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A Gigantic Heterogenous Silent Corticotroph Pituitary Neuroendocrine Tumour With Posterior Fossa Cyst and a Fibrotic Wall—Diagnostic Mimicry and Route-Tailored Staging: A Case Report

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Case Report

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Abstract

Background: Neurosurgical route selection for treating silent corticotroph pituitary neuroendocrine tumours is complicated because of their nature of mimicking aggressive skull-base disease.

Case presentation: A 21-year-old man with a background of autism spectrum disorder presented with headache, diplopia, right third cranial nerve palsy and visual blurring. A giant heterogeneous sellar–suprasellar mass with cavernous sinus/internal carotid artery involvement and a posterior fossa cyst compressing the brainstem is shown on MRI. He has a normal baseline pituitary test result. Endoscopic endonasal transsphenoidal debulking achieved diagnosis and decompression; histology confirmed a silent corticotroph pituitary neuroendocrine tumour (adrenocorticotropic hormone-positive, limited T-box pituitary transcription factor. Post-operative diabetes insipidus occurred. On surveillance, the cyst enlarged with hydrocephalus. Two Ommaya attempts failed due to a taut, fibrotic wall. Right craniotomy enabled lateral debulking and direct cyst cannulation with effective decompression. Residual cavernous/internal carotid artery-adjacent disease was treated with proton beam therapy (50.4 Gy/28 fractions) without notable acute toxicity. Bulbar function and third-nerve palsy improved post-craniotomy; gait and speech improved after radiotherapy. Imaging showed reduced brainstem compression with a manageable residual; visual acuity remained 6/21 right, 6/6 left.

Conclusions: This case supports route-tailored staging-endonasal decompression, early conversion to transcranial access for fibrotic, catheter-resistant cysts, and consolidative proton therapy-and highlights the value of autism spectrum disorder-adapted pathways and multidisciplinary team oversight.

Background

Pituitary neuroendocrine tumours (PitNETs) constitute roughly fifteen percent of intracranial tumours [1]. The 2022 World Health Organisation (WHO) classification emphasises lineage-based diagnosis, and silent corticotroph PitNETs are recognised for a greater propensity for invasion and recurrence than many non-functioning tumours [2]. With the aetiology of PitNETs/Pituitary Adenomas remaining unknown, most are sporadic, as familial cases comprise only 5% of all pituitary tumours [3], with more than fifty percent of those being attributed to multiple endocrine neoplasia type 1 syndrome [4].

Case Presentation

We report a case of a 21-year-old male who initially presented with a month's history of right-sided headache, diplopia and third nerve palsy as well as blurred vision in the right eye. Considering the history of autism spectrum disorder, the consultations and consent process were tailored according to his needs. Baseline pituitary tests, including prolactin and morning cortisol, were in the normal range. Table 1 provides a chronological summary of the patient's diagnostic timeline, staged operative interventions, and adjuvant radiotherapy.

Table 1
Comprehensive summary of patient's diagnostic timeline, staged operative intervention, and adjuvant radiotherapy

