Patankar and Chaudhary
Egyptian Journal of Neurosurgery (2022) 37:21
https://doi.org/10.1186/s41984-022-00158-w

Egyptian Journal of Neurosurgery

CASE REPORT Open Access

An entirely suprasellar Rathke's cleft cyst: a rare case report with review of literature

Amey P. Patankar[1*] and Shivani Chaudhary[2]

Abstract

Background: A Rathke's cleft cyst is a benign intrasellar cystic lesion, with suprasellar extension reported in a few cases. An entirely suprasellar Rathke's cleft cyst without any intrasellar component, though previously reported, is rare.

Case presentation: We report a case of an entirely suprasellar Rathke's cyst in a 25-year-old female presenting with headache and visual complaints. The patient was treated surgically by drainage of the cyst and removal of the cyst wall by transcranial approach. Histopathology and immunohistochemistry confirmed the diagnosis of Rathke's cyst. We review the relevant literature and discuss its neuroembryologic pathogenesis, pathology and the factors influencing the recurrence of the cyst.

Conclusions: To the best of our knowledge, only 62 cases of an entirely suprasellar Rathke's cyst have been reported till date.

Keywords: Rathke's cleft cyst, Suprasellar Rathke's cyst, Suprasellar tumours, Rathke's pouch

Background

Rathke's cleft cysts are predominantly intrasellar lesions originating from Rathke's cleft in the vestigial pars intermedia of the pituitary [1]. We report a case of an exclusively suprasellar Rathke's cyst and review the relevant literature.

Case presentation

A 25-year-old female presented with headache and blurring of vision since 7 months. CT scan of the brain showed a hyperdense lesion in the suprasellar region (Fig. 1). MRI of the brain revealed a cystic lesion in the suprasellar area, mildly hyperintense on T1 images (Fig. 2), hyperintense on T2 and flair images (Figs. 3, 4) and with only mild peripheral enhancement on post-contrast images (Fig. 5) The lesion was entirely suprasellar without any intrasellar component, as can be seen in the coronal sections (Fig. 6). Pituitary gland was visible separately from the lesion in the sella. Based on the MRI findings, a preoperative diagnosis of Rathke's cleft cyst or craniopharyngioma was considered [2–4]. Perimetry showed bilateral temporal superior arcuate visual field defect (Figs. 7, 8) (Table 1).

The patient underwent surgery by transcranial approach. The patient was operated under general anaesthesia in the supine position. The right frontal craniotomy and sub-frontal approach was used to reach the inter-optic space. The cyst was visible in the inter-optic pre-chiasmal space. The cyst wall was punctured with micro-knife. White gelatinous fluid, thick in consistency, drained out. The cyst wall was removed and sent for biopsy. The pituitary stalk was well visualised and intact. Biopsy from the cyst wall was sent for histopathological examination and immunohistochemistry.

The post-operative period was uneventful, and the patient did not develop diabetes insipidus. The patient

*Correspondence: docapp@icloud.com

[1] Neuron Hospital, Vadodara, Gujarat, India
Full list of author information is available at the end of the article

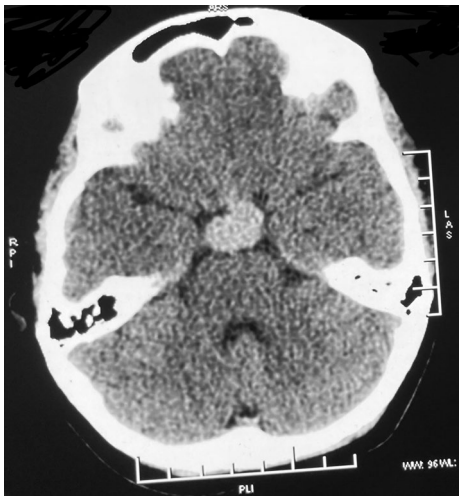

Fig. 1 CT scan of the brain showing hyperdense lesion in the suprasellar area

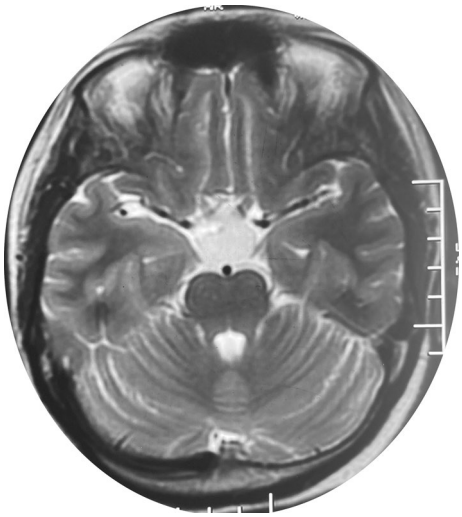

Fig. 3 MRI brain T2 axial image showing the hyperintense suprasellar lesion

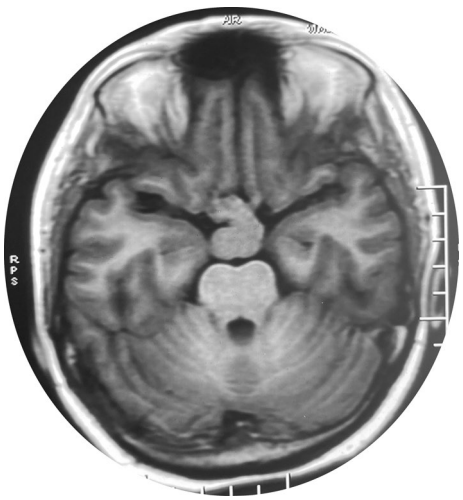

Fig. 2 MRI brain T1 axial image showing slightly hyperintense lesion in the suprasellar area

