ORBITAL MENINGO-ENCEPHALOCOELE AND EXOPHTHALMOS*

BY

A. MORTADA AND I. EL-TORAEI

From the Departments of Ophthalmology and Surgery, Kasr El-Aini Faculty of Medicine, Cairo University, Cairo, Egypt

ORBITAL defects are rare congenital abnormalities and the protrusion of intracranial contents into the orbit is a rare cause of congenital exophthalmos, less than 100 cases having been recorded. Duke-Elder (1952) states "An orbital cephalocoele, wherein a portion of the contents of the skull protrudes into the orbit, is not common; some eighty cases are on record since Brechet's original publication in 1831". The development of the orbit is complicated and is available in text-books of anatomy and embryology. It may be concluded that it is formed from the mesoderm surrounding the eyeball by condensation into bone (Spöndli, 1846; von Kölliker, 1861; Schultze, 1896), and that both membrane and cartilage bone share in its formation. The multiplicity of the embryonic parts sharing in the formation of the bony orbit allows gaps to occur through which cranio-orbital communication or herniation may take place. Herniations occur not only through these defective gaps but also through the natural openings, *e.g.* the optic foramen or the sphenoid fissure.

The defects are of two types, anterior and posterior. The anterior type is the commonest and it is located between the frontal bone, lacrimal bone, cribriform bone and the nasal process of the maxilla. The cephalocoele protrudes forwards "from the inner canthus onto the face or laterally displaces the eye" outwards. Sometimes it is visible at the base of the nose. "It may be bilateral and symmetrical (de Britto, 1904; Rohmer, 1905a,b; Peters, 1917)". The sac is usually connected with the cranial cavity by a pedicle which insinuates itself between one or other of the cranial sutures, thus producing a defect in the bone although occasionally "no communication with the cranial cavity is detected (di Marzio, 1924)". This last form may be called a sequestered meningocele.

The posterior type is less common. Strandberg (1949), who reviewed the literature from 1841 to 1948, reported 31 cases of this type and added one of his own, and Tayebi and Silverman (1956) reported two cases. The cranial contents may protrude through a natural opening, such as the optic foramen, the superior orbital fissure, or the posterior ethmoid foramen, or through a

* Received for publication May 4, 1959.

defect in the bony orbit, usually at the apex, sometimes the roof, occasionally the medial wall, and exceptionally the lateral wall. The defect may be small or very extensive; in a case reported by Tauber (1900), ossification of the roof, floor, and apex of the orbit was so incomplete that the cranial cavity and the maxillary antrum formed a common cavity. One or both wings of the sphenoid may be ill-developed or absent (Jaensch, 1926, 1928, 1941; Scullica, 1927; Strandberg, 1949). Other abnormalities of the skull and brain may coexist. Similarly the eye itself may be normal or abnormal. Of the 31 cases collected by Strandberg, four had microphthalmos and one had coloboma of the optic nerve. Other cephaloceles may occur in the temporal region (Jaensch, 1926, 1928, 1941; Schreyer and Sprenger, 1927; Strandberg, 1949), the occipital region (Jaensch, 1926; van der Hoeve, 1935) or the ear (Jaensch, 1926; Schreyer and Sprenger, 1927). Vascular anomalies may also exist. In Strandberg's case, the carotid artery was peculiarly twisted in relation to the dura mater. Dandy (1929) found a large extradural vein at operation which seemed to have replaced the cavernous sinus, and the internal carotid artery was absent on the affected side. In Tayebi and Silverman's case there was asymmetry of the skull vault. The reported anomalies in the eye include coloboma, microphthalmos, hydrophthalmos, and apparent anophthalmos (Tauber, 1900; Cohen, 1927; Jaensch, 1926; von Hippel, 1906; Borochovic, 1930). Neurofibromatosis associated with orbital defects has also been recorded (Rockliffe and Parsons, 1904; LeWald, 1933; Seaman and Furlow, 1954; Wheeler, 1936; Bruwen and Kierland, 1955; Whiting, 1938). There may also be orbital neurofibromata, but these cases do not belong to the orbital meningo-encephalocele group.

The herniated part may be a meningocele, cephalocele, or hydrencephalocele. In most cases brain substance is included, although this is usually atrophic and may appear oedematous and degenerated as the wall of a cyst. A true meningocele is rare. The dura is fibrous and is adherent to the surroundings or to the skin or it may be defective. The pia-arachnoid tissues are hardly recognizable.

Clinical Appearance

The condition may be evident at birth, but usually develops in infancy and childhood, and is rarely delayed until adult life. Females are affected about twice as often as males. There is no evidence of an hereditary tendency. Trauma to the head may cause the manifestation or accentuation of the condition. Internal hydrocephalus, orbital infections, and neoplasms are rarely exciting factors. It is very unusual to find congenital orbital defects by chance in the routine skull radiographs which are carried out to investigate suspected fractures. Small defects may cause no symptoms.