Timeline stage	Event or intervention	Key clinical and pathological details
Initial presentation (Day 0)	Presentation and baseline imaging	21-year-old male presenting with right headache, diplopia, right third nerve palsy, and visual blurring. MRI showed giant sellar–suprasellar mass with cavernous/ICA involvement and posterior fossa cyst compressing the brainstem. Normal baseline pituitary function.
Primary surgery in 2 months after initial presentation	Endoscopic endonasal transsphenoidal surgery	Central sellar debulking for optic decompression. Inseparable lateral/intracavernous tissue left *in situ*. Pathology confirmed silent corticotroph PitNET (Ki-67 < 3%)
Post-operative inpatient period	Transient diabetes insipidus	Developed postoperative diabetes insipidus managed with desmopressin and discharged at 1 week
First follow up 6 months after initial presentation	Neuro-ophthalmology review	Persistent right-sided visual blurring and confirmed third nerve palsy
Second follow up 9 months after initial presentation	Surveillance MRI	Posterior fossa cyst enlargement with new clinical and radiological hydrocephalus.
Symptom progression 11 months after initial presentation	Clinical deterioration and multidisciplinary team meeting	Rapid onset of gait disturbance and bulbar and speech symptoms over 2 months. MDT recommended stereotactic Ommaya placement.
Minimal invasive attempts 11 months after initial presentation	First Ommaya attempt	Catheter deflected off a taut, fibrotic cyst wall which result in failed cavity enter
9 days after first Ommaya attempt	Second Ommaya attempt	The non-compliant fibrotic wall lead to fail repeat attempt
Second surgery a week after second Ommaya attempt	Right open craniotomy	Open lateral debulking and direct under-vision cyst cannulation. Pathology confirmed stable TPIT corticotroph PitNET (Ki-67 < 3%). Post-operative MRI showed reduced brainstem compression.
Immediately post-craniotomy	Neurological recovery	Improved bulbar function and third nerve palsy and his visual acuity stable with 6/21 in his right eye and 6/6 in his left eye.
Adjuvant therapy 12 to 13 months after initial presentation	Proton beam therapy	Delivered 50.4 Gy in 28 fractions to cavernous/ICA residual without acute toxicity
4 weeks after proton beam therapy	Clinical review shows post-proton beam therapy recovery	Significant improvement in gait and speech clarity

Timeline stage	Event or intervention	Key clinical and pathological details
Final surveillance 20 months after initial presentation which is about 9 months after craniotomy	Follow-up MRI	Marked interval reduction of residual mass and stable decompression of posterior fossa

Initial MRI scan showed a giant enhancing lesion centred at the sella, suprasella and cavernous sinus area with cystic components involving the posterior fossa with brainstem compression (Fig. 1).

Endoscopic Transnasal Approach

A discussion in the dedicated pituitary multidisciplinary team (MDT) meeting raised the possibility of haemangiopericytoma or solitary fibrous tumour, craniopharyngioma and PitNET, with meningioma or chordoma/chondrosarcoma regarded as less likely. It was decided in MDT to pursue a possible staged approach to this lesion. Two months after presentation, the patient underwent an endoscopic endonasal transsphenoidal approach with intra-operative frozen section and measured debulking, leaving behind the lateral and intracavernous segments. Histology reported a PitNET with synaptophysin and adrenocorticotropic hormone (ACTH) immunoreactivity but limited TPIT expression.

The patient first underwent an endoscopic endonasal transsphenoidal decompression because the dominant disease burden was sellar–suprasellar with optic apparatus compromise. After tumour debulking, the sellar dura was reconstructed with a Neuro-patch underlay and Tisseel sealant, and no intra-operative cerebrospinal fluid leak was encountered. This is relevant, as subsequent postoperative diabetes insipidus is more plausibly attributed to manipulation of the pituitary–stalk region than to a reconstruction-related leak. Intraoperatively, the lesion appeared heterogeneous, with a softer central/sellar component and firmer, more adherent tumour laterally and in the cavernous region, consistent with the decision to leave intracavernous disease in situ for staged management. The more indurated elements also correlate with later difficulty in percutaneously traversing the cyst wall.

Post-operatively, the patient developed Diabetes Insipidus, necessitating desmopressin and was discharged a week later with ongoing right-sided blurry vision and third nerve palsy. This was confirmed in a Neuro-ophthalmology review four months after the initial endoscopic procedure. Seven months after the endoscopic procedure, follow-up MRI showed that the posterior fossa cyst had grown, and the patient was showing signs of hydrocephalus, radiologically and clinically (Fig. 2).

Cyst Decompression with an Ommaya Reservoir

Over the next two months, the patient's symptoms progressed with speech problems and severe gait disturbance, at which point the MDT recommended cyst decompression with an Ommaya reservoir. Figure 3 shows the increase in cyst size and hydrocephalus over the two months.

