

# Persistent craniopharyngeal canal with an associated sphenoid sinus fistula

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## ABSTRACT

Persistent craniopharyngeal canal (PCC) is a rare congenital anomaly that appears as a linear well-corticated canal running from the sella through the clivus and into the nasopharynx. Case reports of this anomaly have shown it is associated with a range of craniofacial defects, pituitary abnormalities, and meningoencephaloceles. It predisposes patients to bacterial meningitis. In this case a 46-year-old gentleman presenting for preoperative planning for surgical drainage of Potts Puffy tumor was found to have a PCC on CT and MRI. Imaging also demonstrated the presence of chronic inflammation and a fistula extending from the tract into the sphenoid sinus. This unusual presentation of a PCC with a sphenoid sinus fistula broadens the potential clinical presentations of PCC and further emphasizes the ability of this anomaly to serve as a conduit for CNS infection.

## CASE DESCRIPTION

### CASE DESCRIPTION

A 46-year-old man with a history of chronic sinusitis presented with swelling of the forehead and right eye. The swollen forehead was soft and inflamed; the physical examination was most consistent with Pott's puffy tumor. In anticipation of endoscopic surgical drainage of the associated pyomucocele, preoperative planning CT and MRI of the skull base were obtained.

Unenhanced CT of the skull base demonstrated a corticated tract measuring 4mm in diameter extending through the sphenoid bone from the sella into the nasopharynx. Additionally, a fistula between the tract and the sphenoid sinus was evident (Figures 1,2). Findings of chronic sphenoid sinusitis were present, including osteoneogenesis and mucosal thickening. A contrast enhanced MRI of the skull base demonstrated enhancement along the course of the tract consistent with persistent inflammation. The pituitary gland was unremarkable and there was no evidence of intracranial mass or cephalocele. No findings of meningitis were present.

Because of the risk of meningitis, the patient was urgently treated with functional endoscopic sinus surgery. During the course of the procedure, the patient went into atrial fibrillation

which was not responsive to medications, so the surgery was aborted and the patient underwent needle lavage of the frontal sinuses through the anterior table defect. He returned to the OR several days later following a cardiology consult and a second attempt was made at performing endoscopic sinus surgery. During surgery, however, the surgical field was obscured by bleeding from anticoagulation for atrial fibrillation. A stent was placed in the left frontal sinus, but the sphenoid sinuses could not be addressed given the resultant blood loss. Because the sphenoid sinuses could not be addressed, a PICC was placed for administration of long-term antibiotics given the continued risk for meningitis. At a one month follow up visit the patient was found to be recovering well. After this he was lost to follow-up.

### DISCUSSION

#### Etiology and demographics

In fetal life, the craniopharyngeal canal connects the nasopharynx to the sella through the clivus. It usually involutes completely. A persistent craniopharyngeal canal (PCC) results when involution of the craniopharyngeal canal fails to occur. It is thought that PCC may be a remnant of Rathke's pouch or possibly a remnant of a vascular channel [1,2]. During development of the pituitary, the adenohypophyseal pouch (Rathke's pouch) elongates to form the adenohypophyseal stalk. This structure

traverses the sphenoid bone and is eventually obliterated by development of the post-sphenoid cartilage. Failure of the post-sphenoid cartilage to obliterate the adenohypophyseal stalk is one putative mechanism for development of PCC [1].

PCC is associated with a range of craniofacial defects [3,4], pituitary abnormalities/dysfunction, and meningoencephaloceles [5], and also predisposes patients to bacterial meningitis [6-8]. The term PCC is generally reserved for canals measuring greater than 1.5mm with smaller canals being referred to as persistent hypophyseal canals. PCC is often discovered incidentally and is often asymptomatic. In the first months of life, the canal may not have yet fused, and there is an estimated prevalence of 10%. Thereafter, the prevalence falls to 0.42% [2]. A recent classification system developed by Abele et al. divided PCC into three subtypes, with Type 1 being an incidentally found small PCC, Type 2 a moderate sized PCC with ectopic adenohypophysis, and Type 3 a PCC with a larger defect containing encephalocele, tumor, or both [1].

