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Case Report

Tiny Tumor with a Bigger Challenge: A Journey Through Uncertainty Using Neuro-Endoscopy in a Case of Recurrent Cushing Disease

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Abstract:

Cushing disease caused by ACTH-secreting pituitary microadenomas remains a surgical challenge, especially in recurrent cases with distorted anatomy. We report a case of a 35-year-old male with recurrent Cushing disease following unsuccessful initial transsphenoidal surgery. Re-operated using endoscopic endonasal transsphenoidal approach, this case illustrates the complexity of navigating scarred anatomical landmarks and ensuring complete tumor excision. The patient demonstrated marked clinical and biochemical improvement postoperatively, emphasizing the importance of meticulous surgical technique in reoperative endoscopic pituitary surgery(Cappabianca et al., 2014; Losa et al., 2000).

Keywords:

Cushing disease; ACTH-secreting pituitary adenoma; Pituitary microadenoma; Recurrent pituitary adenoma; Endoscopic endonasal transsphenoidal surgery; Reoperative pituitary surgery; Surgical anatomy distortion; Cortisol hypersecretion

Introduction:

Cushing disease, resulting from ACTH-producing pituitary adenomas, presents with systemic metabolic derangements due to hypercortisolemia. While endoscopic endonasal transsphenoidal surgery (EETS) is the gold standard(Cappabianca et al., 2014), recurrent or residual disease poses significant challenges, particularly due to distorted anatomy and fibrotic changes (Wang et al., 2015). Here we present a case of recurrent Cushing disease with complex surgical anatomy successfully treated via revision EETS.

Case Presentation

A 35-year-old male presented with progressive weight gain, generalized weakness, abdominal distension, and dark pigmentation of skin over the past two years. He had been recently diagnosed with type 2 diabetes mellitus and hypertension.

Initial Diagnosis and Management

Initial evaluation at a tertiary care center revealed elevated serum cortisol and ACTH levels. MRI pituitary protocol demonstrated a right-sided pituitary microadenoma.

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The patient underwent endoscopic endonasal transsphenoidal surgery. Despite the surgery, the patient continued to experience clinical features of hypercortisolism.

Initial Diagnosis and Management

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Referral and Re-evaluation

The patient was referred to our neurosurgical service by endocrinology colleagues for further management due to persistent disease symptoms.

Clinical Examination

- Pulse: 80 bpm
- BP: 130/80 mmHg
- BMI: 26.67 kg/m²
- General findings: Generalized hyperpigmentation, buffalo hump, pendulous abdomen with striae, proximal muscle weakness, thin skin, and easy bruising-classic features of Cushing syndrome (Patil et al,. 2008).

Investigations

- Serum Cortisol: 1073.69 nmol/L (elevated)
- ACTH: 124 pg/mL (elevated)
- High-dose dexamethasone suppression test: Positive
- Other pituitary hormones: Within normal range

Magnetic Resonance Imaging (MRI)-Pituitary protocol:

- MRI revealed right-sided pituitary microadenoma abutting the cavernous sinus (Figure 1A)
- There is evidence of previous surgical changes noted: absence of right middle turbinate, both superior turbinates, and posterior nasal septum. (Figure 1C)
- Anterior sphenoid sinus wall and sellar floor is absent (Orange arrow in figure 1B)
- Previous hadad flap and fat graft is well appreciated (Figure 1 B)

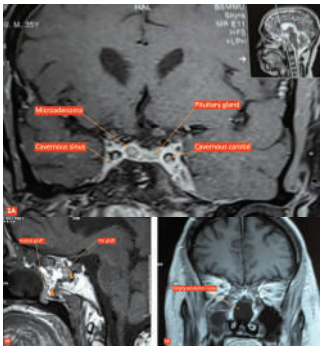

Figure 1: In figure 1A microadenoma is located on the right side of the gland, attached with the right cavernous sinus. Figure 1B and 1C is showing previous surgical changes.

Diagnosis

Recurrent Cushing Disease due to ACTH-secreting pituitary microadenoma with associated hypertension and diabetes mellitus

Management Plan

The case was discussed in a multidisciplinary team with endocrinology, and a decision was made to proceed with revision endoscopic endonasal transsphenoidal resection of the residual tumor.

