CASE REPORT

‘Recurrent pharyngioma’: A rare tale of epidermoid cyst at sellar region

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SUMMARY

Epidermoid cysts (EC) arise from epithelial cells that are retained during closure of the neural tube. ECs are congenital and formed out of aberrant ectodermal cells, which become trapped during the embryogenesis between the 3rd and 5th gestational week. Craniopharyngiomas however, accounts for 6% of brain tumors in children while EC only constitutes only 1 to 2% of brain tumors. Recognizing EC early; being a rare entity and prompt intervention will be able to prevent complications and provide comfort for patients. We would like to share a case of EC in a man who was first treated as craniopharyngioma and subsequently had ‘recurrent pharyngioma’ where subsequent HPE proved to be EC.

INTRODUCTION

Epidermoid cysts (EC), also known as primary cholesteatomas, arise from epithelial cells that are retained during closure of the neural tube. ECs are congenital and formed out of aberrant ectodermal cells, which become trapped during the embryogenesis between the 3rd and 5th gestational weeks.[1] These cysts can grow by accumulating cholesterol and keratin from desquamation of the lining epithelium and potentially encases nearby nerves and arteries.

Intracranial ECs are most often located in intradural, but they can also occur extradurally in the intradiploic space in up to 10% of cases.[2] The most common intradural locations are cerebellopontine angle (60%), fourth ventricle (5%–18%), parasellar area and middle cranial fossa (15%); less frequently, within ventricles and brain parenchyma.

Here we would like to report a rare case of Epidermoid cyst masquerading as a sellar tumour in a patient seen in Hospital Tuanku Ja’afar Seremban (HTJ), Malaysia.

CASE PRESENTATION

Mr. J is a 31-year-old male who worked as a lorry driver. He complained of left eye blurring of vision since 2018. It was associated with headache and loss of weight about three kg in two months. He was referred to a neurosurgical clinic. His blood pressure was 134/89mmHg. On physical examination, his visual acuity on the left eye was 6/18 while his right vision was intact at 6/9. He did not exhibit any sign of hypogonadism. Blood investigations taken in January 2019 showed normal thyroid function test with other hormonal profile.

Magnetic Resonance Imaging (MRI) of the brain was done in July 2018 showed a sellar mass measuring 2.6cm X 2.9cm X 2.3cm. The mass has suprasellar extension and optic chiasm compression. He was advised for surgical intervention for his condition, which he declined at that point of time. He was given a neurosurgical clinic visit to review his symptoms. However, the symptoms persisted and repeated MRI was performed in January 2019 showing increasing sellar mass size measuring 2.9cm X 3.0cm X 2.4cm. The lesion appeared heterogenous, cystic and extends posteriorly which abuts the basilar artery. He was treated for craniopharyngioma, and a date was given for surgery.

He then underwent transsphenoidal excision of tumor (TSS) in March 2019 at Hospital Tuanku Ja’afar Seremban. Pre-operative blood investigations showed low FT4 and serum cortisol (as shown in Table I). Hence, Mr. J was prescribed maintenance of oral levothyroxine 50mcg daily and oral hydrocortisone 10mg in morning and 5mg in afternoon respectively. No cortisol or thyroid function test were repeated post-operatively. He was then given a 2-month endocrine clinic visit for insulin tolerance test (ITT) at medical daycare. As ITT result showed adequate cortisol response, his oral hydrocortisone was off by June 2019 and plans were given to regularly monitor his hormonal profile. Unfortunately, Mr. J did not come for any follow-up during COVID-19 pandemic. He did not take the thyroxine supplement during this period as he was asymptomatic.

In February 2023, Mr. J presented to neurosurgical team again for the complaint of headache and reduced right vision for period of 2 months since January 2023. An urgent computed tomography (CT) of brain was done and showed a well-defined lobulated isoechoic mass measuring 3.7cm X 3.0cm X 3.9cm. The mass was in the suprasellar region with mass effect (as shown in Figure 1). Again, he declined surgical intervention via open craniectomy for recurrent craniopharyngioma.

Subsequently, a new MRI brain was done on 20th April 2023 which showed larger lobulated sellar mass measuring 3.0cmX 3.3cmX 3.5cm. It was compressing to 3rd ventricles, optic chiasm and adjacent left midbrain. With the MRI brain findings and new onset of right sided body weakness for 1

This article was accepted: 11 December 2024
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Table I: Table shows blood investigations taken prior surgery and one-week post-surgery.

