

Case Report

# Pseudotumor cerebri comorbid with meningioma: A review and case series

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## Abstract

**Background:** Pseudotumor cerebri (PTC), which has a prevalence in the general population of 1 to 2 out of 100,000, presents with raised intracranial pressure (ICP) but generally lacks a space occupying lesion.

**Case Description:** Patient 1 is a 32-year-old woman with a history of multiple meningiomas. Upon presentation to our institution, her clinical exam was notable for a right sixth nerve palsy. An integrated diagnosis of PTC was made and shunting for the cerebrospinal fluid (CSF) diversion was recommended. Approximately 6 weeks after surgery, the patient exhibited complete symptom resolution and discontinued all medications. Patient 2 is a 40-year-old woman with history of meningioma causing partial obstruction of the right transverse sigmoid sinus. She agreed to undergo surgery for the left ventriculoperitoneal (VP) shunt placement, for management of her PTC. Postoperatively, the patient reported that her vision significantly improved. Patient 3 is a 49-year-old woman with history of meningioma who presented with left visual field cut. A right frontal VP shunt was recommended for the treatment of PTC. Postoperatively, the patient reported significant symptom improvement and resolution of visual complaints.

**Conclusion:** This case series demonstrates that it is important to keep PTC in the differential diagnosis even when mass lesions such as meningiomas are discovered. Although PTC, as the name indicates, is classically diagnosed in patients without intracranial tumors, it is critical that this not be used as an absolute exclusion criterion. Finally, this case series supports the hypothesis that venous obstruction can result in PTC.

**Key Words:** Idiopathic intracranial hypertension, meningioma, pseudotumor cerebri, venous outflow obstruction

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## INTRODUCTION

Pseudotumor cerebri (PTC), which has a prevalence in the general population of 1 to 2 out of 100,000,<sup>[24]</sup> presents with raised intracranial pressure (ICP) but generally lacks a space occupying lesion.<sup>[1-3,6,7,9-11,14,15,20-25,27,29-31,33,35,36,39]</sup> The exact mechanism of this condition is still subject to great debate.<sup>[6,9,10,14,20,21,23,24,26,29,38,39,41]</sup> We present three

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cases of PTC comorbid with meningiomas seen at our institution and discuss the potential causes underlying the mechanism of this condition.

## CASE SERIES

### Patient 1

The patient is a 32-year-old woman (body mass index [BMI] of 24.8 kg/m<sup>2</sup>) with a history of multiple meningiomas. She initially complained of severely painful intermittent headaches that lasted approximately 20 seconds. Over the course of a year, these headaches increased in frequency to multiple times per day. Workup of the headaches at an outside hospital (OSH) included a brain magnetic resonance imaging (MRI) that demonstrated multiple lesions, mostly like meningiomas. One of the masses exerted mass effect on the superior sagittal sinus [Figure 1]. Gamma knife was recommended for the treatment, but after a second opinion, she agreed to undergo a craniotomy for resection of the largest lesion. Pathology confirmed a diagnosis of World Health Organization (WHO) Grade I meningioma. For approximately 7 months, she was asymptomatic and reported doing well, after which her headaches returned. She denied any visual symptoms. The headaches were refractory to Diamox, Topamax, and steroids. Six months after the headaches returned, she presented to our institution where a lumbar puncture was performed that revealed high normal opening pressure of 20 cm H<sub>2</sub>O. Routine cerebrospinal fluid (CSF) lab studies were unremarkable; however, her clinical exam was notable for a right sixth nerve palsy. Given the high ICP and sixth nerve palsy, an integrated diagnosis of PTC was made and a ventriculoperitoneal (VP) shunt for CSF diversion was placed. Approximately 6 weeks after surgery, the patient exhibited complete symptom resolution and discontinued all medications. She reported feeling the “best she had in the past 2 and a half years.” The patient did agree to eventually undergo gamma knife treatment for the meningiomas.