Progressive enlargement of the posterior fossa cyst and evolving hydrocephalus prompted an attempt at minimally invasive control by stereotactic Ommaya reservoir placement. This was undertaken twice in nine days. The catheter was advanced to the expected depth but met firm resistance at the presumed cyst wall and deflected tangentially on both occasions. Thus, failing to enter the cavity. Despite accurate stereotactic targeting, this behaviour does not permit easy catheter penetration, which is typical of long-standing or low-compliance cysts with a fibrotic lining.

This supports an operative principle for similar lesions: failure of catheter entry due to a taut or fibrotic wall should be regarded as an early threshold for abandoning percutaneous drainage and proceeding to direct/open cannulation rather than persisting with repeated stereotactic attempts.

The cyst was growing, and the MDT therefore recommended an open approach.

Open Right Craniotomy

A week later, the patient underwent a right craniotomy for lateral tumour debulking and direct cannulation of the posterior fossa cyst, with good decompression. Histology also confirmed a PitNET after this operation. Post-operatively, the patient's bulbar function and third-nerve palsy improved, whilst his visual acuity remained 6/21 on the right and 6/6 on the left. Post-op MRI showed reduced brainstem compression with 59 × 37 × 35 mm residual (Fig. 4).

Pathology

Histopathological assessment at two distinct operations confirmed a stable T-box pituitary transcription factor (TPIT)-lineage corticotroph PitNET. The first specimen (obtained 2 months after initial presentation) showed scant to patchy TPIT expression, diffuse synaptophysin, predominant ACTH immunoreactivity, Ki-67 < 3%, and patchy cytokeratin. Scattered pituitary-specific positive transcription factor 1/growth hormone–positive cells were best regarded as entrapped normal adenohypophyseal tissue rather than evidence of plurihormonal differentiation. The second specimen (obtained during the open craniotomy) demonstrated an essentially identical profile—scattered TPIT, ACTH positivity, Ki-67 < 3% — indicating no interval dedifferentiation and no rise in proliferative activity. This pattern fits within the 2022 WHO description of corticotroph tumours that may behave invasively despite modest proliferation.

Interpretation, however, is constrained by several factors. Sampling was necessarily limited at both procedures and cannot absolutely exclude focal areas of higher grade; and a low Ki-67 in a corticotroph tumour does not, by itself, guarantee indolent behaviour when there is already cavernous or clival involvement. In large and topographically complex silent corticotroph PitNETs, histology should be read alongside imaging and operative findings, and repeat tissue confirmation at the time of transcranial or alternative-route surgery is worthwhile to demonstrate biological stability.

Proton Beam Therapy

The patient underwent proton beam therapy to 50.4 Gy in twenty-eight fractions, and notable acute toxicity is absent. The patient was symptomatically better with a steadier gait and clearer speech four weeks after completion.

Follow-up

Immediate post-craniotomy MRI demonstrated reduced brainstem compression and a smaller posterior fossa component. Surveillance MRI approximately nine months after craniotomy and following delivery of proton beam therapy showed further interval reduction of the heterogeneous enhancing mass, consistent with treatment effect.

Discussion and Conclusions

This case shows that a large pituitary lesion with challenging skull-base anatomy may not be amenable to a single-stage treatment. The mass was centred in the sellar–suprasellar region and produced chiasmal compression, so an endoscopic endonasal transsphenoidal operation was selected first to obtain histology and decompress vision, consistent with standard practice for symptomatic pituitary tumours [1]. Histology later confirmed a pituitary neuroendocrine tumour, silent from a clinical standpoint but in keeping with established pituitary tumour biology and the spectrum of non-functioning lesions described in endocrine series [2].

