# Giant craniopharyngioma in an adult presenting with new onset seizure

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## DESCRIPTION

A 43-year-old man presented to the emergency department after experiencing a first episode seizure. No further information was available at presentation as the patient was confused on his arrival. On physical examination, vital signs were normal. The patient was confused, and his Glasgow Coma Score was 12(E3V4M5). Initial workup included complete blood count, electrolytes and ECG were normal. Thyroid function tests, insulin growth factor-1 and plasma random cortisol level were within the normal range. Testosterone levels were low, and luteinising hormone was inappropriately normal. CT demonstrated a cystic mass with peripheral calcification measuring 6.0×4.5×3.9 cm. The lesion originated from the pituitary fossa and expanded superiorly distorting the third ventricle and the left lateral ventricle causing hydrocephalus (figure 1). A pituitary MRI confirmed these findings (figure 2). The patient underwent a craniotomy with mass resection. The procedure was uneventful. Pathology demonstrated nodular whorls and irregular trabeculae of well-differentiated squamous epithelium, bordered by palisading columnar cell consistent with an adamantinomatous craniopharyngioma (CP) (WHO grade I) (figure 3). Postoperatively, the patient developed panhypopituitarism, and on close monitoring, he was noted to have hypothalamic dysfunction manifesting with hypothermia, hypoglycaemia and adipsia.

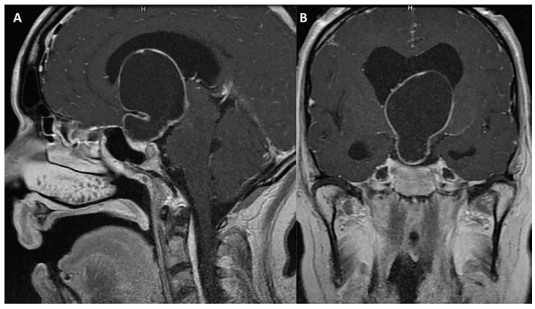


**Figure 2** Sagittal (A) and coronal (B) sections of T1 MRI of the head demonstrates a cystic mass with minimal enhancing solid components and calcifications (white arrow) in the sella region with suprasellar extension. The mass measures 6.01×4.52×4.07 cm and extends superiorly into the third ventricle, the body of the left lateral ventricle and right parasellar causing hydrocephalus.

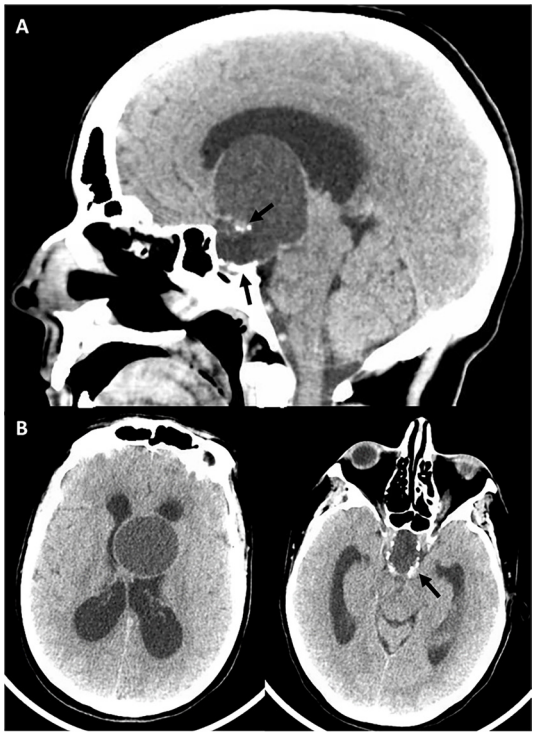


**Figure 1** Sagittal (A) and axial (B) sections of CT scan of the head demonstrate a cystic mass lesion with peripheral calcification (black arrows), measuring 6.0×4.5×3.9 cm, originating from the pituitary gland, expanding into the suprasellar cistern, interpeduncular cistern and distorting the floor of the third ventricle with significant compromise of foramen of Monroe, resulting in bilateral ventricular dilatation.

CPs are benign, slow-growing epithelial tumours that derive from ectodermal remnants of Rathke's pouch. They represent 2%–5% of all primary intracranial tumours with an incidence rate of 1–2 cases per million a year. About 30%–40% of cases present during adolescence, while around 25% are diagnosed in patients over the age of 25.[1] Over 60% of cases arise from within the sellar and presenting symptoms result from compression to the surrounding structures. Clinical manifestations include visual defects, pituitary insufficiency, ocular motor nerve palsies and symptoms of increased intracranial pressure. Two pathological types of CP are recognised: adamantinomatous and papillary CP. Adamantinomatous CPs can be seen in patients of all ages while papillary CP are more common in adults.[1] On imaging, calcification of a tumour is seen in 60%–80% of CP. The mean average tumour size at the time of presentation is 3 cm in adults[2] and 5 cm in children.[3]

Giant CPs, described as a tumour larger than 5–6 cm on their largest axis, are more common in children. They are usually found as adamantinomatous CP and can range from 5 to 11 cm.[2] In adults,



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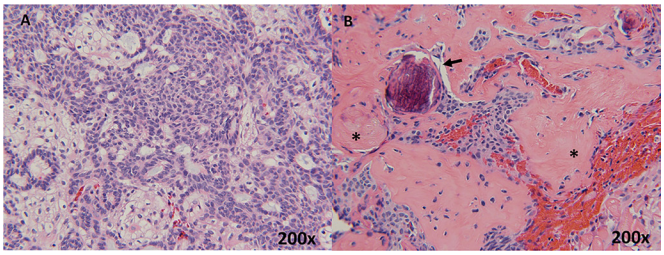


**Figure 3** H&E stain of pituitary mass demonstrated (A) nodular whorls, lobules and irregular of well-differentiate trabecular squamous epithelium bordered by palisaded columnar epithelium. (B) There are multiple nodules of wet keratin (black asterisk) and stellate reticulum (black arrow) which are classic histological feature of the adenomatous craniopharyngioma.

giant CPs are extremely rare and infrequently reported in the literature. Large tumours are associated with worse prognosis due to severe mass effect and residual neurological sequelae. Seizures as a manifestation of pituitary lesions are rare, but more commonly seen with large tumours. The mechanism triggering seizures is not fully understood. It has been suggested that it may relate to mass effect exerted over the suprasellar and/or temporal areas, eliciting an epileptic discharge. Regardless of the tumour size, CPs rarely undergo malignant transformation.[1] Treatment includes resection and possible adjuvant chemotherapy.

## Learning points

- Craniopharyngiomas are usually benign and their morbidity usually results from damage of surrounding structures due to compression.
- In adults, the average tumour size is 3 cm, and giant craniopharyngiomas are rare.
- Larger tumours are more likely to compress surrounding structures and associated with worse outcomes.

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