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Case Report

Management of concurrent symptomatic tuberculum sellae meningioma and idiopathic intracranial hypertension: A case report

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ABSTRACT

Background: Coexisting intracranial pathologies of distinct etiology which require intervention are rare. Only a handful of cases have been reported in the literature. The effects of each treatment option on both pathologies need to be considered during management. We describe the first report of the management of a patient with concurrent symptomatic tuberculum sellae meningioma (TSM) and idiopathic intracranial hypertension (IIH).

Case Description: A 58-year-old male presented with 2 weeks of vision loss and 3 months of headaches. He was found to have an inferior hemi-field deficit in the left eye and bilateral papilledema. Imaging studies revealed bilateral transverse sinus stenosis and a TSM abutting the left optic nerve. The opening pressure was 40 cmH_2O. An expanded-endoscopic endonasal approach was performed for mass resection. Intraoperatively, a lumbar drain was placed to aid skull base repair integrity before definitive treatment was obtained. On postoperative day 9, a right transverse-sigmoid sinus stent was placed for IIH treatment. The patient was discharged the following day.

Conclusion: Our management of this patient targeted the etiologies of each symptomatic pathology. Stenting provided treatment for the IIH and mass resection for the vision loss. Both the order and approaches to treatment were felt to maximize patient benefit while minimizing harm.

Keywords: Case report, Idiopathic intracranial hypertension, Symptomatic, Tuberculum sellae meningioma

INTRODUCTION

Careful consideration of all treatment options available is essential in the management of complex cases. We recently treated a patient with concurrent tuberculum sellae meningioma (TSM) and idiopathic intracranial hypertension (IIH) who presented with headaches, papilledema, and focal visual defect. We describe the rationale and considerations for the management of this patient.

IIH is a diagnosis of exclusion with an unclear etiology.[22] IIH classically presents with recurrent headache, visual loss secondary to papilledema, and an elevated opening pressure >25 cm water on lumbar manometry.[9,23] IIH is thought to be due to an imbalance in cerebrospinal fluid (CSF) absorption, with a growing pool of evidence attributing it to stasis of venous outflow at the transverse-sigmoid junction.[22] In mild cases without visual disturbances, conservative

management with weight loss and medications such as acetazolamide should be considered as the first line of treatment. In refractory or more severe cases, surgical intervention is indicated and should be tailored to the patient's pathophysiology. Surgical options for IIH include optic nerve sheath fenestration (ONSF), venous sinus stenting (VSS), CSF diversion, and bariatric surgery.[17,18,23]

TSMs are a subset of typically benign intracranial tumors that arise from the meningeal layer of the tuberculum sellae. Depending on the size and growth pattern of the tumor, patients may be asymptomatic and have headaches or visual field deficits.[15] Management options include observation, surgical intervention, and radiosurgery. Patients found to have visual deficits from compression of the optic apparatus should be offered surgical intervention. Surgical resection of TSMs can be achieved from either extracranial or intracranial approaches taking into consideration the pathology and neuroanatomy on a case-by-case basis.[2,15] Both approaches offer high rates of gross total resection but come with their own set of potential complications.[3,6,11]

While both entities have a unique pathophysiology, their presenting symptoms can overlap, which complicates the development of an optimal therapeutic plan. Concurrent intracranial pathologies of distinct etiology requiring surgical intervention are rare, with only a handful of cases reported in the literature.[12,19,20] Here, we describe the first reported case of the management of a patient with concurrent symptomatic IIH and TSM.

CASE DESCRIPTION

We present a 58-year-old male with class I obesity (body mass index [BMI] 31.0), a history of renal cell carcinoma in remission status post gross total resection, hypertension, and diabetes mellitus type 2, evaluated as an outpatient by an ophthalmologist for a 2-week history of vision loss in his left eye and 3 months of mild frontal headaches. He was found to have inferior nasal and temporal field defects in his left eye with bilateral papilledema. His visual acuity (VA) was 20/25 on the right and 20/30 on the left. He was sent to the emergency department for further work up. Initial head computed tomography (CT) was suspicious for bilateral transverse sinus stenosis (TSS) and a mass in the suprasellar region. A CT venogram was obtained and confirmed bilateral TSS. Magnetic resonance imaging (MRI) confirmed the presence of a homogeneously enhancing, dural-based tuberculum sellae mass with involvement of the left optic nerve and proximal segment of the left anterior cerebral artery [Figure 1]. The MRI also demonstrated mild dilation of the distal optic nerve sheath bilaterally, as well as flattening of the posterior globe [Figure 2]. A high-volume lumbar puncture was performed, revealing an opening pressure of 40 cm of water. Neurosurgical assessment of the clinical