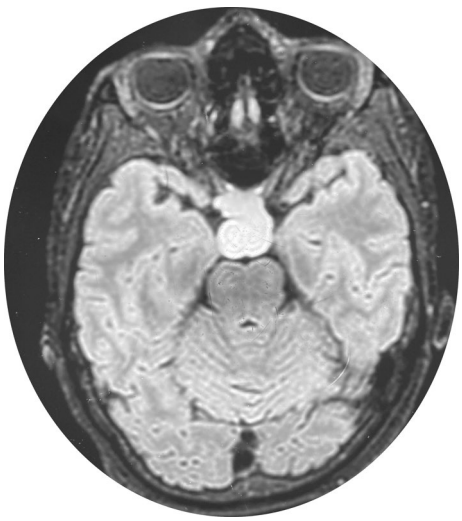

Fig. 4 MRI brain flair sequence showing the hyperintense suprasellar lesion

was discharged on the fifth post-operative day. Sutures were removed on the tenth post-operative day.

Post-operative MRI confirmed the completeness of cyst removal (Figs. 9, 10). Histopathology showed columnar and cuboidal epithelium lining the cyst wall with empty cyst spaces (Fig. 11). Squamous metaplasia of the epithelium lining was seen at some places (Fig. 12). Immunohistochemistry showed positivity for pankeratin (Figs. 13, 14) and epithelial membrane antigen (EMA) (Figs. 15, 16) supporting the diagnosis of Rathke's cleft cyst. Overlapping histological features between Rathke's cyst and craniopharyngioma has led

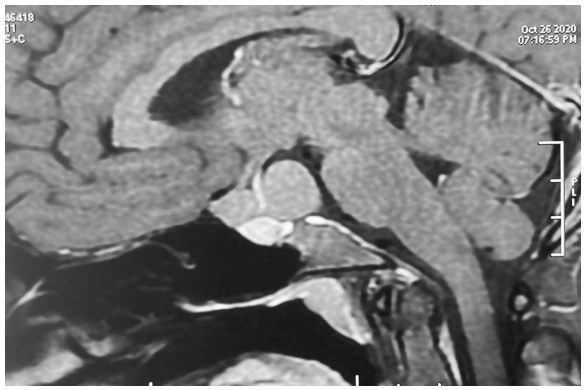

Fig. 5 MRI brain T1 contrast image showing mild peripheral enhancement of the lesion

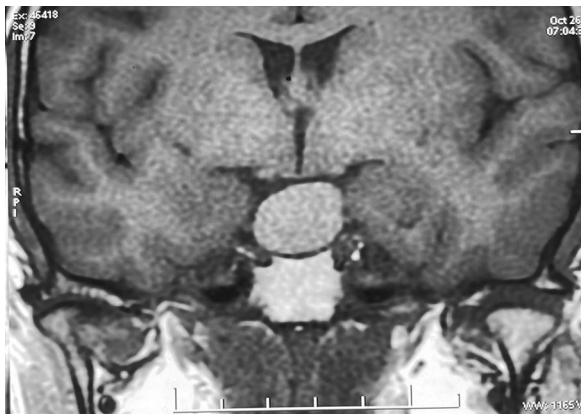

Fig. 6 MRI brain T1 coronal image showing the entirely suprasellar location of the cystic lesion

the pathologists to believe that these lesions constitute the two ends of a continuum of a spectrum of ectodermal derivatives.

Follow-up perimetry after 6 months showed complete resolution of the scotomas in both the eyes (Figs. 17, 18).

Discussion

Though Rathke's cysts are found in about 13–33% of unselected post-mortem cases [5], they account for only 2–9% [6] of patients undergoing transsphenoidal surgery for symptomatic sellar lesions. This discrepancy is because the majority of the patients with asymptomatic lesions do not require any intervention throughout their life.

The peak incidence of the cyst is between 30 and 50 years of age, with a female predominance in the ratio of 2:1 [7]. The female predominance may be secondary to an increased awareness of disturbed endocrinological function manifest as menstrual irregularities. No racial or genetic predilection exists in the pathogenesis of Rathke's cleft cyst (RCC).

The cyst contains mucoid or gelatinous material encapsulated in a thin cyst wall of simple or pseudo-stratified cuboidal or columnar epithelium.

Neuroembryologically, Rathke's cleft cysts are thought to be derived from Rathke's pouch, which appears in the 3rd or 4th week of embryonic life as an outgrowth of stomodeum which elongates dorsally to form the craniopharyngeal duct. Proximal end obliterates by 11th week, and the cranial end comes in contact with infundibulum which is a downgrowth of neuroepithelium from the diencephalon (forms the posterior lobe of pituitary). Anterior wall proliferates to form anterior lobe of pituitary, and posterior wall forms pars intermedia. The residual lumen is reduced to a narrow cleft and regresses later. Persistence and enlargement of this cleft cause symptomatic Rathke's cleft cysts [8]. Rathke's pouch cells predominantly spread through sellar and suprasellar region, but few cells can be located outside the sella turcica. Failure of ectopic pouch remnants beyond the pituitary fossa to regress may lead to formation of a symptomatic Rathke's cyst outside the sella.

The incidence of DI following surgery for Rathke's cyst is reported to be between 3 and 19% and is more likely when radical cyst wall excision is attempted. The incidence of DI decreased from 42 to 9% when less radical cyst wall excision was done [9].

