

# A rare epidermoid cyst occurring in an young female: a case report and literature review

Roxana Dumitriu (✉ [roxanaeu@gmail.com](mailto:roxanaeu@gmail.com))

National Institute of Endocrinology C. I. Parhon <https://orcid.org/0000-0003-4568-2714>

Roxana Dușceac

National Institute of Endocrinology C. I. Parhon

Anda Dumitrașcu

National Institute of Endocrinology C. I. Parhon

Cătălina Poiană

National Institute of Endocrinology C. I. Parhon

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## Case Report

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# Abstract

## Background

Epidermoid cysts are very rare benign lesions of epithelial origin, slow growing extra-axial tumours and they accord for 1% of all intracranial tumours. They frequently occur at the cerebellopontine angle and parasellar regions insinuating brain structures. They are often associated with a high rate of residual tumor and recurrence.

## Clinical Case

We present the case of a 18 year old patient who was admitted in the our clinic for secondary amenorrhea. The CT scan showed a large median mass with a mixed structure (calcareous and cystic) resembling a craniopharyngioma. Preoperatively the hormonal profile showed deficiency of all anterior pituitary hormones. Transsphenoidal surgery was performed and the histopathological exam established the diagnosis of sellar epidermoid cyst with parasellar extension. Postoperatively the patient developed iatrogenic Cushing syndrome (due to large amounts of dexamethasone given) and diabetes insipidus.

## Conclusion

Epidermoid cysts are challenging tumours because of the extension along the subarachnoid spaces around delicate neurovascular structures therefore they are difficult to be completely resected. The particularity of this case stands in the rare occurrence of this types of tumours and the fact that although in these patients with hypopituitarism diabetes insipidus is present primarily our patient developed postoperatively.

# Background

Epidermoid cysts are lesions which develop from the neuroectodermal epithelial cells. They are benign lesions rarely undergoing malignant transformation (1). The intracranial location is uncommon, it can occur only in 1.5% of all epidermoid tumours and 0.3–1.8% of all intracranial tumours (2). In the majority of the cases the initial presentation is related to its local mass effect such as: hearing impairment, trigeminal neuralgia, facial palsy, diplopia, headaches, seizures etc (3). Surgical total resection remains the first line therapy, however this may be not possible as the cyst wall can be adherent to neurovascular structures. the most common complications after resection are: chemical meningitis, hydrocephalus, infectious meningitis and cranial nerve palsies (4). They can be found in the subarachnoid space, at the cerebellopontine angle and parasellar space (4). Only 3 epidermoid cysts involving the pituitary infundibulum have been reported in the literature ( 5,6,7). We report a case of a cerebral epidermoid cyst of the median line resected by a transsphenoidal approach. The patient, a young female, was admitted in our clinic for amenorrhea.

# Case Presentation

A 18 year old female, non-smoker, presented for secondary amenorrhea (lack of menses for 1 year and 4 months). The patient had no family history and no relevant medical history, particularly no cranial trauma.

At the clinical exam the patient was overweight (BMI = 28,1 kg/m<sup>2</sup>), normotensive neurologically intact excepting right visual field impairment confirmed by the ophthalmological examination.

Laboratory evaluation revealed mild hyperlipemia and the hormonal profile revealed the presence of panhypopituitarism - hypothyroidism, hypogonadism. The visual field chart of the right eye showed temporal field loss and bitemporal relative scotomas.

Differential diagnosis was made with: arahnoid cysts, dermoid cysts (often fat density due to sebum, located along the midline), neurocysticercosis (smaller, may be multiple), cystic tumours (acoustic schwannoma or craniopharyngioma), neuroenteric cysts or Rathke's cleft cyst.

