Acta Neurologica Indonesia 2025; 03(04)

Acta Neurologica Indonesia
Department of Neurology, Faculty of Medicine Universitas Indonesia

ISSN 3032-6303

Case Report

MANAGING ELEVATED INTRACRANIAL PRESSURE AS THE COMPLICATION OF ANGIOMATOUS MENINGIOMA

Adriana Damayanti[1], Tiara Aninditha[1], Henry Riyanto Sofyan[1], Irma Savitri[1], Rahmad Mulyadi[2], Mohamad Saekhu[3]

[1]Department of Neurology, Faculty of Medicine, Universitas Indonesia - Dr. Cipto Mangunkusumo Hospital, Jakarta, Indonesia; adrianamarpaung@gmail.com
[2]Department of Radiology, Faculty of Medicine, Universitas Indonesia - Dr. Cipto Mangunkusumo Hospital, Jakarta, Indonesia
[3]Department of Neurosurgery, Faculty of Medicine, Universitas Indonesia - Dr. Cipto Mangunkusumo Hospital, Jakarta, Indonesia

ABSTRACT

Introduction Meningioma is the most common central nervous system primary tumor, which can grow at the cranial base. Angiomatous meningioma, with a prevalence of 2.1%, is classified as the World Health Organization (WHO) grade I meningioma. Although considered benign, it has high Vascular Endothelial Growth Factor (VEGF) activity.

Case Summary A 41-year-old woman presented with clear nasal discharge upon sitting, progressive headaches, blurred vision, and right-sided hemiparesis following prior surgeries for angiomatous meningioma. Initial symptoms included episodic headaches, later worsening with visual impairment and motor deficits. She underwent decompressive craniectomy, VP shunt placement for hydrocephalus, and cranioplasty but had a functional decline. Examination revealed decreased consciousness, anisocoria, right facial nerve palsy, spastic hemiparesis, and pressure ulcers. Imaging confirmed tumor progression with uncal herniation and hydrocephalus. After infection management, VP shunt revision improved her condition, and tumor resection was planned. The prognosis remained poor regarding survival, function, and recovery.

Discussion Although considered benign, it has high activity of Vascular Endothelial Growth Factor (VEGF). Massive peritumoral edema may lead to complications by increasing intracranial pressure (ICP). The management of elevated ICP includes tumor resection, cerebrospinal fluid (CSF) diversion, and administration of anti-edema agents. Due to its challenging access and proximity to vital structures such as cranial nerves and blood vessels, determining an appropriate management strategy poses a significant challenge to avoid disability.

Conclusion Skull base meningioma in women aged 40–70 poses a high risk of complications and disability if mismanaged, especially angiomatous meningioma with high vascularity despite being WHO Grade I.

Keywords: angiomatous meningioma, elevated intracranial pressure

INTRODUCTION

The skull base separates intracranial and extracranial spaces and houses vital structures. About 25% of meningiomas occur here, primarily in women aged 40–70. Due to high vascularization and VEGF secretion, complete removal is difficult, with

WHO Grade I meningiomas having a 30–50% recurrence rate if only partially resected. Skull base meningiomas, often treated with decompression, biopsy, or subtotal resection (Simpson Grade V), have an 88.9% 10-year recurrence rate. Angiomatous meningiomas are particularly challenging due to high VEGF activity and severe peritumoral edema, requiring careful management of intracranial pressure.[1,2]

CASE REPORT

A 41-year-old woman presented with a three-month history of clear nasal discharge and a two-year history of intermittent left-sided headaches (NRS 4–5) that worsened, causing bilateral blurry vision. A CT scan suggested meningioma, but the patient declined surgery.

Twelve months before admission, she developed right hemiparesis, worsening headaches, and persistent blurry vision. A follow-up brain CT scan showed disease progression, and surgery was deemed necessary.