The reported recurrence rate of Rathke's cyst varies from 3 to 33%. The factors which influence the recurrence rate include the enhancement pattern of the cyst wall, the presence of squamous metaplasia, chronic inflammation or stratified epithelium, the aggressiveness of cyst wall resection and the insertion of an abdominal fat graft. Some authors advocate that the histology is a major predictor of recurrence rather than the surgical technique [10].

In the series of 118 patients, Aho and colleagues [9] reported a recurrence rate of 18% with 32% radiological recurrence in 38 patients with squamous metaplasia, while only 11% of those without metaplasia showed recurrence.

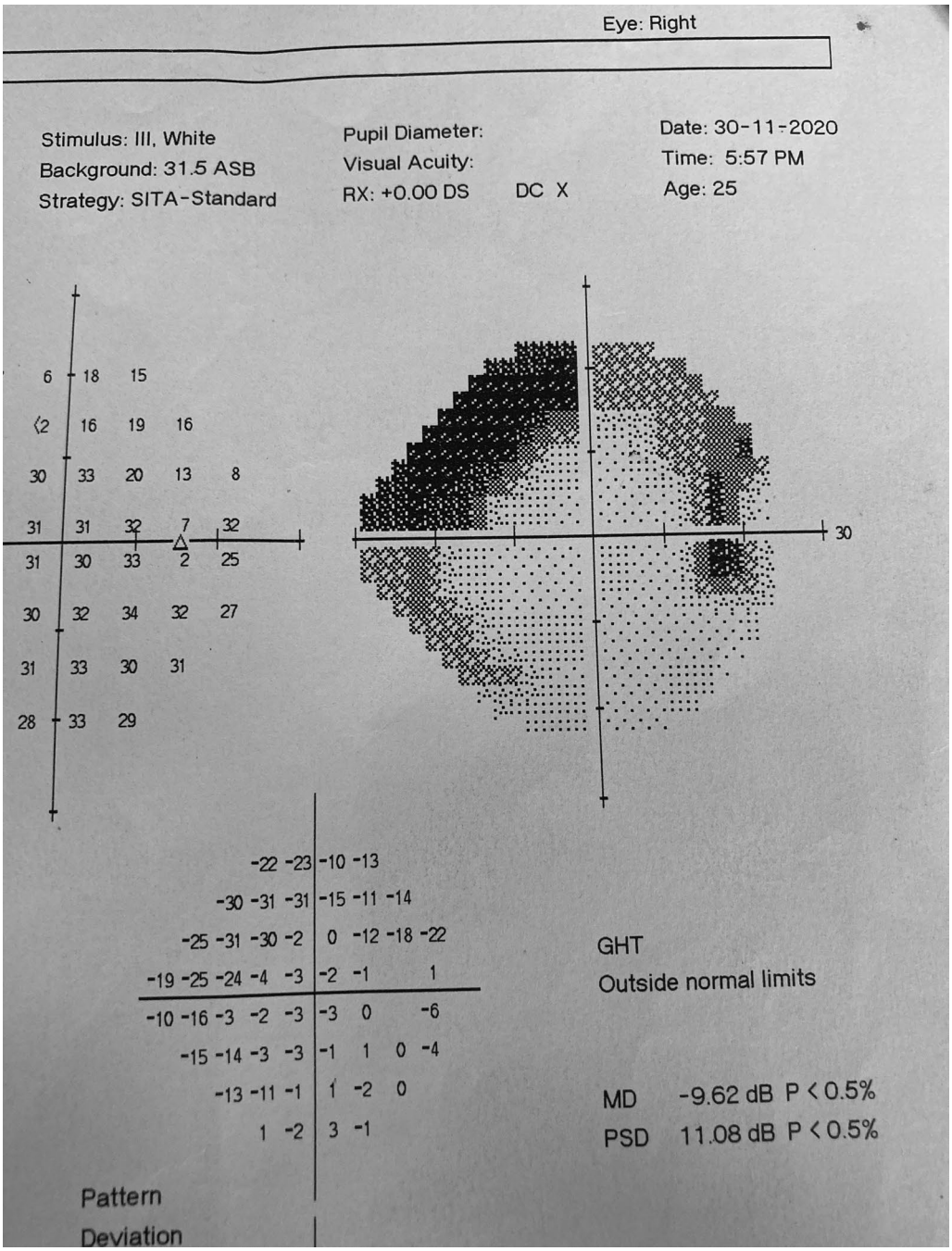

Fig. 7 Perimetry of right eye showing superior temporal arcuate scotoma

For surgically treated patients, MRI is recommended at the 3-month follow-up point and then on a yearly basis for 5 years. After this, imaging follow-up may be performed every 2–3 years if patients are clinically and endocrinologically stable, with overall follow-up for at least a decade after the operation.

Rathke's cyst at ectopic locations [11–18], other than the sellar suprasellar area, have been reported.

Exclusive suprasellar Rathke's cleft cyst is rare, and to the best of our knowledge, only 62 cases have been reported till date [19–33], and ours being the 63rd case.