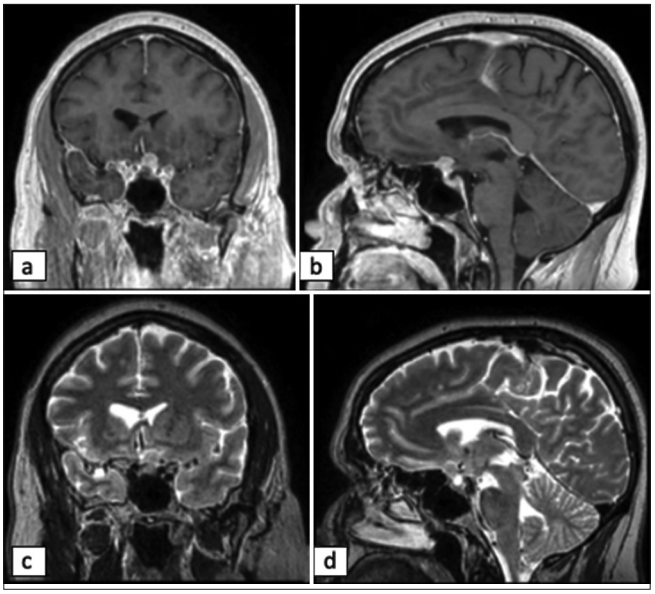

Figure 1: Preoperative T1 with contrast (a) coronal and (b) sagittal sequences demonstrating a tuberculum sellae meningioma eccentric to the left. Preoperative T2 (c) coronal and (d) sagittal sequences demonstrating compression of the optic apparatus.

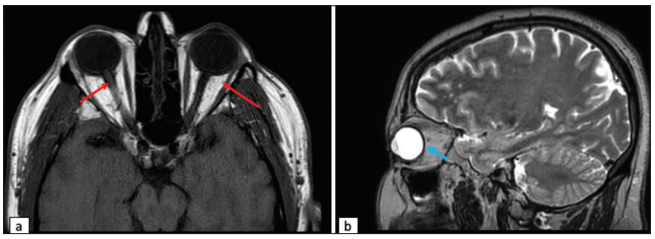

Figure 2: Preoperative axial T1-weighted image of the orbit (a) demonstrating distal dilation of the optic nerve bilaterally (red arrows). Preoperative sagittal T2-weighted image of the brain (b) demonstrating flattening of the posterior globe (blue arrow).

and radiographic findings concluded that the etiology of the patients presenting symptoms was due to compression of the left optic nerve by the TSM and IIH secondary to TSS.

The patient underwent an expanded endoscopic endonasal approach for surgical resection of the mass. Intraoperatively, a lumbar drain was placed. Skull base repair was done, implementing both an inlay and overlay graft to create a dural substitute. A pedicle of the nasal septal flap was kept intact to maintain its blood supply. The vascularized flap was placed on top of the overlay and covered with surgical sealant. Postoperatively, the patient reported a slight improvement in vision. The lumbar drain was removed on postoperative day 7. On postoperative day 8, the patient was loaded with 650 mg of aspirin and 300 mg of clopidogrel. On postoperative day 9, the patient was taken to the interventional radiology suite to undergo VSS. The angiogram confirmed bilateral TSS, worse on the left than the right; however, the right side appeared to be

the dominant sinus. A right transverse-sigmoid sinus stent was subsequently placed with the pressure gradient decreasing from 11 mm Hg to 3 mm Hg. There was no evidence of complications [Figure 3]. The patient tolerated the procedure well and was discharged the following day on 325 mg of aspirin and 75 mg of clopidogrel daily for a planned duration of 3 months.

