Persistent central ACTH-dependent hypercortisolemia following pituitary stalk tumor resection: Case lesson

Patrick Toyota, MD[1]; Kristen Marciniuk, MD, PhD[1]; Devon Houdek, MD[1];
Neil Arnstead, MD[1]; Amit Persad, MD[1]; Luke Hnenny, MD[1]
[1]University of Saskatchewan

Abstract

Background

Infundibular lesions are rare entities with variable clinical manifestations. Their detection during workup for endocrinologic abnormalities represents a decision-making challenge. We present a patient with ACTH-dependent hypercortisolemia found to have a stalk lesion, which was treated surgically.

Results

A 57F underwent workup for Cushing's syndrome, demonstrating evidence of ACTH-dependent hypercortisolemia. Imaging demonstrated a 4.7mm infundibular nodule. We elected to proceed with endoscopic endonasal approach for resection of the infundibular lesion with goal of biochemical cure.

A satisfactory technical and radiographic resection of the infundibular lesion was achieved. However, the patient's hypercortisolemia failed to resolve. Histopathologic analysis identified the lesion as a granulocytoma. Inferior petrosal sinus sampling further demonstrated evidence of ACTH-dependent central hypercortisolemia. She then underwent bilateral adrenalectomy for management of her persistent hypercortisolemia.

Conclusions

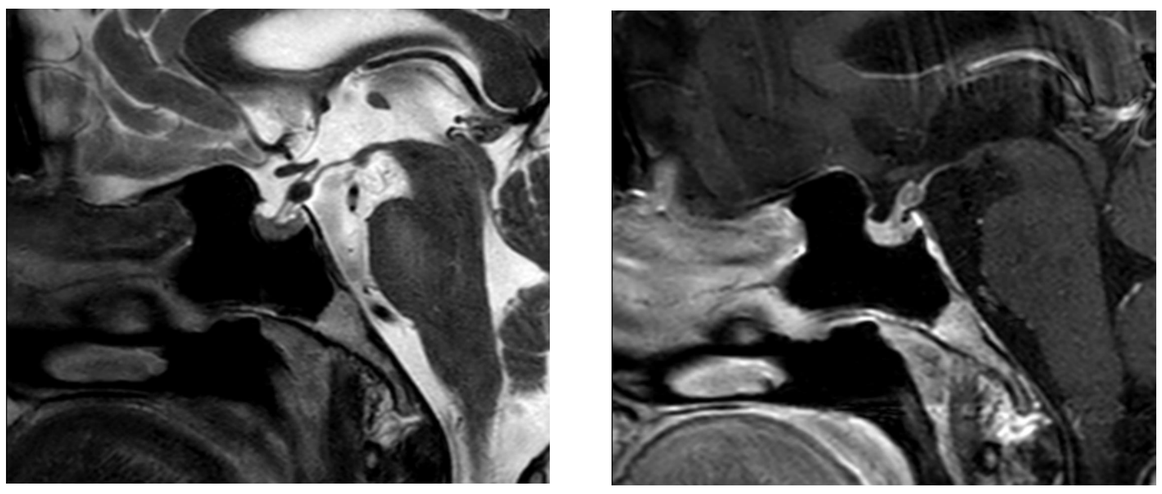
This demonstrates a complex clinical picture in which our patient presented with biochemical results suggesting central ACTH-dependent hypercortisolemia with no identifiable glandular lesion. The presence of an infundibular lesion led to surgical intervention which unfortunately did not result in biochemical cure despite adequate technical results. The authors believe this case illustrates a challenging clinical conundrum which emphasizes the uncertainty that should be associated with management of stalk lesions.

Background

Pituitary stalk lesions are rare entities with variable associated clinical manifestations. Their detection during work-up for endocrinologic abnormalities represents a challenging decision with respect to management. We present the case of a 57-year-old female with central ACTH-dependent hypercortisolemia who was discovered to have a pituitary stalk lesion treated surgically.