The essential symptom is a slowly progressive proptosis showing pulsations synchronous with the heart, usually unchanged with the compression of

one carotid but reduced on pressing both arteries. Coughing or straining usually increases the protrusion of the eye. Slight pressure on the globe usually reduces the proptosis to some extent and may produce some giddiness and nausea. On the other hand, however, the exophthalmos may not pulsate at all.

In the interior type the cephalocoele appears at the inner angle of the eye and displaces the globe downwards and outwards; but the cyst may appear above the globe (Whiting, 1938), in the neighbourhood of the lacrimal gland (Zeidler, 1926), in the floor of the orbit (van Duyse, 1920), or in the lateral wall displacing the eyeball in the opposite direction. The displacement is sometimes so marked as to cause congenital luxation of the eyeball (Speciale-Cirincione, 1923). These cysts may pulsate and may bulge more on straining; sometimes this sign is very marked and at other times it is negligible or absent. The pulsation is reduced by bilateral carotid pressure and the size is reduced by pressure and by withdrawal of cerebrospinal fluid. Pressure may produce cerebral symptoms such as slowing of the pulse, convulsions, or even coma. In our case we noticed that the size increased when the patient's head was bent down, and after the injection of air into the thecal space. The overlying skin may be normal but more frequently it is hyperaemic and it may be very thin. Oedema of the lids is common, especially in the upper lid. Ulceration rarely occurs; it is usually initiated by trauma leading to very serious infection.

Diagnosis

This is usually possible when the above signs are present, but is sometimes difficult. Aspiration of cerebrospinal fluid from the cyst or after injection of phenolphthalein in the lumbar theca is diagnostic. Radiological features are important. A defect in the orbit will correspond to the site of the abnormality, which may be of variable size, and the orbit itself will be enlarged. When the defect is big with prominent cerebral herniation, the anterior cranial fossa is reduced in size as evidenced by displacement of the sphenoid ridge forward and upward. At the same time the middle fossa is increased, as is evidenced by a forward bulge in the floor in the coronal and sagittal projections. In the lateral views the usual parallelism of the orbital roofs and of the floors of the middle fossa is lost. The line of the orbital roof in the lateral view is shorter on the affected side, and when the roof is lacking only one line is seen instead of two. In the posterior type of lesion the apex of the orbit may be lacking, either alone or with the adjacent part of the roof and side walls. The wings of the sphenoid may not be identifiable, and the orbital fissure may be enlarged compared with the other side. The orbital foramen may be normal or enlarged. Other defects may be present in the skull and in the maxilla. Pneumo-encephalography may show subarachnoid air in the orbit, especially by means of lateral horizontal

beam examination in the "brow up" position. The ventricular system is usually normal, but there asymmetry or ipsilateral dilatation or deformity may be caused by the herniation. Associated hydrocephalus may also be detected. Cerebral angiography may show vascular anomalies in association with or conforming to the cerebral herniation. The condition has to be differentiated from other causes of exophthalmos.

Treatment

The treatment of orbital cephalocoele, especially the posterior variety, is difficult and the mortality is high because of the age of these patients and the development of such complications as meningitis and cerebrospinal fistulae. Old measures, such as ligation of the carotid or the injection of irritants, are to be condemned. The transorbital route is difficult and very unsatisfactory. The transfrontal technique, which was successfully tried by Dandy (1929) for a case of orbital meningocele, is the best. Birch-Hirschfeld (1915) found only five successful cases out of 43. If the cyst is a meningocele sac, it is dissected and ligated and then excised. If the defect is large it is closed by a bone graft, generally taken from the skull vault, by a dural substitute (fascia lata or pericranium), or by gel film. Synthetic material (such as polyethylene) and metals (such as tantalum or vitallium) have not been used. Enucleation of the eye is not indicated except when the globe is destroyed or congenitally abnormal. Partial recurrences of the proptosis may occur in large defects and may need a second surgical intervention.

Case Report

A female child 18 months old was admitted to hospital on January 14, 1959. She had been born at full-term by forceps delivery, and proptosis of the left eye had been present since birth. The mother had had two other deliveries also by forceps, and one of these babies had died, and she had also had three abortions. There was no similar abnormality in any member of the family. The mother had had no fever or illness during pregnancy but she had had a non-adherent leucoma in the left eye for many years.

Examination.—The child was of normal weight and in good general condition. The size of the skull was normal.

The left eye was proptosed and was displaced downwards and laterally. The oedematous eyelids hid the eyeball (Fig. 1). Examination of the eye by speculum retraction showed no anomaly. On the nasal side of the orbit there was a cystic swelling covered by thin skin showing dilated veins. It was compressible and slightly pulsating, and increased in size when the child was crying. Firm pressure on the swelling caused the child to faint. Compression of both carotids stopped the pulsation.

The right eye was normal. Ear, nose, and throat, heart, chest, and abdomen were normal. The blood, urine, and cerebrospinal fluid were normal.

The child was not mentally backward. X-ray of the skull showed a wide left orbit, wide orbital fissure, thin orbital roof, and a defect in the nasal wall of the orbit (Fig. 2).

Encephalography showed some air in the left orbit (Fig. 3) and some dilatation of the anterior horn of the lateral ventricle (Fig. 4).