	31/1/19	1/2/19	20/3/19	21/5/19	19/6/2023	29/6/23	2/7/23	3/7/23	4/7/23	11/7/23
Na (135-145)						141	150	155	148	139
K (3.5-4.5)						3.8	3.9	4.0	3.9	4.1
Urea (3.2-8.2)						4.5	4.5	3.5	3.3	3.4
Creatinine (62-115)						60	90	110	90	80
Prolactin uiu/ml (86-390)	304									
LH Iu/L (1.5-9.3)	3.7			3.2	8.1					
FSH Iu/L (1.5-12.4)	7.7			8.9	12.1					
Cortisol nmol/L	254.6	516								340.6
T4 (11.5-22.7)	11.1			12.4	12					12.9
TSH (0.55-4.78)	0.59			1.67	0.26					<0.01
Testos nmol/L (6.85-23.23)	28.64			34.25	22.9					<0.24
U. Osmo (Osm/kg)	246	550	169			618	211	106	538	
Sr Osmo (Osm/kg)		284	293			303	306	311	296	
UFEME SG						1.015	<1.005	<1.005	1.010	

Na = Sodium; mmol/L, K = Potassium; mmol/L
LH = Luteinizing Hormone, FSH = Follicle-stimulating hormone, Testos = Testosterone
T4 = Thyroxine, TSH = Thyroid Stimulating Hormone

month since May 2023, Mr. J agreed for surgical intervention during neurosurgical clinic visit on June 2023. He underwent open decompressive craniectomy surgery on 27th June 2023 for mass excision without many complications. Intra-operatively, the tumor was noted to be yellowish and has thick capsule surrounding the core. It has three main lobules which have fat-like substances in consistency. Capsulectomy was done without any major adverse event.

On 2nd July 2023, post operation day 5 (D5), Mr. J had polyuria of four liters in the span of 24 hours. Several paired serum osmolality and urine osmolality samples were taken and consistent with cranial DI (as shown in Table I). At the same time, his potassium level showed an increase from 3.8mmol/L to 4.0mmol/L. Creatinine was also noted to rise from 60 mg/L to 110mg/L. To prevent further polyuria, Mr. J was started on oral Minirin® 0.5mcg BD. He responded well to the oral Minirin®. He was discharged home well with oral Minirin® 0.5mcg BD and oral hydrocortisone 10mg BD on 11th July 2023. The Histopathology examination (HPE) result showed the sellar mass to be consistent with epidermoid cyst (as shown in Figure 2).

In following clinic visits, Mr. J recovered well. He responded well with hormone replacement oral Minirin® 0.5mcg BD and oral hydrocortisone 10mg BD. Intramuscular Testosterone replacement 250mg monthly was initiated in clinic visit as serum testosterone remained low. However, his visual field remains the same - left eye was 6/18 while his right vision was intact at 6/9. His repeated MRI Brain in September 2023 also showed no recurrence of Epidermoid Cyst. Previous right sided body weakness had also improved. He can ambulate with a walking frame and perform simple daily chores.

DISCUSSION

Epidermoid cysts (EC) account for approximately 1%-2% of all brain tumours.[3] ECs form from the accumulation of keratin and cholesterol, which desquamate into a pearly material within their walls. ECs grows in the cisternal spaces and remaining asymptomatic for years due to the absence of initial mass effect. They are commonly seen in males during the 3rd–4th decade but turn malignant predominantly in females.[4]

