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CASE REPORT - PEDIATRIC NEUROSURGERY

Bipartite craniopharyngeal canal with a lipoma and cephalocele: a previously unreported entity

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Abstract

A 13-year-old male child was evaluated for headache and visual deterioration; he underwent routine MRI imaging which revealed a large craniopharyngeal canal, divided by an abnormal bony septum giving a bipartite appearance of the canal, with a lipoma and cephalocele on either side of the septum. The child had undergone a previous surgery for cleft palate repair at the age of 7. The child had normal pituitary function inspite of nonvisualization of pituitary gland in MRI. To best our knowledge, this is the first case with such a variation. We have also discussed the possible embryological hypothesis for this previously unreported entity. Knowledge about this rare variant might have surgical relevance in selected cases.

Keywords Canal · Bipartite · Lipoma · Cephalocele · Postsphenoid center

Abbreviations

CPC	Craniopharyngeal canal
MRI	Magnetic resonance imaging
CSF	Cerebrospinal fluid
CT	Computed tomography
FLAIR	Fluid attenuation inversion recovery

Background

Persistent craniopharyngeal canal (CPC) is a well-corticated bony defect which extends from the midline of the sphenoid body to the roof of nasopharynx. It is a rare congenital bony defect involving the skull base [1]. The prevalence of persistent craniopharyngeal canal is reported to be 0.42% [1, 2]. CPC has been classified into three major categories based on the size of the canals which has clinical and prognostic importance [1, 3]. Most common clinical presentations of persistent CPC are headache, diminution of vision, visual field defects, endocrine dysfunction-like hypopituitarism and meningitis due to cerebrospinal fluid (CSF) leak [1–5]. Various midline anomalies like cleft palate and tumors (nasopharyngeal and intracranial tumors) have associations with this entity [1]. We describe a rare variant of craniopharyngeal canal with lipoma and cephalocele on either side of the bony septum giving a bipartite appearance. We also discussed the possible embryological hypothesis of this rare anomaly. To the best of our knowledge, this is the first report of such a condition.

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Case report

A 13-year-old male child with normal birth and developmental history was brought to our hospital with complaints of holocranial headache for the past 1 year. The headache was mild to moderate in intensity and intermittent in nature. It was decreased by the use of oral analgesics. No history of any postural or diurnal variation of headache. He underwent surgery for cleft palate at 7 years of age. He also complained of decreased vision bilaterally with a corrected normal visual acuity on glasses. Visual field examination showed bilateral attenuation of visual fields in the temporal quadrants. Fundus

examination was also normal. Clinical examination at the time of presentation revealed normal higher mental functions. His motor, sensory, and cerebellar examinations were also normal. All the baseline hematological and biochemical investigations were normal so as the hormonal profile. No evidence of pituitary dysfunction was noted. He underwent neuroimaging to rule out any structural cause for the headache. Magnetic resonance imaging (MRI) and correlative CT (computed tomography) images revealed a large persistent craniopharyngeal canal (Fig. 1) with an abnormal bony septum dividing the canal into two hemi-canals (Fig. 2). The abnormal bony septum was seen in continuity with the basisphenoid (posterior sphenoid body). MRI showed T2 and FLAIR hyperintense, lobulated, well-circumscribed non-enhancing lesion in the right side of canal occupying both sellar and suprasellar region (Fig. 3). The lesion measured 3.5 × 2.4 × 4.3 cm (APXTRXCC) in size. Predominant part of the lesion appears hyperintense on T1 and showed signal suppression on fat suppressed images, suggestive of a lipoma (Fig. 1). CT images showed fat attenuation of the lesion (-45 HU) located in the right side of the canal. Left side of the canal showed herniated CSF cavity with soft tissue within, suggestive of a cephalocele (Figs 1 and 2). Pituitary gland and posterior pituitary bright spot were not separately visualized from the lesion.

Neuroimaging findings were consistent with a persistent large craniopharyngeal canal divided by an abnormal bony septum, containing both lipoma and cephalocele on either side of the septum.

In view of evolving visual field constriction, patient was planned for surgery. He underwent neuronavigation guided extended endoscopic approach (EEA). During the procedure, a bony spur was seen causing direct compression of the optic chiasm. The base of the spur was attached to the floor of the sella, and the tip of the spur extended into the third ventricle. A soft tissue mass, later confirmed to be a lipoma was seen on the right side of the bony spur. Anterior wall of canal and part of planum were drilled and widened. The lipoma was dissected away from the adjacent microvasculature and excised. The bone spur was drilled and excised completely. Histopathological examination confirmed the fat lesion as lipoma. Postoperative period was uneventful.

