

## Symptomatic intracranial tumors in pregnancy: an updated management algorithm. Illustrative case

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**BACKGROUND** Intracranial tumors are infrequently encountered during pregnancy, and their diagnosis and management require a multidisciplinary approach to ensure the best possible outcomes for the mother and fetus. The pathophysiology and manifestations of these tumors are influenced by hormonal changes, hemodynamic modifications, and alterations in immunological tolerance that occur during pregnancy. Despite the complexity of this condition, no standardized guidelines exist. This study aims to highlight the key points of this presentation, along with the discussion of a possible management algorithm.

**OBSERVATIONS** The authors report the case of a 35-year-old woman who presented during the third trimester of pregnancy with severe signs of increased intracranial pressure (ICP) due to a posterior cranial fossa mass. The decision was made to stabilize the patient by placing an external ventricular drain to temporize her increased ICPs until the baby could be safely delivered via cesarean section. The mass was resected via suboccipital craniectomy 1 week postpartum.

**LESSONS** In considering treatment modalities and their timing in patients presenting with intracranial tumors during pregnancy, each patient should be managed on the basis of an individual treatment algorithm. Symptoms, prognosis, and gestational age should be taken into account to optimize the surgical and perioperative outcomes of both the mother and fetus.

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**KEYWORDS** aneurysmal bone cyst; brain tumor in pregnancy; external ventricular drain; EVD; hemodynamic change; intracranial pressure; ICP

Intracranial tumors during pregnancy are an infrequent condition that requires a unique multidisciplinary approach to ensure the best possible outcome for both the mother and the fetus.<sup>1,2</sup> Essentially, pregnancy does not pose an increased risk of developing intracranial tumors.<sup>3</sup> Nevertheless, it influences the pathophysiology and manifestations of these tumors due to hormonal changes, immunological tolerance alteration, and hemodynamic modifications, which occur in a female's body during pregnancy.<sup>3,4</sup> These systemic changes may also cause preexisting silent tumors to clinically manifest for the first time during pregnancy.<sup>5</sup> Despite their complexity, there are no standardized guidelines for the management of this rare clinical circumstance.<sup>1</sup> This study details the case of a pregnant 35-year-old female who presented during the third trimester with severe signs of increased intracranial pressure (ICP) due to a posterior cranial fossa tumor. Her symptoms

were stabilized, and the tumor was resected after delivery. A literature review was also conducted to research and discuss a possible management algorithm.

### Illustrative Case

A 35-year-old pregnant female at 32 weeks of gestation presented to our institution with persistent headache, nausea, vomiting, and progressive right-sided vision loss. Further investigation revealed that the patient had initially started having symptoms 3 weeks earlier and had sought care at an outside hospital emergency room. Brain magnetic resonance imaging (MRI) was ordered at that time, which demonstrated a large right posterior fossa mass with fluid levels and features of intratumoral hemorrhage. She was transferred to our tertiary care center for further workup and monitoring. The maternal and

**ABBREVIATIONS** C-section = cesarean section; CT = computed tomography; CTV = computed tomography venography; EVD = external ventricular drain; ICP = intracranial pressure; MRI = magnetic resonance imaging.

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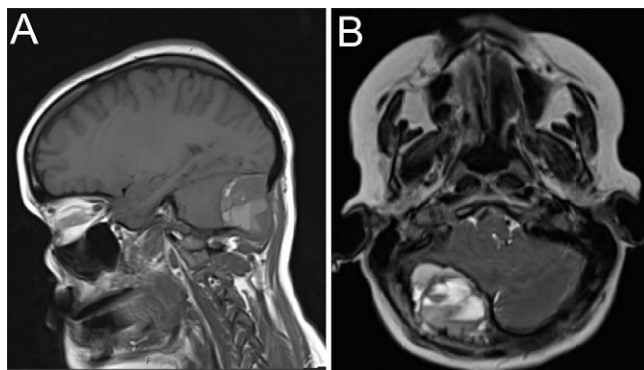
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fetal vital signs were stable throughout this process. Ophthalmology consultation revealed bilateral disc edema with marked visual field abnormalities. Other than the presenting symptoms, the patient was neurologically intact.