Posterior fossa involvement arising from a pituitary lesion is unusual and is more often described in very large adenomas that expand backwards and ultimately require an open cranial route because the posterior fossa allows little room for minimally invasive manoeuvres [5]. In the present case, the posterior component was a distinct, relatively non-compliant cyst in continuity with the sellar tumour. A further point is that silent corticotroph tumours have been reported to invade laterally, inferiorly and into clival or sphenoidal compartments more than other non-functioning subtypes, so an extension along less typical pathways is biologically plausible in this lineage [6].

Once the posterior fossa cyst became the main driver of morbidity — hydrocephalus, gait disturbance, bulbar symptoms — stereotactic Ommaya placement was attempted. This choice reflects experience with cystic sellar and parasellar lesions, where a thin, compliant wall allows repeat aspiration and occasionally intracystic therapy and can defer craniotomy [7]. In this patient, the catheter twice met a taut, probably fibrotic wall and failed to enter the cavity, so the reservoir never functioned. When catheter passage is prevented in this way, reports of open or supraorbital approaches for Ommaya placement show that direct visualisation of the wall permits deliberate entry and reliable drainage [8]. Reviews of cystic sellar lesions also point out that thicker or long-standing cyst walls are less amenable to percutaneous drainage and are better managed through a surgically controlled access route than by repeating stereotactic passes [9].

Because of persistent posterior fossa mass effect, a right craniotomy was undertaken to provide lateral access for tumour debulking and direct cyst cannulation under vision. This relieved brainstem compression and improved bulbar function and the third-nerve palsy, although a subtotal resection was expected because tumour remained around the cavernous sinus and internal carotid artery (ICA). At that stage, the aim changed from complete removal to durable control. Proton beam therapy to 50.4 Gy in 28 fractions was delivered to consolidate control of the skull-base residual close to the optic apparatus and brainstem, a strategy that is consistent with the 2022 WHO update, which recognises pituitary neuroendocrine tumours that, despite modest proliferation, benefit from multimodal management and close surveillance [2]. It is also in line with endoscopic series on giant pituitary adenomas that acknowledge the need for complementary routes or adjuvant treatment when extension is lateral, posterior or irregular and cannot be cleared safely through a single corridor [10]. Procedural reports on Ommaya use in other intracranial cystic lesions further support this escalation pattern: when a catheter cannot traverse the wall, durable control is most often achieved after definitive access followed by radiosurgical or radiotherapeutic treatment [11].

Read this way, the sequence is linear. Start endonasally for a sellar–suprasellar epicentre and visual compromise [1, 10]. If a posterior cystic component becomes dominant and stereotactic access fails because of a rigid wall, convert to a transcranial route rather than repeating ineffective percutaneous attempts [8, 9]. Once the residual tumour is fixed to cavernous or ICA structures, plan adjuvant conformal or proton radiotherapy early to secure long-term control in accordance with current pituitary tumour classification and skull-base experience [2, 10, 11].

This report delineates a rare presentation of a silent corticotroph PitNET with a posterior fossa cyst whose fibrotic wall defeated stereotactic catheterisation. A route-tailored sequence proved decisive. Endonasal decompression secured the diagnosis and relieved chiasmal pressure. Failure of percutaneous entry signalled low cyst compliance and justified timely conversion to a transcranial corridor for direct cannulation and lateral debulking. Proton therapy then consolidated control at the cavernous and carotid interface, where further resection would have risked unacceptable morbidity. The patient improved neurologically and radiologically, with a durable reduction in posterior fossa mass effect.

The clinical value lies in the decision point it defines. When accurate stereotactic targeting still cannot traverse a tense cyst wall, repeated attempts add risk without benefit. Surgeons should treat that finding as an early threshold for open or direct access, rather than an invitation to persist. Invasive behaviour in silent corticotroph disease may reflect anatomy more than proliferation, so multimodal planning should be explicit from the outset. Applied thoughtfully, this strategy preserves function, curbs delay and provides a reproducible pathway for similarly complex skull base tumours.