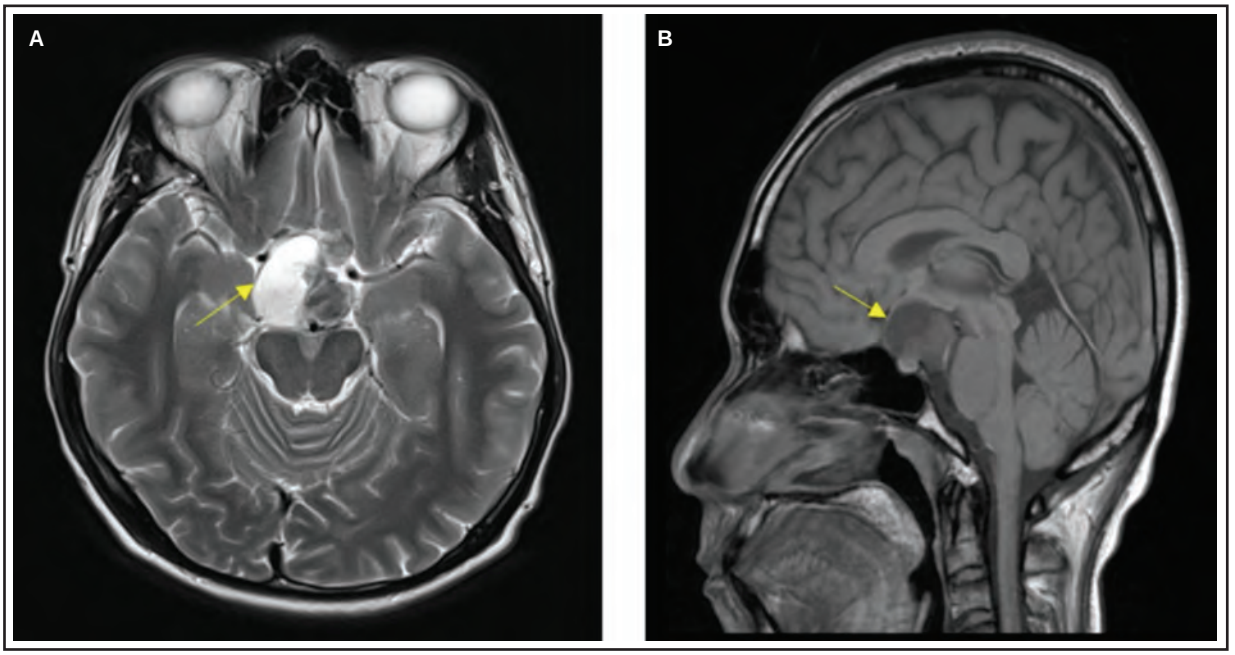

Fig. 1: Picture on the left (a) showing T2 weighted MRI brain axial view. The mass can be see via the yellow arrows. Picture on the right (b) featured MRI brain sagittal view FLAIR.

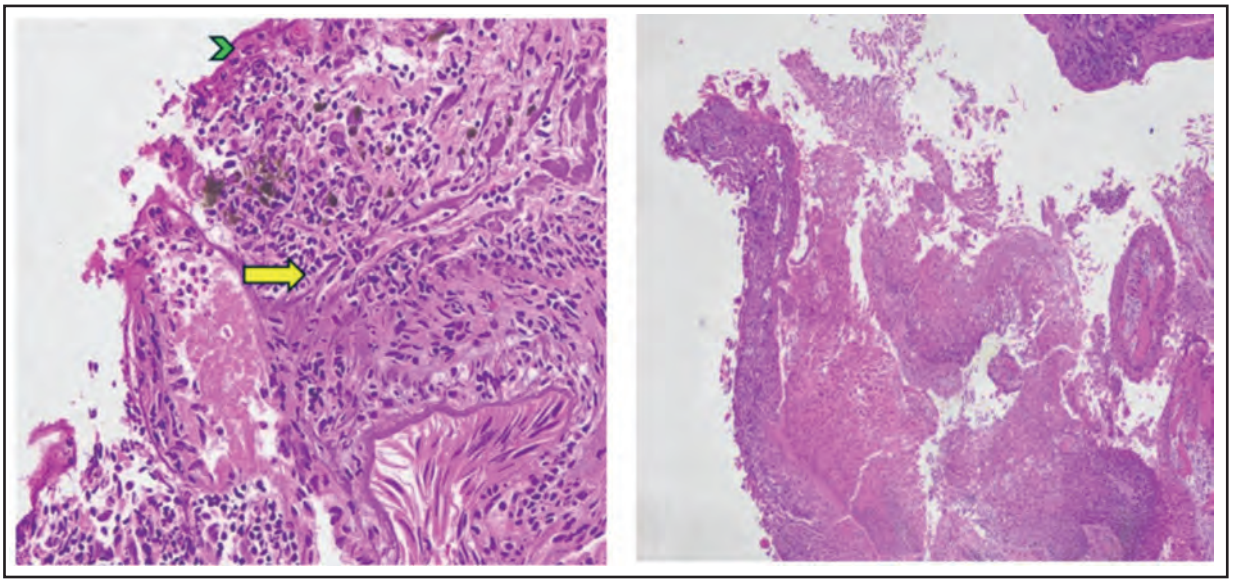

Fig. 2: Section shows severely crushed tissue composed of fibrocollagenous tissue lined by stratified squamous epithelium with granular cell layer ( ). In areas, the cyst lining is replaced by granulation tissue formation ( ). The surrounding stroma tissue show dense lymphoplasmacytic infiltrate. Some hemosiderin laden macrophages are also identified. No stellate reticulum, papillary structure or wet keratin is seen. Adjacent pituitary tissue with glial tissue is also observed.
Immunohistochemistry: The stratified squamous epithelium is positive for CK5/6 and negative for CK7, CK20, S100, GFAP, synaptophsin and chormograninA.

