Endoscopic Endonasal Approach to Sellar Decompression and Biopsy for Lymphocytic Hypophysitis

Camille K. Milton, MD[1]; Emal Lesha MD[1]; David G. Laird BS, BA[1]; Delaney M. Graham BS[1], Alexandra H. Kramer BS[1], Christopher P. Golembeski MD, MBA[2]; Samuel Smith MD, MPH[3], L. Madison Michael MD[1]

1.University of Tennessee Health Science Center-Semmes-Murphey Clinic Department of Neurosurgery; 2.Pathgroup; 3.University of Tennessee Health Science Center Department of Otolaryngology

Abstract

A 21-one-year-old woman presented 6 days postpartum with progressive headache, blurred vision, and left hemianopia. Imaging demonstrated a sellar lesion with mass effect on the optic chiasm. After endoscopic endonasal approach to sellar decompression, the patient's vision returned to baseline. Lymphocytic hypophysitis is a rare autoimmune condition affecting the pituitary gland that commonly occurs during pregnancy and postpartum. Our case demonstrates that neurosurgical intervention should be considered for cases with symptomatic mass effect.

Clinical Presentation

A 21-year-old female with history of migraines presented to the emergency department 6 days postpartum with progressive left sided vision loss and headache. These episodes began during her third trimester and significantly worsened after delivery. She endorsed peripheral vision loss in the left eye with blurry vision.

Laboratory: Endocrine panel within normal limits with exception of low free T4 (0.49)

Exam: GCS15, nonfocal neurological exam with exception of ophthalmologic exam significant for decreased visual acuity and left hemianopia without papilledema

Neuroimaging

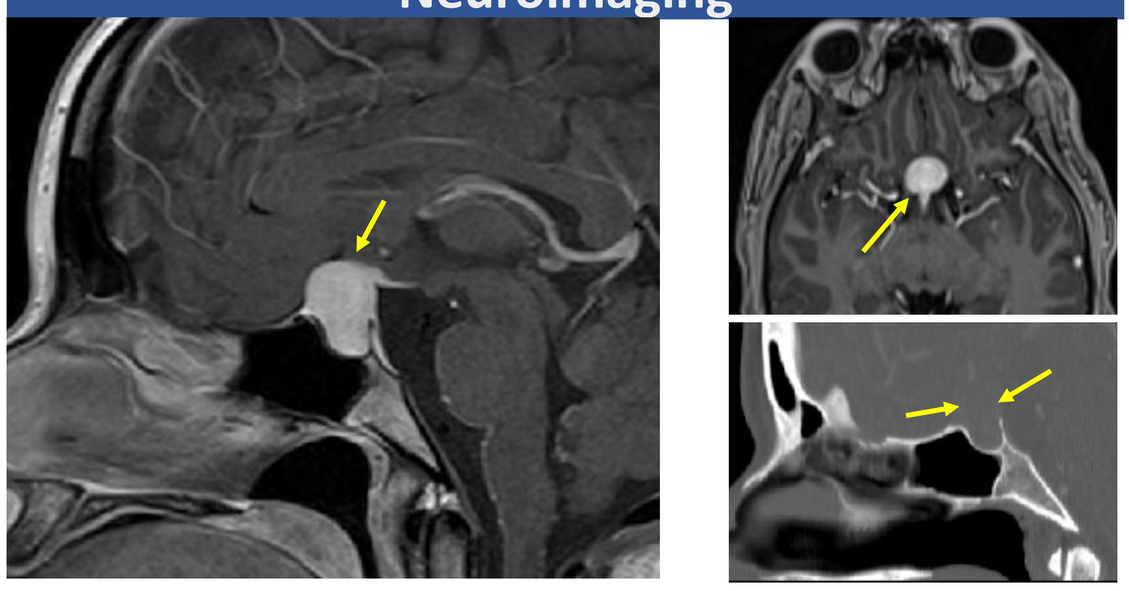

Figure 1. MRI and CT imaging showed a 14 x 18 x 21 mm homogeneously enhancing sellar and suprasellar mass with thickening and enhancement of the infundibulum, and without significant expansion of the sella.

.

Rationale for Procedure

She was admitted to the hospital for further workup and evaluation by Endocrine, Ophthalmology and ENT services. Steroids were initiated with moderate improvement in visual complaints.

As the patient had a large sellar mass and acute vision loss, surgery was chosen as an option to obtain diagnosis and allow decompression of the sella and the optic apparatus.