Pathology was consistent with a World Health Organization grade 1 meningioma; therefore, no plans were made for postoperative radiation. He has been followed with serial imaging. Radiographic outcomes have demonstrated gross total resection of the TSM without evidence of recurrence and patency of the right sinus stent [Figure 4]. Clopidogrel was discontinued at 3 months as planned, and he is to continue taking aspirin indefinitely. Clinically, the patient reports mild headaches and significant improvement in his vision. A formal ophthalmologic examination at 7 months postoperatively demonstrated an improving blind spot with only a partial outer inferior nasal visual defect of the left eye. His VA on the left improved to 20/25, with no change in VA on the right.

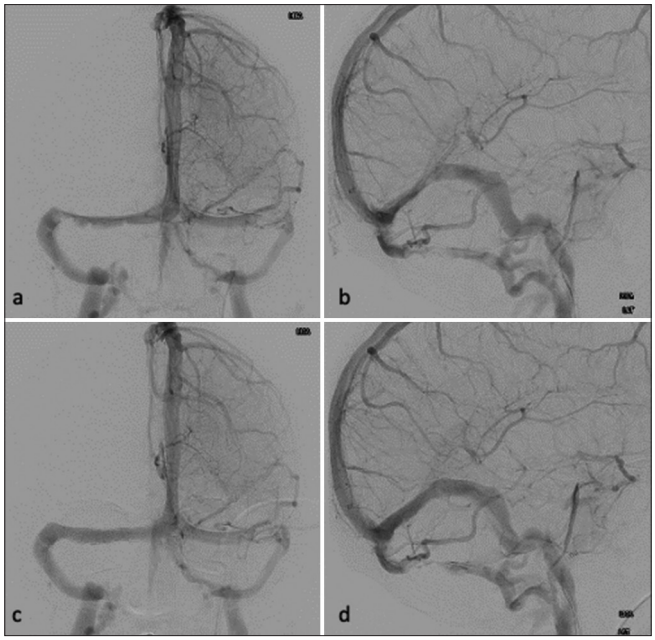

Figure 3: Preoperative (a) left anteroposterior (AP) internal carotid artery (ICA) and (b) left lateral ICA injections demonstrating bilateral transverse sinus stenosis. Postoperative (c) left AP ICA and (d) left lateral ICA injections demonstrating improved stenosis of the right transverse sinus after venous sinus stent placement.

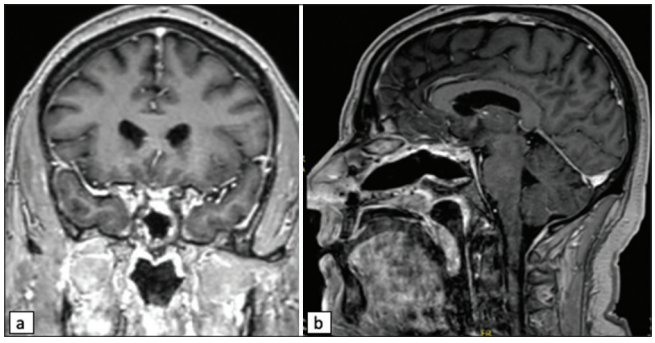

Figure 4: Postoperative T1 with contrast (a) coronal and (b) sagittal sequences showing postoperative changes of an endonasal approach to the tuberculum without evidence of residual or recurrent tumor.

DISCUSSION

The management of patients with concurrent intracranial pathologies presents unique challenges to developing an optimal treatment plan. Careful consideration must be given to both while prioritizing the treatment to address the patient's symptoms.[12,19,20] In this case, the ophthalmologic findings of left eye inferior nasal and temporal field cuts were attributed to the TSM. Although the radiographic findings of a small tumor technically excluded the patient from a diagnosis of IIH, it was not felt to be a contributing factor to the findings of elevated opening pressure, bilateral papilledema, and evidence of transverse sinus stenosis. Delay in treatment for either pathology would have risked worsening vision loss, and therefore devising an optimal plan required addressing both pathologies quickly and safely. The optimal treatment for the elevated intracranial pressure in this patient was determined to be a VSS.[7,13,14,17] Acceptable alternative options would be ONSF or CSF diversion; however, ONSF would not address the headaches and given the radiographic findings of bilateral TSS, CSF diversion would not have addressed the underlying pathophysiology. In addition, this would have condemned the patient to potential lifelong complications associated with shunting, which may be more prevalent in the IIH population.[10]

The treatment for IIH was not thought to address the visual field cut, and undergoing a venous stent procedure would require the patient to be on dual anti-platelet therapy (DAPT). This would have delayed a possible surgical decompression of the left optic nerve for at least 3–6 months after stent placement, during which time there would have been a high concern for deterioration of the patient's vision. In the case of deterioration, the patient would have faced worsening vision with no intervention or would have risked in-stent thrombosis secondary to the cessation of DAPT to perform surgery. Contrarily, it was felt that VSS with DAPT therapy could safely be initiated within a week after surgical resection, avoiding the risks mentioned above.