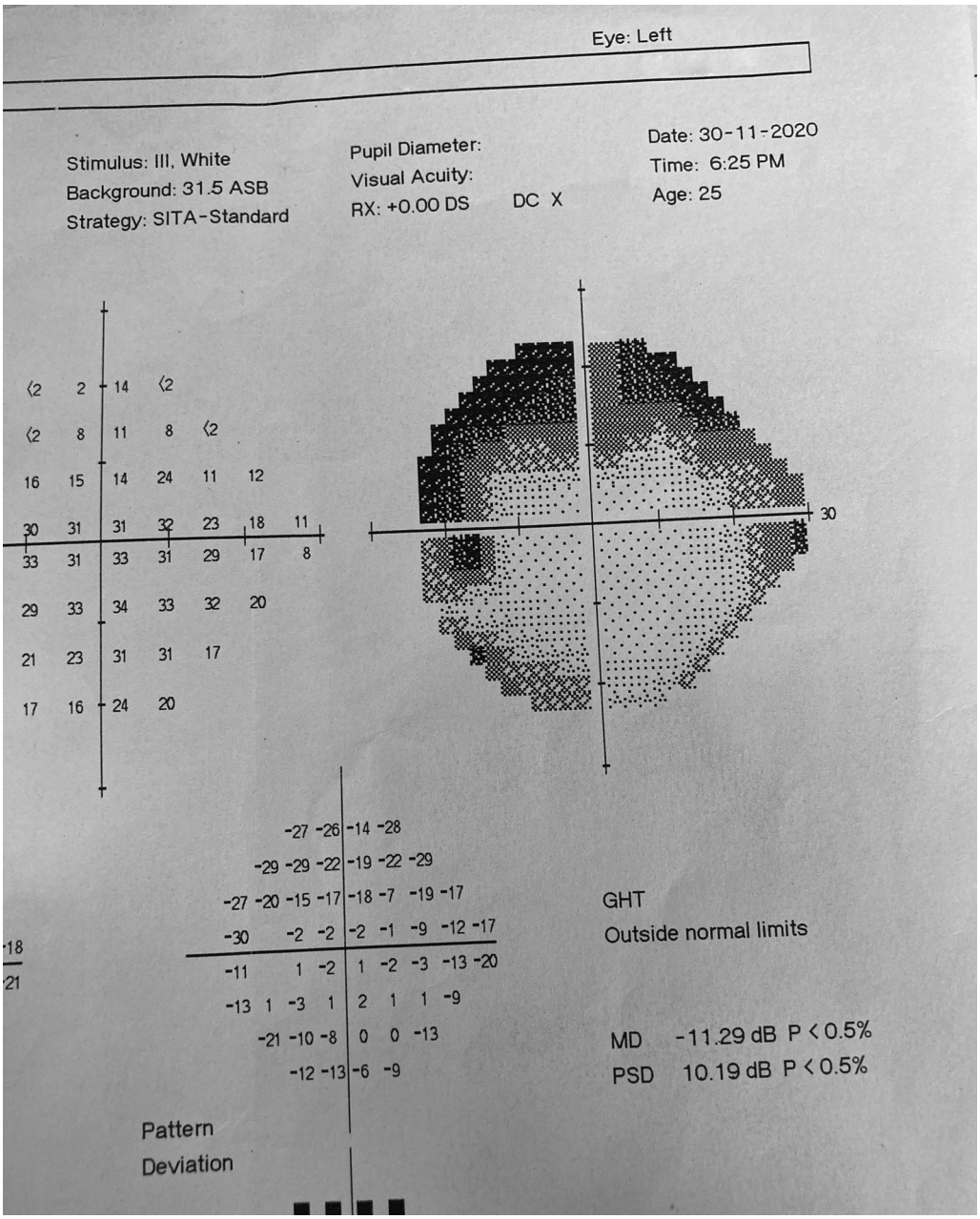

Fig. 8 Perimetry of the left eye showing superior temporal and nasal arcuate scotoma

Conclusions

Rathke's cyst, though an intrasellar lesion, can arise in the suprasellar area and other ectopic locations.

Though benign, the lesions showing squamous metaplasia have a higher tendency to recur. Hence, a regular follow-up, to detect recurrence at the earliest, is advisable.

Table 1 Hormonal profile

Hormone	Patient value	Normal range
S prolactin (ng/ml)	44.88	Less than 20
S TSH (mIU/ml)	14.72	0.35–4.5
S cortisol (ûg/dl)	5.84	10–20
S growth hormone (ng/ml)	0.094	0.4–10

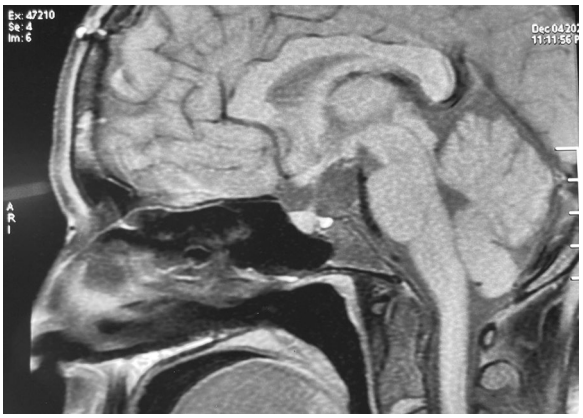

Fig. 9 Post-operative MRI sagittal section showing complete removal of the lesion

