic intervention to optimise maternal outcome.

On the other hand, these three cases underscore the critical importance of contraception during the evaluation and follow-up of reproductive-age women with CNS manifestations.

CONCLUSION

Despite their rarity, CNS tumours may contribute to

neurological symptoms during pregnancy. Suspicion is crucial, and an MRI is recommended for an early diagnosis. The management of CNS tumours is determined by maternal neurological symptoms and foetal maturity. Most importantly, early multidisciplinary commitment and cooperation are critical, often demonstrating improved patient outcomes.

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