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# Persistent Craniopharyngeal Canal (Type 3C) with Vertebrobasilar Dolichoectasia and Bilateral Sclerochoroidal Calcification

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## Abstract

The persistent craniopharyngeal canal is a rare, well-corticated midline congenital bony defect through the sphenoid bone between the sellar floor and the nasopharyngeal roof. The prevalence of persistent craniopharyngeal canal is reported to be 0.42%. A 42-year-old male was evaluated for nasal discharge and progressive vision loss; and underwent computed tomography and magnetic resonance imaging, which revealed a large craniopharyngeal canal with ectopic pituitary, lipoma, encephalocele, deformed globe with sclerochoroidal calcification and vertebrobasilar dolichoectasia. The presence of orbital and optic tract malformation, craniofacial anomalies, and tumors can be associated with the craniopharyngeal canal.

**Keywords**
- cephalocele
- lipoma
- craniopharyngeal canal

## Introduction

The persistent craniopharyngeal canal (PCC) is a rare congenital neural crest cell anomaly in the anterior skull base. The prevalence of PCC is reported to be around 0.42%,[1] and commonly presents with headache, diminution of vision, visual field defects, endocrine dysfunction-like hypopituitarism, and meningitis due to cerebrospinal fluid (CSF) leak.[1,2] The association of microphthalmia with colobomatous cyst of the eye, optic chiasm abnormality, ectopic bipartite pituitary, cleft palate, and tumor with various anomalies has been reported with PCC. PCC has been classified into three categories based on the size of the canals which has clinical and prognostic importance.[1,2] Our case of PCC is associated with bilateral sclerochoroidal calcification with vertebrobasilar dolichoectasia, which is very rare.

## Case Presentation

A 42-year-old male with normal birth and developmental history, presented to our department with a history of headache for 15 years, progressive reduction in visual acuity for 14 years, and nasal discharge for 8 years. On examination, vision in his right eye was PL-ve and in his left eye was movement close to his face. A computed tomography scan shows a well-corticated, oval-shaped defect in the sphenoid body measuring 11 mm in anteroposterior (AP) diameter extending from the floor of the sella turcica to the nasopharynx. Sella was widened with fat density −70 Hounsfield units mass lesion with few areas of soft tissue density seen in the nasopharynx, which was communicated with intracranial cephalocele. Calcification was seen along the sclera-choroidal region and within the globe bilaterally,

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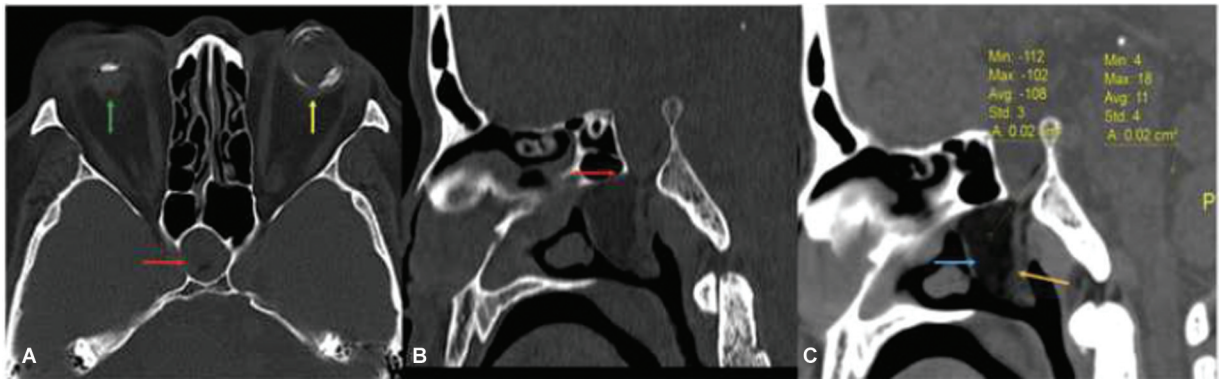


