ORIGINAL ARTICLE

Eur J Anat, 16 (1): 68-73 (2012)

A sphenoid with multiple developmental variations

Kumud Dharwal

Department of Anatomy, Shri Guru Ram Das, Institute of Medical Sciences and Research, Amritsar, India

SUMMARY

In a study of 156 skulls the interior of one particular skull showed many additional foramina on the sphenoid bone, one large foramen almost on the centre of sella turcica (0.64%) (the large craniopharyngeal canal); a second foramen on the inferomedial angle of the optic canal (the lateral craniopharyngeal canal), which were also present in 2 more skulls (2.56%); and two large foramina Vesalii on each side. In addition the sphenoid showed some morphological changes such as a hypoplastic left lesser wing; the upper parts of the greater wings were formed by separate bones; all sutures of the sphenoid were coarse, irregular and had tiny sutural bones. This skull also had another large foramen at the confluence of sinuses, also seen in 11 more skulls (7.7%), a large foramen caecum, a large posterior condylar foramina and a foramen in the left sigmoid sinus groove. Knowledge of these additional foramina and variations are important for radiologists, endocrinologists, neurosurgeons and anthropologists.

Key words: Skull – Sphenoid – Accessory foramina – Craniopharyngeal canal – Development

INTRODUCTION

The sphenoid bone forms the medial part and some of the lateral parts of the middle cranial fossa. In the particular skull above mentioned the sphenoid bone showed additional foramina, and some architectural changes. The large foramen in the body of the sphenoid at the floor of the sella turcica is the craniopharyngeal canal (Bergman et al., 1988; Hughes et al., 1989), the one near the optic canal is the lateral craniopharyngeal canal or Sternberg's canal (Schick et al., 2000; Nayak, 2008). The aetiopathology of these additional foramina is described and being either due to a vestige of a craniopharyngeal canal (Currarino et al., 1985) or as a remnant of a vascular channel (Arey, 1950). There should be some correlation between it and altered intrauterine environments during the development of this bone (Sadler, 2005). These large foramina can lead to dura or / and brain herniation (Coticchia, 2006; Currarino et al., 1985; Bendersky et al., 2011) or cerebrospinal rhinorroea (Hooper, 1971), which are life-threatening conditions.

MATERIAL AND METHODS

The study was conducted on 156 dry macerated skulls of unknown age and sex of north

Submitted: December 03, 2011
Accepted: January 30, 2012

Corresponding author:
Dr Kumud Dharwal. Dharwal clinic, Cheel mandi, near Ramgarhia School, Amritsar (Pb.), 143001 India. Phone: +919872737679. E-mail: kdharwal@gmail.com

Indian origin from a pool of the bone bank of the department and a collection of the students. Each skull was scrutinized for 150 different morphometric and foramina interrelation parameters on each side. One particular skull showed a few variations in the interior of the skull especially in the middle cranial fossa.

Observations

Out of 156 dry macerated skulls studied, one particular skull showed a few variations in the interior of the skull.

1. A large foramen in the large craniopharyngeal canal 'CPC' was present (Fig. 1, large red arrow) on the sella turcica almost at its middle but just to the right of the midline, opening into the sphenoidal air sinus. The foramen was almost circular - 3.24 mm x 3.48 mm - and had smooth margins.

2. A small foramen in the lateral craniopharyngeal canal 'LCPC' was seen at the inferomedial margin of the right optic canal, communicating with the sphenoidal air sinus (Fig. 1, small red arrow).

3. There were two large foramina Vesalii 'FV' of open type on each side; the left ones bigger -anterior 1.2 mm and posterior 1.88 mm- in diameter than the right foramina anterior - <1 mm and posterior 1.22 mm-, in diameter, opening at the scaphoid fossa at the base of skull (Fig. 1, blue arrows).

4. The left lesser wing was hypoplastic and quite irregular (Fig. 1, yellow arrow).

5. All the suture lines of the sphenoid in the interior of the skull, especially along the greater wings, were found to be coarse and irregular.

