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Case Report

Isolated fourth ventricle craniopharyngioma: Representative case illustration and review of literature

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ABSTRACT

Background: Sella and suprasellar areas are frequently affected by craniopharyngiomas. In this article, eight occurrences have been documented. One is new, and the remaining seven are from previously published articles. Their prevalence in the posterior fossa without expansion from the suprasellar area is unusual.

Case Description: We present a case of a primary 4th ventricular craniopharyngioma of the posterior fossa in a 16-year-old male with no association with Gardner's syndrome. He presented with sudden deterioration of consciousness level and was diagnosed as having a homogeneously contrast enhancing lesion occupying the 4th ventricle with obstructive hydrocephalus. The patient underwent emergency placement of external ventricular drain followed by complete removal of the tumor on the next day by midline suboccipital craniotomy with telovelar approach. Histopathological features were consistent with the adamantinomatous variety of craniopharyngioma. He had complete neurological recovery and no evidence of tumor recurrence in 1-year follow-up.

Conclusion: The craniopharyngioma in our case was distinct because it was a solid tumor with no cystic component, exhibited homogeneous contrast enhancement in neuroimaging, developed in the fourth ventricle, and reached the level of foramen magnum, features that had rarely been documented previously. We also reviewed the literature on reported cases of 4th ventricular craniopharyngioma to strengthen knowledge in this area and highlight the embryological basis of ectopic craniopharyngioma.

Keywords: Craniopharyngioma, Ectopic craniopharyngioma, Fourth ventricle, Primary posterior fossa

INTRODUCTION

Craniopharyngiomas make up 1.2–4.6% of all intracranial tumors. They are uncommon, benign, extra-axial, largely cystic, and slow-growing epithelial tumors of the central nervous system. They often start in the suprasellar region of the brain's infundibulohypophysial axis and spread to affect the hypothalamus, optic chiasm, cranial nerves, third ventricle, and blood arteries.[1,12]

Although few craniopharyngiomas are seen in the infrasellar, intrasellar, or anterior regions, the tumor is often suprasellar in position. There are two possible types of posterior fossa craniopharyngiomas: recurrence/extension and the exceedingly uncommon initial tumor (*de novo*). A relatively high 50% recurrence rate is seen in craniopharyngiomas. In addition, it has significant rates of survival (65–100%: 10-year survival, 83–96%: 5-year survival) as well as morbidity.[4,7]

In this article, we have described a rare example of a *de novo* fourth ventricle craniopharyngioma that reached the foramen magnum and was completely excised. We also showed the schematic picture of development of craniopharyngioma [Figure 1]. The metencephalon grows into the cerebellum and pons, containing the 4th ventricle can be a possible site for the development of craniopharyngioma due to the abnormal migration of neural crest cells [Figure 2]. In an effort to raise neurosurgeons' knowledge of this unusual occurrence, the goal is to document a very rare case of craniopharyngioma in the mentioned location and to review the literature on this area.

CASE REPORT

History and physical examination

A 16-year-old man who had previously been in good condition arrived at the hospital's emergency with a 30-h history of altered consciousness. He experienced a headache and had trouble walking for four weeks before the presentation. The occipital area was the site of a persistent headache accompanied by nausea, vomiting, and diplopia. His Glasgow Coma Scale was 12 out of 15, and he did not have facial paralysis, papilledema, or any other neurological impairments.

Neuroimaging

A solid lesion measuring 5 cm in the craniocaudal plane and 4.5 × 3.5 cm in the axial plane was detected using magnetic resonance imaging (MRI) [Figures 3a and b]. The lesion extends toward the left cerebellopontine cistern in the axial plane and the foramen magnum in the craniocaudal plane. On T1-weighted imaging, the lesion is heterogeneously hyperintense with a well-defined margin, which becomes mixed intensity in T2-weighted imaging. There are no intrinsic or marginal flow voids within the lesion. After administration of contrast, there was avid homogenous contrast enhancement [Figure 3c]. Mass effect is evident by compression of the brainstem anteriorly, cerebellar hemisphere, and middle cerebellar peduncle laterally and compression and displacement of part of the 4th ventricle toward the right.