The patient underwent decompressive craniectomy and tumor biopsy. Histopathological results suggested angiomatous meningioma WHO Grade I. Postoperatively, her headaches and vision improved. Two weeks after surgery, she experienced worsening diffuse headaches and difficulty communicating. A follow-up brain CT scan showed hydrocephalus, leading to the insertion of a ventriculoperitoneal (VP) shunt. After the second surgery, her headaches improved, but other symptoms persisted.

One month post-second surgery, her right-sided weakness worsened, rendering her immobile. She became bedridden. Two weeks later, she underwent cranioplasty.

Three months before admission, there was clear rhinorrhoea while being repositioned into a sitting position.

Additionally, the family reported that she frequently coughed and had recurrent fevers.

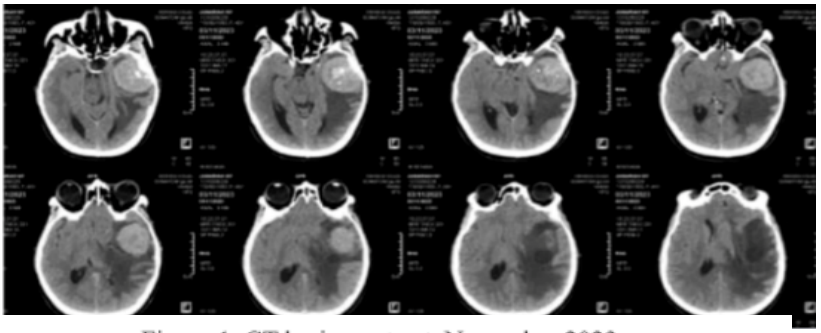


Figure 1. CT brain contrast, November 2023

She was hospitalized due to a suspected lung infection. No seizures or involuntary movements were observed.

On physical examination, her respiratory rate was 22 times per minute, she was febrile, and her oxygen saturation was 97% with 3L/min nasal cannula oxygen. There was a bulging postoperative wound at the left frontotemporal region, with palpable underlying bone. Pulmonary auscultation revealed bilateral rhonchi. A sacrogluteal examination showed multiple pressure ulcers.

Neurological examination findings included a Glasgow Coma Scale (GCS) score of Eye 3 Motor 5 Verbal global aphasia. No meningeal irritation signs were present. Pupils were anisocoric with bilaterally decreased light reflexes. Cranial nerve examination showed right central facial nerve paresis and signs of papilledema on fundoscopy. Increased deep tendon reflexes were noted on the right extremities. Motor examination showed a right-sided weakness with spasticity. The Karnofsky Performance Scale (KPS) was 50. Laboratory finding was hypoalbuminemia (2.7 g/dL). The patient was diagnosed with angiomatous meningioma with suspected Cerebrospinal Fluid (CSF) leakage, pneumonia, pressure ulcer grade 4, and hypoalbuminemia.

Before VP shunt revision surgery, the patient received dexamethasone (5mg twice daily), Which improved GCS improvement (Eye 4 Motor 5 Verbal global aphasia). After resolving the infection, a VP shunt was revised due to shunt malfunction. Postoperatively, the pupils were equal and reactive to light, but motor deficits remained unchanged. Dexamethasone was discontinued after surgery. The patient was discharged and scheduled for gross tumor resection.

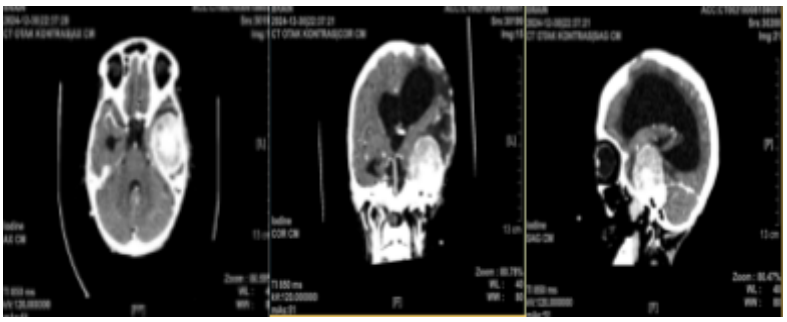


Figure 2. CT brain contrast, December 2024

DISCUSSION

Meningioma

Meningiomas are the most common benign CNS tumors, making up 54.3% of primary benign CNS tumors and 37.6% of all primary CNS tumors. Angiomatous meningiomas are rare, comprising 2.1% of meningiomas, and are distinguished by their highly vascular composition (>50%).[3,4] 88.9% of patients with angiomatous meningioma experience severe peritumoral edema due to hypervascularity, increased vascular permeability, and higher levels of VEGF compared to other meningiomas.[4]