6. The greater wing sutures showed many tiny sutural bones, 'SB', in them (Fig. 1).

7. The sphenoid greater wings, 'GW', had separate upper ends, 'S', on the exterior of the skull (Fig. 3).

8. Another large foramen, 'IOP' (Fig. 2, red arrow), - 3.46 mm x 1.68 mm- was present on the internal occipital protuberance just to the

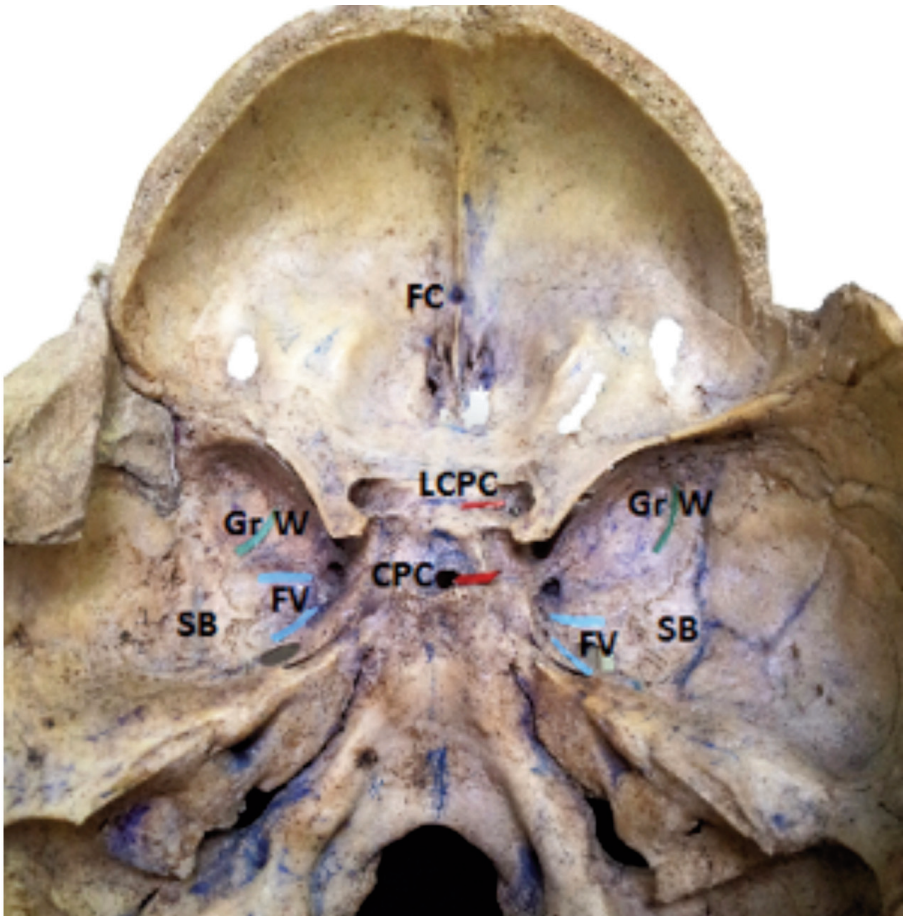

Fig. 1. Interior view of the skull showing a large foramen on the sella turcica, the craniopharyngeal canal (CPC, large red arrow), a small foramen in the right optic canal, the lateral craniopharyngeal canal (LCPC, small red arrow), Foramina Vesalii (FV, blue arrows), sutural bones (SB) along greater wing of sphenoid (GW), separate apexes of the greater wing of the sphenoid (green arrow), the lesser wing of the hypoplastic sphenoid (yellow arrow), and a large foramen caecum (FC).

left of the midline at the confluence of the sinuses communicating to the exterior.

9. The posterior condylar foramina were quite large: right 5.67 x 4.99 mm, left 5.36 x 3.92 mm.

10. A large foramen caecum 'FC' (Fig.1) was present.

11. A large foramen, 'SS', at left sigmoid sinus groove (Fig. 2) 3.4 x 3.1 mm was present.

Apart from this, the skull showed 4 more sutural bones and change in the shape of the foramen ovale. Since the foramina were noted in a dry skull, their contents could not be predicted with precision.