**Fig. 1** Persistent craniopharyngeal canal type 3C computed tomography (CT): Axial (**A**) and sagittal (**B**) reconstructions of the bone window of head noncontrast CT (NCCT). An oval-shaped defect (red arrow, **A**) is seen in the sphenoid body at the sellar floor, measuring 11 mm in anteroposterior (AP) diameter. Widened sella is seen, communicating with the nasopharynx (red arrow, **B**). Bilateral globes show calcification (yellow arrow, green arrow), with phthisis bulbi of the right eye (green arrow). A well-defined, fat-density mass lesion (blue arrow, **C**) is seen in the nasopharynx (−108 Hounsfield units [HU]) along with soft tissue density on the posterior aspect (orange arrow, **C**), consistent with dermoid.

and associated with microphthalmia of the right eye (►**Fig. 1**). On a 3T magnetic resonance imaging (MRI) of the sella, an enlarged and empty sella turcica filled with CSF was seen. A vertical canal extending from the floor of the sella turcica to the nasopharynx was noted. The pituitary gland was located posteriorly and inferiorly in this canal, and a part of it, into the nasopharynx (►**Fig. 2**). The normal T1 hyperintensity of the neurohypophysis was not seen. The MRI findings were consistent with the large craniopharyngeal canal (CPC) associated with the nasopharyngeal extension of the pituitary gland. Membrane-bound CSF-containing sac with pituitary tissue was extending in the nasopharynx, suggestive of a cephalocele. Additionally, a $25 \times 20$ mm well-defined, T1-T2 hyperintense mass lesion was noted in the nasopharynx, attached to the cephalocele, along with a few soft tissue components along the periphery. This mass lesion showed signal suppression on the fat-saturated sequence and no diffusion restriction. Findings