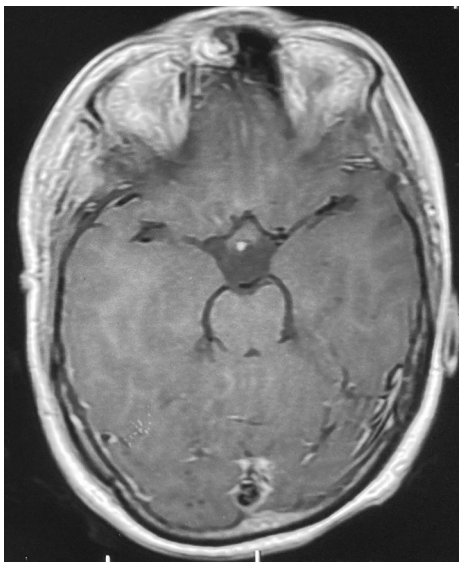

Fig. 10 Post-operative MRI axial section showing complete removal of the lesion

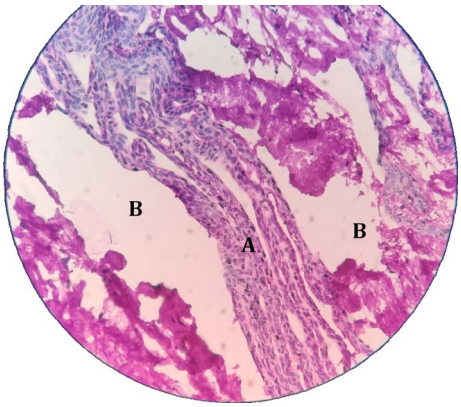

Fig. 11 H&E stain showing Rathke's cleft cyst wall lining. **a** Cuboidal to columnar epithelial lining of the cyst wall. **b** Empty space within the cyst

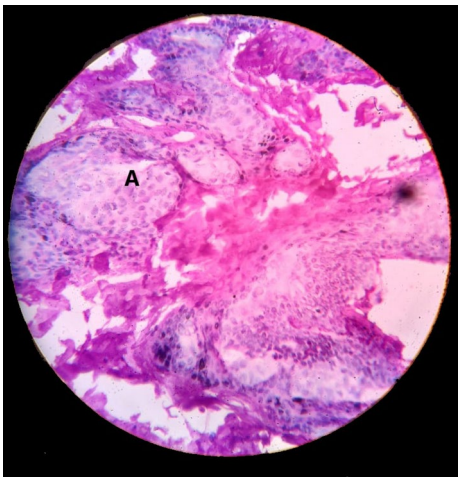

Fig. 12 H&E stain of Rathke's cleft cyst showing squamous metaplasia within cyst wall lining as 'A'

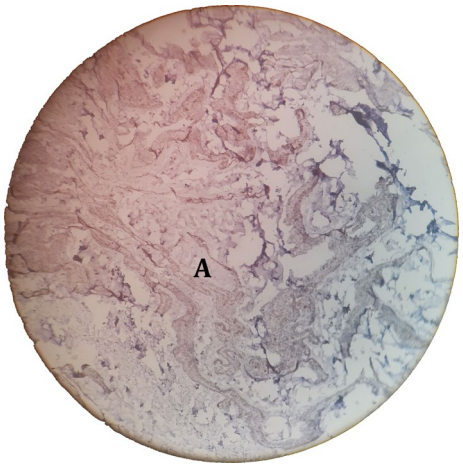

Fig. 13 IHC × 40 image; showing pankeratin positivity seen in cyst wall lining epithelium shown as 'A'

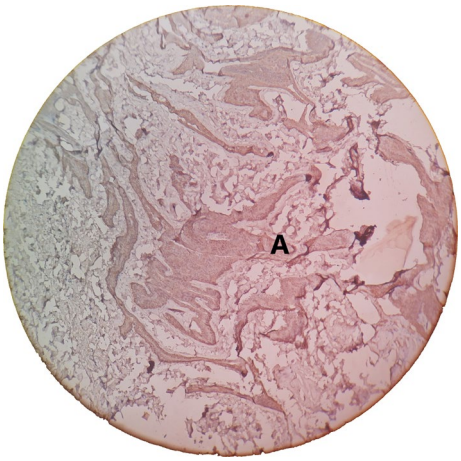

Fig. 15 IHC × 40 image; showing EMA positivity in epithelial cell lining of cyst wall as 'A'

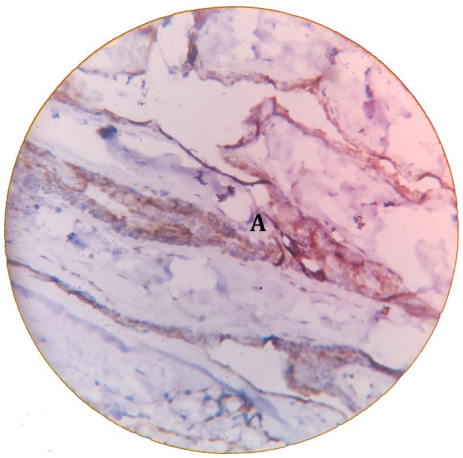

Fig. 14 IHC × 20 image; showing pankeratin positivity seen in a cyst wall lining epithelium shown as 'A'

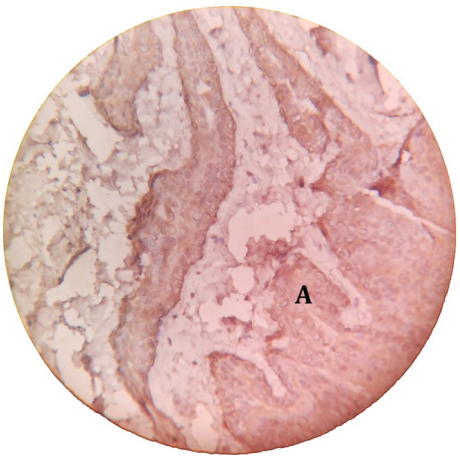

Fig. 16 IHC × 20 image; showing EMA positivity in epithelial cell lining of cyst wall as 'A'

