Case Report

A rare case of giant cystic adamantinomatous craniopharyngioma in an adult

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A B S T R A C T

Craniopharyngioma is an uncommon intracranial tumor that primarily occurs in the sella turcica. Giant cystic craniopharyngioma is rare in general and extremely rare in adults. We report a rare case of giant cystic craniopharyngioma in the anterior pontine cisterna and suprasellar cisterna. A 27-year-old man presented with double vision, and craniocerebral MRI revealed cystic masses in the anterior pontine cisterna and suprasellar cisterna. The masses were removed surgically and diagnosed as large cystic craniopharyngiomas by pathology and MRI. Giant cystic craniopharyngioma is rare in adults. Through this case report, we hope to increase awareness of this disease among various clinicians, including radiologists.

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Introduction

Craniopharyngioma is a rare intracranial tumor that mainly originates from residual epithelial cells in Roethke’s pouch and is mostly found in the sella turcica [1]. Giant posterior fossa cystic craniopharyngiomas (GPFCCPs) are rare and more common in children than adults [2-4]. Clinical symptoms of GPFCCPs mainly include increased intracranial pressure, headache, vomiting, and visual impairment [5]. Here, we report the case of a 27-year-old man with a brain tumor and visual impairment.

**Abbreviations:** MRI, magnetic resonance imaging; CT, computed tomography; WHO, World Health Organization; CNS, central nervous system; GPFCCPs, giant posterior fossa cystic craniopharyngiomas; DWI, diffusion-weighted imaging; ACP, adamantinomatous craniopharyngioma; PCP, papillary craniopharyngioma.

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Case report

A 27-year-old male came to the hospital complaining of “dou-
ble vision for 1 month with aggravated symptoms for half
a month.” Cranio-cerebral MRI at our hospital showed cystic
masses in the anterior pontine and superior sella turcica cisterns; these masses were approximately 52 mm × 26 mm × 61
mm, with an irregular shape and a clear edge (Fig. 1). The
T1-weighted imaging and diffusion-weighted imaging signals
were low, and the T2-weighted imaging signal was high; no
significant enhancement was observed in the enhanced scan,
and the vertebra basilar artery and branch arterioles were vis-
ible in the interior.

Intraoperatively, the tumor was localized to the slope of the
anterior pontine cistern, with the top of the mass at the bot-
tom of the third ventricle, the bottom at the anterior margin
of the foramen magnum, and the sides at the level of the inner
auditory canal. After the tumor capsule was cut, yellow fluid
containing cholesterol crystals flowed out.

Postoperative pathology was performed (Fig. 2). The tumor
was cystic, covering a thin layer of epithelium. Some sections
were solid and lobulated, with palisade, polyhedral epithelial
basal cells in the middle; some of these cells were arranged
in a circular fashion. “Wet” keratin nodules, focal calcifica-
tion, hyperemia, and edema mesenchyme were also observed,
along with considerable lymphocyte infiltration. The initial
diagnosis based on occupation of the right brainstem was
adamantinomatous craniopharyngioma (ACP), and the addi-
tion of right brainstem imaging data led to a diagnosis of giant
cystic ACP.

Discussion

Craniopharyngioma is a rare intracranial epithelial tumor
with an estimated incidence of 0.13 cases per 100,000 per
year. The age distribution is bimodal, peaking in childhood
(5-14 years) and late middle age (50-74 years) [5,6]. Cranio-

Fig. 1 – (a) Low sagittal T1WI signal (white arrow); (b) High coronal plane T2 flair signal (thick white arrow); (c) No enhancement in the cross-section T1 enhancement (black arrow); (d) Low DWI signal (thick black arrow). DWI, diffusion-weighted imaging; T1WI, T1-weighted imaging.
pharyngioma grows along the midline, mainly in the sella turcica, with some involvement of the suprasellar region; it can also present simultaneously in both the sellar and suprasellar regions [7]. Occasional cases have been reported in the nasopharynx, third ventricle, and lateral ventricle [8,9]. In this case report, we describe a young male with a lesion in the anterior bridge pool. Both the age at diagnosis and the tumor location are rare.