Discussion

The sphenoid bone normally shows a garland of foramina along the basi- ali-sphenoid junction, the foramen rotundum, the foramen ovale and the foramen spinosum. Some rare foramina, foramina Vesalii and canaliculus innominatus are also seen in relation to the sphenoid bone. In Fig. 1 the large foramen 'CPC' seen on the floor of sella turcica on body of sphenoid can be observed, despite it is rather rare, except for two cases of persistent craniopharyngeal canal reported by Hughes et al. (1999), which LIE between the pre- and post-sphenoid. The prevalence of this foramen has been reported to be 0.3% (Bergman et al., 1988) and 0.42% (Arias et al., 2009), while it was 0.64% in the present study. The foramen at the inferomedial angle of the optic canal 'LCPC' opening in the sphenoid sinus is somewhat similar to the case reported in one study (Nayak, 2008) and also in another by Schick et al. (2000), where it was named a lateral craniopharyngeal canal (Sternberg's canal), which is between the pre – ali- post sphenoids. The prevalence of this foramen is 4% (Bergman et al., 1988), and was found in 4 cases (2.56%). According to their positions, the abnormal foramina of this skull can be labeled, although with some reservations,

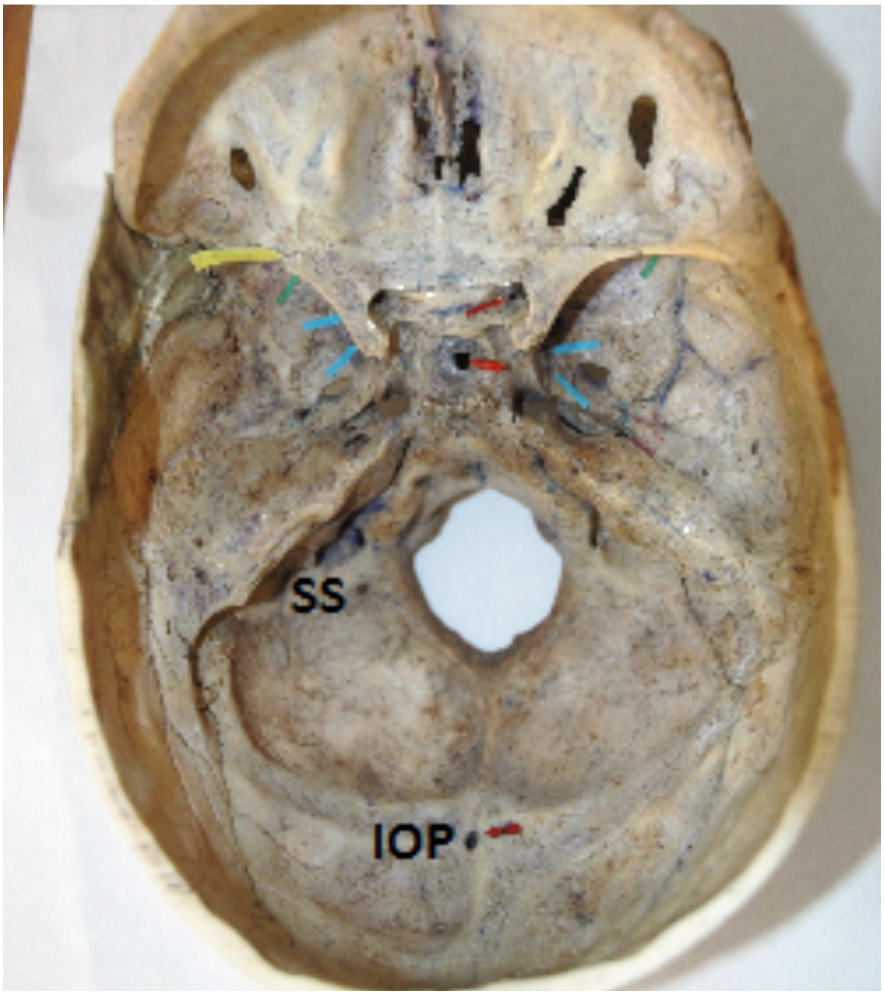

Fig. 2. Interior view of the skull showing a large foramen on the left sigmoid sinus groove (SS), and a large foramen at the internal occipital protuberance (IOP, red arrow).

Fig. 3. Exterior of the skull showing separate upper ends (S) of the right greater wing (GW) of the sphenoid.

