Median defect in the skull

Singh R, Bandyopadhyay M

ABSTRACT

A median defect in the region of the root of the nose, in between the two orbits, was discovered in the dried skull of a 44-year-old female cadaver, during routine undergraduate teaching. The two small nasal bones articulated with each other and the cribriform plate of the ethmoid. The lacrimal bones and frontal processes of the maxillary bones were also deformed. We propose that the median defect was due to abnormality at the fonticulus frontalis, the prenasal space and the interorbitonasal part of the nasal capsule, as well as defects in the ossification of the maxilla, lacrimal and frontal bones.

Keywords: encephalocoele, frontal bone, lacrimal bone, median skull defect, nasal bone

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INTRODUCTION

The skull is composed of the viscerocranium and the neurocranium. The viscerocranium is derived from the frontonasal and the first arch mesenchyme. The facial and the vault bones are formed from the mesenchymal condensation, which fuse and may either ossify by intramembranous ossification or via endochondral ossification. There are recognised spaces between these structures: the fonticulus frontalis, the prenasal space, and the foramen caecum,[1] formed before the mesenchymal condensation commences to fuse. The fonticulus frontalis is the space between the frontal and nasal bones. The prenasal space is between the nasal bones and the nasal capsule (the precursor of the septum and nasal cartilages).[1] As the development of the foetus proceeds, these spaces fuse and ossify. Abnormal development of these structures is thought to be involved in the formation of congenital midline masses, such as the dermoids, gliomas, and encephaloceles of the nose.[1] One such encephalocele is the sincipital, which presents as a mass over the nose, glabella, or forehead.

CASE REPORT

A dried specimen of the skull was discovered during an undergraduate teaching session. The skull was obtained from a female cadaver, 44 years of age, donated to the Department of Anatomy, Calcutta National Medical College, India. (Fig.1). The donated body did not have any recorded, visible abnormality in the face. Evaluation of the skull revealed that it was dolichocephalic, with a cranial index 64.92%. A circular, smooth outlined gap was found, as an upper median craniofacial cleft, at the root of the nose, in between the two orbits. It occupied the region of nasion without forming the true nasion. The maximum

Fig. 1 Photograph shows the norma basalis of the skull; the united basi-sphenoid and basi-occiput indicate the adult age of the skull.

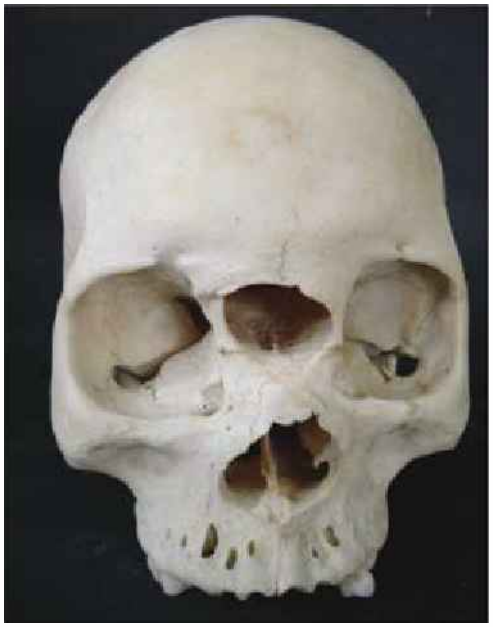

Fig. 2 Photograph shows a smooth outline defect between the orbits in the frontal bone.

Department of Anatomy, Calcutta National Medical College, 32 Gorachand Road, Kolkata 700014, West Bengal, India

Singh R, MD, DNB
Assistant Professor

Bandyopadhyay M, MS
Associate Professor

Correspondence to:
Dr Royana Singh, B-115 Brij Enclave, PO Sunderpur, Varanasi 221005, Uttar Pradesh , India
Tel: (91) 5422 317 438
Email: singhroyana@rediffmail.com

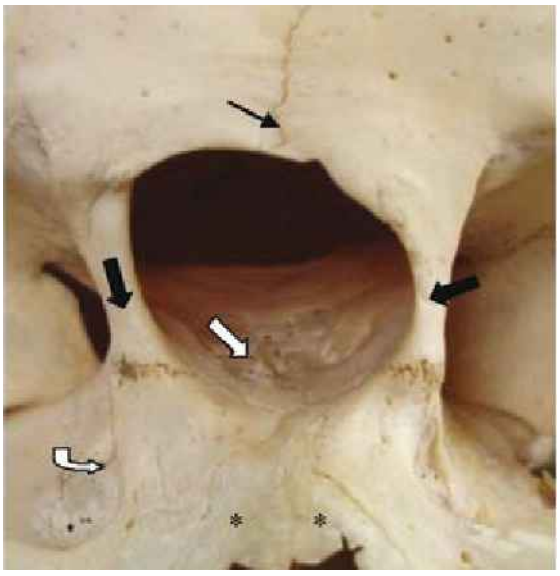

Fig. 3 Photograph of the skull shows the defect in the frontal bone, the metopic suture (thin black arrow), the floor of the outline showing the cribriform plate and the long maxillary process of the frontal bone articulating with the small frontal process of the maxilla (bold arrows), the lacrimal bone at the junction between the nasal bones, the maxilla and the ethmoid (curved white arrow), and the two nasal bones articulating with each other (asterisks).

