Intra-infundibular Epidermoid Cysts- A Rare and Distinct entity

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Introduction

Epidermoid cysts (EC) located within the pituitary infundibulum are a rare disease with only seven reported cases. They are anatomically and embryologically distinct from suprasellar ECs. Preoperatively, they are usually assumed to be craniopharyngiomas or Rathke's cleft cyst. However, all these entities have different management strategies and morbidity profiles. In this study, we have described our experience with three cases of intra-infundibular epidermoid cysts (IEC) operated on at our institute and performed a systematic review of literature highlighting its distinguishing features on imaging, treatment nuances and morbidity profile.

Case description

Case 1: A 51-year-old male patient complained of visual blurring and diplopia. His MRI brain (Figure 1) showed a 2.2x1.4x1.9 cm cystic lesion in the suprasellar cistern abutting the anterior third ventricle and displacing the optic tracts. The patient had a normal ophthalmological and hormonal evaluation. He underwent an endoscopic endonasal trans-tubercular resection of the lesion. An intraoperative endoscopic ultrasound (USG) was used, and the lesion appeared heterogeneous but mostly hyperechoiec (Figure 2). The infundibulum was incised, and pearly white fluid escaped. All the fluid and keratin debris were carefully dissected off the stalk, hypothalamus, and third ventricular walls except a small plaque firmly adherent to the stalk. Postoperatively the patient did well with subjective improvement of vision and a brain MRI showed complete tumor resection (Figure 3). He did not suffer any endocrinological deficits and appeared healthy and asymptomatic in his last follow-up (8 months since surgery). Histopathological examination showed the presence of stratified squamous epithelium with keratin debris which was consistent with EC.

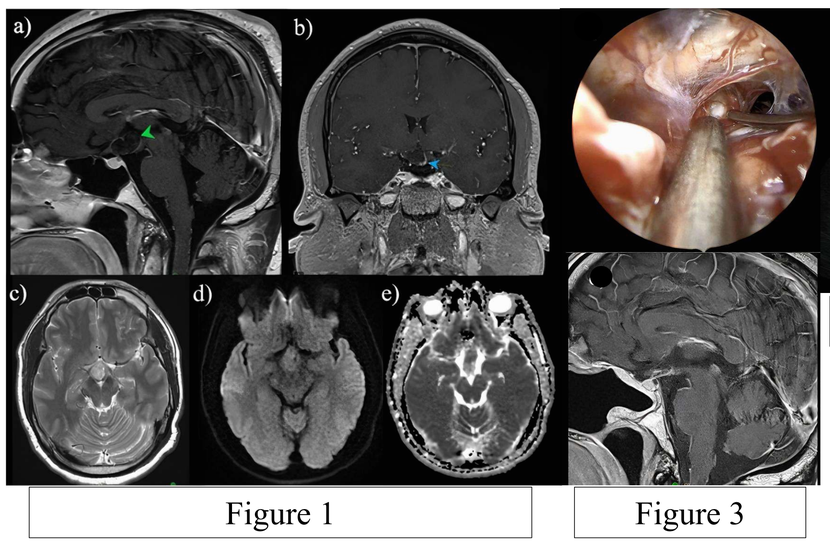

Figure 1

Figure 2

Figure 3

Case 2: A 45-year-old female presented to the clinic with complaints of headache, dizziness, and blurring of vision with diplopia. She also reported increased thirst and urination which was confirmed to be central diabetes insipidus. Her endocrine evaluation also revealed low serum thyroid stimulating hormone (TSH) and free T4 for which she was started on levothyroxine supplementation. A brain MRI (Figure 4) revealed a 1.1x1.2x1.8 cm lobulated suprasellar midline mass arising from the infundibulum extending from the sella into the suprasellar compartment inseparable from the hypothalamus and the floor of the third ventricle and causing mass effect over the optic chiasm
A working diagnosis of craniopharyngioma was made and the patient underwent surgery through an expanded endoscopic endonasal trans-tubercular approach. Dura was incised widely, and the infundibulum was incised at its most expanded portion (towards the right) and thick whitish fluid extravasated. On further exploration, flakes of pearly white material evacuated, and the optic chiasm was completely freed of tumor. A complete resection was achieved along with an intact pituitary stalk. Postoperatively patient reported complete resolution of headache and visual problems. Her postoperative MRI (Figure 5) showed complete resection of the tumor, and she continues to be on desmopressin and levothyroxine. Histopathological examination was consistent with EC. In her last follow-up (3 years since resection) she remains asymptomatic without recurrence.