Key Surgical Steps

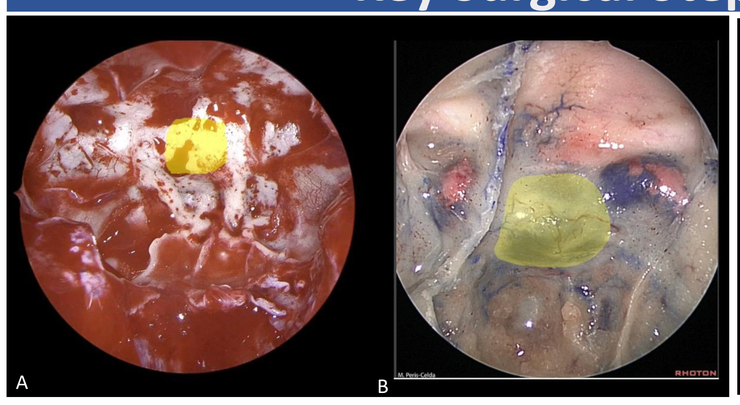

Figure 2. Step 1. A) Wide sphenoid and posterior ethmoid exposure. Sella highlighted in yellow. B) Courtesy of the Rhoton Collection, American Association of Neurological Surgeons (AANS)/Neurosurgical Research and Education Foundation (NREF).

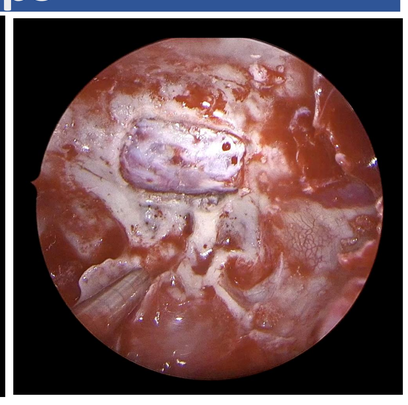

Figure 3. Step 2. Bony removal of sellar floor

Key Surgical Steps (Continued)

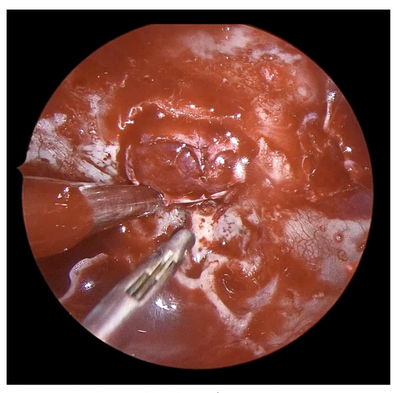

Figure 4. Step 3. Dural opening

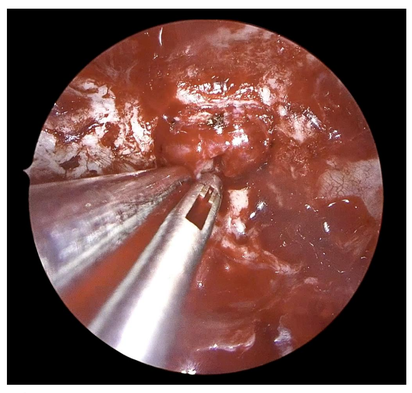

Figure 5. Step 4. Biopsy

Discussion

Lymphocytic hypophysitis is a rare autoimmune condition characterized by inflammation of the pituitary gland. While the exact cause of this disease is unclear, genetic and environmental factors are believed to play a role.

Neurosurgical management with surgical decompression or stereotactic radiosurgery have been previously described as a treatment for lymphocytic hypophysitis and are generally reserved for cases in which inflammation leads to significant mass effect causing visual disturbances, severe headaches, or pituitary apoplexy. Surgical intervention may also be needed when medical management with corticosteroids or other immunosuppressive agents do not adequately reduce the inflammation.

The most common surgical approach in patients with LH is the transsphenoidal approach, with rare reports of patients requiring craniotomy.

Clinical Outcome

Postoperatively, the patient's visual acuity was 20/20. The left sided visual field defect had normalized and returned to full baseline at two months post-operative evaluation by Ophthalmology.

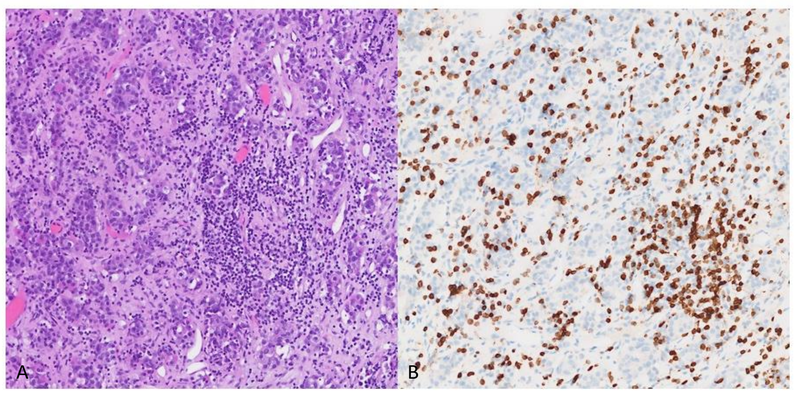

Figure 6. Pathology A) H&E stain demonstrates well organized nests of anterior pituitary cells with morphologies consistent with benign gland with focal chronic hypophysitis. No adenoma was identified. B) The majority of inflammatory cells in lymphocytic hypophysitis should be T cells, as highlighted by the CD3 stain