Pre and post contrast scans were performed including dynamic imaging through the pituitary fossa. A complex cystic lesion is seen in the suprasellar region - a mixed structure (calcarous and cystic) with a maximum diameter of 2.76/1.95/2.17 cm, resembling a cranyopharingioma (Fig. 1). The patient was discharged with the recommendations for neurosurgery and resection of the tumour, treatment with levothyroxine 50 micrograms daily and 5 mg of prednisone. After the neurosurgical exam an MRI (magnetic resonance imaging) exam was performed which confirmed the intracranial mass in the sellar and suprasellar space of 23/18/23 mm with extension at the third ventricle. The transsphenoidal surgery was performed with the removal of the cerebral mass. The histopathological exam confirmed the diagnosis of epidermoid cyst. The high suspicion that had arisen after the CT scan was the main key of the diagnosis. The first suspicion was of a craniopharyngioma and only after surgery and histopathological exam the diagnosis could be established.

# Postoperative Evolution

Two weeks after surgery at follow-up the patient had polyuria and polydipsia (liquid intake of 6 l per day), therefore the therapy with desmopressin was initiated (60 micrograms twice per day). After two months the patient was readmitted in the endocrinology department for severe fatigue, weight gain (18 kg in 2 months), polyarthralgia, red stretch marks which appeared on the skin of the hands, armpits, thighs, inferior abdomen, hirsutism, palpitations. On clinical exam the patient was obese (BMI = 32,81 kg/m<sup>2</sup>), moon face, normotensive, elevated heartbeat of 114 beats per minute. Laboratory test showed hyperlipemia and mild hepatic cytolysis. The hormonal confirmed the Cushing syndrome most probably iatrogenic due to large amount of dexametazone given postoperatively in the neurosurgical department and panhypopituitarism with adequate thyroid substitution for hypothyroidism with levothyroxine 50 micrograms daily, hypogonadism. (Table 2).

Table 1  
Hormonal profile – before surgery

<b>freeT4</b>	<b>9.14</b>	<b>9–19 pmol/l</b>
TSH	2.84	0.5–4.5 microUI/ml
ACTH	11.90	3–66 pg/ml
8.00 am Cortisol	1.52	6.7–22.6 microgr/dl
estradiol	< 20	pg/ml
FSH	0.46	mUI/ml
LH	< 0.2	mUI/ml
Prolactin	76.77	3.4-26.72 ng/ml
IGF-1	114.80	117–436 ng/ml

Table 2  
Hormonal profile – after surgery

<b>freeT4</b>	<b>11.4</b>	<b>9–19 pmol/l</b>
ACTH	2.74	3–66 pg/ml
8.00 am Cortisol	0.294	
estradiol	< 20	pg/ml
FSH	< 0.2	mUI/ml
LH	< 0.2	mUI/ml
Prolactin	50	3.4-26.72 ng/ml
IGF-1	189.1	117–436 ng/ml

The cause of the amenorrhea was the secondary hypogonadism so estrogen-progestative treatment was initiated, the prednisone was stopped and the treatment with levothyroxine and desmopressin for the diabetes insipidus was continued. The optic chiasmatic syndrome was partially resolved. Two years after surgery the cerebral IRM showed a tumour recurrence, a mass of 21/16/14 mm.

## Discussion

Epidermoid cysts are congenital lesions resulting from sequestration of embryonic components of the epithelial tissue. 37,3% are located in the cerebellopontine angle (the most common location) followed by parasellar, middle fossa and the spinal channel. Long-term survival is good, the rate of malignancy is very low.

One meta-analysis which included 508 reports identified only 11 pertinent cases (5). Only one case was located in the pituitary stalk underwent endoscopic endonasal transsphenoidal approach of the suprasellar region without a complete resection due to the adherence to the optic chiasm and optic nerves. This is the first case of an epidermoid cyst of the pituitary stalk with successful complete removal with a combined suprasellar and infrasellar approach. In the same meta-analysis the most common clinical presentations were: amenorrhea, diplopia or vision loss, The most common imaging findings showed on the CT scan hypodense lesions and the presence of calcifications (Table 3).