Discussion

Persistent CPC is a well-corticated bony channel of the midline sphenoid body extending from the floor of the sella turcica to the roof of the nasopharynx. Abele et al. classified

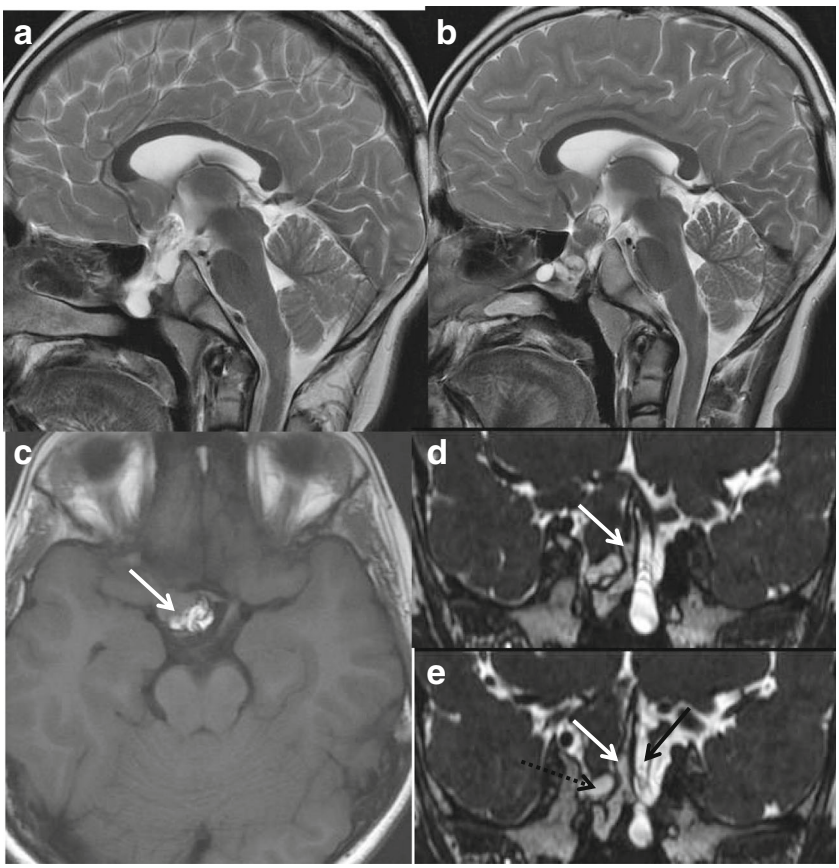

Fig. 1 **a**–**e** Sagittal T2 MRI at midline section shows large craniopharyngeal canal with herniated CSF cavity with some soft tissue structures within suggestive of cephalocele. **b**, **c** Sagittal T2 and axial T1 image shows large craniopharyngeal canal containing well defined lobulated T1 hyperintense lesion suggestive of lipoma (arrow). **d**, **e** 3D Coronal CISS images shows abnormal bony septum (white arrow) dividing the craniopharyngeal canal into two, resulting in the appearance of bipartite CPC. Right side of the canal contains lipoma (dashed arrow) and left side of the canal contains cephalocele (black arrow). The abnormal bony septum is seen in continuity with posterior sphenoid body