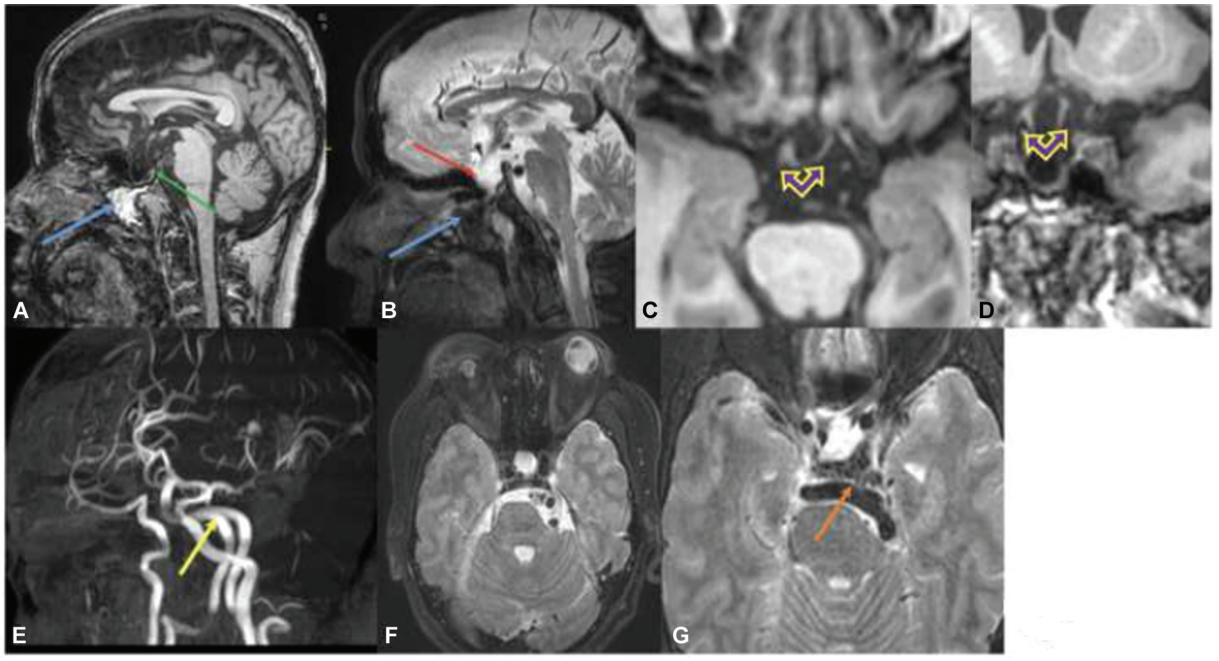


**Fig. 2** Type 3C persistent craniopharyngeal canal magnetic resonance imaging (MRI), showing sagittal T1 turbo spin echo (Sag-T1-TSE) (**A**), sagittal T2 fat suppression (Sag-T2-FS) (**B**), axial T1 (Ax-T1) (**C**), coronal T1 (Cor-T1) (**D**), magnetic resonance angiography (MRA) (**E**), and axial T2 fat suppression (Ax-T2-FS) (**F**, **G**). Midline sections in **A** and **B** show widened sella containing cerebrospinal fluid (CSF) (red arrow), and an elongated pituitary gland (green arrow) with extension into nasopharynx. T1 hyperintense lesion is seen in the nasopharynx, suppressed on fat-saturated sequence (blue arrow) with isointense soft tissue in the posterior wall of the sac, consistent with dermoid. Abnormal redundant partial fused atrophic optic chiasm (double violet arrow) is seen. MRA (**E**) shows dolichoectatic vertebra-basilar system (yellow arrow) with high basilar bifurcation (orange arrow, **G**).

 

were consistent with lipoma. The bilateral vertebral artery V4 segment and basilar artery were dolichoectatic and tortuous, with high basilar bifurcation. Asymmetrically, partially fused, atrophic, and inferiorly displaced optic chiasma was seen (►**Fig. 2**). The patient was counseled about his disease and the future prognosis. Surgery was advised to repair the craniopharyngeal defect. The patient was primarily concerned about his vision, but his vision will not recover after surgery. The patient has not undergone surgery to date. So, we described the imaging finding of such a rare case.

## Discussion

The PCC is a rare, well-corticated defect of the midline sphenoid body extending from the floor of the sella turcica to the roof of the nasopharynx. CPCs can be classified into three types: small incidental canals (type 1), medium-sized canals with ectopic pituitary tissue (type 2), and large canals containing cephaloceles (type 3A), tumors (type 3B), or both (type 3C) (►**Fig. 3**).[1] Hori et al[3] evaluated nasopharyngeal pituitary tissue of fetal specimens and showed adenohypophyseal tissue in a PCC. Type 1 (small size canals), which were delimited by cortical bone and measured (0.7–1.1 mm) in AP diameter. Type 2 (medium size canal) (3–5 mm in diameter) development is associated with ectopic adenohypophysis. In a study by Abele et al,[1] two of seven type 2 CPCs had pituitary dysfunction. A case of type 3A large CPC containing cephalocele and ectopic pituitary tissue was first described by Klinkosch in 1764,[4] in which a neonate had a 5-mm wide passage extending through the middle of the sphenoid that contained prolapsed dura and an ectopic pituitary gland. Recent case reports of large CPCs with cephaloceles containing ectopic pituitary tissue describe canals measuring 10 to 13 mm in diameter.[5] This finding underscores the importance of recognizing cephaloceles and ectopic pituitary tissue in type 3A CPCs to avoid CSF leak and/or pituitary resection during nasopharyngeal mass surgery. As in type 3 CPCs with cephalocele, identifying intracranial extension of a dermoid, teratoma, or glioma into a type 3 CPC is important for surgical planning to avoid inadvertent CSF leak and potential meningitis. PCC is associated with CSF rhinorrhea, meningitis, panhypopituitarism, accidental removal of hypophysis mimicking a midline nasal polyp and hydrocephaly,[6] Rathke's cleft cyst, intrasellar craniopharyngioma, holoprosencephaly, cephalocele, sphenoid teratoma,[7] ectopic pituitary tissue, hyperprolactinemia, hypothalamic hamartoma, corpus callosal dysgenesis,[6] and other anomalies of the skull base. Duplication of the pituitary gland with oropharyngeal teratoma, cleft palate, and other craniofacial abnormalities is associated, termed "syndrome of pituitary duplication – plus."[8] The association of microphthalmia with colobomatous cysts with various anomalies has been reported. PCC results from the defect in the neural crest cell, which has a broad disease association with the coexistence of multiple anomalies including dysraphism.[9] Some patients had congenital anomalies including PHACE (posterior fossa malformations, hemangiomas, arterial abnormalities, cardiac defects, eye abnormalities) syndrome,[10] Blake remnant, duplicated pituitary gland, and congenital cleft lip or cleft palate. In our case, the bilateral eyeball shows calcifications along the wall. The bilateral V4 segment of the vertebral artery and basilar artery is dilated and elongated crossing the midline from left to right with the high bifurcation of the basilar artery.