Table 3  
Literature review of intrasellar and suprasellar epidermoid cysts (6)

Authors & Years	Age	Presentation	Imaging Characteristics	Operative Approach	Preoperative Visual Fields	Postoperative Visual Fields	Preop. Hormone Status	Postop. Hormone Status
Costa et al	27, F	Amenorrhhea, Galactorrhea, polyuria, polydipsia	Mixed signal, bilobate, suprasellar cystic mass	Endoscopic extended transphenoidal approach	Decreased acuity in the left eye	Return to baseline	Hyperprolactinemia	Not reported
McCormack et al	36, F	Headache, diplopia, nausea/vomiting	Loculated suprasellar cystic mass	Endoscopic endonasal extended transphenoidal approach	Bilateral enlarged blind spots	Not reported	Not reported	Transient diabetes insipidus (resolved by day 7 postop)
Khan et al	55, M	Vision changes	Mixed signal, lobulated suprasellar mass	Endoscopic endonasal approach	Bitemporal hemianopsia (left > right)	Subjective improvement	Normal	Panhypopituitarism
Nakassa et al	54, F	Headache, visual disturbance	Mixed signal lobulated suprasellar mass	Endoscopic endonasal approach	Decreased acuity (left > right)	Subjective improvement	Diabetes insipidus, mild hyperprolactinemia	Irreversible diabetes insipidus

Cystic epithelial tumours of the sellar region is a continuum of disease resulting from an abnormal Rathke pouch development. In the literature we found 4 epidermoid cysts that had mixed histopathologic features similar to craniopharyngioma (7). Our patient had imaging characteristics similar to a craniopharyngioma.

Chronic endocrine disturbances can be the presenting complaints of a suprasellar epidermoid cyst. The suprasellar cistern is a cerebrospinal fluid-filled space below the third ventricle with a floor formed by the dura of the diaphragm sellae.

Generally suprasellar epidermoid cysts have been excised by transfrontal craniotomies, but in the last years a less invasive transnasal endoscopic approach has been adopted. The first craniotomy for a suprasellar epidermoid cyst was reported by Sadeh et al (8) when a subtotal resection was performed. A systematic review (Huo et al) showed that in the majority of cases reported in the literature of epidermoid cysts treated by combined endoscopic endonasal and endoscopic transphenoidal technique the short term outcomes included improvement vision, but in some cases visual loss persisted (Prasad et al), another patient remained on corticosteroid and thyroid replacement therapy (Oge et al), pituitary insufficiency remained but vision acuity was recovered (Eliash et al) – transfrontal surgical approach at in the majority of cases there was no recurrence identified.

Comparing different surgical approaches open craniotomy versus the endoscopic approach: the transnasal approach provides a direct view of the optic apparatus and avoids potential damages of the cranial nerves and arteries, produces good patients outcomes with reduces complications rates, but versus the first method the success can be used only in small tumours without extensive adherence to adjacent structures, it may not allow maximal exposure and no complete resection, and it can be associated with a higher tumour recurrence rate.

No other cases where reported in the literature, as we know, that describe suprasellar epidermoid cysts with pituitary insufficiency, diabetes insipidus and iatrogenic Cushing postoperatively.

Intracranial epidermoid cysts are benign lesions of epithelial origin that most frequently present with symptoms of mass effect. They are rarely resected completely due to the adherences to other anatomical structures and tumours that involve the pituitary stalk are very challenging due to the high risk for postoperative endocrinopathies.

## Conclusions

We report a rare case of a young female with a sellar epidermoid cyst with suprasellar extension with a typical clinical presentation: amenorrhhea and visual impairment. This case is unique in all literature due to the association of pituitary insufficiency, diabetes insipidus and postoperative iatrogenic Cushing syndrome. Overall the evolution of the patient was favorable.

## Abbreviations

CT  
computed tomography  
MRI  
magnetic resonance imaging

## Declarations

Ethics approval and consent to participate

We, the authors, confirm that informed consent has been obtained from the patient for participation for the case report: A rare epidermoid cyst occurring in a young female: a case report and literature review. The presented case was approved by the ethics committee of the National Institute of Endocrinology C. I. Parhon, Bucharest.