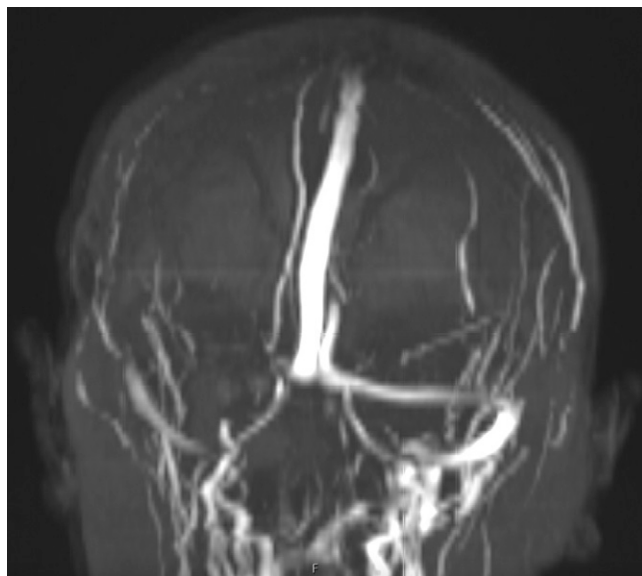
Brain MRI and computed tomography (CT) venography (CTV) were ordered. MRI demonstrated the presence of a 5.6-cm right occipital bone mass with blood-fluid levels extending into the right posterior fossa and no invasion into the underlying brain parenchyma, consistent with an aneurysmal bone cyst. Regional mass effect on the right cerebellar hemisphere, effacement of the fourth ventricle, and tonsillar herniation were noted (Fig. 1). Mass effect on the right transverse sinus was observed but incompletely characterized. CTV confirmed the MRI findings and demonstrated a truncated right transverse sinus (Fig. 2). These findings favored classification of the lesion as chronic.

Given the signs and symptoms of increased ICP, likely due to chronic transverse sinus compression, and the complexity of the clinical circumstances, a multidisciplinary meeting was held to establish the most appropriate management. A cesarean section (C-section) delivery was scheduled for the 34th week of gestation with a plan of resecting the likely benign mass after delivery. In the interim, the decision was made to place an external ventricular drain (EVD) to temporize her increased ICPs. After EVD placement, the patient was admitted to the intensive care unit. Her vision improved, and her ICPs permanently remained within normal values.

The baby was delivered successfully via C-section as scheduled, and the patient was deemed stable for resection of the aneurysmal bone cyst 1 week after delivery. A right-sided suboccipital craniectomy was planned. The mass immediately became apparent with a natural plane. Great care was taken to avoid injuring the underlying dura and transverse sinus at the superior extent of the operative field. The mass was compressive with significant mass effect on the underlying posterior fossa dura; however, there was no infiltration. Preliminary pathology results confirmed that the mass was an aneurysmal bone cyst, and gross-total resection was achieved. The final histopathology report demonstrated a multicystic lesion lined by fibroblast-like spindle cells and partly filled with blood. No atypical cells were identified to suggest malignancy. The postoperative CT scan showed no complications, with a stable ventricular caliber, and the EVD was



**FIG. 1.** Sagittal (A) and axial (B) noncontrast MRI showing a 5.6-cm right occipital bony mass with blood-fluid levels and no invasion into the underlying brain parenchyma, consistent with an aneurysmal bone cyst. Imaging also demonstrates regional mass effect on the right cerebellar hemisphere, effacement of the fourth ventricle, and tonsillar herniation.



**FIG. 2.** CTV showing a truncated right transverse sinus secondary to mass effect from the aneurysmal bone cyst.

removed 2 days following surgery after a successful clamp trial with normal ICPs. The patient was discharged to home in good condition.

## Discussion

### Observations

#### Pathophysiology

Multiple theories have been proposed to explain the connection between pregnancy and intracranial tumors. One of the most accepted theories is based on the effects of physiological changes during pregnancy on the tumor.<sup>3,4</sup> These changes are benign, and their purpose is to provide an adequate environment for the growing fetus.<sup>6</sup> Nevertheless, they influence the pathophysiology and the manifestations of intracranial tumors, sometimes resulting in their initial presentation during pregnancy.<sup>3,4</sup>

Hormonal changes during pregnancy are one of the most important physiological modifications. Increased levels of hormones and growth factors have been directly linked to the progression of intracranial tumors due to the presence of tumor cellular receptors such as estrogen and progesterone receptors, epidermal growth factor receptor, fibroblastic growth factor receptor 2, platelet-derived growth factor receptor B, and vascular endothelial growth factor receptor.<sup>7–11</sup>