ECs presents similarly like the rest of intracranial space occupying lesions. Common features like headaches, cranial nerve deficits, cerebellar symptoms, seizures, raised intracranial pressure are usually seen.[5] Conventional MRI sometime cannot reliably be used to distinguish epidermoid tumours from arachnoid cysts since both lesions are very hypointense relative to brain parenchyma on T1-weighted MR. images and very hyperintense on T2- weighted images. In some literatures, ECs have been described along as cystic craniopharyngiomas.[6] Mr. J's lesion in MRI appears as suprasellar and cystic, hence initially impression was craniopharyngioma.

Histopathology examination (HPE) remains mainstay of diagnosis of ECs. The diagnosis of epidermal cyst made in this case due to presence of predominant cyst lining by stratified

squamous epithelium with prominent granular layer. The surrounding tissue shows features of foreign body granuloma possibility due to ruptured epidermal cyst. Usually there is a keratin layer within the cyst but due to long standing disease the cyst might be ruptured, and the keratin layer has been eaten up by the macrophages. No obvious stellate reticulum, papillary structure or wet keratin is identified which therefore consistent with the findings of ECs. Macroscopically epidermoid cyst appears as white caseous-like material which is like Mr. J's intraoperative findings.

Craniopharyngioma on the other hand, appears as dark 'motor-oil' fluid. Histologically, craniopharyngioma is composed of cords, lobules, nodular whorls, and trabeculae of well differentiated squamous epithelium bordered by palisading columnar epithelium cells surround looser plumper cells called stellate reticulum.

Surgical approach for ECs is generally determined by the location and the extent of the lesion.[7] Meticulous discussion and planning must be done for complete tumour resection to prevent recurrence. However, adherence of the capsule to the important neurovascular structures in and around the brain stem often leads to its incomplete removal.[8] Therefore, surgical debulking with capsule removal is a definitive treatment. Open craniotomy is the preferred choice in the case, as the lesion is large and compressing the optic chiasm and left midbrain. The surgical plane will be bigger, and lesion can be approach by the surgeon which avoiding major neurovascular structures as ECs have high capsular adherence to its' surroundings. Complications from surgery include CSF leak, aseptic meningitis, vision loss, nerve injury, diabetes insipidus (DI) and panhypopituitarism.

The risk of postoperative hypopituitarism varies according to case series and the aetiology. Surgeon's experience, the size and consistency of the tumour, the extension of surgical manipulation, and surgery for recurrent disease play a role in the occurrence of hypopituitarism. Evaluation for the anterior pituitary function should be performed approximately 4–6 weeks after pituitary surgery. Mr. J developed episodes of polyuria which prompted the screening for anterior pituitary function during postsurgical recoveries. As this is a second surgery for him, much emphasize was given to detect hypopituitarism. Early recognition enabled Mr. J to receive hormonal replacement and aided in his postoperative recovery.

Even though surgical option remains the mainstay treatment option, only 50- 80% of patients have complete removal of EC. Therefore, the recurrence rate for intracranial epidermoid cysts was stated at 24%.[9]

CONCLUSION

Identifying the nature of sellar and parasellar lesions remains challenging because of the complexity of anatomical structure of the skull base.[10] The extensive variations in pathology that one may encounter, and the similar imaging appearance and clinical presentation of some entities continue to test the acumen of clinicians. Physical findings like worsening visual field without apparent reason and compressive symptoms like headache should prompt clinicians even at primary care level to investigate further. It is also interesting to note that despite the compressive symptoms, post-surgery ITT was fairly normal. Only when the EC recurred, and Mr. J went for a second surgery did he develop cranial DI and hypopituitarism. The site of the lesion at suprasellar may mimic craniopharyngioma which may cause difficulty in diagnosis. Moreover, craniopharyngiomas accounts for 6% of brain tumors in children while EC only constitutes 1 to 2% of brain tumors – highlighting diagnosis challenge. In summary, revisiting HPE sample when encountering diagnosis dilemma and prompt intervention will be able to prevent complications and provide comfort for the patients.

ACKNOWLEDGEMENT

The authors would like to thank the patient for agreeing to publish this case.

DECLARATION

The authors have no conflict of interest to disclose.

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