OPHTHALMOLOGICAL ASPECTS OF FRONTO-ETHMOIDAL MENINGOENCEPHALOCELES FROM THE SOUTH EAST ASIAN REGION

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Abstract

Fronto-ethmoidal meningoencephaloceles for reasons unknown, are far more common in South East Asia than in most other regions of the world, and account for 25% of overseas referrals to the South Australian Cranio-Facial Unit. We reviewed twelve consecutive cases presenting to our Unit. All patients were of Malay or Indian origin, no cases were familial, but mean paternal age at the child's birth was above normal. Cases were assessed by clinical examination, X-rays, with subsequent confirmation at surgery. In all cases the encephalocele exited via the foramen caecum, with entry on to the face being one of three types; naso-frontal, naso-ethmoid, and naso-orbital. There was considerable medial canthal displacement, moderate globe displacement, and minimal lateral canthal displacement. All cases showed an elongated face. The encephalocele appears to act as a foreign body displacing the eyes laterally, and the mid-face downwards, acting quite differently to normal facial clefts. Ocular abnormalities included hypertelorism, telecanthus, non-functioning lacrimal apparatus, orbital dystopia, and squint. Binocular vision was usually present. Surgery involved shifting one or both orbits, or medial orbital walls only, to correctly position the eyes. Complications were minimal, but included convergent squint, which usually resolved without treatment.

Key words: *Encephalocele, hypertelorism, cranio-facial surgery, facial deformity.*

Fronto-ethmoidal meningoencephaloceles are exceptionally rare deformities in most regions of the world, but for reasons which are currently obscure, they appear to be much more common in South East Asia and adjacent regions of the world, though not in the European population of Australia. Only three of 41 cranial encephaloceles reported in one series from Australia were of the fronto-ethmoidal variety and two of these were Aborigines. However, in Thailand the ratio is 100 fronto-ethmoidal encephaloceles for every 15 of all other types of encephaloceles combined. Not surprisingly, these lesions which cause a major disruption to the face represent a significant percentage of referrals to the S.A. Cranio-Facial Unit for correction. Approximately 25% of all cases referred to the Unit from South East Asia have fronto-ethmoidal meningoencephaloceles as the major problem. This paper reviews our experience with this lesion, based on the first 12 consecutive cases.

CLINICAL MATERIAL

Twelve cases of fronto-ethmoidal meningoencephalocele were referred to our unit over a two and a half year period, of which ten were children of Malay descent, and two children of Indian descent. All cases were sporadic. There were no familial cases, and none of the families had relatives with other neural tube defects. The mean position of the child in the family was fifth, and there were an equal number of females and males. In all cases the pregnancy was apparently normal. Parental ages at time of birth were noted for all cases, and the paternal age mean was 36 years, maternal age mean 28. This indicates a significantly higher mean paternal age than might