### CLINICAL AND IMAGING FINDINGS

On imaging, this rare congenital anomaly appears as a linear well-corticated canal running from the sella through the clivus and into the nasopharynx [1]. This anomaly is also associated with a range of craniofacial defects, pituitary abnormalities, and meningoencephaloceles which can also be seen on imaging in association with PCC. In rare cases, PCC can result in development of bacterial meningitis, but is most often asymptomatic.

In this paper we report an unusual case of a PCC with evidence of chronic inflammation and a fistula into the sphenoid sinus. To our knowledge, a sphenoid sinus fistula in the setting of PCC has not been reported previously. On imaging in this case, there is persistent inflammation along the course of the PCC even in the absence of signs of meningitis. Bacterial meningitis is a well-known rare complication of PCC with the canal acting as a conduit for infection from the nasopharynx. It is unclear whether chronic inflammation of the PCC resulted in development of a fistula to the sphenoid sinus, or whether this communication was part of the original anomaly and therefore raises the intriguing possibility that errors in development could result in formation of an additional fistula between the PCC and sphenoid sinus.

In this case, the PCC was discovered incidentally and the patient was asymptomatic. He had no known history of pituitary dysfunction or CNS infection despite evidence of inflammation on imaging. This reinforces that although PCC has the potential to result in CNS infection, this is a rare complication of a rare anomaly and most case reports describing this complication have been in children [6,8,9].

### TREATMENT AND PROGNOSIS

Generally, PCC are treated when there is a history of bacterial meningitis, CSF rhinorrhea, or airway compromise due to nasopharyngeal mass, although controversy exists as to indications for treatment of asymptomatic PCC [9]. When treatment is indicated, PCC are surgically treated using

endoscopic repair with obliteration of the tract. In this report, the patient was unable to undergo obliteration of the tract due to excessive intraoperative hemorrhage. The presence of chronic inflammation within the tract as well as sphenoid sinusitis would argue in favor of treatment given the potential risk for development of meningitis.

### DIFFERENTIAL DIAGNOSIS

There are several entities which can potentially mimic the diagnosis of a PCC: these include sphenoid-occipital synchondrosis, transsphenoidal meningoencephalocele, and a clival canal or canalis basilaris medianus.

### SPHENO-OCCIPITAL SYNCHONDROSIS

One entity which can potentially be mistaken for PCC is sphenoid-occipital synchondrosis. The sphenoid-occipital synchondrosis is a cartilaginous joint which is present at birth. It is located at the juncture of the basisphenoid and basiocciput and generally fuses in late adolescence. In contrast to PCC, sphenoid-occipital synchondrosis is not seen in adults and will not involve the sella. It will also present with varying stages of closure on imaging depending on the age of the individual.

### TRANSSPHENOIDAL MENINGOENCEPHALOCELE

Transsphenoidal meningoencephalocele is a rare congenital malformation that presents as a bony defect in the sphenoid with herniation of an ependyma-lined sac containing CSF and brain tissue [10]. Meningoencephaloceles have been reported to be an associated finding in PCC [5]. Transsphenoidal meningoencephalocele may be considered a part of the spectrum of PCC presentations in which a larger canal defect is present, along with herniated brain tissue. According to Abele et al.'s classification scheme, Type 3 PCCs may present with cephaloceles containing CSF, dura, and ectopic adenohypophysis [1].

### CLIVAL CANAL

A clival canal or canalis basilaris medianus is an anatomic variant that presents as a bony defect in the basiocciput. Typically, this anatomical variant travels through the clivus and presents with either single or multiple openings or as a deep recess (fossa navicularis magna) that does not traverse the full extent of the clivus [11]. In contrast to PCC there is no involvement of the sella.

