

Hydranencephaly

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A case report of hydranencephaly is presented with discussion upon morphology and etiology. The diagnosis was made by autopsy examination upon the corpse of a six day old male infant.

Introduction

The term hydranencephaly was introduced for the first time by Spielmeyer in 1904.¹⁷ However, Breschet was the first to describe the condition in 1823.¹⁰ Later in 1929 Cruveilhier described it as anencephalie hydrocephalique.⁵ The term hydranencephaly designates a congenital malformation of the central nervous system in which cerebral hemispheres are reduced to fluid (C.S.F) filled membranous sac with a relatively normal sized cranium.

Case report

Six days old male baby was admitted to St Luke's Hospital with inertia, bradycardia and hypothermia. Suction and muscle reflexes were absent. Pupils reacted to light. The baby developed arrhythmia and died on the same day.

The baby was born to a twenty-year-old primigravida after 38 weeks of gestation. Mode of delivery was through forceps application. Post mortem examination revealed the corpse of a baby weighing 2 kg, the head circumference being 30 cm. Cracial cavity contained a fluid filled sac weighing 15 grams. The fluid was clear. No cerebral hemispheres were seen. Cerebellum was present. Spinal cord was developed normally. Carotid and basilar arteries were present. There were no other developmental abnormalities. Microscopic examination of the membranous sac revealed gliosis and no neurones

were seen. Sections of cerebellum and spinal cord revealed normal structure. The lungs showed oedema, congestion and pneumonia.

Etiology

The condition may develop any time between third month to full term utero.⁷ There have been only two cases reported wherein the condition developed post nately. Benda, in 1945, described a case in which the child developed hydranencephaly at the age of two years. The other case is described by Smith in 1967 where hydranencephaly developed subsequent to meningococcal meningitis in a child at the age of two years.¹⁸

Several etiological factors have been proposed in causation of hydranencephaly. Prenatal occlusion or agenesis of carotid artery and jugular vein was considered as essential pathogenetic mechanism in hydranencephaly by Norman in 1958.¹⁵ Myer in 1968 and 1969 produced hydranencephaly in monkeys by bilateral carotid and jugular ligation and by incomplete placental abruption.^{13,14} Wildi,²⁰ Haymaker⁹ and Booth³ considered Herpes Simplex and other viral encephalitis to be responsible for production of hydranencephaly in utero.

Halsey⁷ in 1971 considered infective process to be responsible for liquefactive necrosis of the brain and thus production by hydranencephaly. Despite the fact that Miller in 1963¹² and DeMyer in 1963⁶ found chromosomal abnormalities and suggested a causative contribution in production of cerebral malformation, there have been only three recorded familial instances where hydranencephaly developed in twins.^{8,16,17}

After perusal of the literature all these suggestions seem to be valid and well documented. Therefore one may conclude that hydranencephaly results from a variety of etiological factors and that pathogenetic mechanism may not be the same in all cases destructive process following vascular occlusion or agenesis, and infective process being the most likely pathways leading to production of hydranencephaly.

Morphology

Morphologic changes occur in the brain and eyes mainly. Cerebral atrophy, thalamic atrophy and absence of medullary pyramids are common to all cases. Cerebellar abnormalities, though not a constant feature, are a common occurrence.

The histologic structure of the hydranencephalic sac is usually predominantly astrocytic. It is usual for the ependymal lining to be lost in encephaloclastic process, though occasionally some fragments may be preserved. Neurones are absent. Absence of neurones is an important distinguishing feature, albeit posthumously, from severe hydrocephalus.

Changes in the eye consist mainly of gliosis of the inner retinal layer, complete absence of the ganglion cells, rods and cone cells and absence of retinal vessels.

There are only two instances where hydranencephaly was associated with changes in organs other than brain and eyes. Halsey et al⁷ reported a case in 1971 in which hydranencephaly was associated with polycystic kidneys. The other instance was reported by Bauer et al¹ in 1977 when they reported a case of hydranencephaly in association with gyrate scalp.

Diagnosis

Clinical features include hypothermia, bradycardia, physical inertia, feeding difficulties, incoordinated eye movements, light reflex may be absent, strabismus and nystagmus. Occasionally, jaundice is present. Positive transillumination of the cranium is always obtained. Diagnosis can be made antenatally by ultrasound.¹⁹

Prognosis

Prognosis in hydranencephaly is extremely poor and dependent upon the integrity of hypothalamus, corpus striatum and brain stem tegmentum. Most of the patients die within first month of life. Longest survival has been reported by Crome and Sylvester⁴ when the child lived up to 4½ years. However, the child had spasticity, convulsions and blindness.

An interesting case has been reported by Lorder¹¹ where a child with hydranencephaly had a relative normal development. The author fails to furnish any valid explanation for this phenomenon and admits this case to be an enigma.

Summary

Hydranencephaly is a congenital condition in which cerebral hemispheres are replaced by a thin sac containing C.S.F. The sac wall consists of a pia and arachnoid overlying the glial layer. The condition may result from a variety of developmental or destructive abnormalities any time in utero, e.g. vascular occlusion or agenesis of carotid artery and jugular vein and infections. Atrophy of cerebral hemispheres, atrophy of thalamus and absence of medullary pyramids are characteristics of all cases.

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