Abbreviations

• ACTH

Adrenocorticotropic hormone
• CT
Computed tomography
• Gy
Grey
• ICA
Internal carotid artery
• MDT
Multidisciplinary team
• MRI
Magnetic resonance imaging
• PitNET
Pituitary neuroendocrine tumour
• TPIT
T-box pituitary transcription factor
• WHO
World Health Organisation

Declarations

Ethics approval and consent to participate

Ethical approval was waived as this study describes a single case report with patient consent for publication.

Consent for publication

Written informed consent for the publication of his clinical data and images was obtained from the patient.

Competing interests

The authors declare no competing interests.

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Author Contribution

B.R. wrote the original draft. O.C., A.S., S.C., J.P., D.D., and A.G. are all involved in the editing of the manuscript. A.S. also involve in editing manuscript and supervision as well. All the authors read and approved the final version.

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Availability of data and materials

All data generated or analysed during this study are included in this published article.

References

1. Molitch ME. Diagnosis and Treatment of Pituitary Adenomas. JAMA: The Journal of the American Medical Association 2017; 317: 516–516.
2. Casar-Borota O, Burman P, Lopes MB. The 2022 WHO classification of tumors of the pituitary gland: An update on aggressive and metastatic pituitary neuroendocrine tumors. Brain Pathology; 35. Epub ahead of print 2024. DOI: 10.1111/bpa.13302.
3. Vandeva S, Jaffrain-Rea M-L, Daly AF, et al. The genetics of pituitary adenomas. Best Practice & Research Clinical Endocrinology & Metabolism 2010; 24: 461–476.
4. Tichomirowa MA, Daly AF, Beckers A. Familial pituitary adenomas. Journal of Internal Medicine 2009; 266: 5–18.
5. Shkarubo AN, Shishkina LV, Serova NK, et al. Endoscopic endonasal surgical treatment of large pituitary adenoma, spreading into the posterior fossa. Zhurnal voprosy neĭrokhirurgii imeni N N Burdenko 2015; 79: 85–85.
6. Himstead AS, Wells AC, Kurtz JS, et al. Silent Corticotroph Adenomas Demonstrate Predilection for Sphenoid Sinus, Cavernous Sinus, and Clival Invasion Compared with Other Subtypes. World Neurosurgery 2024; 191: e41–e47.
7. Chen A, Ai M, Sun T. Advances in the treatment of Adamantinomatous craniopharyngioma: How to balance tumor control and quality of life in the current environment: a narrative review. Frontiers in Oncology; 13. Epub ahead of print 2023. DOI: 10.3389/fonc.2023.1326595.
8. Greuter L, Richards OL, Malik N, et al. Supraorbital minicraniotomy for open Ommaya reservoir placement in pediatric craniopharyngiomas: a case series and technical report. Journal of Neurosurgery: Pediatrics 2023; 32: 421–427.

9. Schmutzer-Sondergeld M, Quach S, Niedermeyer S, et al. Risk-benefit analysis of surgical treatment strategies for cystic craniopharyngioma in children and adolescents. Frontiers in Oncology; 14. Epub ahead of print 2024. DOI: 10.3389/fonc.2024.1274705.
10. Koutourousiou M, Gardner PA, Fernandez-Miranda JC, et al. Endoscopic endonasal surgery for giant pituitary adenomas: advantages and limitations. Journal of Neurosurgery 2013; 118: 621–631.
11. Lv J, Wu Z, Wang K, et al. Case Report: Clinical and Procedural Implications of Ommaya Reservoir Implantation in Cystic Brain Metastases Followed by Radiosurgery Treatment. Frontiers in Surgery; 9. Epub ahead of print 2022. DOI: 10.3389/fsurg.2022.901674.