Meningiomas have several predilection sites that can lead to space-occupying lesions. The prevalence of intracranial meningioma locations includes parasagittal/falcine (25%), convexity (19%), sphenoid ridge (17%), suprasellar (9%), posterior fossa (8%), olfactory groove (8%), middle fossa/Meckel's cave (4%), tentorial (3%), lateral ventricle (1-2%), foramen magnum (1-2%), and orbit (1-2%). In this patient, the meningioma is located in the middle fossa.

Meningiomas originate from arachnoid and dural border cells. Chromosome 6 mutations increase VEGF levels, causing neovascularization in angiomatous meningiomas. NF2 gene mutations (chromosome 22) disrupt MERLIN protein function, leading to meningioma development. Angiomatous meningiomas are often mistaken for hemangioblastomas or hemangiopericytomas. Imaging shows iso/hypodense lesions on CT and T1 iso/hypointense, T2 iso/hyperintense features on MRI, with intense contrast enhancement and possible dural tail sign. Severe peritumoral edema and high vascular activity make angiography essential for evaluation.[3] Histopathologically, vascular components predominate over meningothelial cells. Two types of vascular diameters can be observed in histopathological findings: macrovascular, with 50% of blood vessels having a diameter greater than 30 µm, and microvascular, with 50% of blood vessels having a diameter smaller than 30µm.[5]

Peritumoral edema in angiomatous meningioma

Peritumoral edema affects 40–78% of meningiomas, raising intracranial pressure and worsening postoperative outcomes. Tumor growth triggers ischemia and increased intratumoral water content. Ischemia induces VEGF

secretion, increasing vascular permeability and fluid leakage, which steroids help inhibit. Water content compresses venous drainage, causing blood stasis and the buildup of angiogenic factors, enhancing vascularization and worsening fluid leakage into surrounding brain tissue.[6]

Ultimately, this entire process results in peritumoral edema, which can elevate intracranial pressure and worsen neurological symptoms. Therefore, the main mechanisms of peritumoral brain edema in angiomatous meningioma involve ischemia, blood stasis, increased vascular permeability, and the recruitment of new blood vessels. Hence, therapies such as steroid administration aim to suppress the effects of VEGF to reduce vascular permeability and prevent further progression of severe edema.[6]

Several risk factors contribute to the development of peritumoral edema: (1) Tumor size, larger tumors are more prone to ischemia due to limited blood supply (2) Tumor location, skull base meningiomas can compress large veins or venous sinuses, disrupting venous drainage and leading to blood stasis (3) Tumor type, angiomatous meningioma is a histopathological subtype characterized by high vascularization and VEFG expression (4) Hormone receptors, rogesterone receptors play a role in regulating angiogenesis and vascular permeability, (5) Biochemical analysis, VEGF increases vascular permeability, leading to fluid extravasation into the brain tissue surrounding the tumor, (6) Patient age, older patients are more susceptible to severe peritumoral edema due to reduced vascular elasticity, impaired vascular autoregulation, and a heightened inflammatory response to tumor growth.[6]

Space-occupying lesions

Space-occupying lesions cause symptoms based on location: frontal lobe (motor deficits, executive dysfunction), temporal lobe (language, memory, hearing issues), parietal lobe (sensory deficits), and occipital lobe (visual disturbances).