Many factors were considered when choosing the optimal surgical approach. An endonasal approach was indicated here since the tumor was small and located between the carotid arteries and below the optic apparatus.[2] An open craniotomy, typically through a pterional or cranio-orbital approach, is a more invasive procedure with lower rates of

CSF leak but greater manipulation of brain parenchyma to reach a midline lesion and places important neurovascular structures between the surgeon and the underlying tumor.[1,11,15] In addition, the small size of the tumor and underlying IIH limits the surgical corridors created by opening subarachnoid spaces, using gravity retraction, and taking advantage of the space created by larger tumors. Due to these reasons, open approaches can be more technically challenging and place the patient at higher risk for neurovascular compromise. While an endonasal approach would allow for direct access to the tumor with minimal risk to the brain parenchyma, optic apparatus, and vascular structures, it would place the patient at higher risk for CSF leak.[1-3,6] Furthermore, the underlying IIH would increase the already known risk of CSF leak with endonasal anterior skull base approaches to intradural lesions.[5] However, in cases of spontaneous CSF leak, endoscopic endonasal surgical repair alone can reach success rates of up to 90%.[24] It follows that in cases where a primary CSF leak has not occurred, endoscopic repair following an intervention that leaves a dural defect should provide adequate prevention. To the best of our knowledge, there are no studies in patients with documented IIH who undergo endoscopic skull base surgery for a separate pathology. Elevated BMI is associated with an increased risk for CSF leakage after endoscopic endonasal approaches.[5,8] In these high-risk patients with elevated BMI, Michael *et al*. found that implementing a vascularized flap in the repair of spontaneous CSF leaks reduced the risk of recurrence.[16] Cohen *et al*. found that placing an intraoperative pre-resection lumbar drain may help reduce the risk of postoperative CSF leak in patients with elevated BMI.[4] This is consistent with a study by Teachey *et al*., who found that preoperative intervention, both medical and procedural, of elevated ICP increased the success rate of endoscopic repair of spontaneous CSF leak.[21] In a randomized study, perioperative lumbar drainage used in the context of a vascularized nasoseptal flap for closure was associated with a significantly reduced rate of postoperative CSF leaks.[25] Weighing all the pros and cons of the different approaches for our patient, an expanded endonasal transsphenoidal/transtubercular approach with intraoperative placement of the lumbar drain was recommended. Gross total resection was achieved, and postoperatively, the lumbar drain was used to assist with healing the skull base reconstruction in the setting of known increased intracranial pressure.[21] Lumbar drainage also offered the secondary therapeutic benefit of CSF diversion in a patient with IIH and papilledema. To reduce the risk of CSF leak from increased ICP in the setting of IIH and an anterior skull base reconstruction of a large defect, the lumbar drain was left for 7 days. We implemented the use of the vascularized flap and intraoperative lumbar drain to decrease the risk of postoperative leak in our patient with class 1 obesity and documented IIH.

For our patient, treatments were selected to target a specific pathologic sign/symptom. Tumor resection was aimed primarily at vision loss due to compression of the optic apparatus, while VSS aimed to relieve the headache and papilledema. The order of the treatment plan was chosen based on minimizing potential deterioration of the patient's vision and complications associated with surgical management. The threat of increased vision loss in this patient presenting with a subacute visual deficit was felt to be the most important clinical factor in our decision.

CONCLUSION

The treatment plan for this patient was complex due to the presence of two distinct pathologies potentially contributing to overlapping symptoms. The diagnostic workup for these two pathologies and treatment options available when these illnesses occur simultaneously is no different than when they occur in isolation. A sophisticated treatment plan was necessary to address both pathologies individually, as the management of one pathology complicated that of the other and vice versa. Most importantly, the finalized treatment plan should minimize harm to the patient.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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