Figures

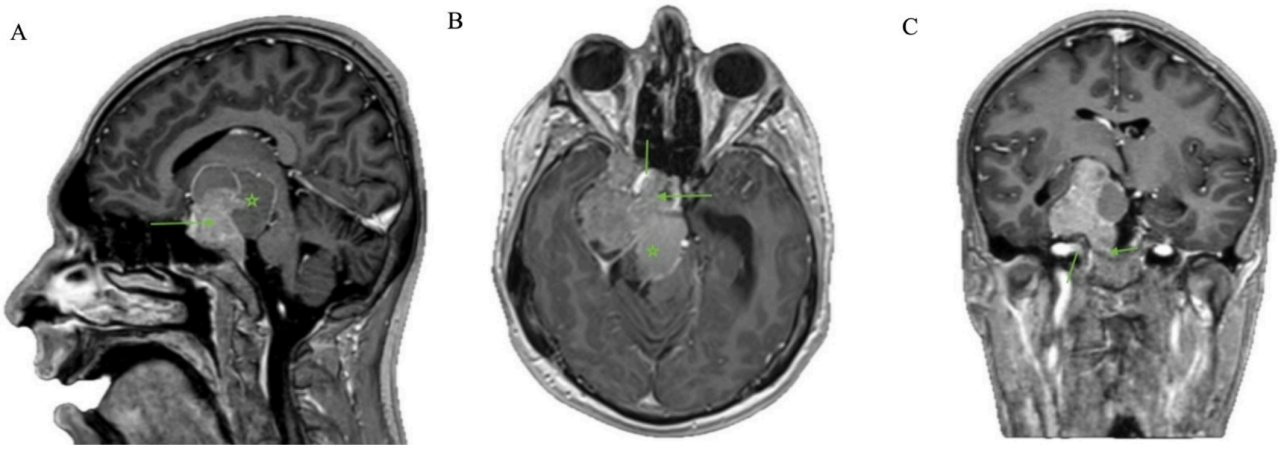


Figure 1

MRI showing initial morphology of pituitary lesion ((a) – sagittal, (b) axial and (c) coronal)

