ADAMANTINOMA PRESENT AT BIRTH

SUMMARY

A case of a large hemispherical cyst of unknown pathology is reported. It presented with anomalous clinical features. It was treated by simple intraventricular drainage. Encephalograms made 8 years later show the communication still in existence.

CASE REPORT OF AN ADAMANTINOMA PRESENT AT BIRTH

C. G. SUBRAMANI IYER, M.D.*

Neurological Unit, Boston City Hospital, and the Department of Neurology, Harvard Medical School, Boston, Massachusetts

(Received for publication October 23, 1951)

A perusal of the available literature has not revealed any instance of the presence of a pituitary adamantinoma at birth. The following remarkable case is therefore placed on record.

Clinical History. The patient M., a girl, was the sixth child born to a mother whose prenatal history was not remarkable. The mother's blood was Rh positive. X-rays before birth revealed a very large fetal head. The mother was admitted to Boston City Hospital in labor on Mar. 1, 1951 with a B.P. of 140/100 and edema of the ankles. Urinalysis revealed 2+ albumin. In order to facilitate birth a lumbar puncture needle was inserted through the top of the presenting head into the lateral ventricle and xanthochromic fluid was permitted to drip out for about 2 hours. This reduced the head to approximately a third of its previous size and delivery was effected by low transverse caesarean section.

Examination. At birth the baby breathed normally and was noted to be in good condition. The heart rate and rhythm were normal and color of the skin was good. The head was markedly enlarged, somewhat asymmetrically with a parietal bulging on the left. The left parietal and temporal bones were extremely thin. Bilateral proptosis and lateral nystagmus were noted. The Moro reflex was sluggish; the deep tendon reflexes were all obtained and there was fanning of the toes on stroking the feet. The cry was high-pitched. The weight of the child at birth was recorded as 8 lbs. 14½ oz.

Laboratory Data. Hb. was 14.5 gms./100 cc.; RBC 7.6 million; WBC 6550/c.mm., with 48 per cent neutrophilic leucocytes, 46 per cent lymphocytes and 11 per cent eosinophils. Occasional nucleated erythrocytes were seen in the blood smear. The platelets appeared normal. The CSF 5 days after birth was slightly xanthochromic; protein was 40 mg. and chloride 690 mg. per cent. There were 260 red cells and no leucocytes/c.mm. of fluid. The sugar value was reported as too low to read. The ventricular fluid was also xanthochromic, with 2000 mg. of protein and 642 mg. of chloride/100 cc. There were 320 red cells and no leucocytes/c.mm. of fluid. The sugar was again reported as too low to read. Radiographs of the head showed a marked expansion of the skull in all dimensions, consistent with hydrocephalus. In one film (anteroposterior projection) a round patch of increased density to the right of the midline was observed.

Course. A few days after birth the baby became deeply jaundiced but this cleared up rapidly. Subsequent ophthalmoscopic examination revealed pale optic discs bilaterally; the disc margins were well defined and there was no trace of papilledema. During the following weeks there was a progressive enlargement of the head and all four extremities became spastic. Throughout the patient’s life, all of which was spent in hospital, the temperature was normal, except for occasional elevations to 100°F. and depressions to 95°F. The child died at the age of 3 months.

* Rockefeller Foundation Fellow from Indian Council of Medical Research, Neuropathology Unit, Tata Memorial Hospital, Bombay, India.
C. G. SUBRAMANI IYER

Postmortem Examination. Autopsy was performed approximately 24 hours after death. The body was poorly developed and emaciated and was in striking contrast to the enormous size of the head. The circumference of the head was 59 cm. Scanty hair was present over the head and the fontanelles were found to be unusually wide and bulging. The pupils were equal and measured 5 mm. The gums were edentulous and the navel was well healed. The length of the body was 55 cm. and the approximate weight was 15 lbs. There were no significant abnormalities outside of the central nervous system. Microscopic examination of the liver and spleen revealed large amounts of hemosiderin in the Kupffer cells and the histiocytes respectively. A few cells containing the same pigment were found in the interstitial tissues of the kidneys.

Neuropathological Findings. The dura mater was unusually thick and white over the superolateral surface of the brain. The cerebral hemispheres were large and finely convoluted. Over the occipital and temporal poles the cortex was paper-thin. At the base of the brain there were several moderately dense fibrinous adhesions between the dura and arachnoid. A firm mass presented on the inferior surface of the cerebral hemispheres in the midline encroaching upon the orbital plates of the frontal bones and obscuring from view the vessels and nerves at the base. It was found to arise from the region of the sella turcica and was firmly embedded in the base of the skull. The pituitary body was not found; it had apparently been destroyed by the mass. At the base of the brain (Fig. 1) the mass extended from the posterior third of the orbital surfaces of the frontal lobes to the anterior border of the pons.