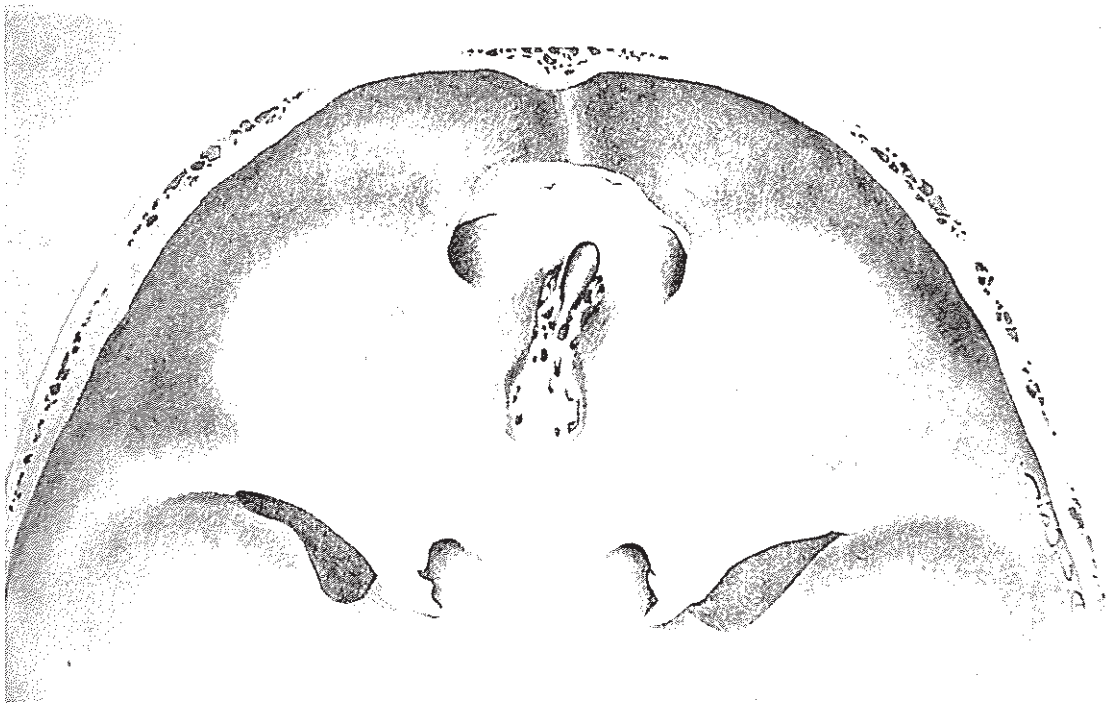

Figure 1: Medical artists rendition of the defect in the anterior cranial fossa in the region of the foramen caecum where the meningoencephalocele exits towards the face, based on the appearance of x-rays and this region at surgery.

be expected in the community, based on the Australian experience, but detailed figures for Malaysian society are not available, although it would appear that there is a tendancy to marry young.

All patients underwent routine extensive cranio-facial workup prior to surgery which included a complete ophthalmological examination including measurements of distances between the medial canthi, mid-pupil, and lateral canthi. Anatomy of the lesion was determined using cephalometric x-rays, conventional tomography, and CT scans. Following investigation all cases underwent cranio-facial surgery to remove the encephalocele, rebuild the nose, and correct any orbital displacement.

RESULTS AND DISCUSSION

All of our cases presented with a significant facial deformity secondary to the herniating meningoencephalocele. In all cases the encephalocele extruded through the foramen caecum, displacing the eyes laterally, the nose inferiorly, causing a varying degree of telecanthus and hypertelorism with a long midface (Figure 1).

Suwanwela has reviewed a series of fronto-ethmoidal meningoencephaloceles presenting in Thailand, and has provided a sub-classification of this deformity based on autopsy findings in those patients which died in childhood. Our studies confirmed this classification.

The first subgroup is the naso-frontal type where the skull defect presents at the root of the nose, pushing the nasal bone inferiorly. The cribriform plate, crista galli, and associated anterior cranial fossae are tilted downward resulting in a deep cranial fossa centrally.

The second subgroup is the naso-ethmoid type, where the nasal bones remain attached to the frontal bone, and the encephalocele presents between the nasal bones above and the nasal cartilage and septum below.

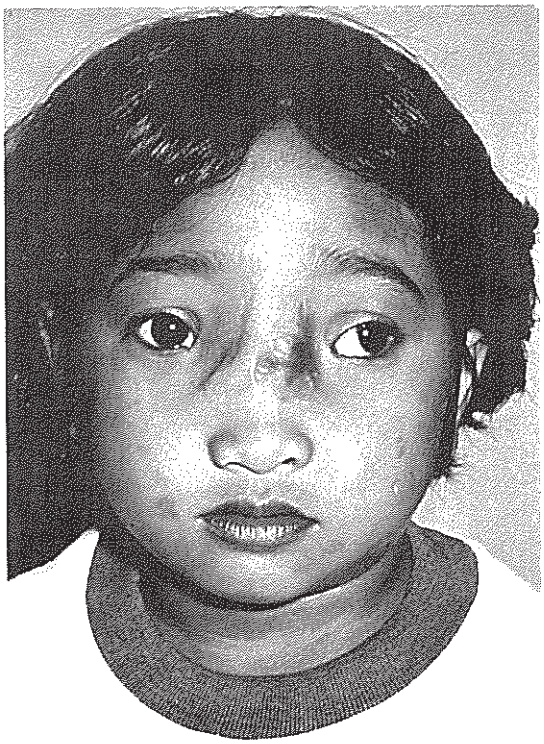

Figure 2: Malaysian child with fronto-ethmoidal meningoencephalocele showing moderate hypertelorism, with telecanthus, and displacement of the midface downwards causing a long face.