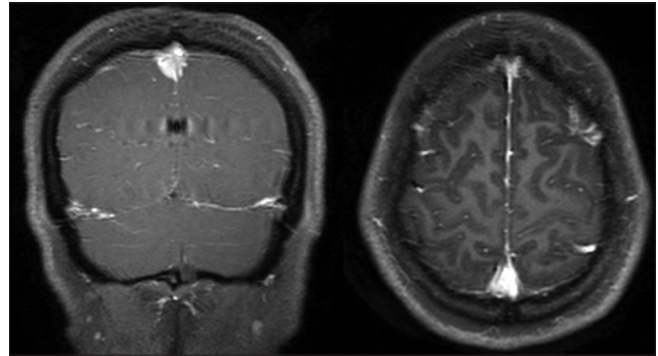
### Patient 2

The patient is a 40-year-old woman (BMI of 31.31 kg/m<sup>2</sup>) with history of meningioma causing partial obstruction of the right transverse sigmoid sinus, with no evidence of hydrocephalus [Figure 2]. Her left transverse sigmoid sinus appeared hypoplastic. She presented with nausea and headaches that were responsive to Diamox, Aleve, and Topamax. Ocular examination demonstrated the central and inferior visual field defect in her left eye and papilledema. Lumbar puncture demonstrated an opening pressure of 40 cm H<sub>2</sub>O. Routine CSF studies were unremarkable. She agreed to undergo surgery for left VP shunt placement for management of her PTC. Postoperatively, the patient reported that her vision significantly improved, although she did still have intermittent tinnitus in the left ear and intermittent echoing sensation in the right ear. She also reported having headaches at night. ENT consultation

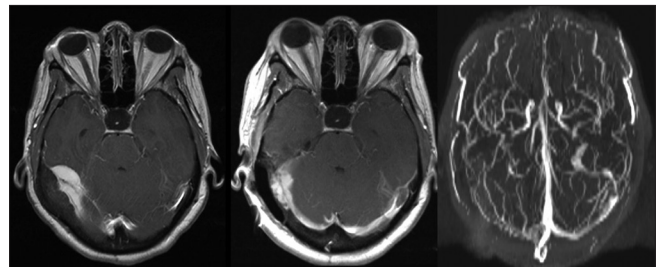
found no peripheral auditory pathology. The patient elected to have the meningioma resected 6 months later due to continued nightly headaches and unusual auditory sounds in the right ear.

### Patient 3

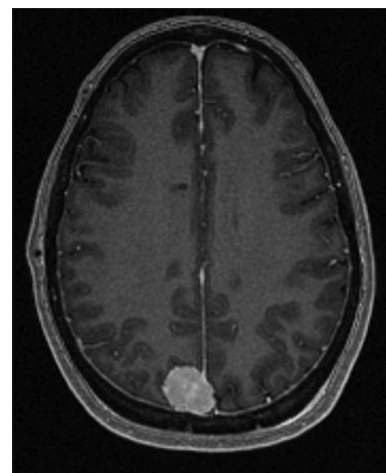
The patient is a 49-year-old woman who presented with left visual field cut (BMI of 27.45 kg/m<sup>2</sup>). Brain MRI revealed right parieto-occipital meningioma that abutted the



**Figure 1:** Coronal (left) and axial (right) T1 postcontrast MRI brain demonstrating meningioma abutting the superior sagittal sinus



**Figure 2:** Left/middle panels: T1 postcontrast axial MRI demonstrating right temporal/right cerebellar meningioma with mass effect on the right transverse sinus before (left) and after (middle) surgical resection. Some residual was noted along the right transverse sinus. Right panel: Axial MR venogram demonstrating R transverse sinus occlusion



**Figure 3:** T1 postcontrast axial MRI demonstrating left frontal and right parieto-occipital meningiomas abutting the superior sagittal sinus

superior sagittal sinus without hydrocephalus [Figure 3]. She had papilledema thought to be caused by venous obstruction. She denied headaches or tinnitus. The patient deferred lumbar puncture. A right frontal VP shunt was recommended for the treatment of PTC. Postoperatively, the patient reported significant symptom improvement and resolution of visual complaints. She denied any headaches, nausea, or vomiting and discontinued all medications. She returned to full-time work without any issues. At the patient's 1 year follow-up, she remained asymptomatic; however, imaging studies showed that the parieto-occipital meningioma increased in size. She elected to undergo gamma knife radiosurgery for treatment of the meningioma.

