An Encephalocele Replacing Nose Completely

Gautam Dasgupta, Chief Specialist. E.N.T.
Tinplate Hospital, Jamshedpur, India, Pin - 831 003

Abstract

Congenital encephalocele presenting in the nasal region is extremely rare. We present here a case of encephalocele completely replacing the nose of a five year old boy and discuss about similar swellings in this region.

Key Words

Encephalocele, Glioma, Meningocele, Dermoids.

Developmental anomalies in the region of nose is very rare. Herniation of brain tissues through bony defect may present as glioma, encephalocele or meningioma. These abnormalities are one tenth as common as neural tube closure defects involving the spine (Haslam, 1996). Encephaloceles are neural tube defects that affect the cranium and result in a midline mass overlying a skull defect (Sanders, 1996). Most occur in the occipital (75%) or frontal region (13%) but some are parietal (12%). They can extend into the nasal and sphenoid areas (Toi et al 1991). We came across such a rare case recently and could not get useful informations from the recent journals. This has prompted us to document this case.

Case Report

G. C. G. a five-year old boy was brought to us by a health worker from a remote village where he went in search of leprosy patients. This boy is the youngest of four brothers and has no history of consanguinity in the family. Father works in brick kiln as a contractor labour and is very poor. This child has a gradually increasing swelling in the nose, replacing the nose completely at the moment. He is a mouth breather with nasal obstruction with occasional headache. Although his speech is normal, he is unable to write because of mechanical obstruction to the eyes by the swelling.

On examination : An otherwise normal boy with an oblong bilobed swelling 15 cm x 8cm in size, skin coloured, originating at the root of nose and replacing the nose completely. Inner canthus of the eyes are

compressed alongwith medial part of both eyes. (fig. 1). The swelling is compressible but has no pulsation and not translucent. Openings are present in anterior nares but hardly anything could be seen through them. Oral cavity and nasopharynx are found to be normal. No other congenital anomalies are present in the body.

Examination of Eyes : Horizontal nystagmus present in the both eyes. Fundi are within normal limits with diminished foveal reflex. Visual acuity could not be tested because of the obstruction at the visual axis. Tensions are normal.

X-Ray Skull : A large soft tissue swelling completely replacing the nose (fig. 2). There is a large destructive lesion in the frontal bone in the region of the frontal sinus (Fig. 3).

Fig. 2. Photograph of X-Ray of lateral view of skull showing soft tissue mass completely replacing the nose

The mass is obviously protruding from within the cranial cavity and is most certainly an Encephalocele.

Discussion

Three developmental abnormalities of the nose that are confused are nasal gliomas, encephaloceles and nasal dermoids.

Nasal gliomas are deposits of glial tissue in an extradural site. Encephaloceles are similar to gliomas but maintain connection to the CNS and contain CSF or brain tissue.

Gliomas can occur extranasally or intranasally. Extranasal gliomas are smooth, firm and non compressible masses. They usually occur along glabella or the lateral wall of the nose off midline. Internasal gliomas may manifest as a cyst or mass within the nose, are polypoid in appearance, do not distend with bending over or with crying and appear quite firm. Encephaloceles when extranasal are generally soft and compressible and one always finds a bony defect present. This is in contrast to gliomas, which are not compressible. Intranasal encephaloceles

Fig. 3. Photograph of X-Ray of Paranasal Sinuses showing a large defect in the frontal bone

are soft and cystic lesions which can be compressed and enlarge with crying or bending over. These lesions are generally pulsatile. Encephaloceles usually require intracranial repair followed by transnasal or extranasal removal of the tumour. When problems arise, it is usually a result of biopsy of this lesion within the nose under the mistaken belief that it is a nasal polyp.

Dermoid cysts are formed by trapped dermal elements external to or between the nasal bones. They are painless swelling found generally in the midline of the nose, often with a small pitlike opening with the skin. The cysts may extend superiorly with sinuses through defects in the frontal bones or nasal bones inferiorly. (DeWeese et al, 1988).

Bailey Love's short practice or surgery (20th edition) has given a picture of meningocele over the occipital bone. This swelling was translucent, differing from the present case of encephalocele. Because of its rarity the present case is being documented. As this case requires neurosurgical intervention facilities of which are not available with us, the father is being persuaded to take the child to a higher centre for further management.

References

1. Bailey and Love's (1989) : short practice of surgery Revised by Harding Rains, A. J. and Mann, C. V. 20th edition H. K. Law's Co London Pages 435-436.
2. Dweese, D. D. Saunders WH, Schullar, DE, Schelenning II A. J., (1988) : Otolaryngology, Head & Neck Surgery, VII edition, The C. V. Mosby Co, St Louis Page 80.
3. Haslam, R. H. A., (1996) : Congenital anomalies of the Central Nervous System in Nelson Text Book of Pediatrics 15th edition-Book-2 (Behrman, R.C., Kliegman, R. M., Arvin A. M., eds) W. B. Sanders Co. USA Page 1979-1980
4. Sanders, R. C. (1996) : Structural Fetal Abnormalities—the total picture—Mosby, St. Louis Page 30.
5. Toi, A., Sanebrai E. E., (1991) The fetal Brain in Diagnostic ultrasound, Vol 2 (Rumack, C.M., Wilson, S. R., Charboneau, JW, eds) Mosby year Book, St. Louis Page 884.