Surgical procedure

The first procedure was the emergency external ventricular drain (EVD) insertion since the patient's state of consciousness quickly declined. The patient recovered consciousness after the EVD, and his condition stabilized. He underwent a posterior fossa craniotomy and complete tumor excision by telovelar approach on the next day. After the opening of the dura in a "Y"-shaped fashion, cerebrospinal fluid (CSF) was sucked from the cisterna magna to relax the brain. On gross inspection, the tumor was brownish red, lobulated with numerous engorged blood vessels mimicking a solid variety of hemangioblastoma [Figure 4]. *Enbloc* resection was done following standard microsurgical technique. At the end of resection, a pulsatile

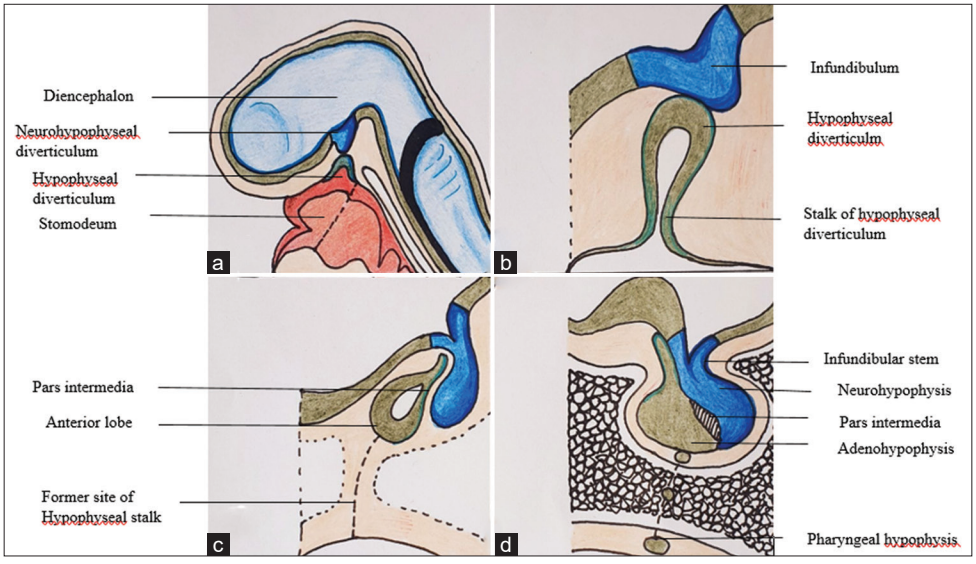

Figure 1: Schematic picture demonstrates (a) development of the pituitary gland from neurohypophyseal diverticulum originating from diencephalon and hypophyseal diverticulum from primitive stomodeum; (b) progressive elongation of the diverticulum gives rise to infundibulum and stalk respectively; (c) after the development of the anterior lobe (adenohypophysis), former site of hypophyseal stalk illustrated; and (d) this remnant of hypophyseal stalk may give rise to ectopic craniopharyngioma.

flow of CSF was observed from the cerebral aqueduct. Hemostasis ensured. Duraplasty is done with a synthetic fabric patch. Bone flap fixed with non-absorbable suture. Layered closure done, leaving no drain tube kept *in situ*. The patient had an uneventful postoperative recovery and was discharged to home on the 5th postoperative day with complete neurological recovery.

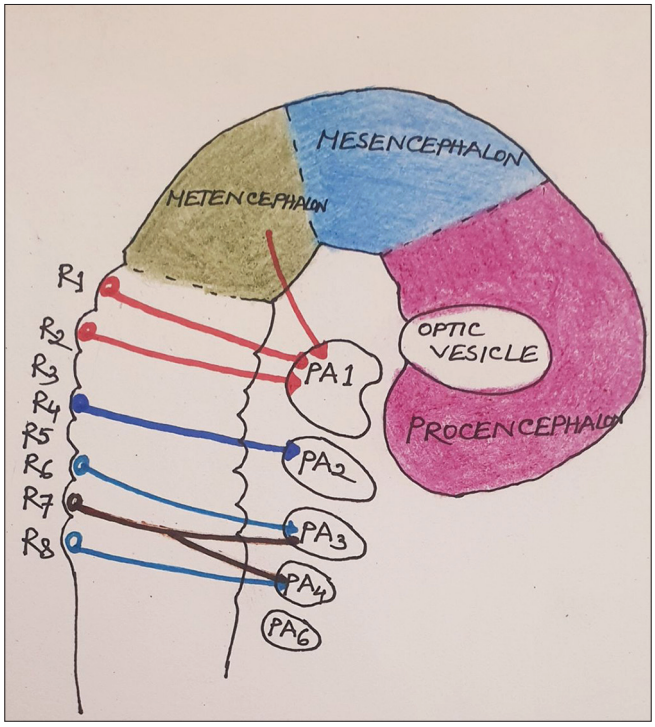

Figure 2: Schematic representation of neural crest cell migration pathway from metencephalon and rhombomeres (R1–R8) toward the pharyngeal arches (PA1–PA6).