Case Description

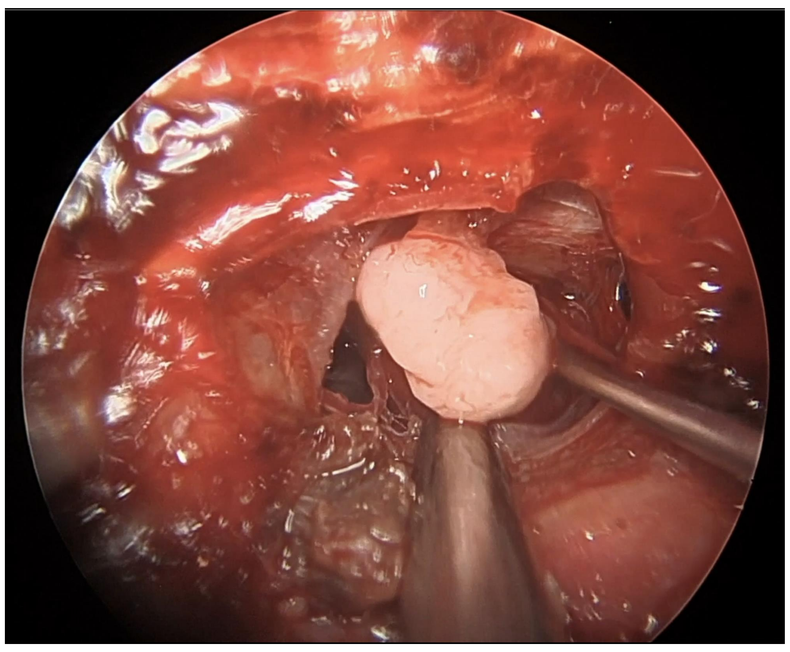
A 57-year-old female was referred to endocrinology for management of her diabetes. She was found to have a Cushingoid appearance and underwent subsequent biochemical workup which
demonstrated evidence of ACTH-dependent central Cushing's Syndrome (Cortisol post-low dose suppression 792 nmol/L; cortisol post-high dose suppression test 324 nmol/L). Imaging demonstrated a 4.7mm nodule involving the pituitary infundibulum.



4.7mm nodule originating from the pituitary stalk.

Management

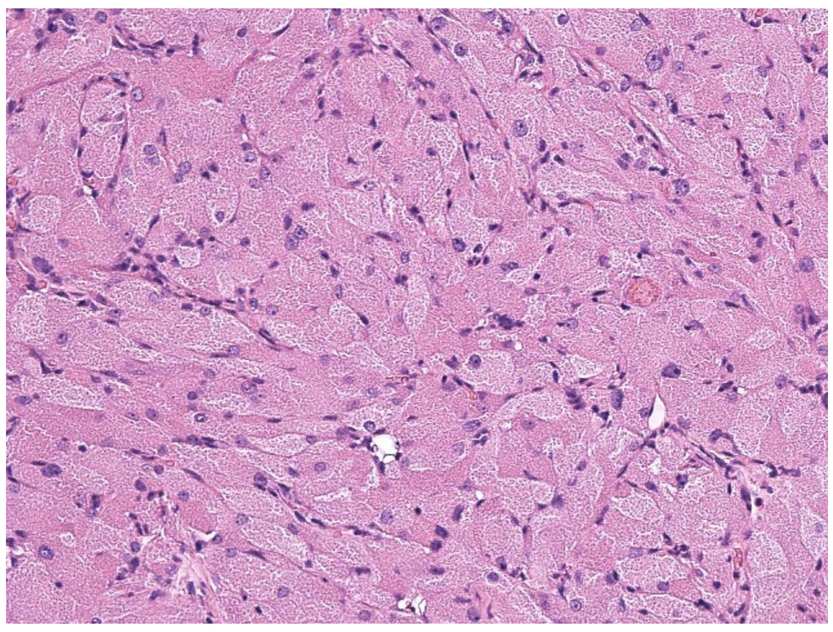
Inferior petrosal sinus sampling and PET-CT was considered, however, given the cortisol suppression seen on HDDST, presence of stalk lesion, and absence of any gland lesion on imaging, glandular pituitary and extracranial ectopic sources were thought to be unlikely. We elected to proceed with transsphenoidal transtubercular resection of the stalk lesion with goal of biochemical cure.



Endoscopic view demonstrating en bloc resection of the stalk lesion.

Outcome

A satisfactory technical and radiographic resection of the infundibular lesion was achieved. However, the patient's hypercortisolemia failed to resolve. Histopathologic analysis identified the lesion as a granular cell tumor. Subsequent inferior petrosal sinus sampling further demonstrated evidence of ACTH-dependent central Cushing's syndrome. Thus, this is a case of MRI-negative Cushing's disease.
With no clear surgical target, the patient was then considered for hypophysectomy as well as bilateral adrenalectomy. After discussion with the patient, they elected to proceed with bilateral adrenalectomy. The patient has continued 6-monthly MRI scans to monitor for growth of a presumed pituitary lesion.



Benign-appearing sheets of densely packed large polyhedral cells, with abundant eosinophilic cytoplasm, consistent with granular cell tumour.

Conclusion

Our case demonstrates a novel clinical picture in which our patient presented with biochemical results suggestive of central ACTH-dependent hypercortisolemia; however no glandular lesion identified on imaging. The presence of an infundibular lesion led to subsequent surgical intervention which unfortunately did not result in biochemical cure despite adequate technical results. The authors believe this case illustrates a challenging clinical picture which emphasizes the diagnostic uncertainty that should be associated with pituitary stalk lesions.

Contact

Patrick Toyota, MD
University of Saskatchewan
patrickrtoyota@gmail.com