'CPC' as the craniopharyngeal or hypophysial canal and the foramen 'LCPC' as the lateral craniopharyngeal canal (Sternberg's canal), large foramina Vesalii 'FV' and 'IOP' at the internal occipital protuberance 'FC' foramen caecum and 'SS; at the sigmoid sinus as the enlarged emissary vein portals.

The CPC foramen reported here is unique in certain respects. The term "craniopharyngeal canal" is generally used to describe a small vertical midline conduit in the skull base that measures <1.5 mm in diameter (Kizilkilika et al., 2005) but this foramen was neither at the midline nor <1.5 mm and hence does not meet either criterion. Moreover this parasagittal foramen extended into the sphenoidal air sinus and not into the nasopharynx, further supporting the likelihood of it being a remnant of the passage channel of Rathke's pouch (the craniopharyngeal cana). Rather, as suggested by Currarino et al. (1985), it should be labeled as a large craniopharyngeal canal.

Aetiopathology -The sphenoid bone represents a complex structure in terms of anatomy and embryology. It develops by the fusion of multiple primordia of different embryonic origins. The complexity of its development and the non-fusion of some of its parts may lead to the formation of abnormal foramina and is non-fusion probably the basis of the presence of these additional foramina in this skull.

Two theories have been proposed to explain the formation of craniopharyngeal canals. The most widely accepted theory was proposed by (Currarino et al., 1985). The development of the pituitary starts during the middle of the 4th week through formation of Rathke's pouch, a diverticulum from the roof of the stomodeum passing upwards between the chondrification centers of the developing presphenoid and postsphenoid (basisphenoid). During 6th week, the connection of the pouch with the oral cavity degenerates. This connection persists as a vertical defect in the basisphenoid in 1% of newborns as a basipharyngeal/ craniopharyngeal canal (Currarino et al., 1985; Moore and Persaud, 2005). Craniopharyngiomas may develop in connection with this remnant in the basisphenoid, most often in and/or superior to the sella turcica (Moore and Persaud, 2005), thereby leading to the persistence of a canalicular bony partial/complete defect between the nasopharynx and the sella turcica. The other theory proposed by (Arey, 1965) is that it would be present at the time of birth due to a remnant of a vascular channel formed during osteogenesis.

The 'LCPC' foramen is a congenitally present lateral craniopharyngeal canal, which can

be produced due to defective ossification of the sphenoid (Currarino et al., 1985). It may be a vascular remnant (Arey, 1950), formed by focal bone atrophy due to pressure from the developing optic nerve at that site (Hooper, 1971), or it may appear after birth due to dehiscence of some weak bones during the formation of sphenoidal air sinuses by bone resorption, under the influence of expanding nasal mucosa (Arey, 1950).

In the present case, the formation of two foraminal CPC and LCPC defects can be explained by the vascular theory, suggesting both defects to be the vestiges of vascular channels formed during osteogenesis. This is also substantiated by the presence of many enlarged additional vascular channel foramina in the particular skull of the present study. The foramina Vesalii (accessory sphenoidal emissary foramina) have been reported to be present in 39% of cases, out of which 23% are present bilaterally (Bergman et al., 1988), although in the present study 55% were found to be present bilaterally and 20% unilaterally, with a size ranging from pinpoint to 3 mm. In our case, not only the foramina Vesalii were present bilaterally but numbered 2 on both sides and were quite large in size, especially on the left side: 1.2 mm and 1.88 mm in diameter. In this skull the morphology of the sphenoid bone is disturbed as is its vascular system, leading to an increase in number as well as in the size of the emissary vascular channels. Functional vascular balance was maintained on both sides by two additional foramina CPC and LCPC with two small foramina Vesalii on the right side and two large foramina Vesalii together with a large foramen on the internal occipital protuberance and on the sigmoid sinus on the left side.

The persistent craniopharyngeal canal has clinical importance as it may lead to sphenoidal meningocoele, encephalocoele (Coticchia, 2006), meningoencephalocele (Currarino et al., 1985; Bendersky et al., 2011) and hamartoma (Ekinci et al., 2003). In recurrent meningitis (Arias et al., 2009), or CSF rhinorrhoea (Hooper, 1971) or precocious puberty (Kizilkilika et al., 2005), this variation should be kept in mind as the prime suspicion. Knowledge of these additional foramina and variations are important for neonatologists, radiologists, endocrinologists, neurosurgeons and anthropologists.