The mechanism of headaches can be caused by both direct and indirect mechanism. Indirect mechanism arise from increased intracranial pressure, while direct mechanism result from distortion or inflammation of pain-sensitive intracranial structures, such as traction of the cortical veins and their branches, traction of the middle meningeal artery, traction of

the large arteries at the skull base, direct pressure on cranial nerves carrying pain fibers (like the trigeminal nerve), and dilation of intracranial arteries. In this patient, the initial left-sided headache was caused by a direct effect, where the mass compressed pain-sensitive structures in the middle cranial fossa, specifically the middle meningeal artery and the trigeminal nerve. Due to increased intracranial pressure, stretching of the dura mater occurs, which contains pain receptors, leading to an indirect effect that manifests as a diffuse headache.[3] The dura mater comprises arachnoid cap cells, originating the most common type of extra-axial intracranial tumor.[7]

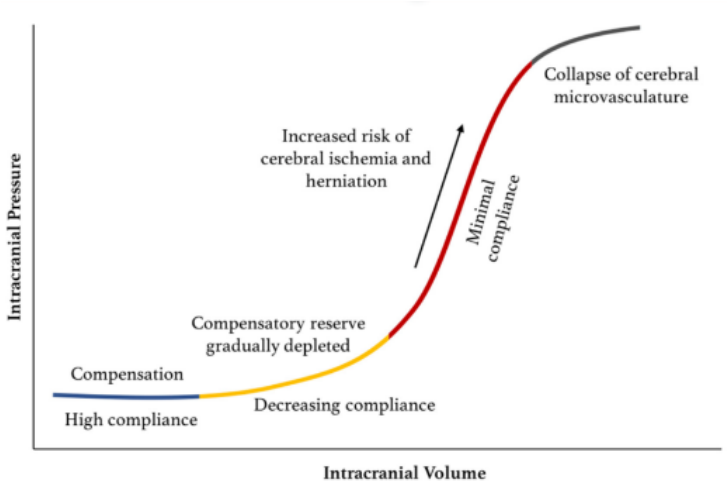
Hydrocephalus

CSF, produced by the choroid plexus, flows through the ventricles into the subarachnoid space. Hydrocephalus is CSF buildup due to impaired production, absorption, or flow. It can be communicating (absorption issues or overproduction) or obstructive (blockage). In this patient, postoperative debris blocked arachnoid drainage, causing hydrocephalus. [8]

Elevated intracranial pressure

Intracranial pressure (ICP) itself is regulated by three components: brain parenchyma (80%), CSF (10%), and blood (10%).

According to the Monro-Kellie doctrine, the total volume within the skull must remain constant. If one compartment's volume increases, the others' volume must decrease to maintain pressure. Cerebral compliance refers to the ability of intracranial components to adapt to volume changes without causing a significant increase in ICP. There are four stages of cerebral compliance, as shown in Figure 3: (1) High compliance zone (Blue): The brain can still compensate for volume increases without a significant rise in ICP (2) Decreasing compliance zone (Yellow): The brain's compensatory capacity begins to diminish, causing a gradual increase in ICP (3) Minimal compliance zone (Red): ICP rises sharply, threatening brain perfusion and increasing the risk of herniation (4) Collapse of Cerebral microvasculature zone (Black): Extreme pressure causes the collapse of cerebral blood vessels, which can be fatal.[9]



Intracranial pressure (ICP) must be managed if it increases above 22 mmHg for more than 5 minutes. This increase presents with clinical manifestations such as papilledema (CSF accumulation around the optic nerve sheath compresses the optic nerve, causing optic disc swelling), visual disturbances (optic nerve swelling disrupts axonal transmission, leading to incomplete visual impulse delivery), and diffuse headache.[9]

ICP elevation due to an obstructive tumor requires immediate management, starting with an analysis of its underlying cause. If the cause is a tumor, surgical removal is indicated. If the problem lies in CSF flow, CSF diversion techniques can be used, such as like ventriculoperitoneal (VP) shunt placement or ventriculostomy, along with carbonic anhydrase inhibitors.