Regarding immunogenic tumors, maternofetal immunological tolerance during pregnancy dampens normal responses to tumor antigens. T-cell apoptosis, upregulation of regulatory T cells, increased secretion of immunoregulatory cytokines (e.g., interleukin-10 or transforming growth factor- $\beta$ ), and overexpression of suppressive ligands (e.g., PD-L1 or CD95 ligands) are all normal safeguard mechanisms that occur during pregnancy to protect the developing fetus from the maternal immune system.<sup>12,13</sup> However, these mechanisms have all been implicated as immunological factors that influence tumor progression, making pregnancy a favorable environment for tumor growth.<sup>12,13</sup>

Hemodynamic changes occurring during pregnancy may also have a significant impact on intracranial tumors.<sup>14</sup> Increased maternal blood volume leads to an increase in cerebral blood flow, which may cause generalized cerebral edema and/or hemorrhage around the

tumor mass.<sup>3,4</sup> The tumor's proximity to vascular structures is of great importance in determining such an effect. In our case, the tumor almost certainly existed before pregnancy; however, it showed no apparent signs or symptoms under normal hemodynamic conditions. With the progression of pregnancy and the associated increase in blood volume, the tumor compressed her dominant right transverse sinus, significantly reducing venous drainage, which led to her symptomatic increase in ICP.

## Diagnosis

Patients presenting with intracranial tumors during pregnancy require specific management regarding not only treatment but also diagnosis. Symptoms of increased ICP, such as headache, nausea, and vomiting, may be falsely attributed as manifestations of normal pregnancy, hampering a prompt diagnosis.<sup>15</sup> An additional hindrance is the tendency to avoid imaging during pregnancy. However, the American College of Obstetricians and Gynecologists recommends the use of CT and MRI scans, with or without contrast, when their potential benefits outweigh their risks to the fetus and are expected to significantly impact diagnosis and overall outcome.<sup>16</sup>