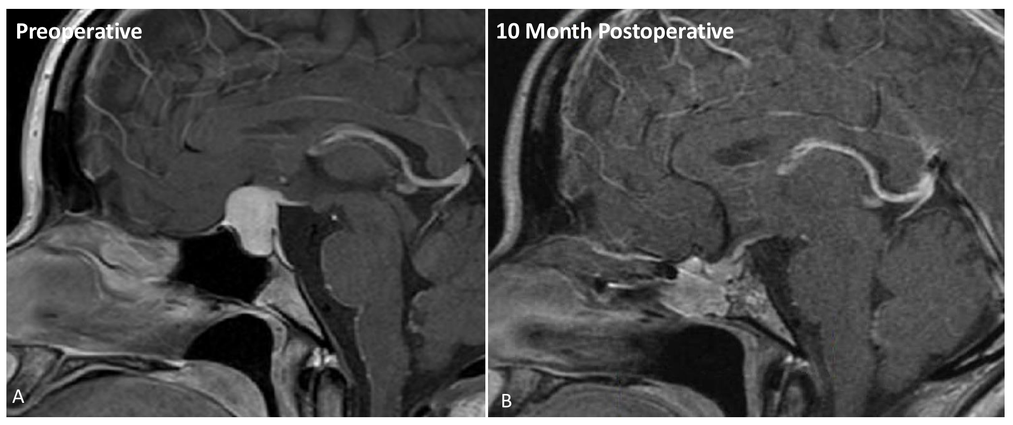

Figure 7. A) Preoperative T1 with contrast MRI B) 10 months postoperative T1 with contrast MRI demonstrating resolution of suprasellar mass

Conclusions

While medical management remains the mainstay of treatment for lymphocytic hypophysitis, cases like ours demonstrate that neurosurgical intervention should be considered for patients with symptomatic mass effect.

Contact

Camille K. Milton
University of Tennessee Health Science Center
Semmes Murphey Clinic
Department of Neurosurgery
847 Monroe Avenue, Suite 427
Memphis, TN 38163

References

1. Yang, C., Wu, H., Bao, X., & Wang, R. (2018). Lymphocytic Hypophysitis Secondary to Ruptured Rathke Cleft Cyst: Case Report and Literature Review. World neurosurgery, 114, 172–177. https://doi.org/10.1016/j.wneu.2018.03.086

2. Ray, D. K., Yen, C. P., Vance, M. L., Laws, E. R., Lopes, B., & Sheehan, J. P. (2010). Gamma knife surgery for lymphocytic hypophysitis. Journal of neurosurgery, 112(1), 118–121. https://doi.org/10.3171/2009.6.JNS08117

3. Pekic, S., Bogosavljevic, V., Peker, S., Doknic, M., Miljic, D., Stojanovic, M., Skender-Gazibara, M., Gacic, E. M., Popovic, V., & Petakov, M. (2018). Lymphocytic Hypophysitis Successfully Treated with Stereotactic Radiosurgery: Case Report and Review of the Literature. Journal of neurological surgery. Part A, Central European neurosurgery, 79(1), 77–85. https://doi.org/10.1055/s-0037-1604079

4. Leung, G. K., Lopes, M. B., Thorner, M. O., Vance, M. L., & Laws, E. R., Jr (2004). Primary hypophysitis: a single-center experience in 16 cases. Journal of neurosurgery, 101(2), 262–271. https://doi.org/10.3171/jns.2004.101.2.0262

5. Iuliano, S. L., & Laws, E. R. (2011). The diagnosis and management of lymphocytic hypophysitis. Expert review of endocrinology & metabolism, 6(6), 777–783. https://doi.org/10.1586/eem.11.74

6. Khaleghi, M., Finger, G., Wu, K. C., Munjal, V., Ghalib, L., Kobalka, P., Blakaj, D., Dibs, K., Carrau, R., & Prevedello, D. (2024). Successful treatment of medically and surgically refractory lymphocytic hypophysitis with fractionated stereotactic radiotherapy: a single-center experience and systematic literature review. Pituitary, 27(2), 213–229. https://doi.org/10.1007/s11102-023-01367-8

7. Guo, S., Wang, C., Zhang, J., Tian, Y., & Wu, Q. (2016). Diagnosis and management of tumor-like hypophysitis: A retrospective case series. Oncology letters, 11(2), 1315–1320. https://doi.org/10.3892/ol.2015.4046