The third group is the naso-orbital type, which was the most common in our series, where the frontal and nasal bones were in their normal relationship, but the frontal process of the maxilla was defective on one or both sides, with protrusion of encephalocele through the medial orbital walls unilaterally or bilaterally.

Analysis of the orbital measurements revealed an interesting situation. The medial intercanthal distance was greater than the 97th percentile for age in all but one of our cases. The interpupillary distance was less dramatic, with most at or just above the 97th percentile for age, but several actually below this figure. The lateral intercanthal distance was the least distorted, with all cases at or below the 97th percentile for age. This indicates a greater degree of telecanthus than actual hypertelorism in nearly all cases. Our interpretation of these figures and other facial features, especially the elongated face, is that the encephalocele acts as a foreign body displacing an otherwise normal anatomy. (Figure 2).

This contrasts with the classic cranio-facial clefts described by Paul Tessier in which there is a wide face, hypertelorism, and maxillary hypoplasia with shortening and retrusion of the middle third of the face.

The ocular abnormalities arise either specifically from the primary anatomical defect

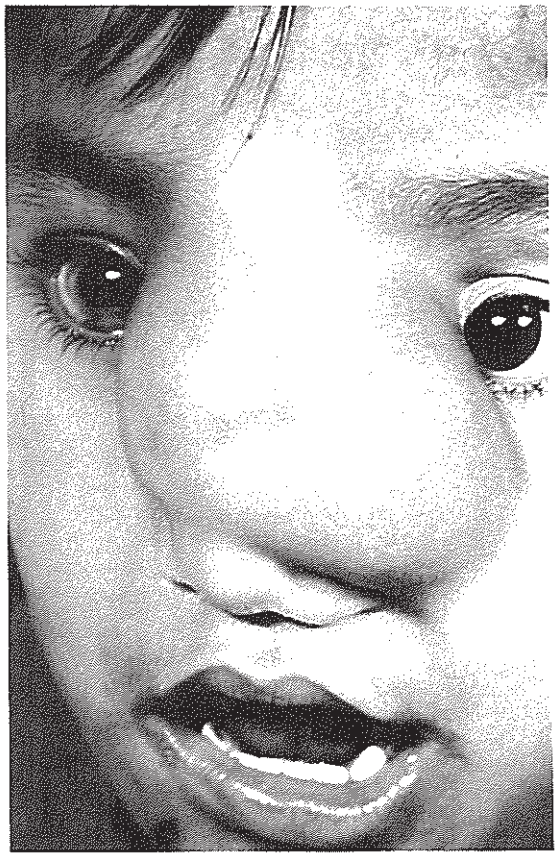

Figure 3: Malaysian child with very large naso-frontal type encephalocele projecting forward as a probosis like structure displacing the eyes laterally and the nose downwards.

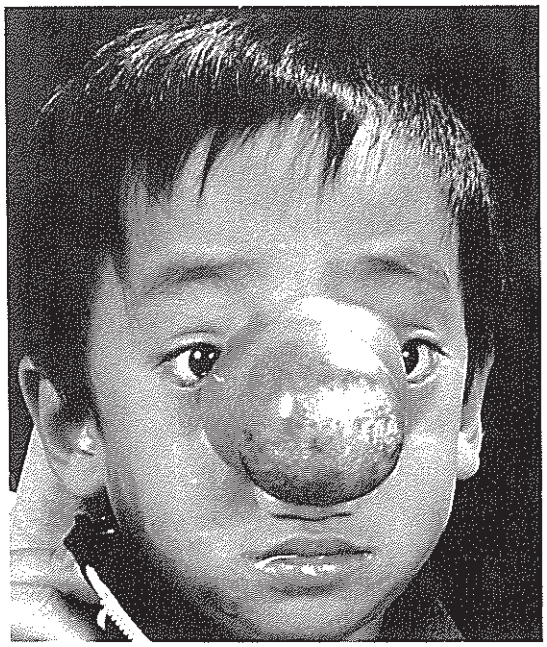

Figure 4: Malaysian child with large encephalocele displacing the orbits, and with the right eye showing a central corneal leucoma and some microophthalmia.