### TEACHING POINT

PCC often presents as an asymptomatic incidental finding of a well corticated linear canal extending from the sella through the clivus and into the nasopharynx and usually requires no intervention. Rarely, however, PCC can serve as a conduit for infection. When findings of inflammation are present on imaging this serves as an indication for treatment to prevent meningitis.

## AUTHOR CONTRIBUTIONS

Chase Dougherty, MD: drafted and edited the manuscript.  
Barton F. Branstetter, MD: original concept, data acquisition, and supervision of drafting of manuscript.

## CONSENT

Exempt study per IRB

## QUESTIONS

- 1) Which of the following is false regarding craniopharyngeal canals?
  - a. often discovered incidentally
  - b. often asymptomatic
  - c. may represent remnant of Rathke's Pouch or vascular channel
  - d. associated with congenital heart defects (applies)
  - e. may require surgical repair
- 2) Which of the following is NOT a potential complication of craniopharyngeal canal?
  - a. Hemorrhage (applies)
  - b. meningitis
  - c. airway compromise
  - d. CSF rhinorrhea
  - e. Migraine (applies)
- 3) Craniopharyngeal canal is a corticated canal which involves which of the following anatomic structures?
  - a. temporal fossa
  - b. ethmoid
  - c. greater wing of the sphenoid
  - d. sella (applies)
  - e. vomer
- 4) Which of the following answer choices would NOT be included in the differential of a craniopharyngeal canal:
  - a. sphenoid-occipital synchondrosis
  - b. transsphenoidal meningoencephalocele
  - c. clival canal
  - d. canal of Sternberg (applies)
  - e. trauma (applies)
- 5) Craniopharyngeal canal is associated with all of the following EXCEPT:
  - a. craniofacial defects
  - b. ectopic pituitary
  - c. meningoencephaloceles
  - d. optic nerve hypoplasia (applies)
  - e. pituitary dysfunction

## EXPLANATIONS

### Explanation 1:

- a. PCC are often discovered incidentally. [It is thought that PCC may be a remnant of Rathke's pouch or possibly a remnant of a vascular channel].
- b. PCC is often asymptomatic. [PCC is often discovered incidentally and is often asymptomatic.]
- c. may represent remnant of Rathke's Pouch or vascular channel. [It is thought that PCC may be a remnant of

Rathke's pouch or possibly a remnant of a vascular channel]

- d. associated with congenital heart defects [incorrect]
- e. may require surgical repair [When treatment is indicated, PCC are surgically treated using endoscopic repair with obliteration of the tract.]

### Explanation 2:

- a. Hemorrhage [incorrect]
- b. Meningitis [PCC is associated with a range of craniofacial defects,[3,4] pituitary abnormalities/dysfunction, and meningoencephaloceles,[5] and also predisposes patients to bacterial meningitis]
- c. airway compromise [Generally, PCC are treated when there is a history of bacterial meningitis, CSF rhinorrhea, or airway compromise due to nasopharyngeal mass...]
- d. CSF rhinorrhea [Generally, PCC are treated when there is a history of bacterial meningitis, CSF rhinorrhea, or airway compromise due to nasopharyngeal mass]
- e. Migraine [incorrect]

### Explanation 3:

- a. temporal fossa [incorrect]
- b. ethmoid [incorrect]
- c. greater wing of the sphenoid [incorrect]
- d. sella [In fetal life, the craniopharyngeal canal connects the nasopharynx to the sella through the clivus.]
- e. vomer [incorrect]

### Explanation 4:

- a. sphenoid-occipital synchondrosis [refer to differential diagnosis section]
- b. transsphenoidal meningoencephalocele [refer to differential diagnosis section]
- c. clival canal [refer to differential diagnosis section]
- d. canal of Sternberg [incorrect]
- e. trauma [incorrect]