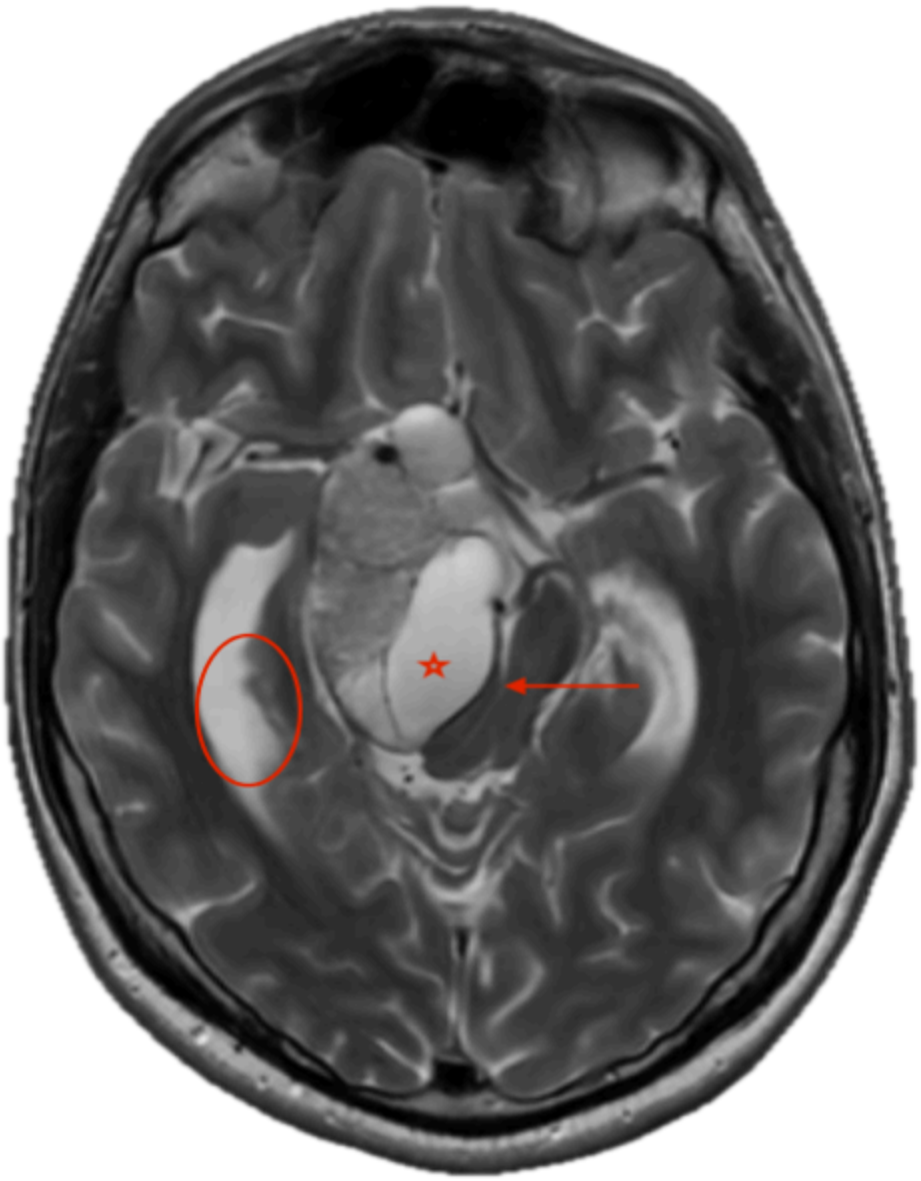


Figure 2

Follow-up MRI showing signs of hydrocephalus

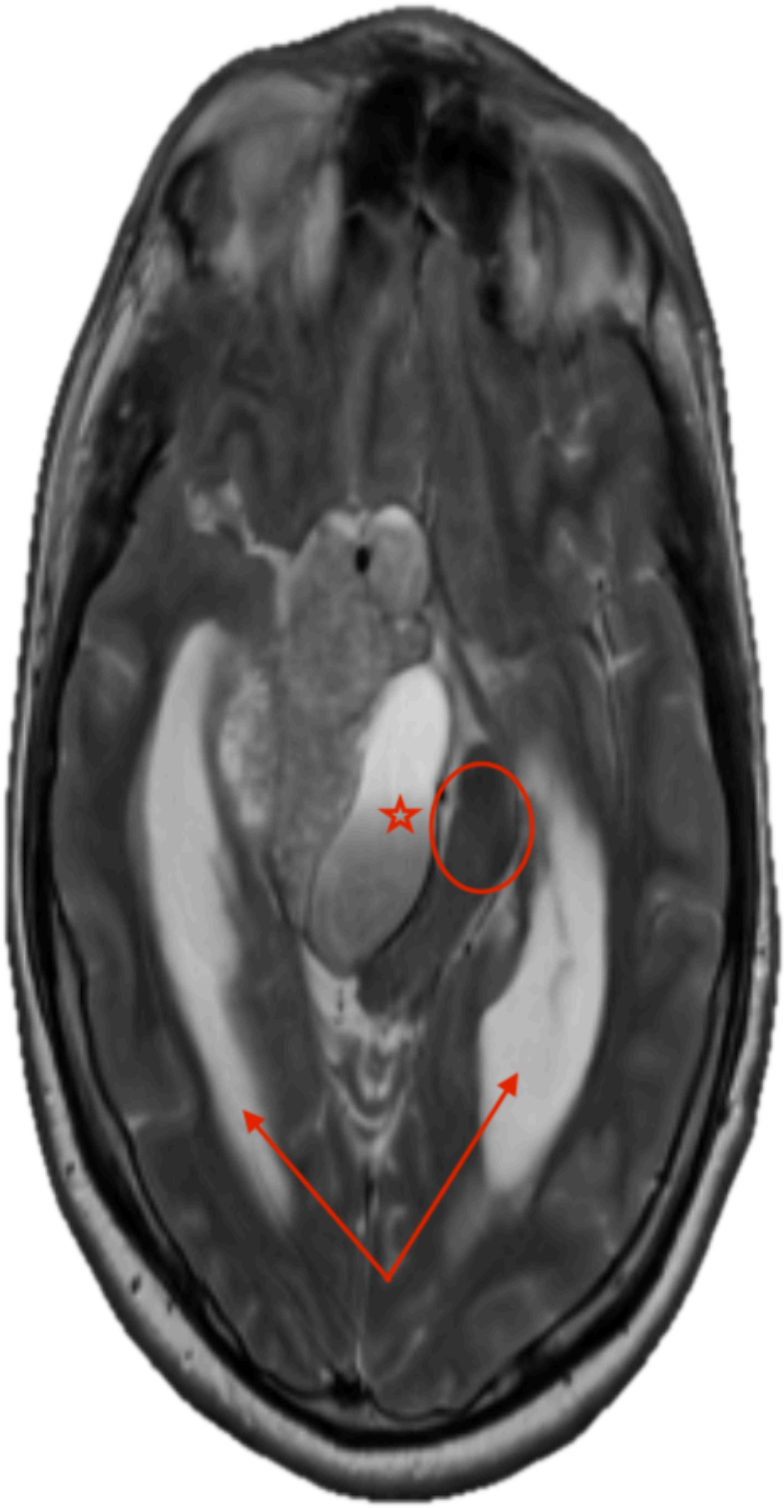


Figure 3

Interval T2-weighted MRI showing an increase in cyst