Dept. Surg., Anaesth. & Radiol.
Fac. Vet. Medicine, Cairo Univ.
Head of Dept. : Prof. Dr. Y Khamis

CONGENITAL ENCEPHALOCELE IN TWO KIDS

(With 4 Fig)

By

S.T. EL-ZOMOR; K.A. FARAG; M.B. MOSTAFA*;
and F.M. FARAG**

(Received at 6/9/1993)

فتاق دماغي وراثي في جديين

سيد الزمر ، بوكات مصطفى ، فؤاد فرج

تم تسجيل حالتين من الفتاق الدماغى المائى فى عدد ٢ جدى.
وأظهرت الصوره الشعاعيه رتق العظام الجبهيه . وباجراء الصفه التشريحيه اتضح اتساع البطينين الوحشى والثالث مع وجود فتحه كبيره فى العظام الجبهيه يخرج منها أنسجة المخ وأغشيته.

*: Dept. Vet. Surgery, Anesthesiology and Radiology, Cairo Univ.
**: Dept. Vet. Anatomy, Cairo Univ.

SUMMARY

Encephalocele associated with hydrocephalus were recorded in two kids. The radiographic appearance showed partial agenesis of the frontal bone. The necropsy finding revealed dilated lateral and third ventricles, large bone cleft in the frontal bone and protrusion of the brain tissues with its meninges through this cleft (Encephalocele).

History and clinical examination:

Two kids 3 days old were admitted to the Surgery Clinic, Faculty of Veterinary Medicine, Cairo University, with a history of swelling at the frontal region. The clinical examination revealed dullness, proptosis of the left eyes, incoordination and trembling of the limbs. The swellings were fluctuating and of an orange size (Fig. 1). Plain radiograph showed a radioopaque swelling at the frontal region. Partial agenesis of the frontal and partial compression of the parietal bones were observed (Fig. 2).

Necropsy findings showed that the swelling measured 3.5 cm height, 4 cm length and 4 cm width and protruded through a defect in the frontal bone. Longitudinal section of the head revealed that the swelling consisted of dilated lateral ventricles filled with cerebrospinal fluid, surrounded by part of cerebral hemisphere with its meningeal investment (Fig. 3).

The lateralventricles were severely dilated and accompanied by dilation of the interventricular foramen (Foramen of Monro) The frontal pole of the cerebral hemisphere was thin walled measuring, 1.5 mm cranially and 1 mm caudally. The occipital pole was abnormally overlaped on the cerebellum and corpora quadrigemina (Fig. 4). The frontal bone defect is ovoid in shape and extended from the frontonasal suture cranially to about 0.5 cm from the frontoparietal suture caudally.

DISCUSSION

The clinical, radiographic and necropsy findings revealed partial bone agenesis (Cranium bifidum) with protrusion of brain tissues with its meninges (Encephalocele). Moreover, the lateral ventricles were dilated and filled with CSF (Hydrocephalus). There is no available record of such case in kids, however, similar cases were previously reported in pups (*CARMICHEAL et al, 1983*) in calves (KHAMIS et al., *1992*), *in pigglets (STEWART et al., 1972; and Vogt et al.1986)*, and in

infants (*NELSON* et al., *1975*). Most studies incriminated hereditary influence on the development of such cases (*STEWART* et al., *1972*; *VOGT* et al., *1986*; *and OJALA* et al., *1992*). However, de *LAHAUNTA* et al. (*1983*) attributed the cause of similar cases in pups to malformation of the mesencephalic aquiduct resulting in a non communicating obstructive hydrocephalus with severe dilation of the third and lateral ventricles. This may be accompanied by a massive expansion of the cranial cavity and occurrence of large nonossified portions of the calvaria. Moreover, *NELSON* et al. (*1975*) mentioned that encephalocele is accompanied with hydrocephalus in approximately two thirds of affected infants. In conclusion, dilation of the lateral and third ventricles with CSF leads to formation of hydrocephalus. The resultant massive expansion of the cranial cavity produced bone cleft in the frontal bone through which herniation of the brain tissues with its meningeal covering would occur forming encephalocele.