### Explanation 5:

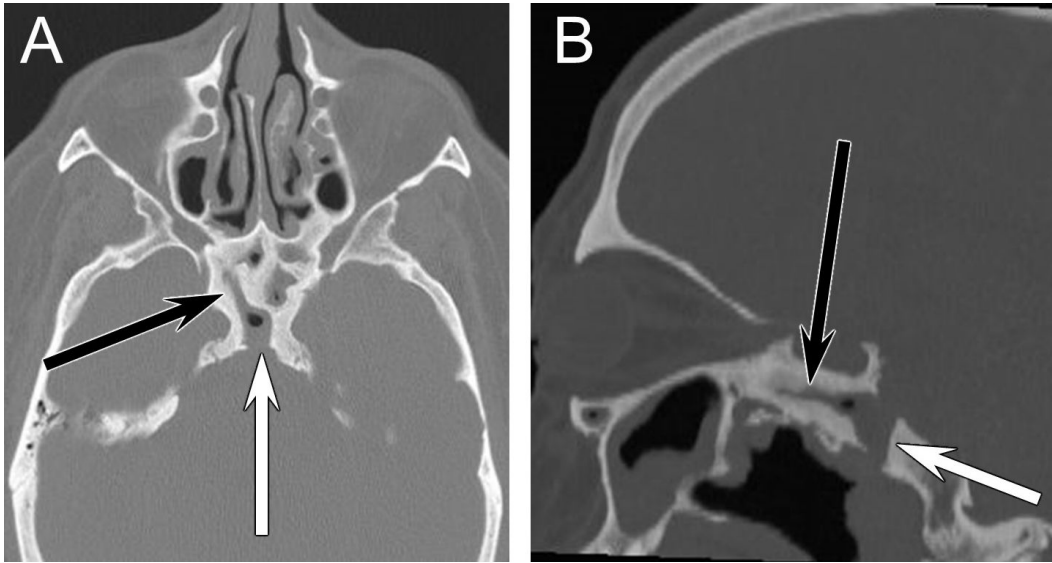
- a. craniofacial defects [PCC is associated with a range of craniofacial defects,[3,4] pituitary abnormalities/dysfunction, and meningoencephaloceles,[5] and also predisposes patients to bacterial meningitis[6-8]]
- b. ectopic pituitary [A recent classification system developed by Abele et al. divided PCC into three types, with Type 1 representing an incidentally found PCC, Type 2 a PCC with ectopic adenohypophysis, and Type 3 presenting with encephalocele, tumor, or both[1].]
- c. meningoencephaloceles [see explanation above]
- d. optic nerve hypoplasia [incorrect]
- e. pituitary dysfunction [see explanation above]

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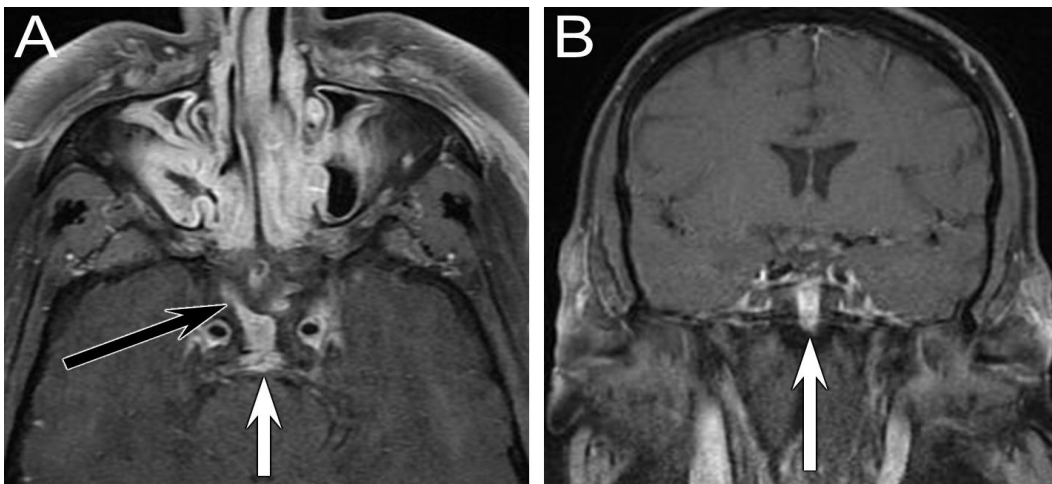
FIGURES



