

Endoscopic Endonasal Resection of a sellar Langerhans Cell Histiocytosis in an 8-month-Old: Technical and Diagnostic Considerations

David Bailey MD, David Goldrich MD, Neerav Goyal MD, Michael McDowell MD
Penn State Milton S. Hershey Medical Center



Abstract

Langerhans Cell Histiocytosis is a rare myeloid neoplastic disease commonly affecting the bone and skin. Central nervous system involvement can involve the sellar region, which can result in cranial nerve and endocrine dysfunction.

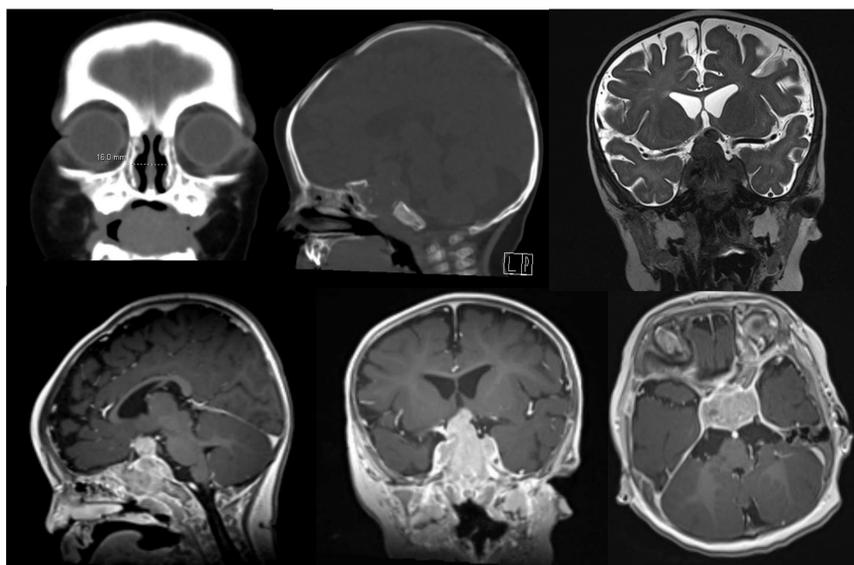
This video illustrates an endoscopic endonasal resection of a sellar mass in an 8-month-old, later diagnosed as Langerhans Cell Histiocytosis (LCH). It highlights the feasibility of this minimally invasive approach in patients under one year of age and briefly reviews key aspects of clinical management for this uncommon pathology.

Case Presentation

- An 8-month-old previously healthy boy with no past medical history presented with fever, progressive irritability, and regression of developmental milestones over the last several weeks.
- Physical exam: Head circumference in the 99th percentile. Left eye ptosis and limited extra-ocular movements. No papilledema. Otherwise, no focal neurologic deficits
- Laboratory examination demonstrated pan-hypopituitarism

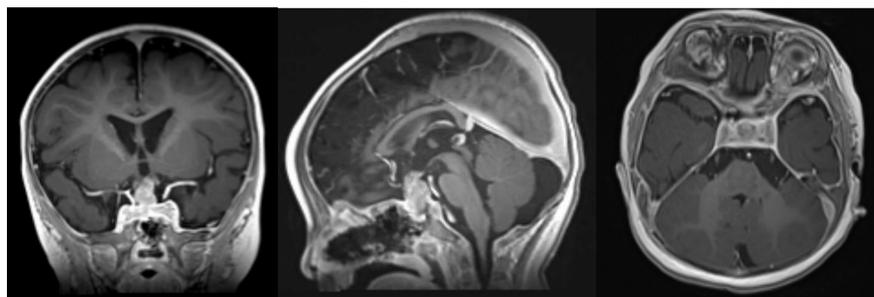
| Test | Result (Normal range) |
|-----------|---------------------------------|
| Cortisol | 2.5 µg/dL (5 – 25 µg/dL (8 AM)) |
| ACTH | 5 pg/mL (10 – 60 pg/mL) |
| Free T4 | 0.49 ng/dL (0.7 – 1.9 ng/dL) |
| TSH | 0.69 µIU/mL (0.5 – 5.0 µIU/mL) |
| LH | < 0.2 IU/L (1.2 – 12.0 IU/L) |
| FSH | < 0.1 IU/L (1.5 – 12.4 IU/L) |
| Prolactin | 33.7 ng/mL (< 20 – 25 ng/mL) |

- Radiographic evaluation:
 - CT with osteoclastic lesion in the sella
 - MRI with T2-hypointense, avidly contrast enhancing lesion extending from the third ventricle to the nasopharynx.
 - No encasement of carotid arteries
 - Differential diagnosis: Pituitary adenoma, craniopharyngioma, meningioma, chordoma, metastatic lesion

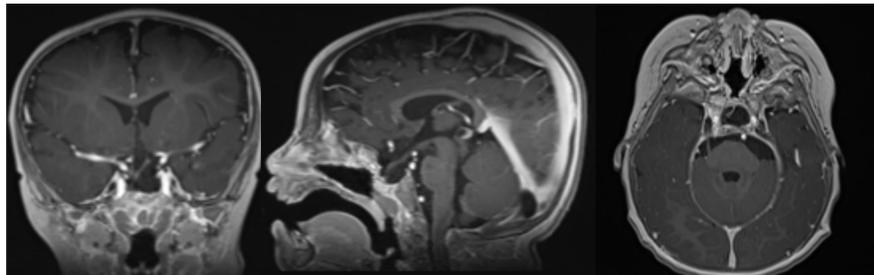


Surgical management

- Decision to complete surgical debulking. Goals of surgery to reduce mass effect exerted on cranial nerves and to achieve tissue for diagnosis
- Surgical plan:
 - Endoscopic endonasal approach with middle turbinectomy, bilateral complete ethmoidectomy and sphenoidotomy
 - Preparation of nasoseptal and reverse nasoseptal flaps
- Surgery:
 - Extra-dural tumor debulked without complication.
 - No cerebrospinal leak encountered
 - Frozen suggestive of high-grade bony neoplasm
 - Decision made to stop, with plans to return pending final pathology
- Post-operative course
 - Uneventful recovery in pediatric intensive care unit
 - Post-operative imaging with successful tumor debulking



- Pathology: Langerhans cell histiocytosis with BRAF mutation
- Treatment: Trametinib (MEK inhibitor)
- Follow-up: 6-month imaging with complete resolution of residual tumor, resolution of cranial nerve deficits, and regaining of developmental milestones. Remained with pan-hypopituitarism



Discussion

- LCH is a myeloid neoplastic disorder which can invade the sellar region, commonly presenting with diabetes insipidus, exophthalmos, and lytic skull lesions (Hand-Schüller-Christian disease)
- Traditional therapy includes chemotherapy, but lifetime BRAF targeting medications offer excellent long-term survival.
- Endoscopic endonasal surgery in infants is feasible and safe
 - Surgical corridor may be limited by piriform aperture width and inter-carotid distance.
 - Non-pneumatized sinus requires careful neuronavigation
 - Reconstruction requires consideration of possible defect size. Nasoseptal flap is a good option in children with an appropriately size defect.

Contact

Michael McDowell, MD
Penn State Milton S. Hershey Medical Center
Mmcdowell1@pennstatehealth.psu.edu

Operative video



References

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