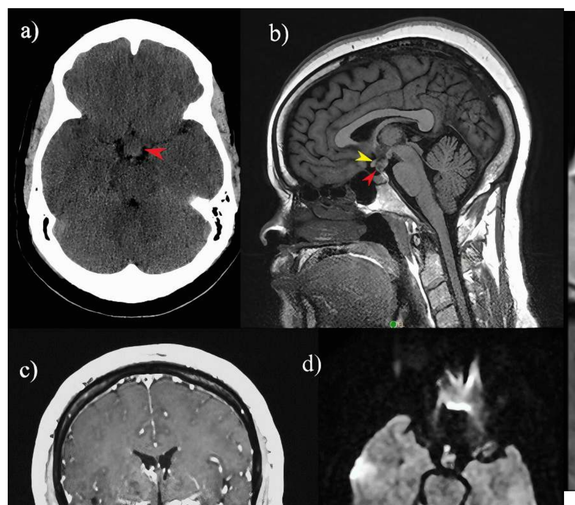

Figure 4

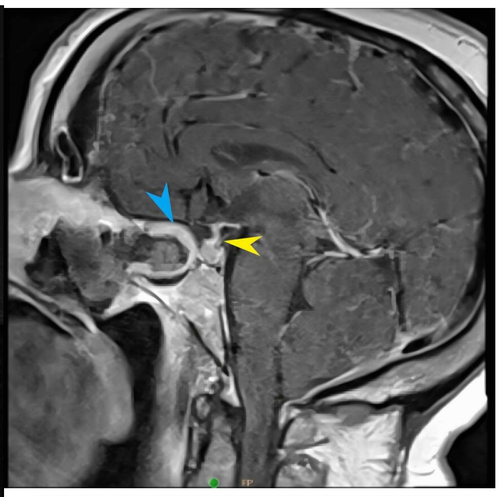

Figure 5

Case 3: A 69-year-old man presented after a subtotal endoscopic endonasal resection of a sellar-suprasellar mass at another institution. A presumptive diagnosis of craniopharyngioma was made due to inadequate tissue sample. He came to us with worsening visual symptoms and a brain MRI (Figure 6) suggesting disease recurrence. He underwent repeat resection of the mass through an extended endoscopic endonasal trans-tubercular approach. Immediately opening the sellar dura, flakes of pearly white semisolid material was encountered. The dural incision was extended, and an incision was made longitudinally along the infundibulum to visualize the cyst (Figure 7). The tumor was progressively dissected from the walls of the hypothalamus until a complete resection was achieved. The histopathology was suggestive of epidermoid cyst. The patient was well for ten months, when a routine surveillance MRI demonstrated reoccurrence of a cystic mass in the suprasellar cistern with avid diffusion restriction. The imaging features and rate of progression suggested an abscess over recurrence and a decision was made to drain the cyst through a right supraorbital approach with evacuation of thick mucoid contents. Postoperatively, the patient developed transient mild left sided weakness which resolved by the ninth postoperative day. Postoperative MRI showed complete evacuation of the abscess (Figure 8), and histopathological examination did not show any evidence of keratin or epidermal cells. Cytology showed the presence of few inflammatory cells, and no organism was grown on cultures.

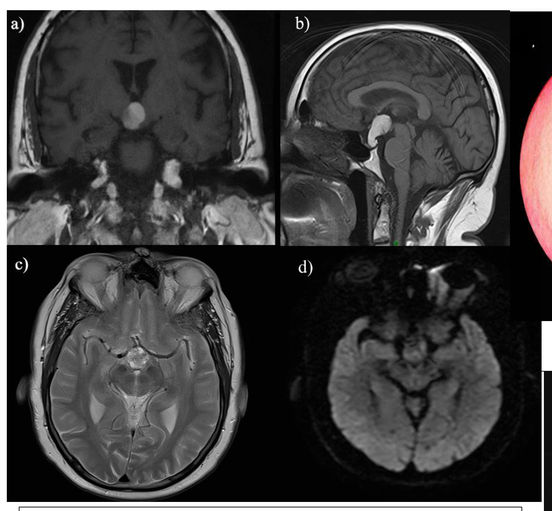

Figure 6

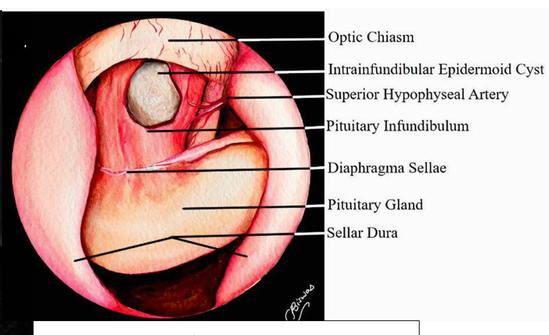

Figure 7

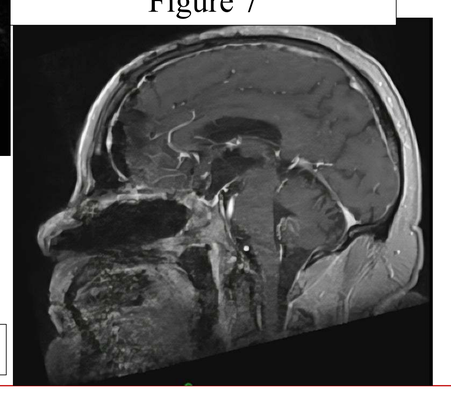

Figure 8

Conclusion

A total of 10 cases (seven from the literature and three of our own) were analyzed. Median age at diagnosis was 53.5 years and six cases were male. Preoperatively, all patients had visual disturbance, 6 out of 8 patients (8 reports mentioned the hormonal profile) had deficiency in pituitary hormones with 3 out of 8 having diabetes insipidus (DI). The average tumor size was 1.62 cm^3 with 7 predominantly cystic lesions. On T1 weighted MRI most lesions had low or mixed signal intensity while one lesion was bright. Diffusion restriction was noted in 4 out of 5 cases. Adhesion of the cyst wall to the stalk and/or the hypothalamus was a common occurrence resulting in residual wall being left behind in 5 cases. Expanded endoscopic endonasal approach (EEA) was utilized in 9 cases and one case underwent fronto-temporal craniotomy with resection via pretemporal approach. All patients except one required postoperative hormonal supplementation. Postoperative chemical meningitis was demonstrated in two cases and a sterile abscess was noted in another case.
In conclusion, IECs have distinct radiological features, treatment strategies and postoperative morbidity profile compared to other cystic lesions in the infundibulum. Thus, it is important to recognize this distinct and rare entity.