#### Consent for publication

We, the authors, confirm that informed consent has been obtained from the patient for publication for the case report: A rare epidermoid cyst occurring in a young female: a case report and literature review, and accompanying images.

#### Availability of data and materials

Not applicable

#### Competing interests

No competing interests

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#### Authors's contribution

Roxana Dumitriu, Roxana Dușceac, Anda Dumitrașcu, Cătălina Poiană had equal contribution to the manuscript (clinical examination, follow-up of the patient, interpretation of data). All authors read and approved the final manuscript.

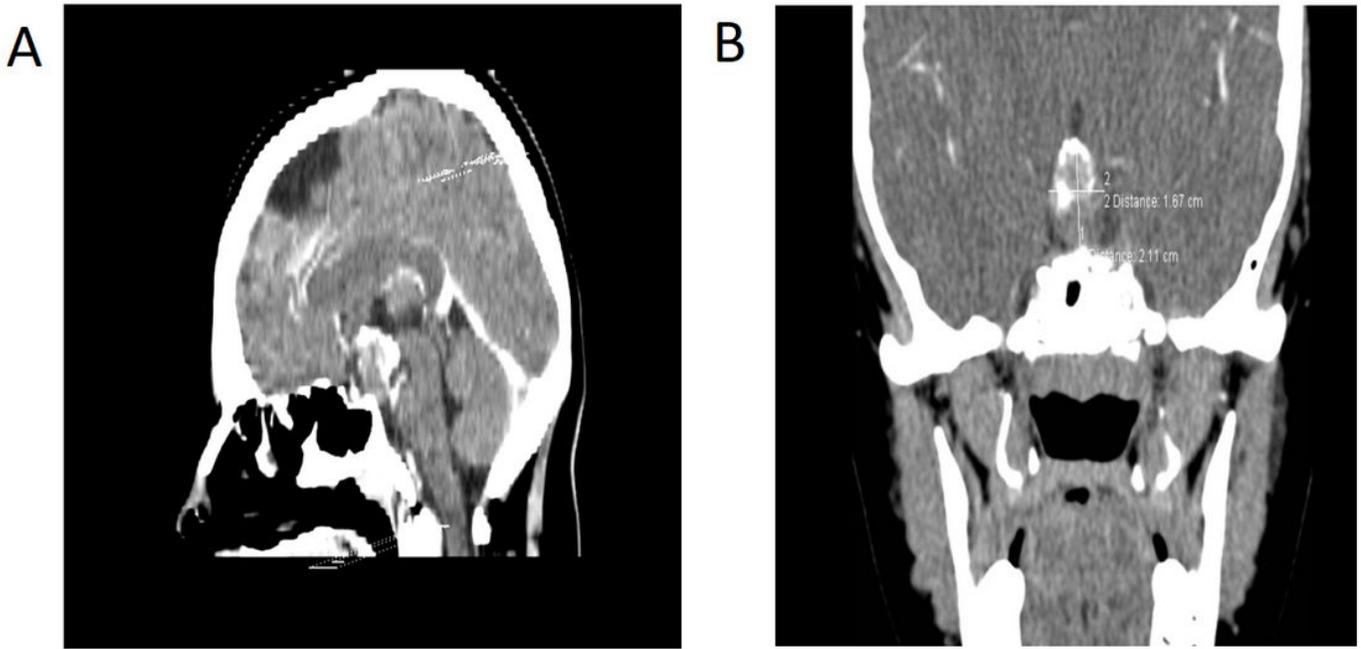
#### Acknowledgements

Nothing to acknowledge.

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## Figures



**Figure 1**  
Pre and post contrast scans were performed including dynamic imaging through the pituitary fossa. A complex cystic lesion is seen in the suprasellar region - a mixed structure (calcarous and cystic) with a maximum diameter of 2.76/1.95/2.17 cm, resembling a cranyopharingioma.

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