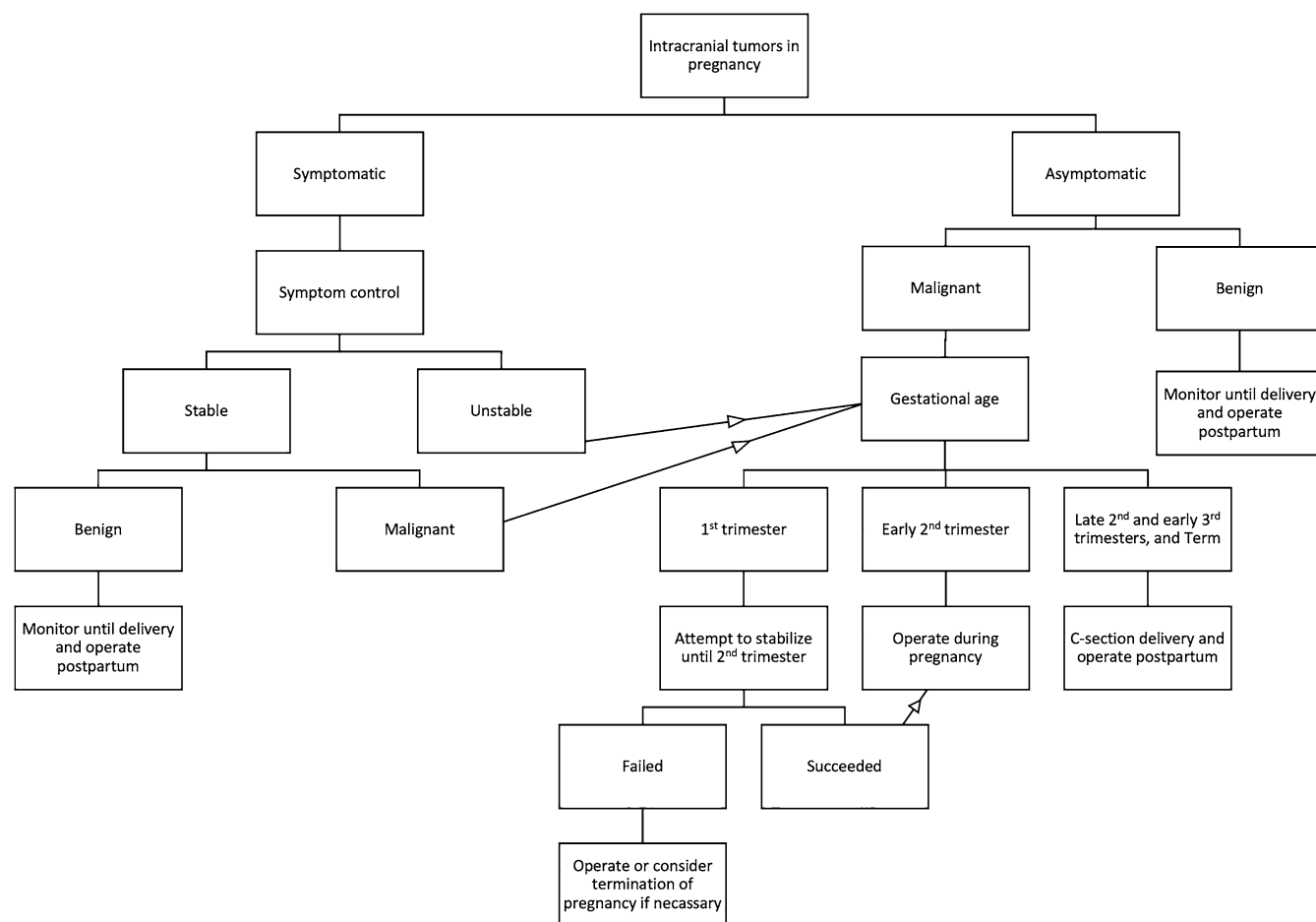
## Treatment

Management of intracranial tumors presenting during pregnancy requires a complex decision-making process in determining whether

to operate as soon as the tumor is discovered or to schedule fetal delivery with delayed surgical management. From a neurosurgical perspective, the decision is impacted by the tumor's pathology, location, growth rate, and size. Malignant and rapidly growing tumors in eloquent areas require aggressive treatment and timely surgical intervention.

Another important factor is the patient's clinical and neurological status. When the patient is stable or there are temporizing interventions (e.g., EVD) or therapeutic modalities (e.g., anticonvulsants) that can safely stabilize the condition, delayed surgical management can be considered with continued close obstetrical monitoring.<sup>17</sup> However, in unstable neurologically deteriorating cases, patients may require urgent (single or staged) surgical intervention or even an immediate C-section delivery, followed by prompt surgical decompression and resection.<sup>1</sup> In these instances, the gestational age of the fetus becomes the primary consideration.<sup>1</sup> Tumors presenting in the first or second trimester of pregnancy are more favorable for surgical intervention than those presenting during the late second and early third trimesters. During the latter, the fetus may be premature and require more complex clinical consideration.<sup>17,18</sup> Therefore, if surgical intervention can be timed, the second trimester of pregnancy is the most favorable.<sup>17</sup>

On the basis of currently available data, there remains little guidance on the management of intracranial tumors during pregnancy. The best recommendation is to consider each case individually with



**FIG. 3.** Management algorithm for stabilization and treatment of intracranial tumors presenting during pregnancy.

its own assessment of the risks and benefits of early versus late surgical intervention. Taking various factors into account, we present the following treatment algorithm as a potential starting point for determining an appropriate management plan in these complex clinical scenarios (Fig. 3).

## Lessons

Although pregnancy has no direct effect on the incidence of intracranial tumors, physiological changes that occur during pregnancy can influence the behavior and clinical manifestations of these tumors. It is also worth noting that the presenting manifestations may be confused with normal symptoms of pregnancy. Finally, when considering treatment modalities and timing, each patient should be managed through a multidisciplinary approach based on a personalized treatment algorithm.

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## Disclosures

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

## Author Contributions

Conception and design: Zohdy, Agam. Acquisition of data: Zohdy, Agam, Jahangiri. Analysis and interpretation of data: Zohdy, Agam, Jahangiri. Drafting the article: Garzon-Muvdi, Zohdy, Agam, Maldonado. Critically revising the article: Garzon-Muvdi, Zohdy, Agam, Maldonado, Pradilla. Reviewed submitted version of manuscript: Garzon-Muvdi, Zohdy, Agam, Maldonado, Pradilla. Approved the final version of the manuscript on behalf of all authors: Garzon-Muvdi. Administrative/technical/material support: Agam. Study supervision: Jahangiri, Pradilla.

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