Surgical Procedure

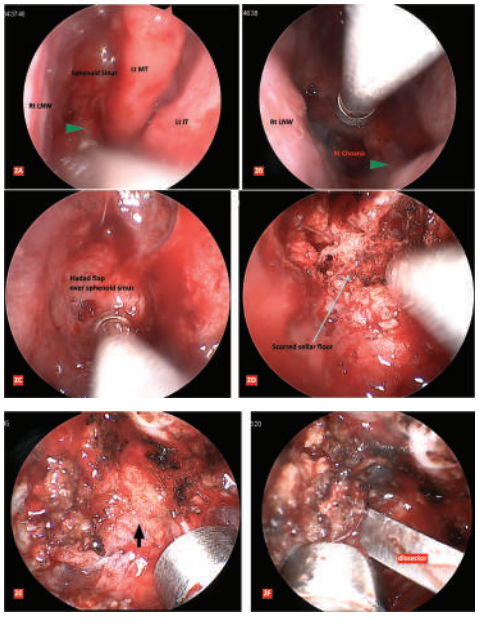

Figure 2 : Here nasal and sellar phases of the surgery have been demonstrated. In figure 2A there is no right middle turbinate and sphenoid ostium is absent. Left middle and Inferior Turbinates are well preserved (Lt MT and IT). Remnants of the posterior nasal septum are marked with green arrows in figure 2A and 2B. Right Choana can be well appreciated in figure 2B. Figure 2C and 2D reveals previous reconstructions with hadad flap and fat grafts respectively. There is scarred fat graft in figure 2E. Meticulous microdissection is demonstrated in figure 2F.

Nasal Phase:

- Anatomical landmarks were distorted from previous surgery(Figure 2A-2C)
- Absent middle turbinate (right), and posterior nasal septum (Figure 2A and 2B)
- We Used left middle turbinate (Lt MT) and arch of choana as orientation landmarks.
- Previous surgical trauma and scarring were evident in line with the findings described in other preoperative cases(Micko et al., 2015)

Sphenoidal Phase:

- Anterior sphenoidal wall, sphenoid osteum and rostrum were absent (Figure 2A, 2C)
- Scarred Hadad flap was encountered and carefully dissected.(Figure 2C)

Sellar Phase:

- Identified fibrous fat graft from prior reconstruction which was meticulously dissected away from the gland and tumour. (Figure 2D-2F)
- Sellar floor bone was absent
- We confirmed and localized cavernous carotids with Doppler which is essential for safety in revision surgery(Cappabianca et al., 2014).

Tumor Phase:

- Fibrotic tissue dissection revealed microadenoma on the right side of the gland, near cavernous sinus. The microadenoma was dirty greyish in color and normal gland was orange-white color(-Figure 3A).
- When an Incision over tumor was made with no-11 blade then tumour tissue started to float in the fluid(Figure 3B and 3C). It was suckable. Biopsy was taken with a pituitary rongeur.
- After complete removal of the tumor there was brisk cavernous bleeding ast the tumour was attached with the medial wall of the right cavernous sinus(Figure 3D)
- Complete tumor removal achieved with a minor arachnoid breach and CSF leak due to severe scarring(Figure 3F) which is a known complication in reoperations (Wang et al., 2015).
- After hemostasis tumor cavity and the normal gland is well appreciated (Figure 3E).

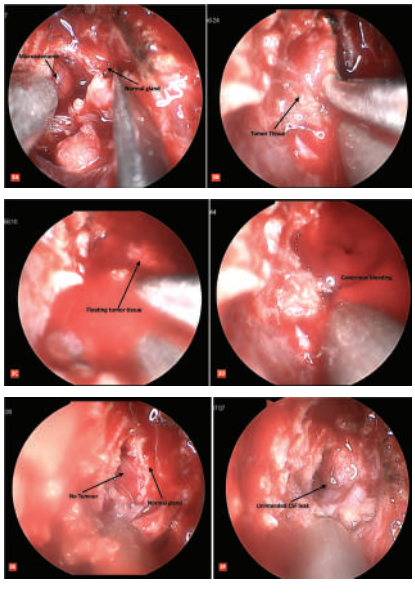

Figure 3: Various stages of tumour removal have been demonstrated in figure 3A-3F.