size with an enlargement of the temporal horns, indicating hydrocephalus

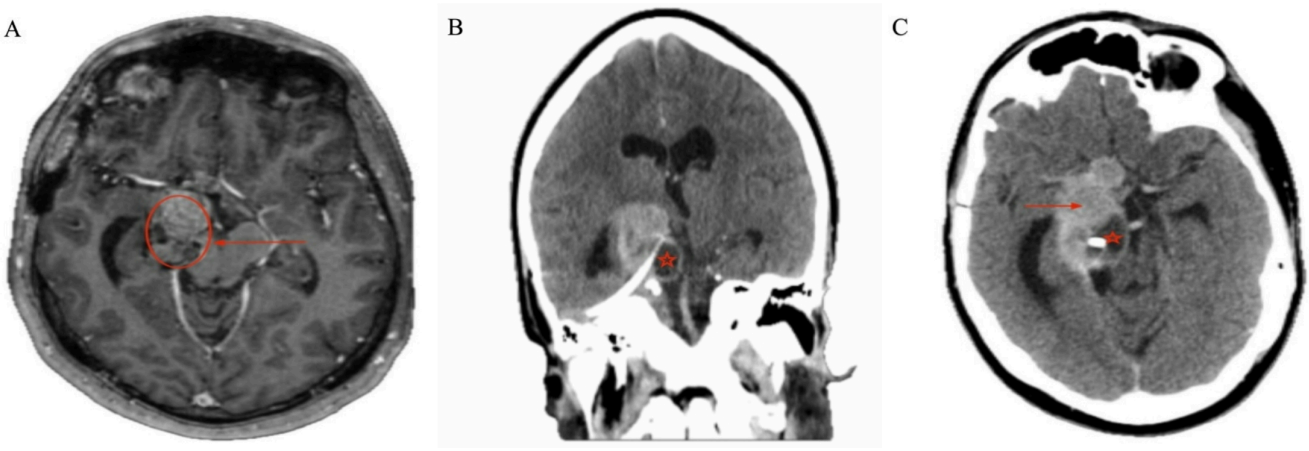


Figure 4

Post-operative craniotomy imaging: (a) axial MRI, (b) coronal CT, and (c) axial CT.