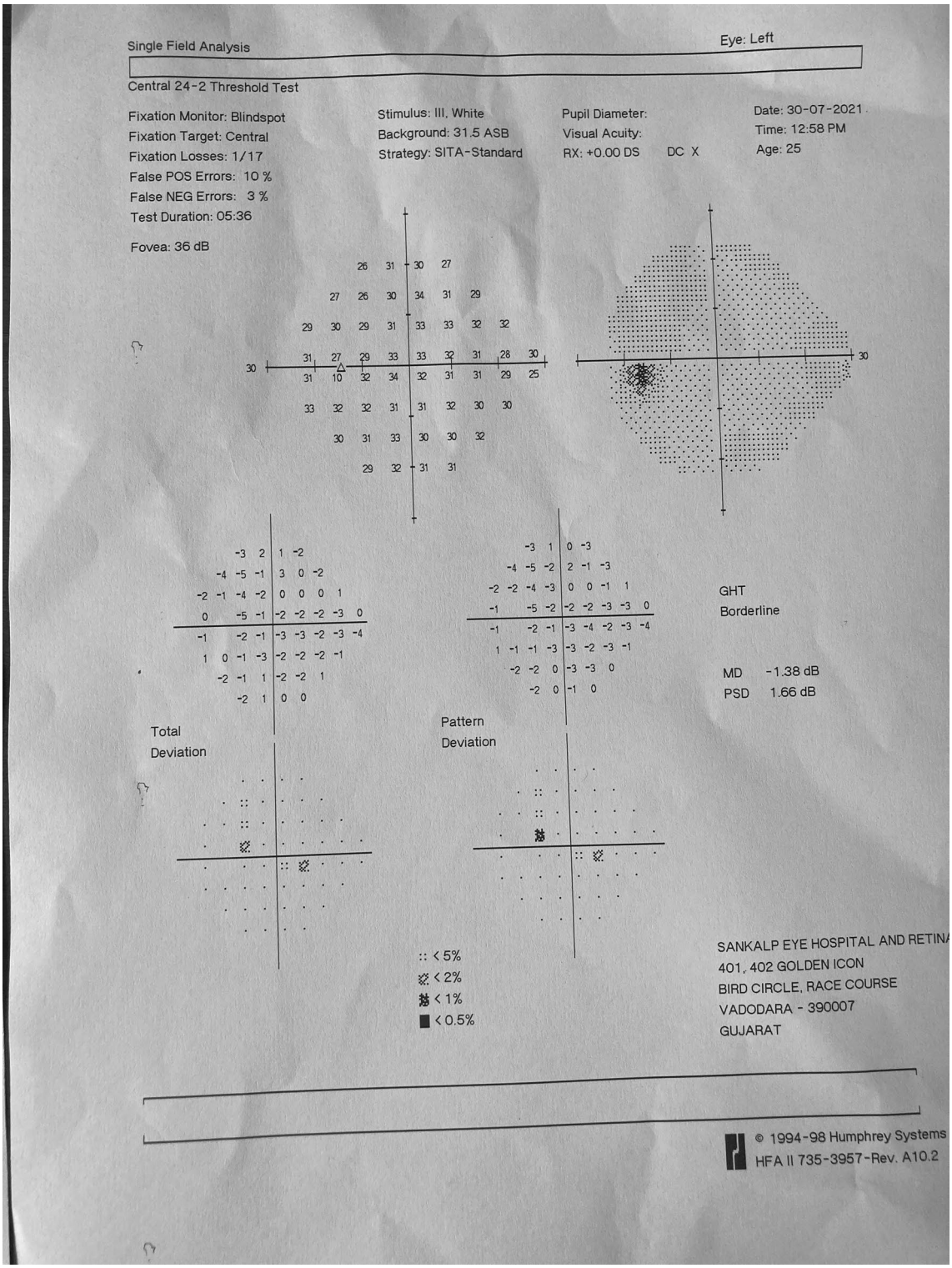

Fig. 17 Perimetry of the left eye after 6 months showing complete resolution of scotoma, with normal visual fields

