CASE NOTE

ORBITAL ENCEPHALOCELE ASSOCIATED WITH ACROCEPHALY*

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ORBITAL encephalocele is a congenital tumour, which is supposed, according to its definition, to be in communication with the intracranial cavity. Its existence suggests the presence of a defect in the bones of the orbit. The theories which pretend to explain the pathogenesis of this anomaly are based on two principal ideas:

- (a) an ossification anomaly of the skull is the primary factor of the encephalocele;
- (b) an hydropic cyst of the cornu ventriculi, with or without added glial proliferation, is formed first, and the bone defect of the skull results from its occupation by the cyst.

Our case seems to corroborate the first idea.

Though there is a great diversity of opinion on this subject, most authors agree that the coexistence of other anomalies of the skull with those of the eye is almost constant. Our case presents two peculiarities: first, the encephalocele had been mistaken for an angioma of the orbit and its diagnosis was ascertained only by the help of various laboratory and radiological examinations; secondly, it was associated with an oxycephaly, the only symptom of which was the characteristic aspect of the skull.

Case Report

M. C., a young man, aged 25, was sent to us by a military doctor from a recruiting office. The patient had never complained of a small tumour at the right of the root of the nose nor of a bigger one occupying the inner part of the left orbit (Figs 1 and 2, overleaf). The medical history revealed little information about the development of these two tumours; according to the patient they had always remained the same size.

The small tumour at the right of the root of the nose, 2 cm. across, had pushed the right eyeball towards the infero-external angle of the orbit without producing a direct exophthalmos. A large vein ran vertically under the skin, on the inner side of this tumour, which was of soft consistency and was totally reducible. Palpation revealed a regular, oval, bony border, the inward part of which seemed to be formed by a prolongation of the spine of the frontal bone, ending near the dorsum nasi, so that a narrow gap was formed between the two. No fluctuation, pulsation, or thrill was noticed in this tumour. There were no pathological findings in the right eye.

* Received for publication October 24, 1951.

FIG. 1.—Front view of the two tumours.

FIG. 2.—Side view from the left.

The tumour on the left had pushed the left eyeball towards the external wall of the orbit. The ciliary portion of the eyelid border seemed shortened and the lacrimal portion considerably lengthened. The rima palpebrarum was narrower than normal. The puncta lacrimalia and the upper and lower fornices were quite normal. A conjunctival fold starting from the inner canthus covered the internal two-thirds of the cornea. The movements of the eyelids and eyeball were normal. The frontal apophysis of the left superior maxilla was more protuberant than that on the right, and this, in addition to the oblique direction of the dorsum nasi, gave to the face a typical asymmetrical appearance. This tumour also was of soft consistency, but could be only partially reduced under pressure. Palpation revealed a regular, oval, bony border, the concavity of which was turned towards the root of the nose. When the patient, pressing his nose, made a forced expiration, the tumour in the right orbit showed a slight expansion, but the left showed no change. Irrigation of the lacrimal canals showed the left one to be obstructed.

Ophthalmoscopic examination revealed a narrowing of the arteries and the presence of a scleral crescent in the right fundus. Since the left cornea was almost totally covered with a conjunctival fold, only a small part of the peripheral retina could be seen, and this was found to be normal.

Vision in the right eye was 20/20; in the left counting fingers at 25 cm.

Wassermann, Mantoux, Casoni, and Weinberg reactions were negative. A puncture was made in the tumour of the left orbit, and 4 to 5 ml. of a transparent fluid, very similar to the cerebrospinal fluid, was obtained. Chemical and microscopical examinations of the fluid revealed the presence of sodium chloride and a few mononuclear cells, which confirmed our presumption that the tumour might be an encephalocele.

Radiological examination helped to corroborate this diagnosis by revealing the presence of a congenital bone defect in the region of the root of the nose. In the antero-posterior radiograph of the skull taken in the nose-forehead position, the bone defect had the shape of an apple (Fig. 3). The left ethmoidal cells seemed more enlarged than those on the right. The existence of the frontal sinuses could not be determined. The two spines of the frontal bone could be seen clearly. The radiographs taken in the optic foramen position showed that the bone defect was in the inner walls of the two orbits

(Figs 4 and 5). Ombredanne (1947) presented a similar case with a bone defect in the same position.

The skull of our patient had the characteristic shape of a tower skull; it was very difficult to decide whether this was a case of ocular hypertelorism (Crouzon's disease): indeed there was no forward protrusion of the eyeballs, no atrophy of the optic nerves, no diminution of the vision, and none of the other secondary signs and

FIG. 3.—Antero-posterior radiograph of skull.

FIG. 4.—Radiograph of right optic foramen.

FIG. 5.—Radiograph of left optic foramen.

FIG. 6.—Radiograph of left sella turcica.

symptoms often encountered in this type of case. The optic foramina and the sella turcica were quite normal (Figs 4, 5, and 6). Only a slight degree of mental deficiency was

apparent such as is quite often seen in cases of encephalocele, and the tower skull was the only symptom.

The encephalocele may impede the manifestation of intracranial hypertension and explain the lack of symptoms. Crouzon's disease is attributed by many authors to an early ossification of the skull, and the coexistence of these two anomalies may suggest that an ossification anomaly was also a pathogenic factor in the development of the encephalocele.

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