CASE REPORT

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Acallosal Brain in Sudden Infant Death Syndrome (SIDS)

REFERENCE: Patel, F., **"Acallosal Brain in Sudden Infant Death Syndrome (SIDS),"** *Journal of Forensic Sciences*, JFSCA, Vol. 37, No. 3, May 1992, pp. 873–875.

ABSTRACT: A definitive explanation of "crib" or "cot" death remains unknown. An unusual incidental autopsy finding of agenesis of the corpus callosum in a case presenting as "near miss" sudden infant death syndrome (SIDS) is discussed. Hitherto, only a single case associated with SIDS has been reported in the literature. The condition may be easily missed outside the interest in neuropathology.

KEYWORDS: pathology and biology, sudden infant death syndrome (SIDS), brain, corpus callosum, agenesis, crib death, cot death

In the sudden infant death syndrome (SIDS) there is usually no specific abnormality to be found at the autopsy [*1*]. This dilemma generates considerable disagreement among forensic/pediatric pathologists as to what is considered adequate as a cause of death, within the definition of SIDS [*2*]. The realm of neuropathology in SIDS may be in the embryonic stages [*3*], but the neuromorphology of the callosal agenesis has been well described [*4–6*].

The congenital absence of the corpus callosum is a rare cerebral malformation that is commonly reported [*4,7*], but only a single case has been associated with SIDS [*8*]. It is usually an isolated incidental finding at neonatal autopsy [*7*]. The diagnosis of a "crib death" or "cot death" is not threatened since it is only when the incidental pathology might reasonably be expected to be lethal that SIDS becomes untenable as a cause of death [*1*].

Case Report

Tm was a 5-week-old boy who was found cold and unresponsive a few hours after being put to bed in his cot. He was rushed to hospital in a coma and put on a life-support machine. There was profound metabolic acidosis, which was easily corrected. He developed hepato-renal failure. There were no seizures but intermittent mouthing and sucking movements were observed. The electroencephalogram (EEG) recorded a low amplitude trace.

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An extensive microbiological investigation was negative and haematological parameters were normal. A chromosomal study was unremarkable. The cranial computed tomography (CT) scan indicated severe cerebral edema, and there was no evidence of a space occupying lesion. The characteristic "wing" shape of the ventricles caused by callosal agenesis could not be discerned.

The medical history of note was typical for SIDS. The provisional clinical diagnosis was ischaemic-hypoxic encephalopathy in a "near miss" SIDS. The absence of brainstem reflexes with irreversible brain damage was established after 36-h post admission.

Postmortem Examination

The external examination showed a well nourished and well cared for male infant. The anthropometric parameters were in the 97 percentile. There was no evidence of violence. The internal examination showed a grossly swollen brain with flattened gyri and generalized softening. The rest of the systems were essentially unremarkable.

The formalin fixed brain weighed 765 g. The interhemispheric surfaces showed complete agenesis of the corpus callosum. There were no identifiable cingulate gyri. There was a characteristic radial pattern of the gyri and sulci at the edges of the lateral ventricles and the roof of the third ventricle (Fig. 1). The white matter was extremely soft. The ventricular system was compressed because of cerebral edema.

There was neuropathological confirmation of the absent corpus callosum by demonstrating an uninterrupted continuation of the leptomeninges with the ependymal lining of the ventricles. The overall feature was that of anoxic-ischaemic encephalopathy and consistent with "near miss" SIDS.

The cause of death was given as SIDS.

Discussion

The cause of the agenesis of the corpus callosum is not known [*5*]. It is most often associated with other cerebral malformations [*4*,*5*,*8*], for example, microencephaly, mi-

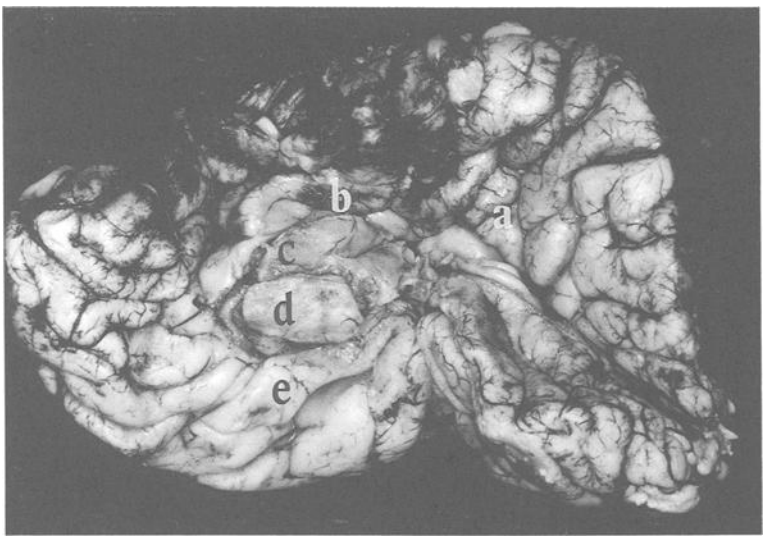

FIG. 1—*Midsaggital plane of acallosal brain hemisphere (left) showing* (a) *radially orientated gyral pattern in the fronto-parietal lobe.* (b) *The cingulate gyrus is absent.* (c) *The anatomical landmarks labelled are thalamus,* (d) *midbrain and* (e) *hippocampal gyrus.*

crogyri, cerebral heterotopia. The callosal dysgenesis has been reported in association with a variety of congenital syndromes, for example, Turner's, Dandy-Walker's. There is also an increased frequency of callosal agenesis in association with several inborn errors of metabolism [*9*], for example, nonketotic hyperglycinemia [*10*]. Although most cases of callosal agenesis are sporadic [*8*] a genetic basis should be sought because of association with chromosomal abnormalities [*4,8*].

An unexplained high incidence of callosal defect indirectly associated with spontaneous hypothermia has been observed [*11*]. There was no clinical evidence of hypothermia in the case reported.

The clinical presentation and natural history vary [*8*]. The cases may be neurologically asymptomatic but most cases present with seizure, mental retardation, or hydrocephalus [*5,7,8*]. This congenital anomaly is compatible with life.

Acknowledgment

The author is grateful to Dr. M. Honavar for the neuropathological examination.

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