Craniopharyngioma is divided into 2 types, ACP and papillary craniopharyngioma (PCP); ACP is more common in children, and PCP is exclusive to adults [10]. This case of ACP pathology in a young male is extremely rare.

MRI scans are complicated to perform, and the results are varied due to the heterogeneous histopathological manifestations of ACP. The typical imaging features of ACP are a high T1 signal, while CT shows calcification. In adults, 70.6% of ACP cases show calcification, and 58.8% have a high T1 signal. Craniopharyngioma with high T1 signal intensity can be secondary to high protein or cholesterol levels, mild calcification or bleeding [11,12]. In this case, MRI revealed a low T1 signal and a high T2 signal. Combined with histopathology, the data indicated a small number of cystic lesions that did not have a high protein or cholesterol concentration; rather, there was only a small amount of protein, so the mass manifested as long T1 and T2 signals.

Craniopharyngiomas are histologically benign (WHO grade I) but considered clinically aggressive due to their tendency to recur [9]. Because most craniopharyngiomas exhibit benign biological behavior, surgery is preferred [13]. According to the location, tumor size, and tumor growth direction, the clinicians can select from several surgical options, including endoscopic transsphenoidal dilatation or endoscopic endonasal approach for pediatric patients or craniotomy [14].

For patients who are candidates for surgery, total resection or subtotal resection plus radiotherapy are usually performed [15]. For craniopharyngioma patients with challenging recurrence or resection, treatment with adjuvant therapy, such as radiotherapy, chemotherapy, and targeted therapy; or palliative therapy, such as vesicu-lar aspiration, is an option [16]. Radiotherapy includes external radiotherapy, such as highly focused methods of intensity-modulated photon and proton therapy, and internal radiotherapy (90Y and 32P) [17,18]. For recurrent cystic craniopharyngioma, chemicals can be injected into the lumen to reduce cystic fluid secretion and control tumor growth [19]; the commonly used drugs are bleomycin and interferon D. It has been reported that tumors with aggressive characteristics have a higher recurrence rate and increased morbidity and mortality. Metabolic syndrome and hypothalamic obesity are the leading causes of death among patients with cystic craniopharyngioma. Hormone replacement therapies, including amphetamine derivatives and oxytocin supplementation, are potential therapeutic options for these patients.

Recently, scholars have detected BRAF V600E mutations in 95% of PCP patients and CTNNB1 mutations in 75%-96% of ACP patients [20]. Treatment with inhibitors targeting BRAF V600E was recently reported to be associated with the shrinkage of recurrent craniopharyngioma. Preoperative treatment with mutant BRAF inhibitors can reduce tumor size and thus facilitate subsequent surgery or radiotherapy. The BRAF V600E mutation can be predicted by the characteristics of craniopharyngioma in MR images. The BRAF V600E mutation was not considered in this case due to the imaging features [21].

The average size of craniopharyngiomas in adults is 3 cm, and GFCCPs >6 cm in diameter or >60 ml in volume are rare and mostly occur in children [17]. Large craniopharyngiomas have been reported to extend forward to the sphenoid sinus and nasopharynx, rarely along the slope and backward to the cerebellopontine angle. Giant craniopharyngioma in adults has been reported only a few times, but giant ACP in adults has not been reported previously. In this case, the lesion was approximately 52 × 26 × 61 mm, indicating an extremely rare adult giant ACP.

**Conclusion**

Giant cystic ACP is rare in adults. The age, imaging manifestations, lesion location, and lesion size in this patient were all rare. It is hoped that this case report will provide more

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**Fig. 2**  Tumor pseudopodia extend deep into the surrounding brain tissue and show wet keratin nodules (white arrows in panels e and f). The basal cells of the tumor epithelium are palisaded (f).
knowledge to clinicians, including radiologists, to improve the accuracy of diagnosing this rare disease.

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