Postoperative period and follow-up

A solid lesion with squamous epithelium and isolated peripheral palisading was identified by histopathology. Extensive fibrosis, cholesterol clefts, ongoing inflammation, calcification, cystic alterations, and isolated osteoid development is all seen on the wall. There were isolated areas of reactive gliosis in the cerebellum. These characteristics were indicative of a craniopharyngioma of the adamantinomatous type. The patient's postoperative period was uneventful. An MRI performed after surgery revealed no signs of tumor residual or hydrocephalus [Figure 5]. The patient is still asymptomatic, and there is no sign of a recurrence at the 1-year follow-up.

DISCUSSION

Other than the present case, we have reviewed seven more cases here published between 1996 and 2022 of primary posterior fossa craniopharyngioma located within the 4th ventricle, as shown in Table 1. Males are more commonly carry craniopharyngiomas compared to females, and also more commonly seen among young people.[1,7] This type of tumor is seen among a wide range of age groups; however, it is commonly seen among young people; seven out of eight cases reviewed here are young. Clinical presentation depends on the location of the craniopharyngioma. However, craniopharyngioma of the 4th ventricle may present with cerebellar dysfunction and cranial nerve palsies. Obstructing the 4th ventricle can give rise to hydrocephalus, and the patient may develop rapid deterioration of the consciousness level with other signs of raised intracranial pressure.[1,2,3,6, 9,11,12]

Management

Gross total resection was the surgical technique that was most frequently employed. However, Bashir *et al.* achieved near-total

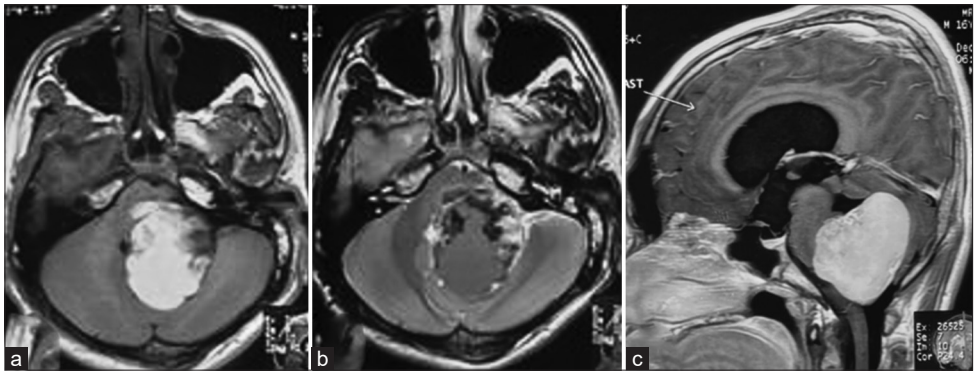

Figure 3: Magnetic resonance imaging of the brain, T1WI axial section demonstrated a 4.5 × 3.5 cm heterogeneously hyperintense lesion occupying the 4th ventricle, extending toward the left cerebellopontine cistern. (a) The mass effect is evidenced by compressing the pons, left middle cerebellar peduncle, and cerebellar hemisphere. (b) The lesion becomes mixed intensity in T2WI, having an irregular hyperintense rim. (c) After administration of contrast, there is intense homogeneous contrast enhancement.

Table 1: Reported cases of isolated 4th ventricle craniopharyngioma to date.

Author	Year	Age/ sex	Association	Presentation	Tumor extension	Extent of resection	Histopathology	Complications	Outcome
Bashir *et al.*[3]	1996	23/M	None	H/A, Papilledema, Cerebellar dysfunction	4th ventricle, Pons, left cerebellopontine angle	NTR	Papillary	None	Complete neurological recovery
Shah *et al.*[11]	2007	12/F	None	H/A, Double vision, Ataxia	4th ventricle , Brainstem, Foramen of Luschka	GTR	Adamantinomatous	None	Complete neurological recovery, including Abducent nerve palsy
Pena *et al.*[9]	2016	20/M	Gardner's syndrome	H/A	4th ventricle	GTR	Adamantinomatous	None	Discharged into subacute rehabilitation, no recurrence at three months follow-up
Alvarez Salgado *et al.*[2]	2017	29/M	Gardner's syndrome	Right-sided facial palsy, double vision	4th ventricle	GTR	Adamantinomatous	None	Not mentioned
Alghatani *et al.*[1]	2018	24/M	None	H/A, double vision	4th ventricle extending down to C1	GTR	Adamantinomatous	None	There is no evidence of recurrence at two years follow-up
Uemura *et al.*[12]	2022	63/F	None	H/A	4th ventricle	GTR	Adamantinomatous	None	No evidence of recurrence at two months follow-up
Present case	2023	16/M	None	H/A, Ataxia	4th ventricle extending down to FM	GTR	Adamantinomatous	None	No recurrence at the one-year follow-up