## DISCUSSION

### Pseudotumor cerebri or idiopathic intracranial hypertension

The term pseudotumor cerebri, which should be differentiated from idiopathic intracranial hypertension (IIH),<sup>[5,23,33]</sup> was coined by Nonne<sup>[20,21,27]</sup> and is thought to be the clinical manifestation of excess CSF production, decreased CSF reabsorption, and/or irregular venous outflow. These abnormal CSF dynamics result in elevated ICP without signs of ventriculomegaly or intracranial tumors.<sup>[4,6,7,10,11,14,15,18,19,23,24,27,30,31,33,35-39]</sup> The importance of differentiating PTC from IIH has been a topic of great discussion.<sup>[4,6,10,12,14,19,20,23,24,29,33,34,38]</sup> PTC includes both primary and secondary PTC.<sup>[6,23,24,33,34]</sup> In the primary PTC, which includes IIH, there is no identifiable cause for the symptoms related to PTC. In the secondary PTC, there is an identifiable cause for the patient's symptoms.<sup>[6,10,12,14,23,24,33,34]</sup>

### Symptoms and incidence

The most common symptoms of PTC are headaches, transient visual obscurations (TVOs), pulsatile tinnitus, and ocular pain.<sup>[2,13,23,24,30,33]</sup> These symptoms are present in 90%-94%, 68%-85%, 58%, and 44% of patients, respectively.<sup>[4,8,15,19,31,37,39]</sup> It is important to note that routine CSF laboratory studies are normal in most cases of PTC.<sup>[19,21,22,29,30,33]</sup> Despite the rare prevalence in the general population (1 to 2 cases out of 100,000),<sup>[33]</sup> there is an increased incidence in females, with a ratio of 8:1 female to male cases.<sup>[13,15,18,19,28,40]</sup> Females who are overweight and at the child-bearing age seem to have the highest risk of development, with a prevalence of 19.3 cases per 100,000 in this patient population.<sup>[7,13,18,19,31,37,40]</sup>

### Diagnosis

Correct and thorough diagnosis is crucial when examining patients with symptoms of PTC. Historically, computer tomography (CT) scans have been used to diagnose intracranial pathology. The advent of magnetic resonance (MR) imaging has

greatly improved our ability to detect intracranial pathology.<sup>[2,6,7,9-11,15,16,23,33,38]</sup> In addition, vascular imaging such as MR venography can detect venous sinus obstruction or flow abnormalities.<sup>[2,6,9-11,15,16,18,19,22-24,31,33,34]</sup> Other criteria for the diagnosis of PTC include CSF opening pressure over 25 cm H<sub>2</sub>O or 28 cm H<sub>2</sub>O for adults and children, respectively.<sup>[5,10,11,22-24,27]</sup> In addition, routine CSF studies, which are normal in PTC, should be sent to rule out other pathologies.<sup>[5,7,10,11,27,38]</sup> The presence of papilledema or sixth nerve palsy is a sign of increased ICP and supports a diagnosis of PTC.<sup>[5,8,10,11,14,22,24,40]</sup> It is important to note that diplopia is also a common symptom and may be the first sign of PTC before papilledema develops.<sup>[5]</sup> In addition, infections such as meningitis, otitis media, and mastoiditis, among others, should be considered in the differential diagnosis.<sup>[17]</sup> If other focal neurological signs are present on diagnostic tests, other diagnoses should be explored.<sup>[7]</sup>

Another important tool used for diagnosis is the Modified Dandy Criteria, which uses a set of criteria to exclude alternative diagnosis similar to PTC.<sup>[2,6,13,14,20,30,33]</sup> The set of guidelines are shown in Table 1. Other than the space occupying masses (meningiomas), all patients in this study met the Dandy criteria, with the sole exception of Patient 1 who had no visual deficits prior to treatment.

### Management of care

Once diagnosed, a treatment plan must be formulated. Although there is no single treatment option for PTC, there are three forms of management (surgical management, medical management, and life style change—namely weight reduction) which aim for symptom resolution through a reduction of CSF production and/or CSF pressure.

Surgical treatment options focus on reducing ICP by diversion of CSF.<sup>[3,6,10,11,14,15,17,18,23,24,27,38,39,42]</sup> Some recommend against surgical treatment as PTC is not life threatening, but the risks and benefits need to be weighed on a case-by-case basis.<sup>[6,10,14,18,23]</sup> Performing a lumbar puncture can aid in diagnosis, and temporary relief of symptoms provides justification for the surgical treatment.<sup>[3,6,11,38-40]</sup>

**Table 1: Modified Dandy Criteria for IIH**

#### Modified Dandy Criteria for IIH

Signs and symptoms related to an increase in intracranial pressure (headache, visual disturbances, papilledema)

No localized neurological symptoms (except for 6<sup>th</sup> nerve palsy) and normal CSF constituents