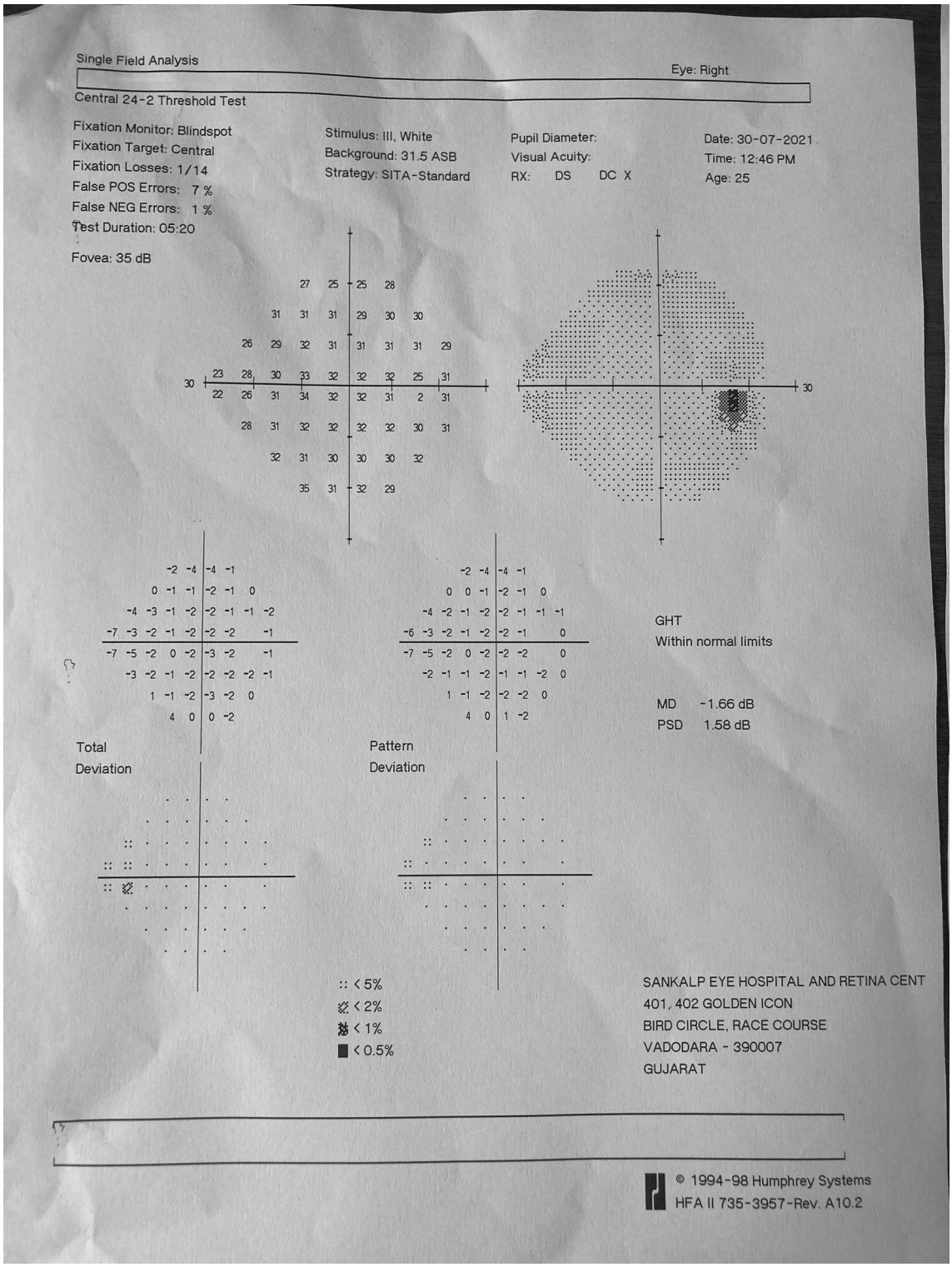

Fig. 18 Perimetry of the right eye after 6 months showing complete resolution of scotoma with normal visual fields

Abbreviations
CT Scan: Computerised tomography scan; MRI: Magnetic resonance imaging; RCC: Rathke's cleft cyst; DI: Diabetes insipidus.

Acknowledgements
Not applicable.

Author contributions
The first/corresponding author, APP, was the operating surgeon involved in the treatment of the patient, preparation of manuscript and its submission. The second author, SC, was the assistant surgeon and was involved in the preparation of manuscript, getting the photographs of the slides and getting the references. Both authors read and approved the final manuscript.

Funding
We have not received any funding for this study from any source.

Availability of data and materials
All the data used and analysed during this study are included in this published article.

Declarations

Ethics approval and consent to participate
The study does not involve any new or experimental procedure. The patient was treated by standard neurosurgical protocols. The study was put up for approval in the Neuron Hospital Ethical committee. After due deliberation, the committee gave approval for the publication of the study.

Consent for publication
The identity of the patient has not been revealed in the study. The patient's consent for publication of the study was taken. We are ready to submit the form of consent for publication whenever required.

Competing interests
The authors declare that they have no competing interests.

Author details
[1]Neuron Hospital, Vadodara, Gujarat, India. [2]Baroda Medical College and SSG Hospital, Vadodara, Gujarat, India.