## Conclusion

Persistence of the CPC is a rare anomaly of the skull base frequently associated with the etiology of transsphenoidal encephaloceles. The surgical indications and approach remain controversial, making each case an individual challenge

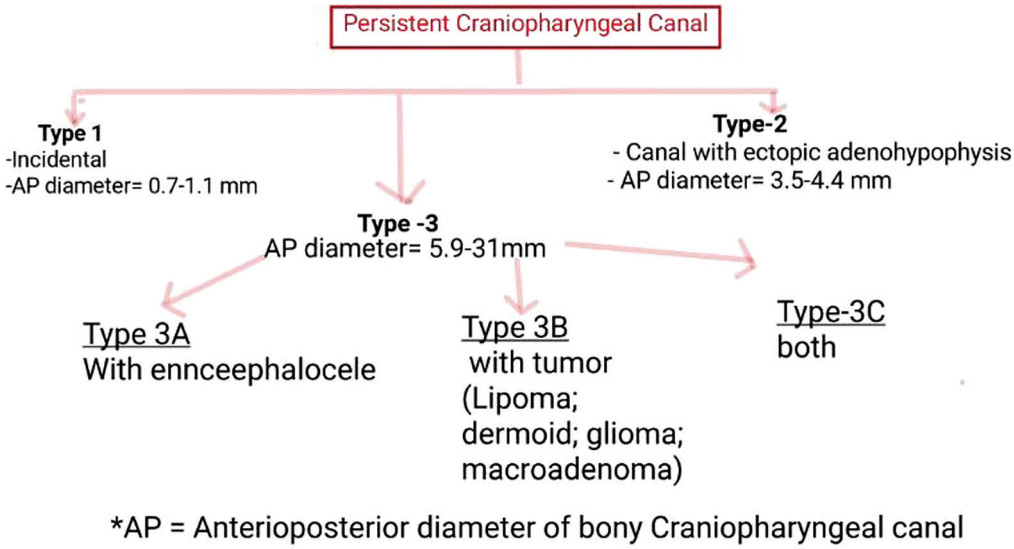


**Fig. 3** Hand-written line diagram demonstrating classification of persistent craniopharyngeal canal (drawn by B.D.C.).

 

according to the anatomical variations, clinical manifestations, and anomalies associated with it.

### Note

A part of the history and images of this patient are published along with other cases series in the original article (DOI: 10.25259/JCIS_87_2023) from the same institute and corresponding author. Here, we want to publish more elaborative literature regarding this anomaly.

### Authors' Contributions

B.D.C., S.J., and S.B.G. contributed to the acquisition, analysis, conception, design, and drafting of the work. All authors have agreed both to be personally accountable for their contributions and ensured that questions related to the accuracy or integrity of any part of the work, even ones in which one was not personally involved, are appropriately investigated, resolved, and the resolution documented in the literature. All authors have read and approved the manuscript.

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Retrospective, consent waiver, adhered to Helsinki guidelines.

### Conflict of Interest

None declared.

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