Cerebral Vasospasm Following Transsphenoidal Hypophysectomy in the Treatment of Lymphocytic Hypophysitis

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Background
Cerebral vasospasm (CV) is a rare entity in the setting of tumor resection, as such patients are typically not monitored for its occurrence nor treated prophylactically for its prevention. They often present to the hospital with a fulminating form that is difficult to treat. With the increased popularity and expanding indications for endoscopic endonasal surgery of the sella and parasellar regions, it is important to recognize this potential complication. There are 11 known cases of CV following transsphenoidal surgery, all associated with the resection of pituitary macroadenomas. We report the first case of vasospasm following transsphenoidal hypophysectomy (TSH) in the treatment of lymphocytic hypophysitis.

Case Presentation
19-year-old female with a four-year history of vague visual complaints. She presented with one-month history of worsening intractable nausea, vomiting, vertigo, and severe new-onset headaches. Hormonal testing was consistent with hypopituitarism. MRI revealed a 1.7 cm x 1.6 cm x 1.4 cm peripherally enhancing pituitary mass with suprasellar extension with mass effect upon the optic chiasm (Fig 1). The patient underwent a stereotactic endoscopic transsphenoidal hypophysectomy. The surgery was uncomplicated and intraoperative blood loss was 230 ml. The patient's visual disturbances resolved postoperatively but headaches persisted. There was evidence of CSF leak and she was discharged on POD 4. Final pathologic analysis of the lesion revealed dense infiltrates of lymphocytes and plasma cells effacing the normal adenohypophysis architecture. Lymphoid follicles with germinal centers and parenchymal fibrosis were also evident, all findings consistent with lymphocytic hypophysitis (LH). On POD 9, the patient presented with altered mental status and slurred speech. An MRI/MDA of the brain showed a small left basal ganglia infarct and vasospasm of the left supraciliary ICA, proximal MCA, and ACA segments. The patient was immediately started on intravenous fluids and nimodipine. Despite initial improvement, the next day she developed acute expressive aphasia, episodic right hemiparesis, and became lethargic requiring intubation for airway protection. Emergent catheter angiography revealed moderate to severe vasospasm involving the left supraciliary ICA, M1 MCA, and A1 ACA segments with restrictive downstream flow (Fig. 2). She was treated with intra-arterial vasodilators, including Nicardipine and Milrinone, with limited improvement. Ultimately, she developed global aphasia and right hemiplegia. MRI showed patchy areas of stroke. At 8 month follow up she has residual hemiparesis and improving residual expressive aphasia.

Discussion
Lymphocytic hypophysitis is rare autoimmune inflammatory disorder characterized by lymphocytic infiltration of the pituitary gland with destruction and fibrosis of glandular tissue. As of 2013, there have only been 390 published cases of biopsy-proven LH. The lymphocytic infiltration of the pituitary gland forms a sellar mass that can expand upward and impinge upon the optic chiasm, dura mater, or cavernous sinus.

The most common symptoms of LH originate from this mass effect and include generalized headaches and visual disturbances. LH is often misdiagnosed as a nonfunctioning pituitary adenoma. In the presence of mass effect, continued deterioration despite medical management or the need for definitive diagnosis, surgery should be undertaken. CV following a transsphenoidal approach continues to be exceedingly rare. All of the reported cases occurred following resection of pituitary adenomas signifying that CV has never been reported following resection of LH, another very rare disease process. In documented cases, the initial signs and symptoms of CV following TSH are changes in mental status, motor deficits, and speech difficulties. The average time to develop clinical CV following tumor resection is 8 days. Patients have often already been discharged prior to symptom onset, so the clinician needs to have a high level of clinical suspicion if the patient returns to the ER with mental status, motor deficits, or speech difficulties.

Conclusions
Cerebral vasospasm is a rare complication of transsphenoidal hypophysectomy but carries with it significant morbidity and mortality. Clinicians should have a high level of clinical suspicion for CV when patients develop altered mental status, motor deficits, or speech difficulties following TSH. While all previously reported cases involved resection of pituitary macroadenomas, our study has revealed that this significant complication also occurs with resection of lymphocytic hypophysitis.

References
8. Rose NR, Mackay IR. The autoimmune diseases. 2004, Elsevier/Academic Press,: Amsterdam; Boston. p. 1 online resource (xiii, 1267 pages)