or as a separate ophthalmological problem in association with it. The first group includes the hypertelorism and telecanthus which have followed from the displacement of the orbits laterally, and the resultant filling of the median defect with dysplastic and usually gliotic non-functioning cerebral tissue (Figure 3).

Five cases in which the encephalocele presented through the medial orbital walls have watering eyes with destruction of the normal lacrimal apparatus. The abnormality was so extensive that generally I was not sanguine of success from such procedures as routine dacrycystorhinostomy or the insertion of a Lester Jones tube, as in all cases the lacrimal anatomy was either absent or grossly distorted at the inner canthi.

In two cases there was a significant degree of orbital dystopia with the encephalocele displacing one eye superiorly. This causes few ocular problems and does not preclude the development of binocular vision and stereopsis provided the visual axes remain parallel.

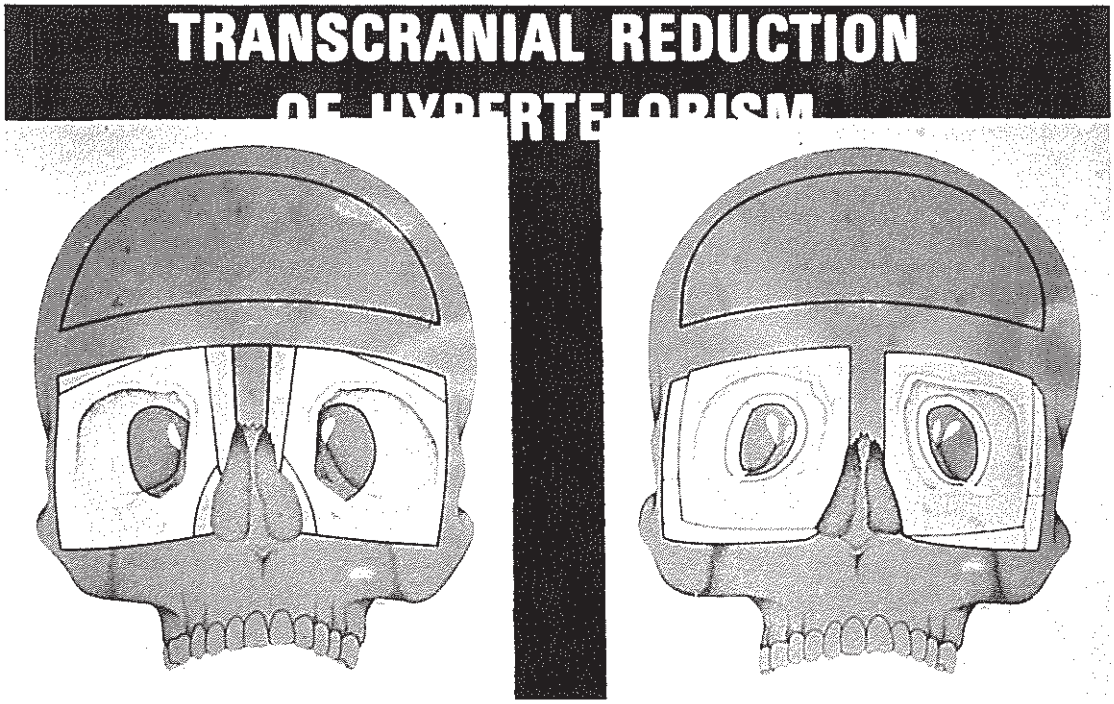

Figure 5: Diagram showing the way bone is removed to allow medial translocation of the orbits for correction of bilateral hypertelorism.