M: Male, F: Female, H/A: Headache, FM: Foramen Magnum, GTR: Gross total resection, NTR: Near total resection

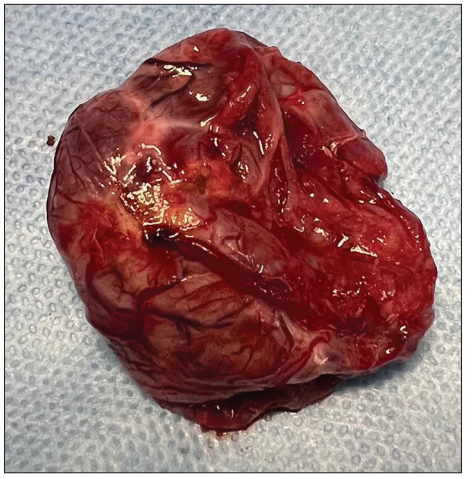

Figure 4: *En bloc* resected specimen showed a lobulated brownish-red tumor with engorged surface blood vessels.

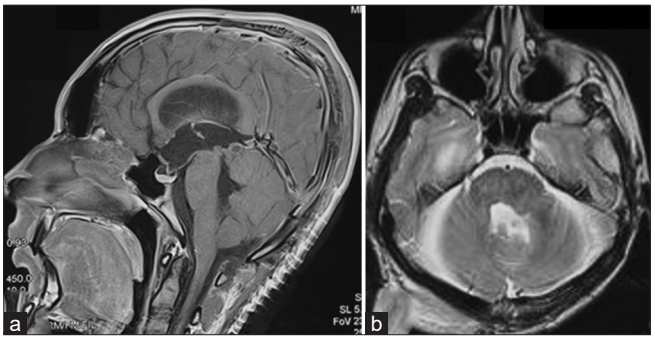

Figure 5: (a) Follow-up magnetic resonance imaging demonstrated complete resection of the tumor with no residual. (b) The tumor mass effect was completely obliterated.

removal of the tumor due to dense adherence of the cyst wall to the brainstem.[3] In this instance, our tumor was devoid of cystic components and not adherent with surrounding neurovascular structures, which facilitated easy peeling of the tumor. With the exception of one papillary example, all of the craniopharyngiomas had an adamantinomatous histology.[1,2,4] In cases that have been documented, the surgical result was outstanding, with complete neurological function recovery. Only three instances included follow-up information and even those where total excision was not possible showed no signs of recurrence in any of the reported cases. Despite the small number of instances that were recorded, we found that fourth ventricular cases had a better prognosis than cerebellopontine angle (CPA) craniopharyngiomas.[1,2,7]

Embryogenesis of ectopic craniopharyngiomas

The origin of craniopharyngioma from the remnant of the obliterated craniopharyngeal duct was first proposed on the basis of the presence of squamous epithelium clusters at the junction of the infundibulum and pituitary in 1904.[8] This

observation explains the embryological basis for the site of ectopic craniopharyngioma in the infrasellar region, including the sphenoid bone and roof of the nasopharynx [Figure 1]. Till now, in the English literature, the reported site of ectopic craniopharyngiomas is the CPA, infrasellar region, petroclival, ethmoid sinus, third ventricle, pineal gland, temporal lobe, frontotemporal area, and orbit.[10] As reported, location involves different regions of the craniofacial area. The proposed hypothesis by John *et al.* denotes that the abnormal migration of the neural crest cells, in addition to mutations of oncogenes and tumor suppressor genes, are responsible for the "*de novo*" origin of ectopic craniopharyngioma.[5,10] From an embryological point of view, neural crest cell migrates from the metencephalon and rhombomeres (R1–R8) toward the pharyngeal arches (PA1–PA6) around the 5th week of gestation following the dorsolateral pathway [Figure 2].[6] During their migration, ectopic cell rest can reside within the metencephalon, dorsolateral pathway, and pharyngeal arches, which later on give rise to ectopic craniopharyngioma. Metencephalon, which later on grows into the cerebellum and pons, containing the 4th ventricle, acts as a primitive location of neural crest cells. After reviewing the remaining seven cases, two cases were adhered with pons,[3,11] whereas all of the cases have a common 4th ventricular location that can explain the basis of "translocation theory."[6]

CONCLUSION

Less than 20 cases of primary posterior fossa craniopharyngioma have been reported, making it an uncommon condition. The rarity of a craniopharyngioma coming from the fourth ventricle is much higher. We have documented the 8th instance of this kind. In the differential diagnosis of CPA and fourth ventricular tumors in adolescent and adult patients, craniopharyngioma should be considered. A good prognosis is linked to complete resection and early diagnosis.

Ethical approval

Not applicable.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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