FIG. 1.—Swelling on the nasal side of orbit with proptosis. The eye is hidden by the oedematous lids. Notice the thin skin over the swelling.

FIG. 2.—Radiograph of the skull, showing left orbit much enlarged, thin defective orbital roof, and thin nasal wall. The orbital fissure is dilated.

FIG. 3.—Encephalogram, showing air in left orbit.

FIG. 4.—Encephalogram, showing dilated anterior horn of lateral ventricle.

Operation.—This was done by one of us (I.T.) under gas-oxygen-trilene anaesthesia with endotracheal intubation. A frontal osteoplastic flap was raised (F. Krause). The dura was opened and the frontal lobe retracted by a spatula. The brain showed a very thin, rather transparent area on the under surface of the frontal lobe about 1 inch in diameter. Further retraction of the brain showed a defect large enough to admit two fingers at the roof of the orbit opposite to the thinned brain area. There was an incomplete sac which was partially removed, and the defect was closed by a pericranial flap taken from the neighbourhood and blocked by a cube of Spongell. The brain was repositioned, the dural opening partially closed, the bone flap replaced and fixed, and the scalp closed with drainage.

Result.—The proptosis receded completely and the post-operative course was uneventful.

Summary

Orbital meningo-encephalocoele is a rare congenital abnormality causing exophthalmos. The classification, aetiology, pathology, clinical appearance, diagnosis, and treatment are discussed. A case treated successfully by operation is the first to be reported from Egypt.

REFERENCES

- BIRCH-HIRSCHFELD, A. (1915). In "Handbuch der gesamten Augenheilkunde", ed. A. Graefe and T. Saemisch, 2nd ed., Band 9, abt. 1, teil 1, p. 539. Springer, Berlin.
- BOROCHOVIC (1930). *Oftal. Z.*, 3, 122.
- BRITTO, V. DE (1904). *Arch. Ophtal.*, 24, 142.
- BRUWER, A. J., and KIERLAND, R. R. (1955). *A.M.A. Arch. Ophthal.*, 53, 2.
- COHEN, M. (1927). *J. Amer. med. Assn.*, 89, 746.
- DANDY, W. E. (1929). *Arch. Ophthal. (Chicago)*, 2, 123.
- DUKE-ELDER, S. (1952). "Text-book of Ophthalmology", vol. 5, p. 4754. Kimpton, London.
- DUYSE, VAN (1920). *Arch. Ophtal.*, 37, 519.
- HIPPEL, E. VON (1906). *v. Graefes Arch. Ophthal.*, 63, 1.
- HOEVE, J. VAN DER (1935). *Ophthal. Valencia*, 1, 237.
- JAENSCH, P. A. (1926). *Klin. Mbl. Augenheilk.*, 76, 433.
- (1928). *Ber. ophthal. Ges. Heidelberg*, 47, 455.
- (1941). *Klin. Mbl. Augenheilk.*, 107, 561.
- KÖLLIKER, A. (1861). "Entwicklungsgeschichte des Menschen und der höheren Thiere". Engleemann, Leipzig.
- LEWALD, L. T. (1933). *Amer. J. Roentgenol.*, 30, 756.
- MARZIO, Q. DI (1924). *Riv. Oto-neuro-oftal.*, 1, 507.
- PARSONS, J. H., and COATS, G. (1906). *Brain*, 29, 209.
- PETERS, R. (1917). *Klin. Mbl. Augenheilk.*, 59, 553.
- ROCKLIFFE, W. C., and PARSONS, J. H. (1904). *Trans. path. Soc. Lond.*, 55, 27.
- ROHMER (1905a). *Arch. Ophtal.*, 25, 329.
- (1905b). *Bull. Soc. franç. Ophtal.*, 22, 359.
- SCHREYER, W., and SPRENGER, W. (1927). *Z. Hals-Nasen-Ohrenheilk.*, 17, 252.
- SCHULTZE, O. (1896). "Grundriss der Entwicklungsgeschichte des Menschen und der Säugethiere".
- SCULLICA, F. (1927). *Ann. Ottal.*, 55, 734.
- SEAMAN, W. B., and FURLOW, L. T. (1954). *Amer. J. Roentgenol.*, 71, 51.
- SPECIALE-CIRINCIONE, F. (1923). *Ann. Ottal.*, 51, 68.
- SPÖNDLI, H. (1846). "Über den Primordialschädel der Säugethiere und der Menschen". Mayer and Zeller, Zürich.
- STRANDBERG, B. (1949). *Arch. Ophthal. (Chicago)*, 42, 254.
- TAUBER, A. (1900). *Arch. klin. Chir.*, 61, 347.
- TAYEBI, H., and SILVERMAN, F. N. (1956). *A.M.A. J. Dis. Child.*, 92, 138.
- WHEELER, J. M. (1936). *Bull. neurol. Inst. (N.Y.)*, 5, 476.
- WHITING, H. M. (1938). *Trans. ophthal. Soc. U.K.*, 58, 58.
- ZEIDLER, M. (1926). *Klin. Mbl. Augenheilk.*, 77, 390.