Received: 4 November 2021 Accepted: 26 April 2022
Published online: 13 June 2022

References

1. Kanter AS, Sansur CA, Jane JAJ Jr, Laws ERJ Jr. Rathke's cleft cysts. Front Horm Res. 2006;34:127–57.
2. Zhang Y, Chen C, Tian Z, Cheng Y, Xu J. Differentiation of pituitary adenoma from Rathke cleft cyst: combining MR image features with texture features. Contrast Media Mol Imaging. 2019;2019:6584636.
3. Choi SH, Kwon BJ, Na DG, Kim JH, Han MH, Chang KH. Pituitary adenoma, craniopharyngioma, and Rathke cleft cyst involving both intrasellar and suprasellar regions: differentiation using MRI. Clin Radiol. 2007;62(5):453–62.
4. Gaddikeri S, Vattoth S, Riley KO, DeHoff GW, Smith CB, Combs JT, Roberson GH. Rathke cleft cyst. MRI criteria for presumptive diagnosis. Neurosciences (Riyadh). 2013;18(3):258–63.
5. Teramoto A, Hirakawa K, Sanno N, et al. Incidental pituitary lesions in 1000 unselected autopsy specimens. Radiology. 1994;193:161–4.
6. Zada G. Rathke cleft cysts: a review of clinical and surgical management. Neurosurg Focus. 2011;31(1):E1.
7. Larkin S, Karavitaki N, Ansorge O. Rathke's cleft cyst. Handb Clin Neurol. 2014;124:255–69.
8. Voelker JL, Campbell RL, Muller J. Clinical, radiographic, and pathological features of symptomatic Rathke's cleft cysts. J Neurosurg. 1991;74:535–44.
9. Aho CJ, Liu C, Zelman V, Couldwell WT, Weiss MH. Surgical outcomes in 118 patients with Rathke cleft cysts. J Neurosurg. 2005;102(2):189–93.
10. Kinoshita Y, Tominaga A, Usui S, et al. The long-term recurrence of Rathke's cleft cysts as predicted by histology but not by surgical procedure. J Neurosurg. 2016;125(4):1002–7.
11. Agarwal V, Palande D, Velho V, Binayke RS. Unusual site of Rathke's cleft cyst: A case report and review of literature. Asian J Neurosurg. 2017;12(1):44–6.
12. Kasliwal MK, Dua SG, Harbhajanka A, Nag S, Jhaveri MD, Moftakhar R. Intrasphenoidal Rathke's cleft cyst. J Clin Neurosci. 2015;22(10):1678–82.
13. Kamei T, Nonaka M, Uemura Y, Yamanouchi Y, Komori Y, Iwata R, Takeda J, Hashiba T, Yoshimura K, Asai A. Enlarging pediatric ectopic Rathke's cleft cyst in the prepontine cistern: case report. J Neurosurg Pediatr. 2017;20(5):480–4.
14. Megdiche-Bazarbacha H, Ben Hammouda K, Aicha AB, Sebai R, Belghith L, Khaldi M, Touibi S. Intrasphenoidal Rathke cleft cyst. AJNR Am J Neuroradiol. 2006;27(5):1098–100.
15. Chuang CC, Chen YL, Jung SM, Pai PC. A giant retroclival Rathke's cleft cyst. J Clin Neurosci. 2010;17(9):1189–91.
16. Ben Hamouda K, Khaldi M, Jemel H, Ben Ismail M, Zemmel I. Rathke cleft cyst of the sphenoid bone and nasopharynx. Case illustration J Neurosurg. 2005;103(6):1095.
17. Fan J, Qi S, Peng Y, Zhang XA, Qiu B, Pan J. An isolated primary Rathke's cleft cyst in the cerebellopontine angle. J Neurosurg. 2014;121(4):846–50.
18. Zhou L, Luo L, Hui X, Chen H, Yu B, Guo G, You C. Primary Rathke's cleft cyst in the cerebellopontine angle associated with apoplexy. Childs Nerv Syst. 2010;26(12):1813–7.
19. Barrow DL, Spector RH, Takei Y, et al. Symptomatic Rathke's cleft cysts located entirely in the suprasellar region: review of diagnosis management, and pathogenesis. Neurosurgery. 1985;16:766–72.
20. Rincon JL, Nunes J, Camuto P, Goodrich I. Intracranial approach to suprasellar Rathke's cleft cyst. Skull base surgery. 1999;9(01):71–3.
21. Sade B, Albrecht S, Assimakopoulos P, Vézina JL, Mohr G. Management of Rathke's cleft cysts. Surg Neurol. 2005;63(5):459–66 (**discussion 466**).
22. Itoh J, Usui K. An entirely suprasellar symptomatic Rathke's cleft cyst: case report. Neurosurgery. 1992;30:581–5.
23. Potts MB, Jahangiri A, Lamborn KR, Blevins LS, Kunwar S, Aghi MK. Suprasellar Rathke cleft cysts: clinical presentation and treatment outcomes. Neurosurgery. 2011;69(5):1058–68 (**discussion 1068–7**).
24. Peng Y, Fan J, Li Y, Qiu M, Qi S. The Supraorbital Keyhole Approach to the Suprasellar and Supra-Intrasellar Rathke Cleft Cysts Under Pure Endoscopic Visualization. World Neurosurg. 2016;92:120–5.
25. Brassier G, Morandi X, Tayiar E, Riffaud L, Chabert E, Heresbach N, Poirier JY, Carsin-Nicol B. Rathke's cleft cysts: surgical-MRI correlation in 16 symptomatic cases. J Neuroradiol. 1999;26(3):162–71.
26. Rout D, Das L, Rao VR, et al. Symptomatic Rathke's cleft cysts. Surg Neurol. 1983;19:42–9.
27. Wenger M, Simko M, Markwalder R, Taub E. An entirely suprasellar Rathke's cleft cyst: case report and review of the literature. J Clin Neurosci. 2001;8:564–7.
28. Mitsuha T, Ikawa F, Ohbayashi N, Imada Y, Kazihara Y, Abiko S, Inagawa T. Symptomatic Rathke's cleft cyst mimicking suprasellar arachnoid cyst. No Shinkei Geka. 2005;33(12):1213–8 (**Japanese**).
29. Ishii T, Yamasaki T, Tanaka J, Tanaka S, Hori T, Muraoka K. Rathke's cleft cyst–Report of three cases. No shinkei geka. Neurol Surg. 1987;15(4):451–6.
30. Yuge T, Shigemori M, Tokutomi T, Kuga S, Nishio N, Yamamoto F, Tokunaga T, Uegaki M, Abe H. Entirely suprasellar symptomatic Rathke's cleft cyst. No shinkei geka. Neurol Surg. 1991;19(3):273–8.
31. Choudhry OJ, Choudhry A, Patel SK, Baisre A, Eloy JA, Liu JK. Giant suprasellar Rathke's cleft cyst mimicking craniopharyngioma: implications for a spectrum of cystic epithelial lesions of ectodermal origin. J Neurol Surg A Central Eur Neurosurg. 2012;73(05):324–9.
32. Sade B, Albrecht S, Assimakopoulos P, Vézina JL, Mohr G. Management of Rathke's cleft cysts. Surg Neurol. 2005;63(5):459–66.
33. Chotai S, Liu Y, Pan J, Qi S. Characteristics of Rathke's cleft cyst based on cyst location with a primary focus on recurrence after resection. J Neurosurg. 2015;122(6):1380–9.

Publisher's Note