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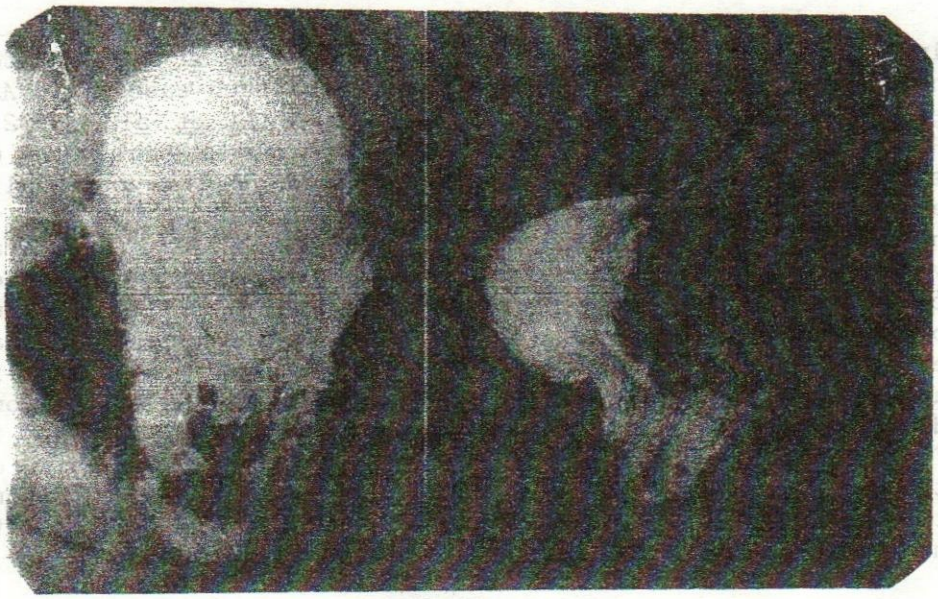

Fig 1: Encephalocele in a kid.

Left : Anterior view showing proptosis of the left eye.

Right: Lateral view showing swelling at the frontal region.

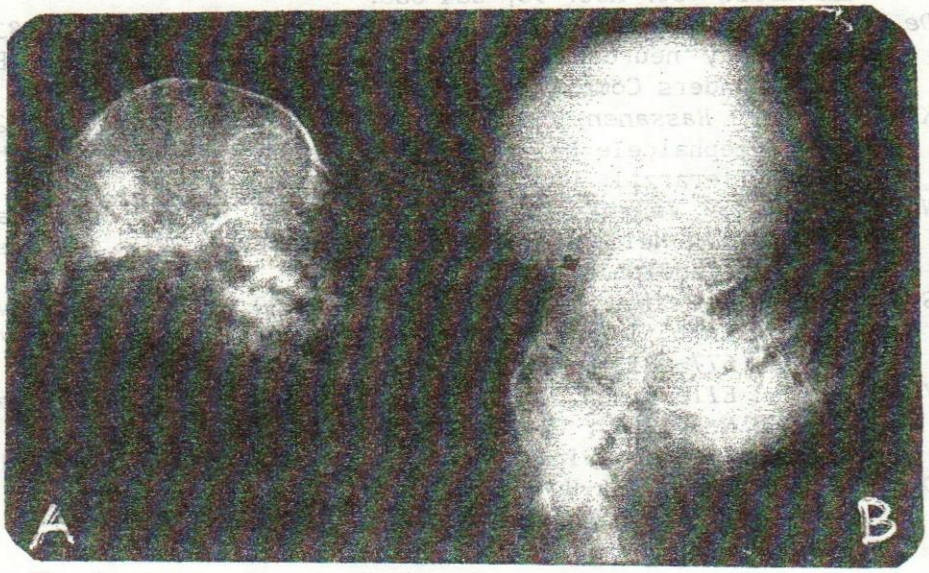

Fig. 2: Lateral plain radiograph of the head.

A: Normal radiograph.

B: Radiograph of kid affected with encephalocele showing radioopaque swelling, compressed parietal bone (short arrow), and partial agenesis of the frontal bone (coarse arrow).