Absence of deformity, displacement, or obstruction of the ventricular system and otherwise normal neurodiagnostic studies, except for increased cerebrospinal fluid pressure

Patient is awake and alert

No other cause for increased intracranial pressure can be isolated

Medical management includes the use of diuretics such as carbonic anhydrase inhibitors, which decrease the production of CSF.<sup>[3,6,10,11,14,17,18,20,23,24,26,28,32,34,40,41]</sup> Other symptoms of PTC, such as headaches, can also be medically managed.<sup>[3,6,10,11,23,28,38,39]</sup>

One of the most important treatments for PTC, however, is the life style change. Specifically, weight management is critical, especially in obese women. Some studies show that a significant decrease in weight resolves major symptoms such as papilledema and headaches.<sup>[3,4,6,7,10,11,15,18-20,22-24,27,38-41]</sup> PTC also has a progressive impact on vision loss, and studies have shown that patients who have higher BMIs should be monitored more closely for decreases in vision.<sup>[4,6,7,10,41]</sup>

### Pathophysiology of PTC

Although the exact mechanism to PTC is still under debate, there have been several potential mechanisms documented in the literature, and decreased CSF absorption is the most commonly proposed mechanism.<sup>[4,6,10,11,21,23,24,27,28,31,35-39]</sup> Some factors that could contribute to this decreased absorption include venous outflow obstruction, defects in the arachnoid villi, increased levels of Vitamin A secondary to obesity, and decreased neurotransmitter production.<sup>[10,11,23,24,35,38]</sup> Defects in the arachnoid villi could trigger a decrease in CSF absorption by increasing villi resistance, even in the presence of other abnormalities, such as intracranial venous hypertension.<sup>[10,18,20,21,23,24,27,28,31,35-39]</sup> One cause of these defects in the arachnoid villi was attributed to high levels of Vitamin A, which has been shown to contribute to increased resistance during CSF absorption.<sup>[10,23,24,38]</sup> Another potential factor is abnormally low neurotransmitter production, specifically serotonin.<sup>[10,11]</sup> This decrease in neurotransmitter production increases CSF production in the choroid plexus, which would ultimately raise the CSF pressure due to an increase in CSF volume.<sup>[6,10,11,20,23,24,27,31,38]</sup> In addition, dural venous stenosis has been attributed to venous outflow obstruction and is exhibited in 14% to 90% of cases.<sup>[6]</sup>

### Meningiomas and PTC

The patients described in this case series met the clinical criteria for PTC and symptomatically improved with CSF diversion. Unique to this case series was the presence of meningiomas in these patients. While the vast majority of PTC patients do not have focal lesions on imaging (and although many include this in the diagnostic criteria), this case series demonstrates that patients with benign mass lesions can have PTC. Primary treatment of the meningioma, as was performed in Patient 1, will not resolve the underlying problem. While headache can be a symptom of meningioma, the actual cause of the headache should be investigated further as was performed in Patients 2 and 3. Only after the treatment of the true

underlying pathology, PTC, did this patient's condition improve. In this case series, it was interesting to note that only one patient had a BMI over 30 kg/m<sup>2</sup> (Patient 2, BMI of 31.8 kg/m<sup>2</sup>). The other two patients (Patients 1 and 3) had a BMI of 24.8 kg/m<sup>2</sup> and 27.44 kg/m<sup>2</sup>, respectively. No other risk factors were documented. Each patient in this case series had a meningioma that abutted a sinus and resulted in obstruction of venous outflow. Patient 2 underwent MR venogram that definitively demonstrated obstruction of venous outflow in the right transverse sinus [Figure 2]. These cases, hence, support venous outflow obstruction as the mechanism for PTC in these patients. This case series allows for unique insight into the mechanism of PTC. Venous outflow obstruction likely plays a role in PTC even in patients without mass lesions.

### CONCLUSIONS

In conclusion, although the prevalence of PTC is quite low in the general population (1 to 2 cases out of 100,000), the strongest incidence is in females who are overweight and at the child-bearing age (19.3 cases per 100,000 in this population). This case series demonstrates that it is important to keep PTC in the differential diagnosis even when mass lesions such as meningiomas are discovered. While PTC, as the name indicates, is classically diagnosed in patients without intracranial tumors, it is critical that this not be used as an absolute exclusion criterion. Furthermore, this case series demonstrates that venous outflow obstruction is a likely cause of PTC.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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