The greater wings show horizontal fissures near the upper ends, probably the apexes are formed by additional separate ossicles. This variation is also cited by Bergman et al. (1988), but no figures regarding prevalence were found in the literature. Hypoplastic lesser wings (Jacquemin et al., 2001) have also been reported earlier, with or without neurofibromatosis. These defective ossifications of the sphenoid; the presence of coarse and irregular sutures and multiple tiny sutural bones clearly indicate a disturbed architecture of the sphenoid bone. Since the sphenoid develops from neural crest cells, these cells are unfortunately a particularly vulnerable cell population and are easily destroyed by the teratogenic effect of simple chemicals such as alcohol and retinoic acid (Sadler, 2005). In addition, the resulting onslaught is multiplied if it is occurs at that critical phase of intrauterine life, which is the development period of that organ: i.e., 4th-6th wk for the sphenoid bone. These variable foramina and the structures passing through them have widespread manifestations so foreknowledge of these variations is important for neonatologists, radiologists, endocrinologists, neurosurgeons and anthropologists.

References

Arey LB (1950) The craniopharyngeal canal reviewed and reinterpreted. *Anat Rec,* 106: 1-16.

Arias DP, Hinojosa J, Esparza J, Muñoz A (2009) Recurrent meningitis and persistence of craniopharyngeal canal: case report. *Neurocirugía,* 20: 50-53.

Bendersky DC, Landriel FA, Ajler PM, Hem SM, Carrizo AC (2011) Sternberg's canal as a cause of encephalocele within the lateral recess of the sphenoid sinus: A report of two cases. *Surg Neurol Int,* 2: 171.

Bergman RA, Afifi AK, Miyauchi R (1988) Compendium of human anatomical variations. Urban and Schwarzenberg, Baltimore, pp 200.

Coticchia J (2006) Transsphenoidal encephalocele in a neonate. *The Free Library*. Retrieved February 23, 2012. From http://www.thefreelibrary.com/Transsphenoidal encephalocele in a neonate.-a0149514654

Currarino G, Maravilla KR, Salyer KE (1985) Transsphenoidal canal (large craniopharyngeal canal) and its pathologic implications. *AJN,* 6: 39-43.

Ekinci G, Kilic T, Baltacioglu F, Elmaci I, Altun E, Pamir MN, Erzen C (2003) Transsphenoidal (large craniopharyngeal) canal associated with a normally functioning pituitary gland and nasopharyngeal extension, hyperprolactinemia, and hypothalamic hamartoma. *Am J Roentgenol,* 180: 76-77.

Hooper AC (1971) Sphenoidal defects-a possible cause of cerebrospinal fluid rhinorrhoea. *J Neurol Neurosurg Psychiat,* 34: 739-742.

HUGHES ML, CARTY AT, WHITE FE (1999) Persistent hypophyseal (craniopharyngeal) canal. *Br J Radiol.* 72 (854): 204-206.

JACQUEMIN C, MULANCY P, BOSLEY TM (2001) Abnormal development of lesser wing of the sphenoid with microophthalmos and microcephaly. *Neuroradiology,* 49: 178-182.

KIZILKILIKA O, YALCINA O, YILDIRIMA T, SENERB L, PAMAKSIZC G, ERDOGANB B (2005) Hypothalamic hamartoma associated with a craniopharyngeal canal. *AJNR – Pediatrics,* 26: 65-67.

MOORE RL, PERSAUD TVN (2005) Nervous system. In: *The Developing Human, Clinically oriented Embryology*, 7th ed, 4th reprint. Saunders Elsevier, New Delhi, pp 446-448.

NAYAK S (2008) An abnormal foramen connecting the middle cranial fossa with sphenoidal air sinus: A case report. *Internet J Biol Anthropol,* 8: 1.

SADLER TW (2005) Central Nervous System. In: *Langman's Medical Embryology*, 10th ed. Wolters Kluver/ Lippincott Williams & Wilkins, New Delhi, pp 267-268.

SCHICK B, BRORS D, PRESCHER A (2000) Sternberg's canal - cause of congenital sphenoidal meningocele. *Eur Arch Otorhinolaryngol.* 257: 430-432.