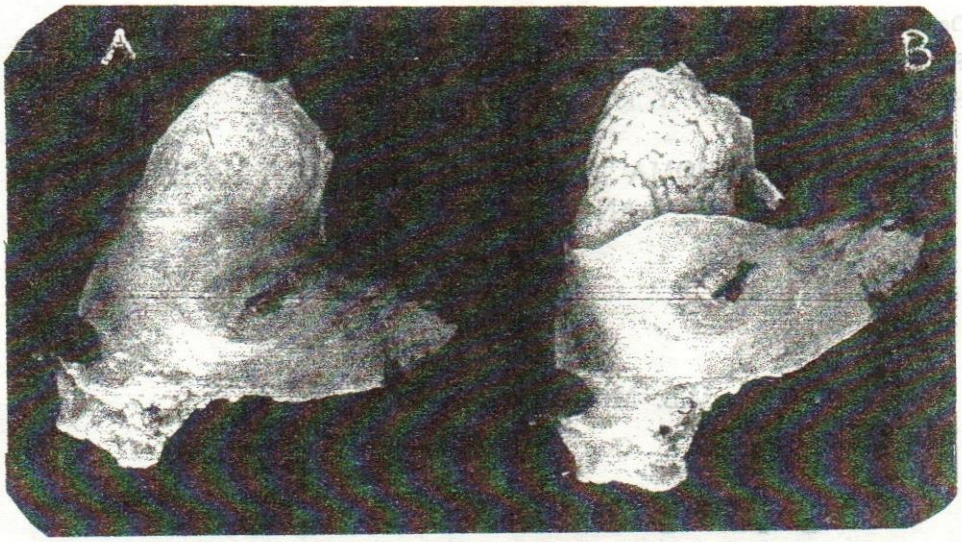

Fig. 3: Lateral view of the right side of the head specimen showing the frontal defect and prolapsed part of the cerebral hemisphere

A: Before removal of the meninges.
B: After removal of the meninges.

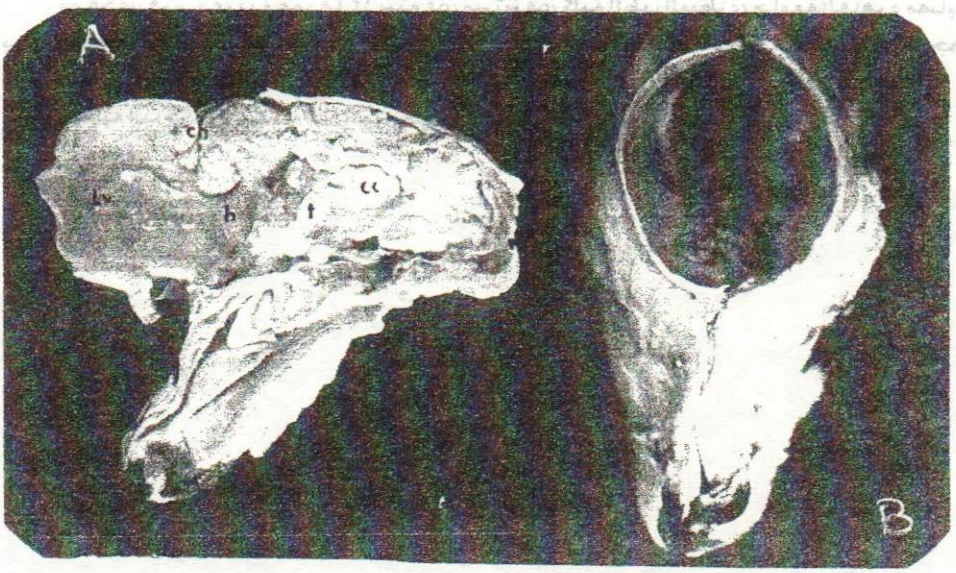

Fig. 4 A: Medial view of the right side of the head specimen.
lv: lateral ventricle ch: cerebral hemisphere
cb: cerebellum h: hippocampus
t: thalamus cq: corpora quadrigemina

B: Dorsal view of the head showing the frontal bone defect.