Reconstruction

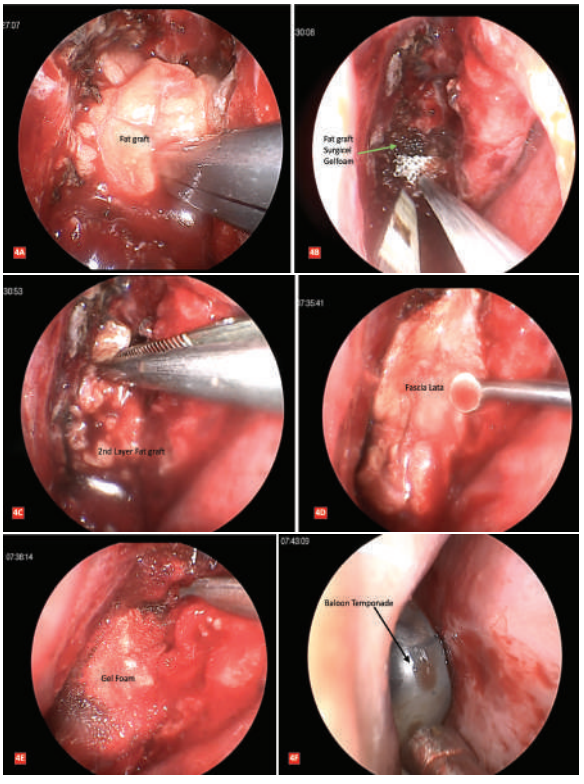

Figure 4: Different stages of sellar floor reconstructions have been demonstrated in figure 4A-4F.

- Hemostasis was achieved with gel foam. Fat graft that was harvested from the right thigh was used for sellar floor reconstruction in 2 layers(Figure 4A-4C)
- Then fascia lata was used to cover the former reconstructions(Figure 4D)
- Subsequently gel foam along with balloon tamponade with a 12FR foley's catheter was used to support the reconstructions(Figure 4E and 4F)
- Nasal cavity was packed with merocele.

Postoperative Outcome

- Recovery was uneventful, with no persistent CSF leak.
- Hormonal evaluation showed improvement with serum cortisol reduced to 335.91 nmol/L and ACTH to 87.32 pg/mL.
- Follow-up MRI showed no residual tumor (Figure 5).
- Patient showed clinical improvement and is under joint follow-up with endocrinology and neurosurgery.

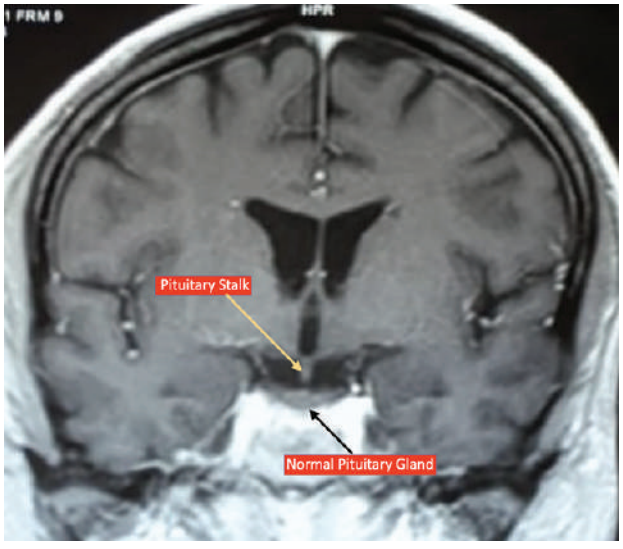

Figure 5: Postoperative MRI after 2nd surgery revealed normal pituitary gland. There is no residual or recurrent tumor.

Discussion

Recurrent pituitary adenomas pose a unique challenge due to anatomical alterations and fibrosis and proximity to critical structures like cavernous sinus (Micko et al., 2015). This case underlines the need for advanced surgical navigation and experience in reoperative endoscopy (Cappabianca et al., 2014). Identifying substitute landmarks and cautious dissection near critical structures like the cavernous carotids and arachnoid, a bloodless field is essential for safe and effective revision surgery (Losa et al., 2000). Although technically demanding, reoperative EETS can yield excellent outcomes in experienced hands.

Conclusion

Even a "tiny" pituitary microadenoma can present a "big challenge" during reoperative endoscopic surgery. This case highlights the technical and anatomical challenges in managing recurrent Cushing disease.With thorough understanding of skull base anatomy, surgical expertise, anatomical knowledge, and multidisciplinary management excellent outcomes are achievable for recurrent Cushing disease (Wang et al., 2015; Micko et al., 2015).

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