**Figure 1:** 46-year-old man with persistent craniopharyngeal canal and an associated sphenoid sinus fistula

**Technique:** Unenhanced CT scan performed on a GE Lightspeed scanner with kVp 120, mA 200, slice thickness 1.25mm in nonhelical axial acquisition (A), then reformatted into sagittal oblique plane at 0.5mm thickness (B).

**Findings:** There is a vertically-oriented linear defect in the clivus (white arrows) extending from the nasopharynx to the prepontine cistern, along the expected path of the craniopharyngeal canal. There is a tract (black arrow) extending anteriorly from the center of the persistent craniopharyngeal canal into the right sphenoid sinus, which shows evidence of chronic inflammation, including thickened septations and osteoneogenesis.



**Figure 2:** 46-year-old man with persistent craniopharyngeal canal and an associated sphenoid sinus fistula

**Technique:** Contrast-enhanced T1-weighted MRI performed on a 1.5T GE Signa HD with TR 600, TE 11, flip angle 90, NEX 2, 3mm slice thickness and 4mm slice spacing using 28mL gadobenate dimeglumine in axial (A) and coronal (B) planes.

**Findings:** There is a vertically-oriented focus of linear enhancement in the clivus (white arrows) extending from the nasopharynx to the prepontine cistern, along the expected path of the craniopharyngeal canal. There is a tract (black arrow) extending anteriorly from the center of the persistent craniopharyngeal canal into the right sphenoid sinus, which shows evidence of chronic inflammation, including thickened septations and mucosal enhancement.

## SUMMARY TABLE

<b>Etiology</b>	• Thought to represent persistence of Rathke's Pouch or a vascular channel
<b>Incidence</b>	• Prevalence is around 10% in first few months of life and drops to 0.42% thereafter
<b>Gender ratio</b>	• No studies have quantified gender ratio
<b>Age predilection</b>	• More common in the first few months of life and very rare in adults
<b>Risk Factors</b>	• No known risk factors
<b>Treatment</b>	• In symptomatic individuals endoscopic repair with obliteration of the tract is performed. Treatment of asymptomatic individuals is controversial.
<b>Prognosis</b>	• The majority of individuals with PCC are asymptomatic although occasionally PCC may act as a conduit for meningitis.
<b>Findings on Imaging</b>	• Seen on CT as a well corticated canal extending from the sella through the clivus and into the nasopharynx.

## DIFFERENTIAL DIAGNOSIS

<b>DDX</b>	<b>Imaging findings</b>
<b>Spheno-occipital synchondrosis</b>	<ul style="list-style-type: none"> <li>• Presents as an area of linear hypodensity between the basiocciput and basisphenoid with varying degrees of ossification depending on the age of patient.</li> <li>• Fuses in adolescence to form the clivus.</li> </ul>
<b>Transsphenoidal meningoencephalocele</b>	<ul style="list-style-type: none"> <li>• Presents as defect in the sphenoid with herniation of an ependyma lined sac containing CSF and brain tissue</li> </ul>
<b>Clival canal (canalis basilaris medianus)</b>	<ul style="list-style-type: none"> <li>• Well corticated canal or canals which extend through the clivus or present as a deep recess in the clivus</li> </ul>

**ABBREVIATIONS**

PC: Persistent Craniopharyngeal Canal

**KEYWORDS**

Craniopharyngeal Canal, Hypophyseal canal, Skull Base, CT, MRI, sphenoid sinus fistula

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