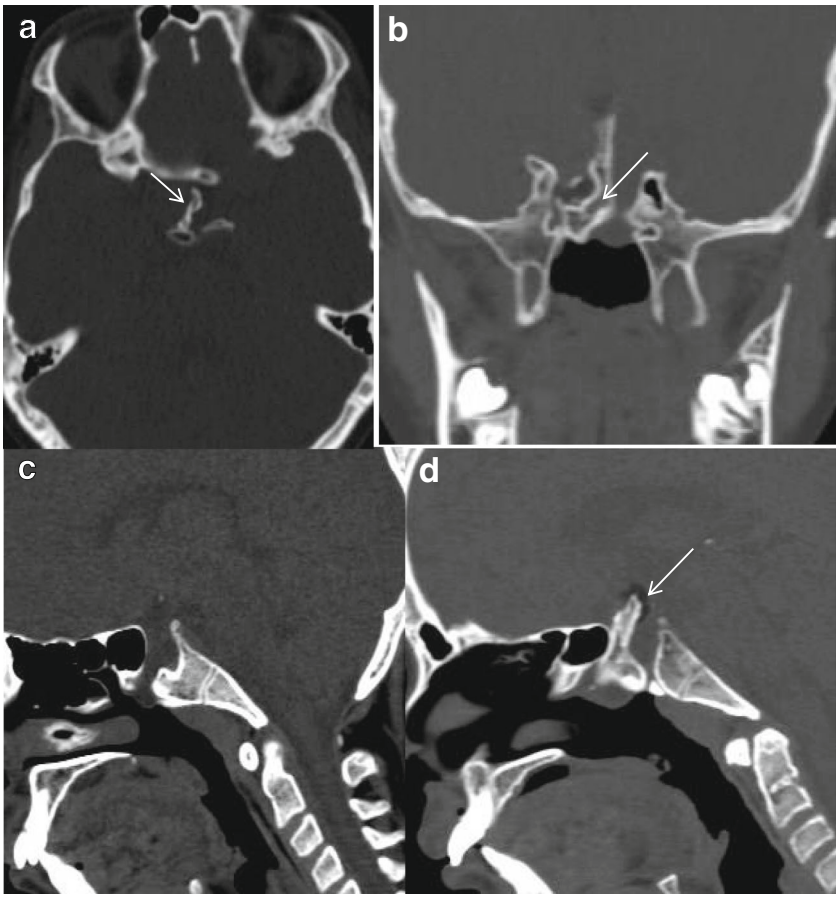

Fig. 2 **a–d**: **a** Axial CT shows bony septum in sellar and suprasellar region (arrow). **b** Coronal reformat shows bony septum seen arising from the part of sphenoid body. **c**, **d** Sagittal CT reformats shows the large bony canal (diameter 5.7 mm) and the vertical bony septum. Tip of the septum shows fat attenuation lesion—lipoma (arrow)

CPC into three types based on the size of the canal and the type of soft tissue lesion within the canal [1]. Type I CPCs (also known as persistent hypophyseal canal) are small, incidental canal with orthotopic, and normally functioning pituitary gland [1, 2]. Type II CPCs are medium sized canal with ectopic adenohypophysis [1, 3]. Type III CPC are large canals and further classified into type 3A (canal contains cephalocele), type 3B (canal contains tumors) or type 3C [1, 4] (tumors and cephalocele). Type I CPCs are associated with PHACE syndrome, duplicated pituitary gland, and cleft lip/palate [1]. Type 2 and 3 CPCs are associated with ectopic adenohypophysis, pituitary dysfunction, and meningitis [1, 4]. Type 3B CPCs usually contains tumors like dermoid, teratoma, craniopharyngioma, and gliomas [1, 4]. These tumors may have suprasellar or infrasellar extension from the canal. Presence of nasopharyngeal mass should raise the suspicion of type 3 CPC [5]. In our case, patient had a midline anomaly, cleft palate for which he underwent surgical repair. CPC in our case was divided by an abnormal bony septum. It contained a lipoma, which was in close relation to the right side of the septum and a cephalocele to the left side of the septum. Our case is similar to type 3C of Abele, but the unique feature is the presence of abnormal bony septum dividing the canal. Another interesting feature in our case was the absence of any pituitary dysfunction although the pituitary gland was not visualized on MRI. It is possible that the child has ectopic functioning adenohypophyseal tissue along with the soft tissue contents of the canal, which is difficult to be delineated by imaging alone [3–5].

Embryology At 3–4 weeks of gestation, neuroectodermal adherence is formed by close contact of roof of the stomoedeum and the diencephalon [6]. At 4–5 weeks, the adenohypophyseal pouch develops and gets elongated to form adenohypophyseal stalk or rathke stalk and during 6–7 weeks, the postsphenoid cartilage develops and fuses which result in obliteration of the stalk. Non-obliteration of the adenohypophyseal stalk results in persistent CPC [6–8].