When ICP elevation is due to edema, steroids or hypertonic fluids can be administered.[8]

In this patient, a VP shunt was placed. Malfunction of the shunt often presents with clinical signs of increased ICP. In this case, the patient developed post-traumatic hydrocephalus, where decompressive craniectomy caused a meningeal defect, and postoperative debris obstructed the subarachnoid space, leading to obstructive hydrocephalus.[9,10] After VP shunt revision, the patient's pupils were isochoric with reactive bilateral light reflexes.

Steroids are the anti-inflammatory treatment for brain tumors because they inhibit inflammatory mediators, stabilize endothelial tight junctions, and suppress vascular endothelial growth factor (VEGF), which helps reduce vasogenic and peritumoral edema. In hydrocephalus, carbonic

anhydrase inhibitors can also decrease CSF production by inhibiting bicarbonate activity in choroid plexus epithelial cells, ion transport, and pH regulation. However, the use of acetazolamide, a type of carbonic anhydrase inhibitor), requires careful consideration due to its numerous side effects, including metabolic disturbances like hypokalemia, hyponatremia, metabolic acidosis from hydrogen ion retention, and gastrointestinal symptoms such as nausea and diarrhea.[10]

Decompressive craniectomy temporarily removes part of the skull to relieve elevated ICP. In neuro-oncology, it's used for ICP elevation due to mass effects, edema, hydrocephalus, or hemorrhage. This patient underwent the procedure for a deep-seated meningioma, midline shift, and neurological deficits.[9] After the procedure, the patient experienced improvements in diffuse headache and visual disturbances.

Failure to manage ICP can cause cerebral herniation, indicated by Cushing's triad: hypertension, bradycardia, and irregular breathing. This patient experienced transcalvarial herniation (brain protrusion through the craniectomy defect) and uncal herniation, leading to decreased consciousness, ipsilateral pupil dilation, and contralateral hemiparesis.[3]

CSF leakage

The patient's chief complaint is CSF rhinorrhea, a condition where cerebrospinal fluid leaks due to a defect in the skull base or meningeal components. This creates a connection between the intracranial space and the nasal cavity, providing external access to the intracranial space and posing a high risk of intracranial infection. CSF rhinorrhea can result from idiopathic (40%), surgical (20%), iatrogenic (26%), trauma (10%), or congenital causes (4%). In this patient, it likely followed decompressive craniectomy, where dura shrinkage and cranioplasty disrupted CSF flow, causing a leak. Management includes bed rest, antibiotics, shunt placement, or dura repair.[11]

In this patient, CSF rhinorrhea likely occurred post-decompressive craniectomy, where the exposed skull made contact with air and the external environment, leading to shrinkage of the dura mater's cellular components. Cranioplasty performed on defective dura can disrupt CSF flow, creating excess CSF pressure, disturbing the CSF flow gradient, and resulting in a

CSF leak. Management can be either conservative (bed rest and high-dose intrathecal antibiotics) or surgical (shunt placement or dura repair).

Angiomatous meningioma management

Angiomatous meningioma management involves total tumor resection to minimize recurrence. Due to high vascularity and skull base location, preoperative trans-arterial embolization (TACE) is recommended to reduce bleeding, ease dissection, and shorten surgery. Subtotal resection requires follow-up radiation, while total resection only needs observation, often using Gamma Knife Radiosurgery.[12,13]

According to the 2024 NCCN guidelines, total tumor removal is the goal for recurrent or progressive meningioma. If only subtotal resection is possible, radiation therapy should follow. Post-treatment brain MRI is recommended at 3, 6, and 12 months in the first year, then every 6–12 months for five years, and every 1–3 years afterward.[14]

CONCLUSION

In women aged 40–70, chronic progressive headaches, visual disturbances, and cranial nerve paresis suggest skull base meningioma. Difficult access, proximity to vital structures, and bleeding risk complicate management and lead to disability. Despite its WHO Grade I classification, angiomatous meningioma is aggressive due to high vascular activity.

ACKNOWLEDGEMENT

We extend our appreciation to the medical professionals involved in the patient's care. Patient identities and specific details have been withheld to maintain confidentiality.

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