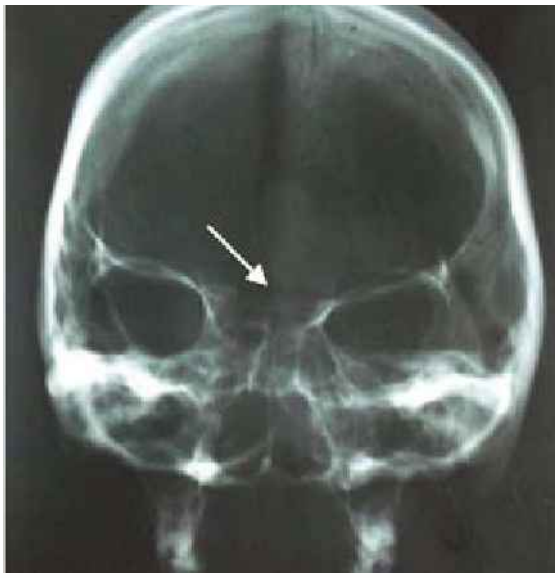

Fig. 4 Radiograph of the skull shows the median defect (white arrow) with no frontal sinuses.

length and breadth of the gap was 20.3 mm and 23.6 mm, respectively, at its widest point (Fig. 2).

The smooth outlined gap revealed at its upper border, an incomplete fusion of the metopic suture, 3.24 cm in length (Fig.3). The lower border of the gap was continuous with the cribriform plate of the ethmoid, onto the interior of the skull. Inferiorly, the two nasal bones, were relatively small and deformed. They articulated with each other in the midline (Fig. 1c, asterisks) and with the short frontal process of the maxilla, but not with the maxillary process of the frontal bone, thus failing to form the nasion. The lateral boundary of the gap was formed by the short frontal process of the maxilla and downward extension of a process from the frontal bone, which could be named as the "maxillary process" of the frontal bone, articulating at a suture, present at the junction of the upper three-fourth and lower one-fourth of the lateral boundary (Fig. 3).

The lacrimal bones were small and deformed. They occupied the site of the nasolacrimal canal and extended above up to the frontomaxillary suture. The nasal cavity was smaller in length with a nasal index of 27.6/24.7 × 100 = 111.74%, but the nasal conchae were normal. The bony components of the nasal septum were also normal. The cleft in the bony wall of the frontal bone had pushed the medial walls of the orbits laterally. The bony orbits were otherwise normal. The anterior cranial fossa was deeper in the central region, where the ethmoid bone had failed to meet the frontal bone, and thus lay at lower level (Fig. 3). The maximum cranial length was 158 mm, maximum cranial breadth 102.7 mm, and cranial height 108.2 mm. As no nasion was discernible, the total facial index, the upper facial index and the gnathic index could not be measured. The orbital index was 34.2/32.2 × 100 = 106.21%. The roentgenogram revealed that there was a cleft with no formation of the frontal air sinuses. The ethmoid bone as well the sphenoid bone were normal (Fig. 4).

DISCUSSION

The margins along the defect on the frontal bone, nasal bone and the cribriform plate are smooth, suggestive of compressive deossification. The smooth margin of the defect, contiguous with the brain in the present case may be due to a less aggressive lesion, such as a benign tumour, or an encephalocele. On the other hand, inflammatory lesions, traumatic deformity, or malignant neoplasm cause destructive bone change which may lead to an irregular margin.

Nasal encephaloceles are very rare, occurring 1:5000,[2] but are exclusively common in Southeast Asia.[3] They are of two main kinds: frontoethmoidal and basal encephaloceles, the former being more common. They can be congenital or an acquired abnormality of the brain in which intracranial contents herniate through a skull defect.[4] Congenital encephaloceles occur when the mesodermal layer between the neural tube and the ectoderm fails to develop and the anterior neuropore remains open. The present case is highly suggestive of frontoethmoidal encephalocele, as the cribriform plate of ethmoidal bone, which normally fuses with the frontal bone at the notch, has not fused with it, but has instead fused with the nasal bones and the maxillary bones. The lower part of the frontal bone has failed to meet at the

midline, due to the protusion of the cranial contents at the fonticulus frontalis, and formed a cleft at the lower part of the frontal bone and a persistent metopic suture. Reverse growth has occurred at the frontomaxillary suture, the frontal bone has extended over to the lateral sides of the cleft to meet the short frontal process of the maxilla in the prenasal area as the "maxillary process" (Figs. 2 & 3). The consequences of the above events lead to the non-formation of the nasion and the frontal sinuses (Fig. 4). The intraorbital cartilage of the nasal capsule, which normally would have met the frontal bone[5,6] at the prenasal area, failed and articulated with the cribriform plate of the ethmoid at a lower level. The nasal bones and lacrimal bones were reduced and deformed. The mass at the region of the fonticulus frontalis hindered in the normal pivotal movement of the positioned lacrimal bone and the surrounding bones, resulting in the widening of the intraorbital space. However, the medial walls of the orbit have maintained their normal orientation.

According to Subramani and Murthy, the cleft may occur as an isolated soft tissue, an isolated skeletal structure, or as a combination of both structures, involving one bone or all the cranial bones.[7] In this case, the defect suggests it to be a sole deformity, as the rest of the skull appears normal with no other bony defect. The frontoethmoid type of encephalocele rarely exists with meningitis,[8] and the present case thus may have survived through to adulthood with little complications, as the skull base suggests. The large gap formed also suggests that it could be the result of a dermoid or glioma. A large dermoid may have formed due to the failure of the regression of the dura in the space, the fonticulus frontalis. The surrounding mesenchyme may have subsequently undergone decompression ossification and non-fusion of the metopic suture. The bones thus formed around it are deformed and may have undergone abnormal growth as described above for the nasal encephalocele. The other possible congenital mass could be the glioma, present as a mass at the root of the nose, with a widened intraorbital distance or hypertetelorism, secondary to the growth.

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