Sphenoid bone derives from multiple ossification centers, up to 19 enchondral and intramembranous ossifications centers have been described [9]. Alisphenoid and orbitosphenoid forms the greater and lesser wings of the sphenoid bone respectively [9]. Sphenoid body anterior to the tuberculum sellae is formed by presphenoid center, whereas the dorsum sellae, sella turcica, posterior sphenoid body, and anterior part of clivus are formed by postsphenoid centers (Table 1) [7–9]. The postsphenoid centers have two medial and lateral

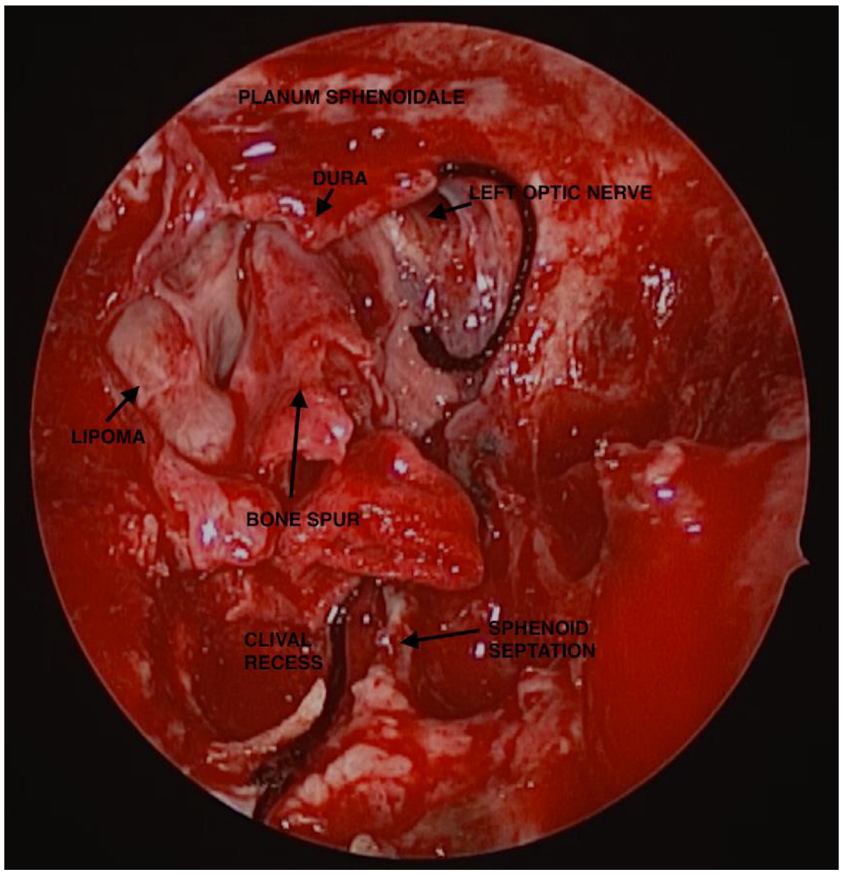

Fig. 3 Endoscopic view after sellar drilling shows bony spur, optic nerve after dural opening, clival recess, and sphenoid septation

ossification centers. Fusion of medial and lateral ossification centers leads to complete formation of sella turcica and basisphenoid [9]. The medial centers usually fuse to form single ossification center. Failure in fusion results in persistence of the adenohypophyseal pouch, leads to formation of CPC [8, 9]. In our case, there is nonvisualization of left half of the sella and basisphenoid with abnormal bony septum seen arising from the dysplastic remnant of basisphenoid. The possible hypothesis in our case would be a defective formation of left medial and lateral post sphenoid ossification centers with formation of accessory ossification centers on the right side, which might have aberrantly developed to form an abnormal bony septum. This would have projecting into the canal and dividing into two hemi-canals. Another possibility would be an abnormal fusion or formation of medial and lateral post-sphenoid ossification centers. This may lead to form an abnormal bony septum within the large CPC resulting in bipartite appearance of CPC.

Conclusion

We describe a previously unreported variant of craniopharyngeal canal, divided by an abnormal bony

Table 1 Embryology of sphenoid body [7–9]

Enchondral ossification centers	Derivative
1. Presphenoid centers	a. Sphenoid bony anterior to tuberculum sella
	b. Crista galli
	c. Perpendicular plate of ethmoid and d. Chiasmatic sulcus
2. Postsphenoid centers (two medial and lateral ossification centers)	a. Dorsum sellae
	b. Sella turcica
	c. Sphenoid bone posterior to tuberculum
	d. Part of clivus

septum resulting in bipartite appearance with Lipoma and cephalocele on either side of the septum. We also discussed the possible embryological hypothesis leading on to this condition. This might have surgical relevance in selected cases.

Authors' contribution S V: Manuscript writing, image analysis, manuscript editing
B T: Manuscript edition and critical revision
J G: Image analysis and description, manuscript edition
S S: Image analysis and description, manuscript edition
C K: Manuscript editing and critical revision
P N: Manuscript editing and critical revision

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Patient consent The patient's guardian has consented to the submission of the case report for submission to the journal.

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